Clinical significance of changes in pB–C2 distance in patients with Chiari Type I malformations following posterior fossa decompression: a single-institution experience

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OBJECTIVE The coexistence of Chiari malformation Type I (CM-I) and ventral brainstem compression (VBSC) has been well documented, but the change in VBSC after posterior fossa decompression (PFD) has undergone little investigation. In this study the authors evaluated VBSC in patients with CM-I and determined the change in VBSC after PFD, correlating changes in VBSC with clinical status and the need for further intervention.

METHODS Patients who underwent PFD for CM-I by the senior author from November 2005 to January 2013 with complete radiological records were included in the analysis. The following data were obtained: objective measure of VBSC (pB–C2 distance); relationship of odontoid to Chamberlain's, McGregor's, McRae's, and Wackenheim's lines; clival length; foramen magnum diameter; and basal angle. Statistical analyses were performed using paired t-tests and a mixed-effects ANOVA model.

RESULTS Thirty-one patients were included in the analysis. The mean age of the cohort was 10.0 years. There was a small but statistically significant increase in pB–C2 postoperatively (0.5 mm, $p < 0.0001$, mixed-effects ANOVA). Eleven patients had postoperative pB–C2 values greater than 9 mm. The mean distance from the odontoid tip to Wackenheim's line did not change after PFD, signifying postoperative occipitocervical stability. No patients underwent transoral odontoidectomy or occipitocervical fusion. No patients experienced clinical deterioration after PFD.

CONCLUSIONS The increase in pB–C2 in patients undergoing PFD may occur as a result of releasing the posterior vector on the ventral dura, allowing it to relax posteriorly. This increase appears to be well-tolerated, and a postoperative pB–C2 measurement of more than 9 mm in light of stable craniocervical metrics and a nonworsened clinical examination does not warrant further intervention.

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KEY WORDS Chiari malformation; brainstem compression; posterior fossa decompression

THE association of Chiari malformation Type I (CM-I) with ventral brainstem compression (VBSC), with or without basilar invagination, has been well-established, with a reported incidence of 4%–31%.^{5,6,8,10,12,13,15,16,18,20,21,25,26,28,31,35} A causal relationship for these so-called “complex Chiari” has not been elucidated to date, but in the treatment of symptomatic patients with both conditions, there is controversy as to what extent the brainstem compression should be surgically addressed, if at all.^{1–3,5–10,14–21,23,24,26,27,29,32,33,35} While symptomatic VBSC must be treated, procedures aimed at or related to allevi-

ating VBSC have substantial risks.^{14,32,35} As such, determining the surgical significance of VBSC before posterior fossa decompression (PFD) is crucial.

Many of the measurements used to track the course of VBSC depend largely on bone landmarks in the posterior fossa and posterior foramen magnum (e.g., the opisthion), which are removed during PFD. Moreover, these traditional measurements do not directly quantify the relationship of the ventral brainstem to the osseous structures compressing it anteriorly. To that end, a previously derived measurement system using ventral cervicomedullary anat-

ABBREVIATIONS CM = Chiari malformation; PFD = posterior fossa decompression; VBSC = ventral brainstem compression.

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omy, pB–C2, is useful in evaluating serial MR images of patients with CM-Is and VBSC.¹⁸

The purpose of this study was to evaluate the relationship between pre- and postoperative VBSC using the pB–C2 measurement in patients undergoing PFD for symptomatic CM-I. Furthermore, we sought to determine the clinical significance of changes in pB–C2 postoperatively to identify the risk of worsening VBSC in patients with PFD without circumferential decompression. We demonstrate our results based on a cohort of patients treated at our institution and discuss our findings in the context of the current literature.

Methods

Study Population

Prior to collection of data, this study was approved by and subsequently conducted in accordance with all institutional and local regulations and guidelines for human research. Departmental records were queried for all patients undergoing PFD for CM-I by the senior author (T.B.M.) at our institution from November 2005 to January 2013, which returned 83 patients. Hospital and clinical records were retrospectively reviewed and data were entered into a database for assimilation and analysis. Patients were excluded if they were presenting for revision of a previous PFD performed by another surgeon (4 patients). Patients were also necessarily excluded if their pre- or postoperative hospital and clinical records or MR images were unavailable (46 patients). Additionally, 1 patient with achondroplasia and 1 patient who upon review actually had CM Type II with myelomeningocele were excluded.

Thus, the study population consisted of 31 patients with complete clinical and radiological records. For these patients, we recorded the following data points: age at operation, sex, presenting symptoms, pre- and postoperative examination findings, pre- and postoperative radiological characteristics, associated conditions (e.g., scoliosis, tethered cord, syringomyelia, and hydrocephalus) and outcome, surgery performed, postoperative complications, outcome of presenting symptoms, and length of follow-up.

T2-weighted MR images were used to determine the objective grades of VBSC via the pB–C2 method, described previously¹⁸ (Fig. 1). We found that these measurements were easily performed on the T2-weighted images, which facilitated identification of the edge of the ventral dura. T1-weighted images were reviewed for cervicomedullary angle;³⁴ distance of tonsillar descent; odontoid tip distance from McRae's, Chamberlain's, McGregor's, and Wackenheim's lines; clival length; foramen magnum diameter; and basal angle (Fig. 2). It should be noted that not all measurements could be made in patients with only spine imaging.

All radiological measurements were performed by 2 authors (A.J.M. and S.S.A.) and were overseen and approved by the senior author. The pB–C2 measurements were performed 3 times each in all patients and were averaged. Measurement precision was excellent: among the sets of 3 pre- or postoperative measurements, the pB–C2 value did not differ by more than 0.3 mm.

It is important to note that pB–C2 may be affected by

FIG. 1. Illustration of the pB–C2 measurement. A line is drawn from the basion to the inferior-posterior aspect of C-2 (B–C2 line). pB–C2 is then measured as the perpendicular distance running through the odontoid tip between B–C2 (thick arrow) and the ventral cervicomedullary dura (small arrow). Reproduced with permission from Grabb PA, Mapstone TB, Oakes WJ: Ventral brain stem compression in pediatric and young adult patients with Chiari I malformations. *Neurosurgery* 44:520–528, 1999.

factors such as the degree of neck flexion or extension during the MRI. We did not directly account for this in our study, assuming that changes in positioning were small enough to not affect the measurements.

Surgical Technique

All surgeries were performed by the senior author with the following operative technique, which did not change during the study period. The patient is positioned prone in a Mayfield pinned head holder and the posterior cranium is shaved, prepared, and draped in the usual sterile fashion. A midline incision is made from the inion to the spinous process of C-2, and the muscles are divided at the midline raphe, exposing the posterior cranium, the posterior arch of C-1, and the superior aspect of C-2. Unless the tonsils are descended down to the level of C-2 (necessitating the removal of the superior aspect of the C-2 posterior arch), the origins of the suboccipital musculature on C-2 are left alone. The arch of C-1 is removed with Kerrison rongeurs. Two bur holes are placed approximately 1.5 cm from the midline, 2 cm above the foramen magnum. An occipital craniectomy is created using a bur and Kerrison rongeurs,

FIG. 2. Other lines and relationships used in the study are shown on sagittal T2-weighted MR images. McRae's (McR), Chamberlain's (Cham), McGregor's (McG), and Wackenheim's (Wack) lines are represented by dashed lines in A–D, respectively. **A:** Odontoid-McRae distance (C2-McR) and tonsillar descent (TD) are shown. **B:** Odontoid-Chamberlain distance (C2-Cham), basal angle (BA), and clival length (CL) are shown. **C:** Odontoid-McGregor distance (C2-McG) and cervicomedullary angle (CMA) are shown. **D:** Odontoid-Wackenheim distance (C2-Wack) is shown. Figure is available in color online only.

to the width of the cervical dura (about 3 cm). The dura is evaluated for pulsations, signifying satisfactory restoration of CSF flow after bone decompression. If the pulsations are inadequate or absent, the dura is then picked up with a 4-0 Nurolon suture and opened in a Y-shaped fashion. Any arachnoidal adhesions are lysed and the obex is visualized to ensure adequate CSF flow from the fourth ventricle and into the central canal. If the tonsils are enlarged or necrotic, they are carefully cauterized using a bipolar device to retract them rostrally and restore good CSF flow. An acellular tissue matrix is used for duraplasty, sutured with 4-0 Nurolon, and a watertight dural closure is confirmed with a Valsalva maneuver. The incision is then closed in layers and dressed in a sterile fashion.

Statistical Analysis

Descriptive statistics were calculated for continuous variables, including pre- and postoperative radiological measures and the change in measurements from pre- to postoperative time points. Frequency tables were created for categorical pre- and postoperative variables. To assess for differences between pre- and postoperative measures, a paired t-test was used after demonstrating normality of data. Some measurements could not be made on all patients, for example, when only spine imaging was available for review. For variables with small numbers of measurements, the Wilcoxon signed-rank test was used. The pre- versus postoperative comparison of pB–C2 was made using a mixed effects ANOVA model, in which a random

patient effect was fit along with a fixed time period effect. The Kruskal-Wallis test was used to test the differences in the mean ranks of continuous measures when making comparisons among more than two groups. Pearson's correlation was used to quantify the strength of the linear association between 2 continuous variables.

Results

Patient Population

Patient demographics are presented in Table 1. The mean age of the cohort was 10.0 years (range 18 months to 43 years), and the mean length of follow-up was 21 months (range 2–47 months). There were 16 male and 15 female patients. The mean tonsillar descent distance was 11.8 mm. The most common presenting symptom was headache (20 patients, 64.5%), followed by clumsiness (7 patients, 22.6%). Seven patients were diagnosed in association with scoliosis (22.6%). Seventeen patients (54.8%) were found to have syringomyelia. Three patients (9.7%) were found to have tethered cords, and 1 additional patient had received prior detethering. Two patients (6.5%) presented with hydrocephalus.

Intraoperatively, the majority of patients (25 patients, 80.6%) underwent duraplasty; in the remaining 6 patients, a dural opening was unnecessary, as tonsillar pulsations were visualized through the dura after bone decompression.

Pre- and Postoperative Craniocervical Relationships

Table 2 presents radiological measurements of craniocervical relationships. Preoperatively, the mean tonsillar descent distance was 11.8 mm (range 1.3–23 mm) and the

TABLE 1. Patient demographics

Characteristic	Value (%)
Sex	
Male	16 (51.6)
Female	15 (48.4)
Age (yrs)	
Mean	10.0
Range	1.5–43
Presentation	
Headache	20 (64.5)
Clumsiness	7 (22.6)
Scoliosis	7 (22.6)
Preop tonsillar descent (mm)	
Mean	11.8
Range	1.8–23
Duraplasty	
Yes	25 (80.6)
No	6 (19.4)
Clinical follow-up (mos)	
Mean	21
Range	2–47
Postop imaging interval (mos)	
Mean	10
Range	3–37

TABLE 2. Radiological craniocervical relationships

Relationship	Preop Mean $\pm$ SD (range)	Postop Mean $\pm$ SD (range)	Mean Change $\pm$ SD	p Value
pB–C2 (mm)	7.5 $\pm$ 1.1 (5.7–9.8)	8.0 $\pm$ 1.4 (5.2–9.9)	0.5 $\pm$ 1.2	<0.0001*†
Cervicomedullary angle (°)	152.1 $\pm$ 8.8 (128.6–165.4)	154.4 $\pm$ 9.5 (136.7–176.0)	2.3 $\pm$ 11.3	0.29‡
Clival-spinal angle (°)	156.1 $\pm$ 12.5 (124.9–179.8)	155.1 $\pm$ 14.0 (127.2–199.1)	–1.0 $\pm$ 10.5	0.77§
Odontoid to Wackenheim's line (mm)	2.8 below $\pm$ 3.7 (11.6 below to 3.6 above)	2.9 below $\pm$ 3.4 (11.9 below to 4.4 above)	0.12 below	0.79§
Odontoid to McRae's line (mm)	7.9 below $\pm$ 2.6 (13.8 below to 3.9 below)	NA	NA	NA
Odontoid to McGregor's line (mm)	2.9 below $\pm$ 3.8 (11.7 below to 2.8 above)	NA	NA	NA
Odontoid to Chamberlain's line (mm)	3.4 below $\pm$ 3.3 (12.0 below to 1.0 above)	NA	NA	NA

Below = below the respective line; NA = not applicable.

* Statistically significant.

† Mixed-effects ANOVA.

‡ Paired t-test.

§ Wilcoxon signed-rank test.

mean foramen magnum diameter was 34.2 mm (range 27.1–42.1 mm). The mean clival-spinal angle was 156.1° (range 124.9°–179.8°), and the mean cervicomedullary angle was 152.1° (range 128.6°–165.4°). The mean distance of the odontoid tip from Wackenheim's line and McRae's line were 2.8 mm below and 7.9 mm below the lines, respectively.

The mean preoperative pB–C2 measurement was 7.5 mm (range 5.7–9.8 mm). On postoperative measurement, the mean pB–C2 measurement was 8.0 mm (range 5.2–9.9 mm). Thus, the mean pB–C2 distance increased between the preoperative imaging and postoperative imaging by 0.5 mm (mixed effects ANOVA, $p < 0.0001$). The relationship of preoperative and postoperative pB–C2s is demonstrated graphically in Fig. 3. In 8 patients, minimal change in pB–C2 was seen (< 0.5 mm). In 15 patients, pB–C2 increased, with 10 of these between 0.5 and 1.5 mm and 5 greater than 1.5 mm. Finally, in 8 patients, pB–C2 decreased between 0.5 and 1.5 mm. Two patients had a pB–C2 measurement greater than 9 mm preoperatively, 1 of whom had an increase of 0.69 mm and the other a decrease of 1.05 mm postoperatively. Ten patients who had a preoperative pB–C2 measurement of less than 9 mm exceeded 9 mm on postoperative measurement.

The mean preoperative cervicomedullary angle was 152.1° (range 128.6°–165.4°), and the mean postoperative cervicomedullary angle was 154.4° (range 136.7°–176.0°). The change in mean cervicomedullary angle was 2.3°, which was not statistically significant via paired t-test ($p = 0.29$).

The mean preoperative clival-spinal angle was 156.1° (range 124.9°–179.8°), and the mean postoperative clival-spinal angle was 155.1° (range 127.2°–199.1°). The mean change in clival-spinal angle was –1.03°, which was not significant via Wilcoxon signed-rank test ($p = 0.77$).

The mean preoperative odontoid tip distance from Wackenheim's line was 2.80 mm antero-inferior (range 11.56 mm antero-inferior to 3.55 mm posterosuperior). The mean postoperative distance was 2.92 mm antero-inferior (range 11.86 antero-inferior to 4.35 posterosuperior). The

mean change in odontoid tip distance from Wackenheim's line was 0.12 mm in the antero-inferior direction, which was not significant via Wilcoxon signed-rank test ($p = 0.79$).

Correlations between variables of interest are demonstrated in Table 3. The change in pB–C2 was moderately negatively correlated with change in cervicomedullary angle (Pearson's correlation coefficient = –0.46; $p = 0.001$). The change in pB–C2 was also negatively correlated with the change in clival-spinal angle (Pearson's correlation coefficient = –0.44; $p = 0.013$).

Patient Outcomes

Postoperatively, 2 patients developed pseudomeningoceles and 1 patient developed a CSF leak. One patient experienced early postoperative hemiparesis that resolved. In the 20 patients presenting with headaches, 70% resolved and another 25% of headaches improved follow-

FIG. 3. Graphical representation of the change in pB–C2 values after surgery in all 31 patients. Figure is available in color online only.

TABLE 3. Intervariable correlations

Relationship	Pearson's Correlation Coefficient	p Value
Change in cervicomedullary angle vs change in pB–C2	–0.46	0.001*
Change in clival-spinal angle vs change in pB–C2	–0.44	0.013*
Change in odontoid to Wackenheim's line vs change in pB–C2	–0.08	0.685

* Statistically significant.

ing treatment. At last follow-up, at a mean of 21 months, no patients required additional procedures for VBSC. All patients were clinically stable or improved.

The rates and outcomes of CM-I associated conditions are presented in Table 4. The majority of the 17 patients with syringomyelia showed improvement or resolution of the syrinx (13 patients and 1 patient, respectively). For 1 patient, the available postoperative radiological studies of the spine were not adequate for syrinx evaluation. Twelve patients had scoliosis preoperatively. For the 7 patients with accessible spine imaging and clinical notes, the mean preoperative Cobb angle was 50.6° (range 12°–105°). In these patients, the degree of scoliosis was unchanged in 2 patients and worsened in 5 patients postoperatively. Two patients had hydrocephalus preoperatively; 1 patient's hydrocephalus persisted after PFD, requiring a ventriculo-peritoneal shunt.

To determine a possible difference in patient outcomes associated with postoperative pB–C2 values, we stratified the variables above by whether the pB–C2 distance was greater than or less than 9 mm (Table 5). According to the Fisher's exact test, no significant differences were found between groups, although the sample sizes were too small to detect anything but large differences.

Discussion

Pediatric patients with CM-I and symptomatic and/or radiological evidence of brainstem compression are often successfully treated with PFD alone.^{12,18,22} For patients who do not benefit from PFD or who display delayed neurological worsening, VBSC may be the cause of their dysfunction.^{4,12} Thus, quantification of VBSC is advantageous in the choice of decompressive approach (i.e., dorsal vs ventral vs circumferential decompression) and to assist in the long-term follow-up clinical picture.^{18,22} This has been previously documented in regard to patients with basilar invagination, but in symptomatic patients with CM-I without basilar invagination, VBSC is not always a diagnostic or prognostic consideration. In our series, only 8 patients

TABLE 5. Patient outcomes stratified by postoperative pB–C2 distance

Characteristic	pB–C2 <9 mm (%)	pB–C2 ≥9 mm (%)	p Value
No. of cases	20	11	
Headaches	12	8	0.16
Resolved	10 (83)	4 (50)	
Not resolved	2 (17)	4 (50)	
Syringomyelia*	9	7	1.00
Improved	8 (89)	6 (86)	
Not improved	1 (11)	1 (14)	
Scoliosis*	5	2	NA
Unchanged	2 (40)	0 (0)	
Worsened	3 (60)	2 (100)	
Hydrocephalus	2	0	NA
Resolved	1 (50)	0	
Shunt required	1 (50)	0	

NA = not applicable (data too sparse).

* Patients with unknown outcome are not listed.

had preoperative radiological evidence of basilar invagination using Wackenheim's line, and none had preoperative basilar invagination using McRae's, McGregor's, or Chamberlain's criteria.^{11,30}

In reviewing our data, we are intrigued by the statistically significant postoperative increase in pB–C2 distance. At first glance, one may attribute this to increased occipitocervical instability from the PFD. The likelihood of this in light of our technique for PFD is low, as we do not transect suboccipital musculature or violate the occipitoatlantal articulation during the procedure. Additionally, the craniectomy is limited to approximately 3 cm in width, the width of the cervicomedullary dura. Objectively, there was no change in the relationship between the odontoid tip and Wackenheim's line postoperatively. Furthermore, there was no correlation between the change in pB–C2 and the change in the distance from the odontoid to Wackenheim's line, indicating that these variables are unrelated. These data lend support to the idea that there is no instability introduced from the PFD, including patients in whom postoperative increases in pB–C2 are observed. Clinically, the majority experienced satisfactory amelioration of their preoperative symptoms after PFD. None of these patients underwent transoral odontoidectomy or occipitocervical fusion, including the 11 patients who had a postoperative pB–C2 measurement greater than 9 mm, suggesting that this post-PFD increase is well-tolerated. However, we acknowledge that our follow-up period of 21 months may not be long enough to predict long-term stability in these patients. For example, in a patient with

TABLE 4. Associated conditions and postoperative outcomes

Condition	No. of Cases	Postop Outcome
Syringomyelia	17	Resolved (n = 1, 5.9%), improved (n = 13, 76.5%), unchanged (n = 2, 11.8%), undocumented (n = 1, 5.9%)
Scoliosis	12	Unchanged (n = 2, 16.7%), worsened (n = 5, 41.7%), undocumented (n = 5, 41.7%)
Hydrocephalus	2	Resolved (n = 1, 50%), unresolved (n = 1, 50%)

craniocervical kyphosis at presentation, decompression may be a risk factor for future instability and such cases warrant close monitoring.

A possible but unproven explanation for the increase in pB–C2 distance after PFD relates to the change in circumferential forces restricting the dural envelope. That is, PFD relieves elements of the posterior compression vector, including suboccipital bone and the dura. We postulate that the increase in pB–C2 is not from instability, but rather a function of the ventral dura relaxing posteriorly due to the decrease in these forces. Of course, it does not follow that all post-PFD increase in VBSC is benign. If there is symptomatic cervicomedullary compression following PFD, the patient may require additional decompressive or stabilizing procedures. However, the presence of slightly increased pB–C2 and an otherwise improving clinical picture should not immediately dictate an additional procedure that may be of no benefit. In our practice, the decision to undergo these additional procedures is based on clinical criteria, rather than the pB–C2 value or other craniocervical measures.

There are several limitations to our study that warrant discussion. Foremost, as a retrospective study, the limits of data availability are inherent and unavoidable. Secondary to this, our study cohort was small, as more than half the patients who underwent PFD for CM-I did not have a full complement of imaging for evaluation and were necessarily excluded. Thus, the potential impact of selection bias should be kept in mind while interpreting these results. Also, as a result of the retrospective nature of our study, we did not have a protocol for patient positioning during MRI, and it is possible that small changes in patient positioning affected our results by chance. Future studies of the change in pB–C2 measurement would ideally be prospective, with a standardized pre- and postoperative MRI protocol, whereby the acquired sequences are tailored to the evaluation of craniocervical metrics including the pB–C2 measurement. Furthermore, this protocol would include guidelines for patient positioning to ensure that all patients were imaged in a neutral position. Additionally, a standardized symptom rating system for both preoperative assessment and postoperative change of symptoms could allow the evaluation of the change in pB–C2 compared with patient symptoms. Finally, a study with extended clinical and radiological follow-up would be ideal, consisting of 60 months or more.

Conclusions

Our results indicate that there is a slight increase in pB–C2 distance in patients following PFD for CM-I. The mechanism of this increase may be a releasing of pressure on the ventral dura from the posterior vector, rather than changes in muscular, ligamentous, or articular relationships. Given that the relationship of the odontoid to the skull base did not change in this study, our data suggest that an increased pB–C2, even above 9 mm, does not necessarily require intervention.

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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: Mapstone. Acquisition of data: Maurer, Safavi-Abbasi. Analysis and interpretation of data: Mapstone, Bonney, Maurer, Duong, Safavi-Abbasi. Drafting the article: Bonney, Maurer, Safavi-Abbasi. Critically revising the article: Mapstone, Bonney, Cheema, Glenn. Reviewed submitted version of manuscript: Mapstone, Bonney, Maurer, Cheema, Glenn. Approved the final version of the manuscript on behalf of all authors: Mapstone. Statistical analysis: Duong, Stoner. Administrative/technical/material support: Bonney. Study supervision: Mapstone.

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