J Neurosurg Pediatrics 10:134–141, 2012

Complex Chiari malformations in children: an analysis of preoperative risk factors for occipitocervical fusion

Clinical article

Robert J. Bollo, M.D., Jay Riva-Cambrin, M.D., M.Sc., Meghan M. Brockmeyer, and Douglas L. Brockmeyer, M.D.

Division of Pediatric Neurosurgery, Department of Neurosurgery, Primary Children's Medical Center, University of Utah, Salt Lake City, Utah

Object. Chiari malformation Type I (CM-I) is a congenital anomaly often treated by decompressive surgery. Patients who fail to respond to standard surgical management often have complex anomalies of the craniovertebral junction and brainstem compression, requiring reduction and occipitocervical fusion. The authors hypothesized that a subgroup of "complex" patients defined by specific radiographic risk factors may have a higher rate of requiring occipitocervical fusion.

Methods. A retrospective review was conducted of clinical and radiographic data in pediatric patients undergoing surgery for CM-I between 1995 and 2010. The following radiographic criteria were identified: scoliosis, syringomyelia, CM Type 1.5, medullary kinking, basilar invagination, tonsillar descent, craniocervical angulation (clivoaxial angle [CXA] < 125°), and ventral brainstem compression (pB–C2 ≥ 9 mm). A multivariate Cox regression analysis was used to determine the independent association between occipitocervical fusion and each variable.

Results. Of the 206 patients who underwent CM decompression with or without occipitocervical fusion during the study period, 101 had preoperative imaging available for review and formed the study population. Mean age at surgery was 9.1 years, and mean follow-up was 2.3 years. Eighty-two patients underwent suboccipital decompression alone (mean age 8.7 years). Nineteen patients underwent occipitocervical fusion (mean age 11.1 years), either as part of the initial surgical procedure or in a delayed fashion. Factors demonstrating a significantly increased risk of requiring fusion were basilar invagination (HR 9.8, 95% CI 2.2–44.2), CM 1.5 (HR 14.7, 95% CI 1.8–122.5), and CXA < 125° (HR 3.9, 95% CI 1.2–12.6).

Conclusions. Patients presenting with basilar invagination, CM 1.5, and CXA < 125° are at increased risk of requiring an occipitocervical fusion procedure either as an adjunct to initial surgical decompression or in a delayed fashion. Patients and their families should be counseled in regard to these findings as part of a preoperative CM evaluation.

(http://thejns.org/doi/abs/10.3171/2012.3.PEDS11340)

Key Words • **Chiari malformation** • **basilar invagination** • **occipitocervical fusion** • **syringomyelia**

The association of CM-I with osseous abnormalities, such as scoliosis and anomalies of the craniovertebral junction (including retroflexed odontoid process, basilar invagination, and platybasia), has long been recognized.[3–5,12,13,16,22] Such patients often present with bulbar symptoms, including vertigo, diplopia, dysphagia, and apnea,[7–9] and demonstrate ventral brainstem compression on MRI.[8,10–12] These symptoms are especially common in younger patients (0–2 years old).[2] Many patients with this constellation of radiographic findings fail to improve after standard suboccipital decompression.[8,11]

Significant variability persists among surgeons regarding specific radiographic criteria used to identify patients for ventral decompression and occipitocervical fusion.[8,9,11,14,27] The pB–C2 line is a clinical measure developed to quantify ventral brainstem compression,[11] and pB–C2 ≥ 9 mm is often associated with clinical symptoms,[11] but not all patients with CM and pB–C2 ≥ 9 mm require ventral decompression. In addition, as many as 12% of patients with CM-I may have connective tissue disorders associated with occipitocervical instability and progressive cranial settling, often associated with a ret-

Abbreviations used in this paper: CM = Chiari malformation; CM-I = CM Type I; CXA = clivoaxial angle; ICC = intraclass correlation coefficient.

This article contains some figures that are displayed in color online but in black-and-white in the print edition.

roodontoid pannus and pathologically flat craniocervical angle without bone ventral compression.[15,25,30] Furthermore, some patients experience cranial settling after posterior decompression, and ultimately require stabilization and fusion of the craniovertebral junction.[8,25]

Recently, the term "CM 1.5" has been used to identify a subgroup of patients with brainstem herniation (defined as the obex below the level of the foramen magnum) in addition to tonsillar herniation.[31,32] Data suggest patients with CM 1.5 frequently present with bulbar signs and symptoms, have distortion of the brainstem on sagittal MRI, and fail standard posterior decompression significantly more frequently than patients with CM without these features.[31] The high failure rate coupled with the presence of brainstem herniation suggests a more complex pathophysiology, possibly including ventral brainstem compression, in the absence of osseous anomalies of the craniovertebral junction.

Chiari malformation represents a spectrum of pathological conditions, with significant anatomical variation among individuals.[1] Patients with complex CM—including brainstem herniation, abnormalities of the craniovertebral junction, and ventral brainstem compression—may ultimately require occipitocervical fusion. The broad spectrum of causes and radiographic appearances makes the early identification of these patients challenging, and their definitive surgical management remains controversial.[7–11] We sought to identify common radiographic risk factors predisposing patients to occipitocervical fusion that were present at the time of diagnosis among children who underwent surgery for CM at a single pediatric neurosurgical center over a 16-year period.

Methods

Data Collection

Prior to data collection, the study was approved by the Institutional Review Board at the University of Utah School of Medicine and Primary Children's Medical Center. A search of the pediatric neurosurgical operative database was conducted for the 16-year study period (1995–2010), and 206 patients were identified who underwent surgical management of CM-I. Patients managed nonoperatively were excluded from the study. Preoperative and postoperative MRI studies were available for review in 101 patients, who comprised the study population. The study period for each patient began at the time of preoperative neurosurgical evaluation. We noted clinical variables, including sex, age at diagnosis, presenting symptoms, and neurological findings; secondary diagnoses, including the presence or absence of scoliosis, age at surgery, and operation performed; surgical complications, including stroke, hemorrhage, new neurological deficit, infection, pseudomeningocele, hardware misplacement, pseudoarthrosis, or new deformity; and clinical follow-up, including duration, neurological status, and requirement for reoperation.

Radiographic variables recorded from MR images obtained at the time of diagnosis and prior to any surgical intervention included CM type (I or 1.5, 1.5 defined as the obex below the level of the foramen magnum on preoperative MRI);[31] distance of tonsillar descent below the level of the foramen magnum; presence of syringomyelia, a medullary kink, basilar invagination, or retroflexed odontoid; and pB–C2 (as described by Grabb et al.)[11] and CXA (as described by Smoker)[30] measured on midsagittal T1- or T2-weighted images.

Surgical Indications and Technique

The indications for decompressive CM surgery were abstracted from the preoperative clinic notes or the operative notes themselves when documented. The indications varied according to surgeon preference and training, but the most common indications included headache, bulbar or myelopathic symptoms, paresthesias, syringomyelia, and scoliosis, which were consistent with published reports in similar populations.[1,3,7,13,21,26] The indications for occipitocervical fusion included prior transoral odontoidectomy, significant ventral brainstem compression, severe unresolved headache or headache exacerbated by prior decompressive surgery, bulbar or myelopathic symptoms, and progressive or unresolved syrinx despite prior decompressive surgery.[17] Patients who met these criteria at the time of initial neurosurgical evaluation underwent occipitocervical fusion at the time of initial decompression. Patients who met these criteria only after an initial CM decompression underwent reexploration of the decompression followed by occipitocervical fusion.

The surgical technique for standard CM decompression consisted of suboccipital craniectomy (including bone removal of the posterolateral foramen magnum), C-1 laminectomy, dural opening with exploration of the fourth ventricle, tonsillar shrinking, and duraplasty. The multistep surgical technique for posterior occipitocervical fusion consisted first of either a standard CM decompression or reexploration of a prior decompression, including a watertight duraplasty. Second, posterior cervical screw fixation was performed bilaterally using polyaxial screws that were 3.5 mm in diameter (Vertex, Medtronic Inc.), inserted under fluoroscopic guidance in either the C-2 or C-3 pars interarticularis, depending on the presence of a congenital segmentation anomaly. Next, bilateral occipitocervical rod-plates (Vertex) were cut to size and contoured to fit between the occiput and C-2 or C-3 on either side of the decompression. Two posterior rib autografts were harvested in all fused patients and shaped to fit the occipitocervical space between the rod-plates. Finally, the ribs were held in place using the combination of a multistranded titanium cable (Songer cable, Medtronic Inc.) and 1.5-mm (diameter) × 12-mm (length) screws in their cranial ends. Demineralized bone matrix (Medtronic Inc.) was placed around the graft at the conclusion of the procedure, and the incision was closed in layers.

Postoperatively, patients who had undergone fusion were maintained in a hard cervical collar for 2–3 months or until radiographic evidence of successful arthrodesis was achieved. No patient was placed in an external halo orthosis after surgery. Radiographic follow-up for each patient requiring fusion consisted of plain radiographs obtained once a month for the first 2 months, followed by a noncontrast CT scan of the occipitocervical junc-

tion with thin-slice sagittal and coronal reconstructions obtained 4 months after surgery to document successful arthrodesis.

Statistical Analysis

Patients were stratified according to the primary outcome of whether they underwent occipitocervical fusion during the study period. For univariate analysis, the chi-square test was used to compare categorical variables, and t-tests were used to evaluate continuous variables in view of the primary outcome. Colinearity was examined between continuous variables using Pearson correlation coefficients. A multivariate stepwise Cox proportional hazards regression analysis was performed using time to fusion for causal outcome and time to follow-up otherwise. A probability value < 0.05 was considered statistically significant.

In addition, the univariate and multivariate analyses were repeated after excluding patients who underwent fusion at the time of their initial surgical decompression. Finally, Kaplan-Meier survival analysis was performed, with patients stratified according to variables demonstrating significance in the multivariate procedure. The resultant model was secondarily tested excluding the subgroup of patients who underwent occipitocervical fusion as part of their initial surgical procedure to ensure the consistency of our findings.

To ensure the reliability and reproducibility of the two novel radiological measurements (CXA and pB–C2), interclass correlation coefficients were used to measure agreement among 3 independent raters with varying levels of neurosurgical training (an experienced pediatric neurosurgeon [D.L.B.], a pediatric neurosurgery fellow [R.J.B.], and an undergraduate student [M.M.B.]). The statistical software package SAS (version 9.2; SAS Institute, Inc.) was used for statistical analysis.

Results

Patient Characteristics

Baseline demographic data are presented in Table 1. For all patients, the mean age at surgery was 9.1 years, and mean follow-up was 2.3 years. Among 101 patients in the study group, 19 required occipitocervical fusion; in 11 patients this was the initial procedure. Three of the 11 patients required a transoral odontoidectomy for irreducible ventral brainstem compression, and 7 presented with atlantooccipital instability in the context of osseous anomalies of the craniovertebral junction. Eight patients required delayed occipitocervical fusion after 1 or more suboccipital decompressions (interval range 1.3–9.2 years, mean 4.1 years), because of either progressive cranial settling or acquired occipitocervical instability. There was a nonsignificant trend toward older age among patients who underwent fusion.

TABLE 1: Baseline demographic data of the study population*

Variable	Decompression Only (n = 82)	Decompression & Fusion (n = 19)
mean age in yrs ± SD (range)	8.7 ± 5.1 (0.7–16.8)	11.1 ± 6.5 (1.9–21.9)
no. of males (%)	46 (56)	8 (42)
mean time to follow-up in yrs ± SD (range)	2.2 ± 1.9 (0.1–7.8)	2.6 ± 2.7 (0.1–9.3)

* None of the comparisons between groups were significant.

Risk Factors for Occipitocervical Fusion

The results of the univariate analysis of risk factors of categorical and continuous variables associated with occipitocervical fusion are presented in Tables 2 and 3, respectively. A significant association between CM 1.5 and occipitocervical fusion was observed ($p < 0.001$). The presence of a medullary kink, retroflexion of the odontoid process, and basilar invagination were also significantly more common in the fusion group. Table 3 demonstrates that the differences in the continuous variables of pB–C2 (mean 10.2 mm in the fusion group vs 7.2 mm in the decompression-only group), CXA (mean 115.2° in the fusion group vs 141.1° in the decompression-only group), and tonsillar descent (mean 16.3 mm in the fusion group vs 13 mm in the decompression-only group) reached statistical significance. The mean age at diagnosis, patient sex, time to follow-up, and the presence of either a syrinx or scoliosis were not associated with the need for occipitocervical fusion.

When the commonly cited pB–C2 cutoff of ≥ 9 mm[11] was used to dichotomize this variable, statistical significance was preserved: a pB–C2 ≥ 9 mm was noted in 24% of patients with decompression alone versus 74% in those who required an additional occipitocervical fusion (Table 3). The relationship between occipitocervical fusion and pB–C2 is demonstrated in Fig. 1. Interclass correlation coefficients demonstrated good agreement between measurements performed by a pediatric neurosurgeon and those by a pediatric neurosurgery fellow (ICC = 0.64,

TABLE 2: Univariate analysis of categorical variables associated with the need for occipitocervical fusion in patients who underwent surgical management of CM-I*

Categorical Variable	Decompression Only	Decompression & Fusion	Univariate p Value
CM 1.5	18 (22)	18 (95)	<0.001
CM-I	64 (78)	1 (5)	
scoliosis	20 (24)	2 (11)	NS
no scoliosis	62 (76)	17 (89)	
syrinx	42 (51)	9 (47)	NS
no syrinx	40 (49)	10 (53)	
medullary kink	21 (26)	15 (79)	<0.001
no kink	61 (74)	4 (21)	
odontoid process retroflexion	30 (37)	13 (68)	0.01
no retroflexion	52 (63)	6 (32)	
basilar invagination	0 (0)	7 (37)	<0.001
no basilar invagination	82 (100)	12 (63)	

* Data presented as number of patients (%) unless otherwise stated. Abbreviation: NS = not significant.

TABLE 3: Univariate analysis of continuous variables associated with the need for occipitocervical fusion in patients who underwent surgical management of CM-I

Continuous Variable	Decompression Only	Decompression & Fusion	Univariate p Value
mean pB–C2 (mm) ± SD	7.2 ± 2.1	10.2 ± 2.3	<0.001
no. w/ ≥9 mm (%)	20 (24)	14 (74)	<0.001
no w/ <9 mm (%)	62 (76)	5 (26)	
mean tonsillar descent (mm) ± SD*	13.0 ± 5.1	16.3 ± 6.5	0.02
no. w/ >15 mm (%)	24 (30)	9 (47)	NS
no. w/ ≤15 mm (%)	57 (70)	10 (53)	
mean CXA (°) ± SD†	141.1 ± 14.6	115.2 ± 17.2	<0.001
no. w/ <125° (%)	9 (11)	15 (79)	<0.001
no. w/ ≥125° (%)	70 (89)	4 (21)	

* Available imaging did not allow us to accurately determine these values for 1 patient in this group.

† Available imaging did not allow us to accurately determine these values for 3 patients in this group.

95% CI 0.35–0.82); however, only 64% agreement (16/25) was observed for the dichotomized pB–C2 (< or ≥ 9 mm). When either were compared with measurements made by an undergraduate student, correlation was only fair (ICC = 0.46 and 0.56, respectively).

We selected a conservative CXA cutoff of 125° to dichotomize this variable, which represented 1 standard deviation above the mean for the entire cohort. Applying this cutoff to our data, 15 (79%) of 19 patients who underwent fusion had a CXA < 125° at diagnosis, compared with 9 (11%) of 82 who underwent decompression alone ($p < 0.001$; Table 3). Interclass correlation coefficients again demonstrated good agreement between measurements performed by a pediatric neurosurgeon and pediatric neurosurgery fellow (ICC = 0.63, 95% CI 0.33–0.81), with 92% agreement (23/25) for the dichotomized variable (< or ≥ 125°). Fair agreement was observed between measurements provided by either neurosurgeon or fellow and an undergraduate student (ICC = 0.37 and 0.22, respectively).

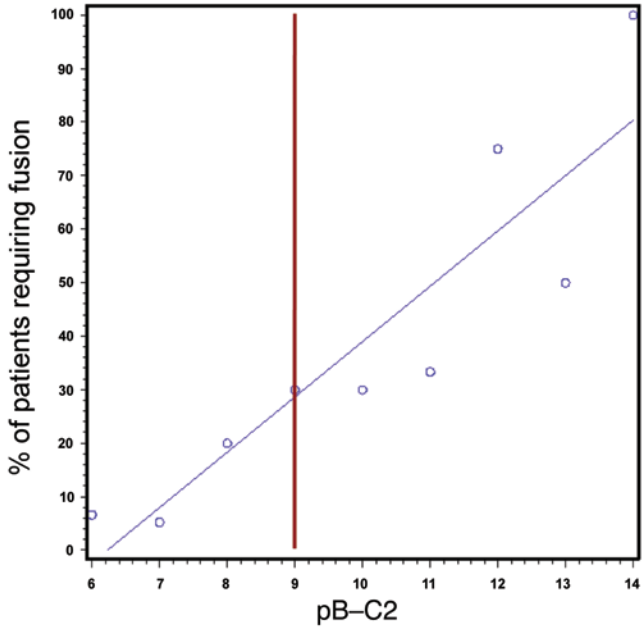

Fig. 1. Scatterplot of occipitocervical fusion according to the pB–C2. While a pB–C2 ≥ 9 mm is frequently cited as indicative of ventral brainstem compression, it was not a robust predictor of fusion in our study population.

All 7 patients with basilar invagination (100%) eventually underwent occipitocervical fusion, compared with 12 (12.8%) of 94 patients without basilar invagination. We used stepwise multivariate Cox proportional hazards analysis to develop a model predicting the need for occipitocervical fusion in our study population (Table 4). Only CM 1.5 (HR 14.7, 95% CI 1.8–122.5), basilar invagination (HR 9.8, 95% CI 2.2–44.2), and CXA < 125° (HR 3.9, 95% CI 1.2–12.6) retained statistical significance in the multivariate analysis. This model was both stable and robust. These relationships are further demonstrated via Kaplan-Meier curves with survival censored at the time of fusion (Figs. 2 and 3). The model was highly discriminatory in our study population (area under the receiver operating characteristic curve [accuracy] = 0.87).

The results of our model remained unchanged in 2 subgroup analyses: first, excluding patients with basilar invagination (n = 7), and second, excluding patients who underwent fusion as part of their initial surgical management (n = 11). Table 5 presents the univariate analysis and Table 6 the multivariate analysis of risk factors for delayed fusion, excluding patients who underwent fusion as part of their initial procedure. The risk factors that predicted delayed fusion (basilar invagination, CM 1.5, and CXA < 125°), present at the time of initial neurosurgical

TABLE 4: Multivariate regression analysis of variables associated with the need for occipitocervical fusion in patients who underwent surgical management of CM-I

Variable	Univariate p Value	Multivariate p Value	HR (95% CI)
CM 1.5	<0.001	0.02	14.7 (1.8–122.5)
medullary kink	<0.001	NS	
retroflexed odontoid process	0.01	NS	
basilar invagination	<0.001	<0.001	9.8 (2.2–44.2)
mean pB–C2 (mm)			
≥9	<0.001	NS	
<9	<0.001	NS	
mean tonsillar descent (mm)			
>15	0.02	NS	
≤15	NS	NS	
mean CXA (°)			
<125	<0.001	<0.001	3.9 (1.2–12.6)
≥125	<0.001	0.04	

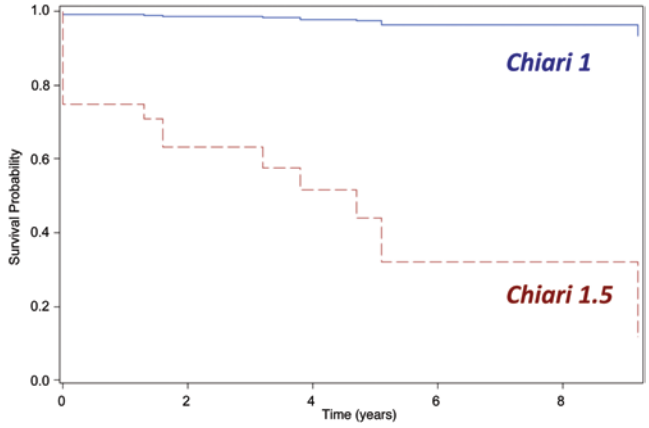

Fig. 2. Kaplan-Meier survival curve illustrating time to fusion stratified by CM type.

evaluation prior to any intervention, match exactly those for the entire fusion group as a whole.

Surgical Outcome

All 19 patients who underwent posterior occipitocervical fusion attained a successful arthrodesis documented on 4-month postoperative CT. Surgical complications in both the fusion and nonfusion groups included 5 patients (5%) who developed pseudomeningocele: 3 underwent reexploration of the wound with or without CSF diversion via lumbar drainage, and 2 were observed and experienced resolution without further treatment. One of the 101 patients developed mild neutropenia in the postoperative period, which was self-limited and resolved spontaneously without further treatment. There were no wound infections, and no patients experienced acute neurological deterioration after surgery; however, 3 patients in the nonfusion group underwent repeated decompression between 1 and 8 months after the original operation because of a persistent syrinx or clinical symptoms. One of these patients underwent placement of a syringoperitoneal shunt as a third intervention because of progressive syrinx enlargement despite repeated decompression. All 3 patients improved after reexploration or shunting of the syrinx.

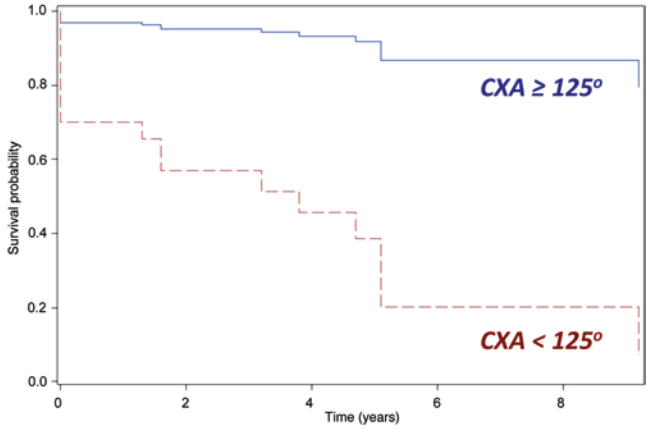

Fig. 3. Kaplan-Meier survival curve illustrating time to fusion stratified by CXA.

Discussion

We have shown that specific radiological features present at diagnosis, prior to any surgical intervention, strongly predicted the need for occipitocervical fusion in a retrospective analysis of children who underwent surgical management of CM at a single institution. Together, CM 1.5, basilar invagination, and CXA < 125° were defining features of complex CMs. All patients with basilar invagination ultimately experienced unsuccessful posterior decompression alone. Among patients with both CM 1.5 and CXA < 125°, 83.3% required occipitocervical fusion. Figure 4 illustrates midsagittal T2-weighted MR images from patients in the high-, intermediate-, and low-risk groups in our study population. Notably, interclass correlation determined good agreement between trained professionals (pediatric neurosurgeon and pediatric neurosurgery fellow), but not between professionals and a layperson (undergraduate student).

Previous authors have recognized the broad spectrum of pathological conditions associated with CM; the association between CM, brainstem compression, and craniovertebral junction anomalies; and the need for individualized treatment, including ventral decompression, posterior decompression with open reduction, and occipitocervical fusion in a select group of complex patients.[6,9–12,19,20,25,33] However, early identification of complex patients by specific radiographic criteria remains challenging. Although patients with CM 1.5 are known to be significantly more likely to fail posterior decompression alone,[31] previous studies have not analyzed the impact of CM 1.5 together with other radiographic indicators of brainstem compression and anomalies of the craniovertebral junction.

Both Greenlee et al.[12] and Goel et al.[9] reported an increased incidence of transoral decompression and posterior fusion among patients with severe basilar invagination. Greenlee and colleagues reported a series of 112 patients with CM, of whom only 44% underwent posterior decompression alone.[12] They noted an increased requirement for ventral decompression among patients with higher degrees of basilar invagination and a higher basal angle. Although they recommended posterior decompression and fusion followed by transoral decompression when indicated in high-risk patients, they did not make specific recommendations based on a statistically validated combination of radiographic parameters.[12] Goel and colleagues reported 190 cases of basilar invagination, of whom 102 had associated CM-I.[9] Overall, 21% underwent transoral decompression, either alone or in conjunction with posterior suboccipital decompression of the foramen magnum; however, only 13% underwent posterior occipitocervical fusion. Among patients with CM undergoing ventral decompression, improvement was observed in 45% who underwent transoral decompression alone, and 57% of patients who underwent transoral decompression in conjunction with posterior decompression of the foramen magnum. It is unclear which patients required occipitocervical fusion, even after both ventral and dorsal decompression.[9] Other authors have stressed the use of posterior decompression and open reduction and fusion

TABLE 5: Univariate analysis associated with the need for delayed occipitocervical fusion, excluding the patients who underwent fusion at the time of initial decompression

Variable	Decompression Only (n= 82)	Decompression & Delayed Fusion (n= 8)	Univariate p Value
mean age in yrs ± SD	9.7 ± 9.1	8.7 ± 5.1	NS
male sex (%)	46 (56)	2 (25)	NS
CM 1.5 (%)	18 (22)	7 (87)	<0.001
presence of scoliosis (%)	20 (24)	2 (25)	NS
presence of syrinx (%)	42 (51)	4 (50)	NS
medullary kink (%)	21 (26)	6 (75)	0.008
odontoid process retroflexion (%)	30 (37)	4 (50)	NS
basilar invagination (%)	0 (0)	5 (62)	<0.001
mean pB–C2 (mm) ± SD	7.2 ± 2.1	9.6 ± 2.8	0.003
pB–C2 ≥9 mm (%)	20 (24)	4 (50)	NS
mean tonsillar descent (mm) ± SD	13 ± 5.1	14.3 ± 4.7	NS
mean CXA (°) ± SD	141.1 ± 14.6	120.4 ± 22.1	<0.001
CXA <125° (%)	9 (11)	5 (62)	0.002

in the context of symptomatic ventral compression resulting from basilar invagination.[19]

Grabb and colleagues[11] analyzed the degree of ventral brainstem compression among 40 patients with Chiari malformation. Specifically, while only 5% of patients in their series had basilar invagination, 30% had a pB–C2 ≥ 9 mm, emphasizing the variable pathophysiology of ventral brainstem compression in the context of CM-I. In contrast to our series, no patients in their study with a pB–C2 < 9 mm experienced a failure of posterior decompression. However, among 12 patients with a pB–C2 ≥9 mm, different treatment strategies were employed, including posterior decompression alone, posterior decompression and occipitocervical fusion, and circumferential decompression together with occipitocervical fusion. No objective radiographic criteria were explicitly outlined as a basis for surgical decision making in this high-risk subgroup.[11]

In addition, the authors discussed potential drawbacks of using CXA as a measurement of ventral compression. Specifically, they noted that it is an indirect measurement of osseous structures only, and they reported variability in the measurement between studies in the same patient, likely because of dynamic changes in flexion and extension.[11] The latter was not the case in our study population. We agree that the pB–C2 is a more direct and less variable measurement of ventral brainstem compression; however, our analysis suggests that a more stringent cutoff of CXA < 125°, rather than 150° as described by previous authors,[30] together with basilar invagination and CM 1.5, were the best predictors of a complex CM requiring an

TABLE 6: Multivariate regression analysis of variables associated with the need for delayed occipitocervical fusion, excluding patients who underwent fusion at the time of initial decompression

Variable	Univariate p Value	Multivariate p Value
CM 1.5	<0.001	0.02
medullary kink	0.008	NS
basilar invagination	<0.001	<0.001
mean pB–C2	0.003	NS
CXA <125°	0.002	0.04

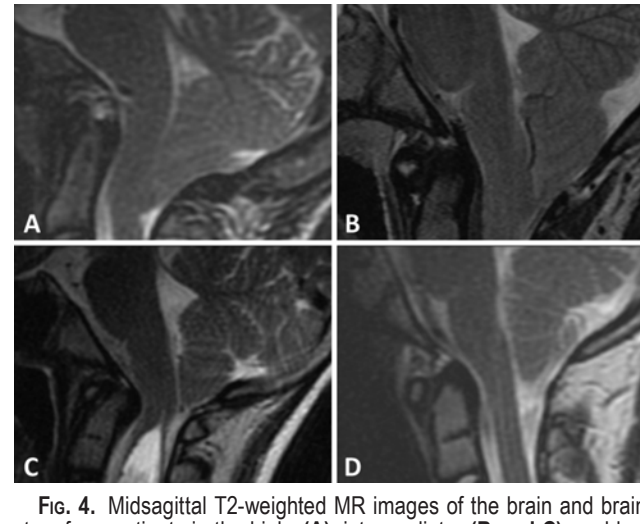

Fig. 4. Midsagittal T2-weighted MR images of the brain and brainstem from patients in the high- **(A)**, intermediate- **(B and C)**, and low-risk groups **(D)** in the study population. **A:** High-risk patient with CM 1.5 and CXA of 100°. Note the clear presence of ventral brainstem compression in the context of retroflexion of the odontoid process. This patient underwent transoral decompression followed by suboccipital craniectomy and occiput-to-C2 posterior fusion as an initial procedure. Within the 1-year follow-up, her symptoms resolved and she has not required further surgery. **B:** Intermediate-risk patient with CM 1.5 and CXA of 135°. This patient underwent suboccipital decompression, but required reoperation 2 months after surgery for recurrent symptoms. He continues to do well after 4 years of follow-up. **C:** Intermediate-risk patient with CM-I and CXA of 124°. Her symptoms resolved and her holocord syrinx completely disappeared 1 year after posterior decompression, and she continues to do well 3 years after surgery. **D:** Low-risk patient with CM-I and CXA of 148°. This patient underwent suboccipital decompression and remains asymptomatic at the 1-year follow-up.

alternative treatment strategy (posterior decompression and occipital cervical fusion rather than decompression alone).

Fenoy et al.[8] proposed a classification system for patients with hindbrain herniation requiring occipitocervical fusion. Specifically, they recognized patients with reducible bone compression (I), irreducible bone compression requiring ventral decompression (II), occipitocervical instability in the absence of osseous anomalies (III), and ligamentous instability (IV). They relied on dynamic MRI and short tau inversion-recovery imaging to identify these groups.[8] Comparatively, our study benefits from applying simple, reproducible measurements to routine static imaging and may identify high-risk patients who require further evaluation with dynamic studies.

Important limitations of our study include the fact that it was a retrospective analysis of the experience at a single pediatric center. Furthermore, the external validity of our analysis is limited because we only had preoperative imaging available for review on 101 of the 206 patients who underwent surgical management of a CM-I during the study period. Finally, definitive conclusions may not be drawn because of the small number of patients with complex CMs requiring fusion in our group. Independent, prospective analysis of a separate study population is an essential next step in validating the clinical significance of our results.

In light of previous work, our series suggests that specific radiographic criteria applied in a simple and robust model may identify patients with complex CM at the time of diagnosis and predict the need for decompression as well as open reduction and occipitocervical fusion. We understand that the underpinnings of these radiographic criteria are grounded in complex embryological, developmental, and biomechanical principles[22,24,29,30] and, furthermore, are highly variable from patient to patient.[10,16,18,23,28] However, the ultimate manifestations of these underlying principles may not be as complex as previously believed. Perhaps studies such as these may help guide further work in determining critical issues important for the early identification and definitive surgical management of patients with complex CMs.

Conclusions

We identified a specific profile of radiographic risk factors among patients with CM, present at the time of diagnosis, which are easily identified, reproducible among professionals, and significantly associated with the need for occipitocervical fusion in this study population. Patients with complex CMs with brainstem compression, anomalies of the craniocervical junction, or both, are identified by CM 1.5, basilar invagination, and CXA < 125°. This is a simple model that may be used to counsel patients, determine surgical treatment, or identify patients in need of careful follow-up; however, this model requires validation in an independent study population.

Disclosure

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 to the study and manuscript preparation include the following. Conception and design: D Brockmeyer. Acquisition of data: Bollo, M Brockmeyer. Analysis and interpretation of data: D Brockmeyer, Bollo, Riva-Cambrin. Drafting the article: Bollo. Critically revising the article: all authors. Reviewed submitted version of manuscript: all authors. Approved the final version of the manuscript on behalf of all authors: D Brockmeyer. Statistical analysis: Bollo, Riva-Cambrin. Study supervision: D Brockmeyer.

Acknowledgment

The authors wish to thank Kristin Kraus, M.Sc., for her help in the preparation of this manuscript.

References

1. Aitken LA, Lindan CE, Sidney S, Gupta N, Barkovich AJ, Sorel M, et al: Chiari type I malformation in a pediatric population. **Pediatr Neurol 40:**449–454, 2009
2. Albert GW, Menezes AH, Hansen DR, Greenlee JD, Weinstein SL: Chiari malformation Type I in children younger than age 6 years: presentation and surgical outcome. Clinical article. **J Neurosurg Pediatr 5:**554–561, 2010
3. Brockmeyer D, Gollogly S, Smith JT: Scoliosis associated with Chiari 1 malformations: the effect of suboccipital decompression on scoliosis curve progression: a preliminary study. **Spine (Phila Pa 1976) 28:**2505–2509, 2003
4. Caetano de Barros M, Farias W, Ataíde L, Lins S: Basilar impression and Arnold-Chiari malformation. A study of 66 cases. **J Neurol Neurosurg Psychiatry 31:**596–605, 1968
5. Chirossel JP, Passagia JG, Gay E, Palombi O: Management of craniocervical junction dislocation. **Childs Nerv Syst 16:** 697–701, 2000
6. Collignon FP, Cohen-Gadol AA, Krauss WE: Circumferential decompression of the foramen magnum for the treatment of syringomyelia associated with basilar invagination. **Neurosurg Rev 27:**168–172, 2004
7. Dyste GN, Menezes AH, VanGilder JC: Symptomatic Chiari malformations. An analysis of presentation, management, and long-term outcome. **J Neurosurg 71:**159–168, 1989
8. Fenoy AJ, Menezes AH, Fenoy KA: Craniocervical junction fusions in patients with hindbrain herniation and syringohydromyelia. **J Neurosurg Spine 9:**1–9, 2008
9. Goel A, Bhatjiwale M, Desai K: Basilar invagination: a study based on 190 surgically treated patients. **J Neurosurg 88:** 962–968, 1998
10. Goel A, Desai K, Bhatjiwale M, Muzumdar DP: Basilar invagination and Chiari malformation associated with cerebellar atrophy: report of two treated cases. **J Clin Neurosci 9:** 194–196, 2002
11. 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
12. Greenlee J, Garell PC, Stence N, Menezes AH: Comprehensive approach to Chiari malformation in pediatric patients. **Neurosurg Focus 6(6):**e4, 1999
13. Hankinson TC, Klimo P Jr, Feldstein NA, Anderson RC, Brockmeyer D: Chiari malformations, syringohydromyelia and scoliosis. **Neurosurg Clin N Am 18:**549–568, 2007
14. Heiss JD, Suffredini G, Smith R, DeVroom HL, Patronas NJ, Butman JA, et al: Pathophysiology of persistent syringomyelia after decompressive craniocervical surgery. Clinical article. **J Neurosurg Spine 13:**729–742, 2010
15. Henderson FC, Wilson WA, Mott S, Mark A, Schmidt K, Berry JK, et al: Deformative stress associated with an abnormal clivo-axial angle: a finite element analysis. **Surg Neurol Int 1:**30, 2010

16. Heusner AP: Platybasia with Arnold-Chiari syndrome. **J Nerv Ment Dis 112:**350–351, 1950
17. Hwang SW, Heilman CB, Riesenburger RI, Kryzanski J: C1-C2 arthrodesis after transoral odontoidectomy and suboccipital craniectomy for ventral brain stem compression in Chiari I patients. **Eur Spine J 17:**1211–1217, 2008
18. Kagawa M, Jinnai T, Matsumoto Y, Kawai N, Kunishio K, Tamiya T, et al: Chiari I malformation accompanied by assimilation of the atlas, Klippel-Feil syndrome, and syringomyelia: case report. **Surg Neurol 65:**497–502, 2006
19. Kim LJ, Rekate HL, Klopfenstein JD, Sonntag VKH: Treatment of basilar invagination associated with Chiari I malformations in the pediatric population: cervical reduction and posterior occipitocervical fusion. **J Neurosurg 101 (2 Suppl):**189–195, 2004
20. Kyoshima K, Kakizawa Y, Tokushige K, Akaishi K, Kanaji M, Kuroyanagi T: Odontoid compression of the brainstem without basilar impression—"odontoid invagination." **J Clin Neurosci 12:**565–569, 2005
21. McGirt MJ, Attenello FJ, Atiba A, Garces-Ambrossi G, Datoo G, Weingart JD, et al: Symptom recurrence after suboccipital decompression for pediatric Chiari I malformation: analysis of 256 consecutive cases. **Childs Nerv Syst 24:**1333–1339, 2008
22. Menezes AH: Craniocervical developmental anatomy and its implications. **Childs Nerv Syst 24:**1109–1122, 2008
23. Menezes AH: Craniovertebral junction database analysis: incidence, classification, presentation, and treatment algorithms. **Childs Nerv Syst 24:**1101–1108, 2008
24. Menezes AH, Traynelis VC: Anatomy and biomechanics of normal craniovertebral junction (a) and biomechanics of stabilization (b). **Childs Nerv Syst 24:**1091–1100, 2008
25. Milhorat TH, Bolognese PA, Nishikawa M, McDonnell NB, Francomano CA: Syndrome of occipitoatlantoaxial hypermobility, cranial settling, and Chiari malformation Type I in patients with hereditary disorders of connective tissue. **J Neurosurg Spine 7:**601–609, 2007
26. Milhorat TH, Chou MW, Trinidad EM, Kula RW, Mandell M, Wolpert C, et al: Chiari I malformation redefined: clinical and radiographic findings for 364 symptomatic patients. **Neurosurgery 44:**1005–1017, 1999
27. Nishikawa M, Ohata K, Baba M, Terakawa Y, Hara M: Chiari I malformation associated with ventral compression and instability: one-stage posterior decompression and fusion with a new instrumentation technique. **Neurosurgery 54:**1430–1435, 2004
28. Quebada PB, Duhaime AC: Chiari malformation Type I and a dolichoodontoid process responsible for sudden cardiorespiratory arrest. Case report. **J Neurosurg 103 (6 Suppl):**567–570, 2005
29. Sgouros S, Kountouri M, Natarajan K: Skull base growth in children with Chiari malformation Type I. **J Neurosurg 107 (3 Suppl):**188–192, 2007
30. Smoker WR: Craniovertebral junction: normal anatomy, craniometry, and congenital anomalies. **Radiographics 14:**255–277, 1994
31. Tubbs RS, Iskandar BJ, Bartolucci AA, Oakes WJ: A critical analysis of the Chiari 1.5 malformation. **J Neurosurg 101 (2 Suppl):**179–183, 2004
32. Tubbs RS, McGirt MJ, Oakes WJ: Surgical experience in 130 pediatric patients with Chiari I malformations. **J Neurosurg 99:**291–296, 2003
33. Zileli M, Cagli S: Combined anterior and posterior approach for managing basilar invagination associated with type I Chiari malformation. **J Spinal Disord Tech 15:**284–289, 2002

Manuscript submitted August 10, 2011.
Accepted March 13, 2012.
Portions of this work were presented in abstract form on December 2, 2010, as proceedings at the 2010 Annual Meeting of the AANS/CNS Section on Pediatric Neurological Surgery in Cleveland, Ohio.
Please include this information when citing this paper: published online June 22, 2012; DOI: 10.3171/2012.3.PEDS11340.
Address correspondence to: Douglas L. Brockmeyer, M.D., Division of Pediatric Neurosurgery, Primary Children's Medical Center, 100 North Mario Capecchi Drive, Suite 1475, Salt Lake City, Utah 84113. email: douglas.brockmeyer@hsc.utah.edu.