Delayed resolution of syrinx after posterior fossa decompression without dural opening in children with Chiari malformation Type I

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OBJECT Chiari malformation Type I (CM-I) is associated with a syrinx in 25%–85% of patients. Although posterior fossa decompression (PFD) without dural opening is an accepted treatment option for children with symptomatic CM-I, many surgeons prefer to open the dura if a syrinx exists. The purpose of this study was to investigate the frequency and timing of syrinx resolution in children undergoing PFD without dural opening for CM-I.

METHODS A retrospective review of 68 consecutive pediatric patients with CM-I and syringomyelia who underwent PFD without dural opening was conducted. Patient demographics, presenting symptoms and signs, radiographic findings, and intraoperative ultrasound and neuromonitoring findings were studied as well as the patients' clinical and radiographic follow-up.

RESULTS During the mean radiographic follow-up period of 32 months, 70% of the syringes improved. Syrinx improvement occurred at a mean of 31 months postoperatively. All patients experienced symptom improvement within the 1st year, despite only 26% of patients showing radiographic improvement during that period. Patients presenting with sensory symptoms or motor weakness had a higher likelihood of having radiographic syrinx improvement postoperatively.

CONCLUSIONS In children with CM-I and a syrinx undergoing PFD without dural opening, syrinx resolution occurs in approximately 70% of patients. Radiographic improvement of the syrinx is delayed, but this does not correlate temporally with symptom improvement. Sensory symptoms or motor weakness on presentation are associated with syrinx resolution after surgery.

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KEY WORDS syrinx; syringomyelia; Chiari; nondural opening; suboccipital decompression; outcomes

CHIARI malformation Type I (CM-I) is associated with syrinx in 25%–85% of patients.^{1,11,12,18,22,26,32,35,42} The descent of the cerebellar tonsils through the foramen magnum in CM-I is associated with abnormal CSF dynamics at the craniocervical junction, and it has been posited that these effects play a role in syrinx formation. A syrinx is considered responsible for motor, sensory, and some pain symptoms as well as scoliosis.^{21,23,43,47}

A syrinx associated with pediatric CM-I is considered by many surgeons an indication for surgical treatment, usually by posterior fossa decompression (PFD).^{4,11,25,26,32,35,42} PFD with dural opening has been associated with improvement in syrinx in 50%–100% of patients, with symptom resolution commonly occurring before syrinx improvement, and scoliosis improvement often occurring after syrinx improvement.^{1–8,10–12,15–17,20,26,27,29–35,42,44}

A variety of methods for PFD have been described for

ABBREVIATIONS BAER = brainstem auditory evoked response; CM-I = Chiari malformation Type I; PFD = posterior fossa decompression; SSEP = somatosensory evoked potential.

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pediatric CM-I with favorable outcomes.^{13,14,26,32} Although pediatric neurosurgeons have a great deal of experience with PFD with duraplasty, both case series as well as intraoperative electrophysiological studies have demonstrated that many patients can experience physiological decompression without opening of the dura.^{2,3,8,15,32,49} As a result, it is controversial whether the dura must be opened for adequate decompression.

Rapid syrinx improvement in these patients has been noted frequently with dural opening,^{1–8,10–12,15–17,20,26,27,29–35,42,44,46} and some pediatric neurosurgeons consider the presence of a syrinx to be an indication to open the dura in PFD.^{17,19,36,39} However, radiographic syrinx improvement has been shown to be unnecessary for symptom improvement,^{6,8,15,26,37,46} and a syrinx may resolve in delayed fashion.⁹ The time course of syrinx resolution after dural opening surgery in pediatric CM-I has been described recently,⁴⁶ but no large series has addressed this after non-dural opening surgery.

The purpose of this study was to determine the clinical outcomes as well as the frequency and time course of syrinx improvement after PFD without dural opening in children with CM-I and a syrinx.

Methods

Following approval by the Institutional Review Board at Columbia University Medical Center/Morgan Stanley Children's Hospital of NewYork-Presbyterian, a retrospective analysis was undertaken of all 68 patients with a syrinx of greater than 2 mm who were younger than 21 years and who underwent surgical decompression of CM-I without dural opening performed by the senior authors (N.A.F. and R.C.A.) between 2003 and 2013. Preoperative and postoperative records from office charts, operative reports, inpatient records, and pre- and postoperative images were reviewed. All patients had undergone preoperative MRI of the brain and spinal cord to evaluate the malformation and syrinx.

Patients were excluded from the study if they underwent a PFD with dural opening. We typically perform PFD without dural opening for children with CM-I regardless of whether there is a syrinx. PFD with dural opening was performed as the primary procedure if any of the following were present: 1) rapidly progressing neurological deficit, 2) rapidly progressing scoliosis with a syrinx and Cobb angle greater than 35°, and 3) tonsillar descent below the caudal aspect of the C-2 lamina on preoperative MRI. Patients who underwent PFD without dural opening were excluded from analysis if they had previously undergone PFD at another institution or if it was believed they had acquired, rather than congenital, Chiari malformation secondary to spinal pathology or spine surgery. Patients with complex craniocervical junction anomalies who required posterior occipitocervical fusion at the time of the PFD were also excluded.

Each patient underwent a bony decompression of the craniocervical junction without opening of the dura as previously described.²⁴ Briefly, under general anesthesia, the patient was placed in 3-point Mayfield fixation, and neurophysiological monitoring of somatosensory evoked

potentials (SSEPs) and brainstem auditory evoked responses (BAERs) was performed with the patient supine and after prone positioning and fixation with neck flexion. SSEP monitoring was performed to ensure safety of positioning with neck flexion,² while BAERs were monitored as a potential indicator of physiological decompression.³ After standard surgical preparation, draping, and administration of local anesthesia, an incision was made from theinion to the upper cervical region. Subperiosteal dissection was performed exposing the occiput, foramen magnum, and C-1, and a generous craniectomy was performed using a Midas drill and rongeurs. A C-1 laminectomy was routinely performed with rongeurs.

Intraoperative ultrasonography was used at this point to confirm that the dural exposure was adequate to reach the caudal aspect of the tonsils. In the rare case that the caudal aspect of the tonsils could not be appreciated above C-2 on ultrasonography, a partial laminectomy or undercutting was performed on the rostral aspect of C-2. No patient underwent complete C-2 laminectomy.

The atlantooccipital ligament was then sharply incised and numerous rostrocaudal scoring incisions were made in the outer layer of the cervical dura (Fig. 1). Ultrasound was repeated to confirm improvement in movement of the cerebellar tonsils and an increase in the subarachnoid spaces dorsal and ventral to the cerebellar tonsils. Meticulous inspection for CSF leak as well as hemostasis were then performed prior to multilayer closure with absorbable sutures.

In general, patients underwent follow-up MRI 1 year postoperatively, unless earlier MRI was clinically indicated. Patients with large multiloculated syringes were more likely to be followed with yearly serial MR images (even if they were doing well clinically) until substantial radiographic improvement of the syrinx was seen.

Statistical analyses were performed using the Student t-test or Fisher's exact test, as appropriate.

Results

Patients

Sixty-eight consecutive patients undergoing PFD without dural opening for CM-I with an associated syrinx by the senior authors between 2003 and 2013 were identified. Forty-three percent (29/68) were boys, and the mean age was 10.9 years (range 3–20 years). Patients presented with a mean symptom duration of 1.9 years. Clinical outpatient follow-up was available for all patients, and the mean follow-up time was 40 months. Five patients had surgery within 1 year of data collection and were being clinically followed but had not yet undergone postoperative imaging, and 6 other patients did not have imaging available for review. Therefore, at least 1 postoperative MRI study was evaluated for the remaining 57 patients. The mean radiographic follow-up for these patients was 32 months.

Presentation

Subjective complaints of headache and neck pain were the most common presenting symptoms (Table 1). Taken together, headache, neck pain, and irritability of a younger patient were present in 62% (42/68) of patients at presenta-

FIG. 1. Intraoperative photograph obtained after bony decompression, with overlaid illustration to indicate the general spatial orientation of the techniques of sectioning the atlantooccipital ligament (arrows) and the vertical scoring incisions in the outer layer of the dura (dashed lines), along with the instrument used for the vertical scoring incisions. Figure is available in color online only.

tion. Hyperreflexia and sensory symptoms were also common in this cohort. Twelve patients were asymptomatic except for a syrinx and CM-I. Mean tonsillar descent was 13.7 mm below the foramen magnum. Sixty-three percent (43/68) of syringes spanned both cervical and thoracic segments, 9 of which (13%) spanned the entire length of the spinal cord. Twenty-two percent (15/68) of syringes were isolated to the cervical region, and the remaining 15% (10/68) were isolated thoracic syringes. Twenty-five percent (17/68) of patients presented with scoliosis with a Cobb angle of at least 10°. On presentation, 4 patients had mild to moderate ventriculomegaly, 1 had neurofibromatosis Type 1, and 1 had a tethered cord.

Intraoperative Findings

All patients underwent intraoperative neuromonitoring with SSEPs and BAERs throughout the surgery, as well as intraoperative ultrasound after suboccipital craniectomy and C-1 laminectomy. No concerning electrophysiological changes were reported during any surgery. Eighty-four percent (57/68) of patients demonstrated improvement in the BAER I-V interpeak latency intraoperatively, and the

TABLE 1. Presenting symptoms and signs

Presenting Symptom/Sign	No. of Patients
Atypical headache	24
Neck pain	22
Occipital/tussive headache	19
Hyperreflexia	20
Sensory symptoms	16
Dysphagia	9
Back pain	9
Motor weakness	7
Ataxia	7
Asymmetric abdominal reflexes	7
Snoring	6
Behavioral symptoms/developmental delay	5
Irritability	4
Nystagmus	3

remaining 11 patients had stable recordings; 8.8% (6/68) of patients underwent a partial superior C-2 laminectomy based on suboptimal decompression of the cerebellar tonsils seen on initial intraoperative ultrasound.

Complications

There were no major complications or deaths. Two patients experienced minor complications, one with a perioperative pneumonia and another with thigh paresthesias due to positioning.

Clinical Outcome

All 56 (100%) of the patients presenting with Chiari symptoms experienced at least some degree of symptomatic improvement within the 1st postoperative year. All 12 (100%) of the patients who had asymptomatic CM-I with syrinx demonstrated syrinx improvement. Eighty-eight percent (60/68) of patients were asymptomatic or minimally symptomatic throughout the follow-up period. Forty-six percent (31/68) were completely asymptomatic and 43% (29/68) remained with subtle, improved symptoms that did not affect their return to school or routine activities. Twelve percent (8/68) of patients required reoperation with dural opening for progression of scoliosis without syrinx improvement or return of Chiari symptoms, 3 of whom demonstrated a concomitantly worsening syrinx. None of these patients were noted to have adhesions or webs obstructing outflow of the fourth ventricle.

Syrinx Outcome

The 57 patients with available images were studied with a mean radiographic follow-up of 32 months, and 70% (40) demonstrated radiographic improvement on MRI. Of these 40 improved syringes, one-third (13) demonstrated near complete collapse and an additional 20% (8) were at least 50% smaller, with 47% (19) showing more modest improvement. All 12 asymptomatic syringes demonstrated radiographic improvement postoperatively.

Eighteen percent (3/17) of patients whose syringes did

not improve required reoperation for persistent or worsening Chiari symptoms: 2 of these patients had demonstrated earlier radiographic syrinx improvement and 1 had remained stable. Despite radiographic improvement of the syrinx, 12% (5/40) of patients underwent a second operation for clinical symptom recurrence.

Age, duration of symptoms at presentation, degree of tonsillar ectopia, extent of cord syrinx, sensory or motor symptoms of the trunk or extremities, need to perform partial C-2 laminectomy based on intraoperative ultrasonography findings, and presence and degree of scoliosis were all assessed for a possible influence on syrinx resolution after PFD without dural opening. The association between motor/sensory symptoms and radiographic resolution of syrinx is determined by the difference between the proportion of patients with these symptoms whose syringes resolve (94%) and the proportion of patients without these symptoms whose syringes resolve (64%) ($p = 0.011$). No other variables were associated with syrinx resolution (Table 2).

For the 17 patients with scoliosis, the median Cobb angle on presentation was 25°. Fourteen patients had appropriate follow-up imaging available for review. Nine patients had improvement of the syrinx and 5 did not. Of the 9 patients with an improving syrinx, 1 had improvement of scoliosis; 6 had stabilization of the Cobb angle (less than 10° change), 1 of whom underwent spinal fusion surgery; 1 demonstrated progression of scoliosis and underwent spinal fusion surgery; and 1 demonstrated progression of scoliosis, underwent reoperation for dural opening PFD, continued to exhibit scoliosis progression, and then underwent spinal fusion surgery. Of the 5 patients whose syringes did not improve, 2 exhibited improving scoliosis; 1 patient's Cobb angle was stable; 1 demonstrated scoliosis progression and underwent spinal fusion surgery; and 1 demonstrated scoliosis progression, underwent reoperation for dural opening PFD, continued to exhibit scoliosis progression, and underwent spinal fusion surgery (Table 3).

Timing of Syrinx Improvement

For the 70% (40/57) of patients who did demonstrate radiographic improvement, the median and mean time to improvement were 21 and 31 months, respectively. Syringes that were not seen to improve were followed radiographically for a mean of 33 months. Some patients, such as the patient in Fig. 2, improved symptomatically early in their postoperative course, but maintained a stable syrinx size for months or years prior to eventual radiographic resolution. Specifically, 19 syringes improved in the 1st postoperative year, 10 in the 2nd year, 4 in the 3rd year, 3

TABLE 3. Scoliosis outcomes by syrinx outcomes

Syrinx	No. of Patients		
	Improved Cobb Angle	Stable Cobb Angle*	Worsened Cobb Angle (also reop for Chiari)
Improving	1	6	2 (1)
Stable or worsening	2	1	2 (1)

* Stable Cobb angle was considered < 10° of change from the preoperative baseline.

in the 4th year, 1 in the 5th year, 1 in the 6th year, 1 in the 7th year, and 1 in the 8th year (Table 4, Fig. 3).

Five percent (3/57) of patients demonstrated worsening of the syrinx, and all 3 had concomitant recurrence of Chiari symptoms and required reoperation. Two of these 3 patients had experienced radiographic and symptomatic improvement at 1 and 19 months, respectively, followed by symptom recurrence and worsening of syrinx at 7 and 35 months, respectively.

Discussion

In this report, we have demonstrated that: 1) 70% of children undergoing PFD without dural opening for CM-I with syrinx will have radiographic improvement in the syrinx, 2) radiographic improvement of the syrinx occurs in a delayed fashion postoperatively, 3) syrinx improvement is not essential for clinical improvement, and 4) patients who present with motor/sensory symptoms are more likely to have syrinx improvement. To our knowledge, this is the largest reported series of outcomes after posterior fossa decompression without dural opening in children with CM-I with syrinx.

Previous studies have shown that CM-I induces alterations in CSF dynamics at the foramen magnum that can cause a syrinx in 25%–85% of patients.^{1,11,12,18,22,26,32,35,42} Children with CM-I have smaller posterior fossae than controls,^{28,38,41,45} and in 1 study, patients with an associated syrinx had yet smaller posterior fossae than CM-I patients without a syrinx.⁴⁰ Syrinx has a strong association with scoliosis as well as motor and sensory symptoms. Many pediatric neurosurgeons consider a syrinx that is associated with CM-I an indication for PFD.^{4,11,25,26,32,35,42} High rates of radiographic syrinx improvement have been reported after PFD with dural opening in pediatric CM-I, with symptom resolution often occurring prior to syrinx resolution and scoliosis improvement often occurring after.^{1–8,10–12,15–17,20,26,27,29–35,42,44} In part because syrinx improvement has been noted to occur in the majority of

TABLE 2. Patients whose syringes improved versus those whose syringes failed to improve

Syrinx*	Total No. of Patients	Radiographic Follow-Up (mos)	Age (yrs)	Symptom Duration (yrs)	Tonsillar Descent (mm)	No. w/ Whole Cord Syrinx	No. w/ Motor &/or Sensory Symptoms	No. w/ C-2 Laminectomy	No. w/ Scoliosis	No. w/ Reoperation
Improving	40	31	10.4	2.3	13.5	6	16	3	9	5
Stable/worsening	17	33	11	1.7	15.8	3	1	2	5	3
p value		0.85	0.73	0.57	0.25	0.99	0.011	0.63	0.74	0.68

* Many presenting signs and symptoms not tabulated here exhibited similar proportions between groups and/or had low numbers of patients.

FIG. 2. Sagittal T2-weighted MR images obtained in a 15-year-old girl who presented with a 6-month history of dizziness and episodes of visual loss with CM-I with 9 mm of tonsillar ectopia on MRI with an associated cervicothoracic syrinx. **Left:** Preoperative MRI showing CM-I and syrinx. **Right:** Image obtained 12 months after nondural opening posterior fossa decompression shows dramatic reduction of syrinx.

patients at an early time point (within a year) after dural opening,^{1–8,10–12,15–17,20,26,27,29–35,42,44,46} a large majority of pediatric neurosurgeons consider the presence of a syrinx to be an indication to open the dura during PFD.^{17,19,36,39} At the 2006 American Society of Pediatric Neurosurgeons meeting, for example, a survey of 50% of the membership demonstrated that for children with a symptomatic CM-I and syrinx, only 4% would perform nondural opening surgery and another 4% would use ultrasound to guide whether to add duraplasty, with the vast majority opting for duraplasty with or without tonsillar resection.³⁶

However, controversy exists over whether the dura must be opened for successful surgery for pediatric CM-I with or without a syrinx, with good clinical outcomes being reported for a variety of methods.^{13,14,26,32} Furthermore, several groups have reported symptom improvement after PFD in the absence of radiographic syrinx improvement,^{6,8,15,26,37,46} and it has been shown in some cases that the syrinx may resolve in a delayed fashion.⁹ The frequency and time course of syrinx resolution after PFD without dural opening has not been previously studied in a large series. This information is important for neurosurgeons when determining whether to perform a dural opening procedure and when deciding whether a prior PFD without dural opening has failed and further surgery is needed.

In the current series of children with CM-I and syrinx, 70% of patients undergoing PFD without dural opening had a radiographic improvement in the syrinx. This rate is similar to previous studies that have reported the proportion of syrinx improvement after PFD with dural opening between 55% and 100%.^{1–3,5,6,11,20,26,35,44} In the largest series of patients undergoing PFD with dural opening for pediatric CM-I, 80% radiographic syrinx improvement was reported.⁴² In the 2 largest series of patients with PFD and mixed dural opening and non-dural opening surgeries, the radiographic syrinx improvement rate was 62% in one study and 29% in the other, with no difference detected between the 2 procedure types.^{27,32} A recent meta-analysis that only included one of the aforementioned studies found that 56% of syringes improved after nondural open-

TABLE 4. Time from surgery to syrinx improvement

Postop Yr	No. of Patients
1	19
2	10
3	4
4	3
5	1
6	1
7	1
8	1

ing surgeries compared with 87% after dural opening, but due to the small number of patients in the literature with a syrinx who underwent PFD without dural opening, this difference was not statistically significant.

Among the patients who exhibited syrinx improvement, the median time to improvement was 21 months and the mean was 31 months, highlighting that some syringes will resolve several years after surgery. This is different from studies of the timing of syrinx resolution in series of dural opening surgery, which demonstrate more rapid radiographic change. One study by Wu and colleagues of 44 pediatric patients undergoing dural opening PFD for CM-I, syrinx, and scoliosis reported syrinx improvement in 82% of patients by 6 months postoperatively, and in 98% of patients over the course of follow-up.⁴⁸ Wetjen and colleagues studied 29 adult patients with CM-I and syrinx after dural opening PFD and found the median time from surgery to greater than 50% reduction in syrinx size to be 3.6 months with a mean of 6.5 months.⁴⁶ By 6 months, 86% of patients had demonstrated radiographic improvement, and all 29 patients demonstrated radiographic improvement during their follow-up. Furthermore, 59% of patients in their series exhibited complete syrinx collapse. Taken together, these data suggest that in patients with CM-I and syrinx, dural opening surgery leads to a more rapid radiographic improvement of syrinx.

Importantly, our results and those of others suggest that syrinx resolution is not essential or directly related to clinical improvement.^{6,9,10,46} Despite only 70% of our patients showing radiographic improvement of the syrinx, 100% had initial improvement postoperatively and 88% remained asymptomatic or with minimal symptoms at last follow-up. These results are similar to the study by Wetjen et al., where at 6 months after PFD with dural opening, 96% of the patients experienced some degree of clinical improvement while only 86% had radiographic improvement of the syrinx; after 2 years, 68% remained mildly symptomatic even though 100% had radiographic improvement of the syrinx. In our study it is possible that with longer follow-up the number of patients with syrinx improvement will increase. This is supported by the fact that the mean time to syrinx resolution was 31 months and the mean follow-up was 33 months, with some patients demonstrating very late improvement up to the 8th postoperative year (Table 4, Fig. 3).

In an attempt to identify preoperative factors that would indicate a higher likelihood of radiographic syrinx

FIG. 3. Proportion of patients whose syringes had improved radiographically plotted against time after nondural opening surgery. Dashed lines indicate 95% confidence intervals. The 3 asterisks indicate the proportion of patients with radiographic syrinx resolution at the 3 time points outlined in the Wetjen et al.⁴⁶ study of dural opening surgeries for comparison.

improvement after PFD without dural opening, we performed multivariate regression analysis. Of all the factors studied, only the presence of preoperative motor or sensory symptoms was significantly associated with radiographic improvement of syrinx postoperatively ($p = 0.011$). All 4 patients with mild to moderate ventriculomegaly experienced syrinx improvement.

Many surgeons routinely open the dura during PFD in patients with syringes to address possible arachnoid veils, webs, or adhesions obstructing outflow of the fourth ventricle that may contribute to syrinx formation. Although these pathological adhesions certainly exist in some patients, we believe that the frequency is very low because we did not see this in any of the 68 patients with syringes in this study. In the 8 patients in this series who underwent reoperation for recurrent symptoms with syrinx, no arachnoid veils, webs, or adhesions obstructing CSF flow were seen.

Our study is limited in its retrospective nature, and due to this, there are no strict criteria governing the timing of imaging for each patient, but we use a relatively consistent algorithm for postoperative imaging as described in *Methods*. The study is further limited by the lack of a control group and rather small numbers from a single center and a relatively short follow-up. However, the favorable clinical outcomes and radiographic improvement reported in this study suggest that the presence of a syrinx should not be considered a contraindication to PFD without dural opening. Furthermore, this study demonstrates that delayed improvement of syringomyelia after PFD without dural opening should be expected and is not an independent indication for reoperation. We look forward to future studies investigating similar questions that will have increased power and granularity generated from collaborative research networks including the Park-Reeves Syringomyelia Research Consortium.

Conclusions

In children with CM-I and a syrinx undergoing PFD without dural opening, syrinx resolution occurs in approximately 70% of patients. Radiographic improvement of the syrinx is delayed, but this does not correlate temporally with symptom improvement. Sensory symptoms or motor weakness on presentation are associated with syrinx resolution after surgery.

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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

Conception and design: Kennedy, Feldstein, Anderson. Acquisition of data: Kennedy, Nelp, Kelly, Phan, McDowell, Feldstein, Anderson. Analysis and interpretation of data: Kennedy, Nelp, Kelly, Phan, Bruce, Feldstein, Anderson. Drafting the article: Kennedy, Feldstein, Anderson. Critically revising the article: Kennedy, Anderson. Reviewed submitted version of manuscript: all authors. Approved the final version of the manuscript on behalf of all authors: Kennedy. Statistical analysis: Bruce. Administrative/technical/material support: Feldstein, Anderson. Study supervision: Kennedy, Feldstein, Anderson.

Supplemental Information

Previous Presentation

The current work has been accepted for presentation to the Congress of Neurological Surgeons Annual Meeting 2015.

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