Chiari malformation Type I (CM-I) is often described as the downward herniation of the cerebellar tonsils through the foramen magnum by 5 mm or greater. While this finding may be acquired as a result of an intracranial mass lesion, cerebral edema, or hydrocephalus, it is believed to most frequently be a congenital condition of the posterior fossa and skull base. Syringomyelia is present in as many as 30%–70% of patients diagnosed with CM-I.8,17,18 CM-I is commonly associated with other structural anomalies of the skull base, includ-

**Analysis and interrater reliability of pB-C2 using MRI and CT: data from the Park-Reeves Syringomyelia Research Consortium on behalf of the Pediatric Craniocervical Society**

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**OBJECTIVE** The distance to the ventral dura, perpendicular to the basion to C2 line (pB-C2), is commonly employed as a measure describing the anatomy of the craniovertebral junction. However, both the reliability among observers and the clinical utility of this measurement in the context of Chiari malformation Type I (CM-I) have been incompletely determined.

**METHODS** Data were reviewed from the first 600 patients enrolled in the Park-Reeves Syringomyelia Research Consortium with CM-I and syringomyelia. Thirty-one cases were identified in which both CT and MRI studies were available for review. Three pediatric neurosurgeons independently determined pB-C2 values using common imaging sequences: MRI (T1-weighted and T2-weighted with and without the inclusion of retro-odontoid soft tissue) and CT. Values were compared and intraclass correlations were calculated among imaging modalities and observers.

**RESULTS** Intraclass correlation of pB-C2 demonstrated strong agreement between observers (intraclass correlation coefficient [ICC] range 0.72–0.76). Measurement using T2-weighted MRI with the inclusion of retro-odontoid soft tissue showed no significant difference with measurement using T1-weighted MRI. Measurements using CT or T2-weighted MRI without retro-odontoid soft tissue differed by 1.6 mm (4.69 and 3.09 mm, respectively, p < 0.05) and were significantly shorter than those using the other 2 sequences.

**CONCLUSIONS** pB-C2 can be measured reliably by multiple observers in the context of pediatric CM-I with syringomyelia. Measurement using T2-weighted MRI excluding retro-odontoid soft tissue closely approximates the value obtained using CT, which may allow for the less frequent use of CT in this patient population. Measurement using T2-weighted MRI including retro-odontoid soft tissue or using T1-weighted MRI yields a more complete assessment of the extent of ventral brainstem compression, but its association with clinical outcomes requires further study.

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**KEY WORDS** Chiari malformation Type I; pB-C2; ventral brainstem compression; interrater reliability
ing basilar invagination, platybasia, and ventral brainstem compression (VBSC)\textsuperscript{1,5,7,10,15}. Although posterior fossa decompression is a highly successful treatment for many children with CMI, immediate or delayed progression of neurological symptoms after posterior fossa decompression in the context of VBSC may occur\textsuperscript{1,6,11}.

Many different measurements have been used to describe the anatomy of the craniovertebral junction (CVJ)\textsuperscript{1,4–16}. The multitude of techniques, however, contributes to inconsistency in the manner with which CVJ parameters are communicated, and no standard measurement has been established. In an effort to improve communication regarding CVJ assessment, we identified a commonly used CVJ measure (pB-C2) and endeavored to assess the reliability of this measurement as completed by different individuals using images that were obtained at different institutions.

In 1999, Grabb and colleagues used sagittal plane MR images to describe the line perpendicular to the basion to C2 line, through the odontoid tip to the ventral dura, termed pB-C2\textsuperscript{6}. From a retrospectively analyzed sample of 40 children and young adults with CMI, they found that no patient with a pB-C2 measurement of less than 9 mm required ventral brainstem decompression or occipitocervical fusion following standard posterior fossa decompression. Patients with pB-C2 measurements equal to or greater than 9 mm had more complex malformations and in some cases required occipitocervical fusion (OCF) with or without ventral brainstem decompression. The authors proposed that pB-C2 provided a more robust assessment of VBSC than alternative measures, such as the clivus–odontoid angle, because it accounted for the presence of retroodontoid soft tissue, which may place direct pressure on the brainstem, and because pB-C2 did not vary with the position of the patient. As a result of this work, pB-C2 has been commonly used in the assessment of VBSC and in neurosurgical decision making\textsuperscript{1,2,9,14}.

More recent analyses have further assessed the role of pB-C2 in clinical practice, with mixed results regarding the application of this measure in the context of risk stratification regarding occipitocervical instability or clinicoangiographic response to posterior fossa decompression\textsuperscript{1,2,12}.

The Park-Reeves Syringomyelia Research Consortium (PRSRC) is a multicenter network dedicated to the study of the etiology, natural history, and treatment of pediatric CM-I and syringomyelia. The PRSRC includes more than 30 contributing centers that have submitted both retrospective and prospective data in a registry format for over 1000 patients. Patient entry criteria include age less than 21 years, cerebellar tonsil herniation ≥ 5 mm, and syrinx ≥ 3 mm in the anteroposterior (AP) dimension. Clinical and imaging data undergo central review prior to inclusion, and imaging studies for each patient are housed in a central neuroimaging data archive (CNDA).

This preliminary study used data from the first 600 patients entered into the PPRSRC database to examine 2 aspects of the pB-C2 measurement: the interrater reliability and the relative magnitude of values obtained using common imaging sequences: CT, T1-weighted MRI, and T2-weighted MRI with and without the inclusion of retroodontoid soft tissue. We hypothesized that different imaging modalities would yield systematically different, but consistent values of pB-C2 and that the interrater reliability would be high. If pB-C2 proved to be a reliable measurement method, we could then use the entire PPRSRC cohort to further study the correlation of the measure with failure of initial posterior fossa decompression.

**Methods**

After obtaining appropriate institutional review board approval at each site, the imaging records of each of the first 600 patients enrolled in the PPRSRC were reviewed for the presence of preoperative CT images. Thirty-one patients who met this criterion were identified. All patient records included MRI with T1- and T2-weighted sequences, as these were required for study enrollment in the PPRSRC registry. Three board-certified pediatric neurosurgeons (T.C.H., G.F.T., and R.C.E.A.) accessed deidentified images for each of the 31 subjects remotely, through the CNDA. Images in the Digital Imaging and Communication in Medicine (DICOM) format were downloaded and converted for standardized measurement using ImageJ, an open-access image-processing program (https://imagej.nih.gov/ij/), under the guidance of the CNDA technical staff. Complete imaging sequences were reviewed. As this study was designed to assess the reliability of pB-C2 measurement by clinicians, each rater independently determined the image and windowing that best demonstrated the midsagittal anatomy. MRI pulse sequence parameters were not standardized across institutions. pB-C2 was measured according to the method of Grabb and colleagues\textsuperscript{6}. The B-C2 line was drawn from the postero-inferior corner of the C2 vertebral body through the posteroinferior tip of the clivus, at the basion, which was usually identified as a sharp angle of hypointense cortical bone on both T1- and T2-weighted MRI. The pB-C2 line was drawn perpendicularly from the B-C2 line, through the odontoid, defining the longest distance from the B-C2 line to the ventral dura. For each patient, 4 measurements of pB-C2 were obtained: 1 from CT, 1 from T1-weighted MRI, and 2 from T2-weighted MRI. One of the 2 measurements on T2-weighted images included all retro-odontoid soft tissue (e.g., pannus), while the other extended from the B-C2 line to the posterior cortex of the odontoid process (Fig. 1). Similarly, measurement using CT extended from the B-C2 line to the posterior cortex of the odontoid process, as the resolution of soft tissue on CT is not as reliable as on MRI. This method was used to help determine the relationship between CT and T2-weighted MRI. Each reviewer independently recorded his measurements. Once completed, the data were submitted for central statistical analysis.

**Data Analysis**

Comparative analysis was done adjusting for variation among patients using general linear models for each CT and MRI measure. For the planned comparison of imaging method the Tukey Studentized Range Test was used. The intraclass correlation coefficient (ICC) was used to measure concordance among 3 independent neurological surgeon raters. SAS statistical software SAS, version 9.4,
was used for statistical analysis; p values less than 0.05 were considered statistically significant.

**Results**

Thirty-one patients with preoperative CT images were identified. All patients had preoperative MRI studies. Image quality precluded pB-C2 measurement on CT in 3 patients, on T1-weighted MRI in 2 patients, and on T2-weighted MRI in 1 patient. These deficiencies did not result in the exclusion of any individual patient from the data set. All measurable data points were therefore included in the statistical calculations.

Mean pB-C2 values measured using CT (4.69 ± 3.04 mm [SD]) and T2-weighted MRI excluding soft tissue (3.09 ± 2.35 mm) were significantly smaller than the values obtained from measurement based on T2-weighted MRI including soft tissue (7.05 ± 2.09 mm) and T1-weighted MRI (7.21 ± 2.22 mm). There was a significant difference in pB-C2 values between CT and T2-weighted MRI excluding soft tissue (bony pB-C2), with values measured by CT being 1.61 mm larger on average. There was no significant difference between pB-C2 measurements made using T2-weighted MRI including soft tissue and T1-weighted MRI (soft tissue pB-C2, Table 1).

Calculation of the ICC demonstrated substantial agreement for each modality, and no modality differed significantly in this regard from any other (Table 2). As assessed using CT, the ICC was 0.75. Using T1-weighted MRI, it was 0.72. Using T2-weighted MRI without and with pannus, it was 0.75 and 0.76, respectively (Table 2).

The percentage of cases in which the measured pB-C2 was greater than 9 mm was equal for all reviewers when measuring using CT or T2-weighted MRI without pannus (bony pB-C2, 3.6% and 0%, respectively). When measuring using T2-weighted MRI with pannus, the proportion of cases with pB-C2 greater than 9 mm ranged from 6.7% to 26.7%. When measuring using T1-weighted MRI, the proportion of cases with pB-C2 greater than 9 mm ranged from 10.3% to 34.5% (Table 3).

**Discussion**

Consistency regarding the metrics and methods employed for the description of CVJ anatomy not only improves clinical communication but also helps establish tools that facilitate the advancement of clinical outcomes research. pB-C2 is a routinely referenced metric, but one with inconsistent application and unclear clinical significance. Prior studies of pB-C2 have come from single institutions.1,2,6,12 For these reasons, we felt that this measure was appropriate for a preliminary reliability study using data from the multicenter PRSRC, potentially leading to more rigorous study of the measurements clinical utility.
This preliminary work demonstrated good correlation of pB-C2 values among pediatric neurosurgeons (ICC 0.72–0.76). Additionally, we demonstrated close correlation between pB-C2 measurements taken using different imaging modalities. Specifically, pB-C2 values obtained using CT or T2-weighted MRI without the inclusion of the retro-odontoid soft tissue correlated very closely with each other, with a mean difference of 1.61 mm. This is clinically relevant because quantifying the expected difference between MRI and CT-based pB-C2 measurement may allow for further reduction in the need for CT scanning in the assessment of CVJ anatomy in children with CM-I. While this preliminary finding requires validation, it may allow for decreased exposure to potentially harmful ionizing radiation3,4 without sacrificing valuable anatomical information. Additionally, measurements obtained using these modalities were systematically lower than those obtained with T1-weighted MRI or T2-weighted MRI that included retro-odontoid soft tissue. This knowledge facilitates comparison among modalities and suggests that the latter 2 modalities may offer a more comprehensive view of the relevant craniovertebral junction anatomy.

In this preliminary study we sought to determine the best modality with which to measure pB-C2 as well as the relationships between different modalities. Subsequent studies will examine the utility of pB-C2 in anticipating clinical, surgical, and radiological outcomes. In their initial description of pB-C2, Grabb and colleagues6 found that 4 (36.3%) of 11 patients with a value of greater than 9 mm required occipitocervical fusion in addition to posterior fossa decompression. Two of these patients also required odontoidectomy. No patient who had a pB-C2 value less than 9 mm required fusion or ventral decompression. Based on these findings, the authors felt that patients with a pB-C2 greater than 9 mm merit special attention preoperatively and in follow-up. Bollo and colleagues1 reviewed 101 cases involving children who had undergone posterior fossa decompression for CM-I at the University of Utah to identify factors associated with the need for concurrent or subsequent occipitocervical fusion (OCF). The factors identified included basilar invagination, CM Type 1.5, and a clivoaxial angle less than 125°. Although pB-C2 was associated with the need for OCF on univariate analysis (both as a continuous variable and a categorical variable, pB-C2 ≥ 9 mm), the relationship with OCF was lost in multivariate analysis. Ladner and colleagues5 found that children with a pB-C2 of 3 mm or greater had a higher likelihood of harboring syringomyelia preoperatively and were more likely to experience improvement in headache symptoms and syrinx size, when compared with those with a pB-C2 less than 3 mm. In their population, only 1 (1.2%) of 78 patients had a pB-C2 of greater than 9 mm and none required ventral decompression. In a population of 31 children who underwent posterior fossa decompression with (n = 25) or without (n = 6) duraplasty for CM-I, Bonney and colleagues2 identified a mean of 0.5 mm increase in pB-C2 at 10 months following decompression (7.5 to 8.0 mm). The direction and magnitude of change in pB-C2 was not found to correlate with the preoperative value, and no patient required ventral decompression or occipitocervical stabilization. Each of these studies measured pB-C2 in the manner defined by Grabb and colleagues6 and undertaken in this study.

Reliability of pB-C2 Measurement

As in our study, Bollo and colleagues1 calculated the ICC of pB-C2 measurement among multiple observers. Using midsagittal T1- or T2-weighted MRI, they documented a good correlation (0.64) between the measurements made by a pediatric neurosurgery fellow and a board-certified pediatric neurosurgeon, with a fair correlation (0.56) when the latter observer was compared with an undergraduate student. In our study, the agreement was strong (ICC 0.72–0.76). This most likely results from the fact that measurements were performed by 3 board-certified pediatric neurosurgeons. This level of agreement was consistent across all 4 modalities, which strengthens the assertion that pB-C2 can serve as a reliable and objective measurement among trained clinicians. More in-depth studies, with measurements obtained by trainees or other specialists, could assess the generalizability of pB-C2 measurement with greater granularity.

Like Bollo and colleagues,1 we found much greater variation between raters when we dichotomized pB-C2 around the 9-mm value (Table 3). They reported a 64% agreement between a pediatric neurosurgery fellow and a board-certified pediatric neurosurgeon (16 of 25 measurements). In our population, when measuring using T2-weighted MRI with retro-odontoid soft tissue, the percentage of our 30 patients with pB-C2 greater than 9 mm ranged from 6.7% to 26.7%. This variation is likely due to the fact that several patients (19.3%–25.8%) on T1-weighted MRI had measurements between 8 and 10 mm, where an extremely small measurement difference resulted in or not crossing the 9-mm threshold in a given case.

Magnitude of pB-C2 Values

Given the range of neuroimaging sequences and modalities that are available, any effort to determine the reliability of a radiographic measurement should attempt to account for inherent variation between modalities. In examining images obtained using T2-weighted MRI, we hypothesized that pB-C2 measurements including the retro-odontoid soft tissue would systematically be larger than those that stop at the dorsal cortical odontoid surface and do not include the soft tissue. In this study, this hypothesis was correct, as the mean value of the former was 7.05 mm, while that of the latter was 3.09 mm. Also as expected, measurements taken using CT correlated closely with those taken using T2-weighted MRI excluding soft tissue, although they were significantly larger. Measurements taken using T2-weighted MRI including soft tissue correlated closely with those taken using T1-weighted MRI. Inclusion of the retro-odontoid soft tissue is consistent with the intent of the measurement, which is to evaluate the degree of ventral impingement on the brainstem and is also consistent with other published methods of pB-C2 measurement.2,6,12 The mean value of 7.05 mm was similar to that reported by Bonney and colleagues,2 which was 7.5 mm. These values are also consistent with those reported by Bollo,1 which were 7.2 mm and 10.2 mm for their patients.
who did not and did require OCF, respectively. Given the referral practice at their institution, it is not unexpected that the mean pB-C2 values identified are slightly higher than those of the more generalized multicenter cohort examined in this study. Ladner and colleagues 12 also used T2-weighted MRI including retro-odontoid soft tissue in their measurements. They, however, reported a substantially smaller mean pB-C2 value in their population (3.5 mm). This could be explained by differences in the population of patients in their practice or in the technique of measurement, which was completed by a single neuroradiologist. As we identified no difference between mean pB-C2 values measured using T2-weighted MRI including retro-odontoid soft tissue and values measured using T1-weighted MRI, we conclude that either of these sequences may be used in the assessment of pB-C2. Our preference is to use T2-weighted MRI because the contrast between the high-intensity CSF and low-intensity adjacent soft tissue allows for greater precision in measurement.

We included CT images in our analysis to understand more completely the relationship between imaging modalities with regard to pB-C2 measurement. As mentioned above, values obtained from this modality are consistently greater (mean difference 1.6 mm) than those obtained by excluding the retro-odontoid soft tissue from measurements taken on T2-weighted MRI. Both measurements were designed to include the full extent of the bone and exclude the adjacent soft tissue. Thus, it is not entirely clear why measurement using CT yielded larger values. While the small number of patients in the PRSRC data set with preoperative CT studies (31 [5.2%] of 600) likely reflects the practice of most physicians to minimize the use of ionizing radiation in children, the recognition that measurements using T2-weighted MRI closely approximate measurements using CT may further limit the indications for CT imaging in this patient population.

Limitations

As mentioned above, only a small subset of the patients within the PRSRC data set (31 [5.2%] of 600) had preoperative CT imaging uploaded into the registry. This limited the number of samples that we had available to study. Furthermore, these patients may have undergone CT scanning because their clinical condition or other imaging indicated a need for detailed comprehension of the osseous anatomy at the CVJ. Accordingly, these patients may be more likely to have a greater degree of VBSC than the majority of the PRSRC population. For the purposes of this study, which intended to assess the reliability of pB-C2 measurement, the potential complexity of the study population’s CVJ anatomy should not impact our results. Furthermore, the similarity of our mean pB-C2 values to those in most previous publications indicates that our study population was reasonably similar to others.

This preliminary radiographic study does not include clinical data and therefore limits our capacity to contextualize our findings. However, our study demonstrates that: 1) pB-C2 can be consistently assessed by multiple neurosurgical observers, and 2) a single measurement of pB-C2 from either T1- or T2-weighted MRI can be used. Therefore, we can now proceed with the next step, in which we plan to investigate the utility of pB-C2 and other radiographic measurements of the CVJ to predict clinical outcomes after suboccipital decompression in CM-I patients utilizing the entirety of the PRSRC data set, which now includes over 1000 patients.

Conclusions

Using a multicenter population and multiple observers, we determined that pB-C2 can be reliably measured in children with CM-I and syringomyelia. Measurement using T2-weighted MRI including retro-odontoid soft tissue yields values that are similar to measurement using T1-weighted MRI and are systematically larger than values obtained with CT or T2-weighted MRI excluding retro-odontoid soft tissue, which are also similar to each other. These data may help reduce the need for CT usage in this population. The clinical significance of pB-C2 remains to be further refined through studies using a larger population with correlative clinical data.

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References


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**Author Contributions**

Conception and design: Hankinson, Limbrick, Anderson. Acquisition of data: Hankinson, Robinson, Limbrick, Park, Anderson. Analysis and interpretation of data: Hankinson, Limbrick, Anderson. Drafting the article: Hankinson, Robinson. Critically revising the article: Hankinson, Robinson, Torner, Limbrick, Park, Anderson. Reviewed submitted version of manuscript: Hankinson, Tuite, Moscoso, Anderson. Approved the final version of the manuscript on behalf of all authors: Hankinson. Statistical analysis: Torner.

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