Child's Nervous System (2021) 37:2891–2898
https://doi.org/10.1007/s00381-021-05280-y

ORIGINAL ARTICLE

Long-term outcomes of posterior fossa decompression for Chiari malformation type 1: which patients are most prone to failure?

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Received: 23 April 2021 / Accepted: 29 June 2021 / Published online: 7 July 2021

Abstract

Purpose The role of an osseous-only posterior fossa decompression (PFD) for Chiari malformation type 1 (CM1) remains controversial. We reviewed *long-term outcomes* for patients with CM1 undergoing a PFD to evaluate if there was any difference for failure when compared to patients undergoing a PFD with duraplasty (PFDD).

Methods Consecutive patients surgically treated at a single tertiary pediatric neurosurgery clinic over a 25-year period with at least 5 years of follow-up were evaluated. PFD patients were compared to those that initially received a PFDD. Demographics, surgical indications, surgical approach, outcomes, and complications were reviewed.

Results A total of 60 patients were included in this study of which 25 (41.67%) underwent PFD and 35 (58.33%) underwent PFDD. Mean age at surgery was 7.41 years (range 0.4 to 18 years) with a mean follow-up of 8.23 years (range 5 to 21 years). Those that received a PFD had a lower rate of radiographic syrinx improvement ($p = 0.03$), especially in the setting of holocord syringes. Failure rate was significantly higher in the PFD group (20% vs 2.90%, $p = 0.03$). However, complications were significantly higher in the PFDD group (17.14% vs 4.0%, $p = 0.04$).

Conclusions PFD provides a safe treatment option with similar clinical improvements and lower post-operative complication rate compared to PFDD, albeit at the cost of greater chance of reoperation, especially in the setting of a holocord syrinx. Patients with a holocord syrinx should be considered for a PFDD as their initial procedure.

Keywords Chiari malformation · Duraplasty · Long-term · Syringomyelia · Syrinx

Introduction

Chiari malformation type 1 (CM1) is defined as a 5 mm or greater caudal displacement of the cerebellar tonsils through the foramen magnum and into the spinal canal [1]. CM1 is an incidental finding on MRI in 0.8–3.6% of the population [1–4]. The clinical presentation varies significantly, and many cases of childhood CM1 are discovered in an incidental fashion [3, 5, 6]. Common presenting signs and symptoms associated with CM1 include occipital headache, paresthesias, motor deficits, nystagmus, hyperreflexia of the lower extremities, hydrocephalus, and gait disturbances [7]. The prevalence of syringomyelia among patients with CM1 ranges from 12 to 23% [1, 3, 6].

The aim of surgical treatment is to adequately decompress the craniocervical junction, re-establish normal flow of cerebrospinal fluid (CSF), and alleviate the CM1 related symptoms [8]. A wide range of surgical approaches exist to treat CM1, ranging from a less invasive osseous-only posterior fossa decompression (PFD) to PFD with duraplasty (PFDD) and PFDD with fourth ventricular stent placement. As PFDD has been associated with increased risk of complications [9–11], there has been a recent trend towards the less invasive extradural PFD approach [12]. Studies have demonstrated comparable efficacy of PFD compared to PFDD with respect to clinical improvement, with PFD offering a shorter operation time, lower rate of post-operative complications, and shorter hospital length of stay [7, 13, 14]. However, others have shown a greater resolution of symptoms with

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PFDD, particularly in patients with syringomyelia [14, 15]. Choosing between PFD and PFDD for surgical management of pediatric CM1 patients remains controversial [12, 16].

Although CM1 has been an active topic of research for years, there is a paucity of literature with respect to studies comparing the *long-term* efficacy of PFD and PFDD for the pediatric population. It is anticipated that forthcoming answers to the long-term failures following these procedures will help guide neurosurgeons in choosing the most efficacious treatment for their patients.

Methods

Study design

This study was conducted using a combination of prospectively and retrospectively collected data for patients evaluated for a CM1 at Children's National Hospital in Washington, DC. Study data were collected and managed using REDCap electronic data capture tools [17]. Institutional review board approval was obtained (Pro00013560). Patients who underwent a Chiari decompression and followed up between 1993 and 2018 were included. Patients were excluded if they were over 18 years old or had less than 5 years of follow-up. Patients' follow-up appointments were only recorded at our own institution, and not collected for those who may have received care at another neurosurgical institution, such as our "lost to follow-up" cohort. For each patient in both surgical cohorts, the decision to open the dura was the surgeon's preference in addition to ultrasound assessment of tonsillar changes after the suboccipital and C1 decompression. The details of both PFD and PFDD surgical techniques have been extensively described in an article by Alexander et al. [12].

Clinical and radiographic evaluation

All patients in this study had a pediatric neurosurgeon perform a comprehensive clinical and radiographic evaluation. All subjects received complete brain and *entire* spinal cord imaging. CM1 was defined as descent of the cerebellar tonsils 5 mm or more below the foramen magnum. Parameters of interest included age, sex, presentation type (*incidental*: defined as indication for imaging was unrelated to investigating CM1, *non-incidental*: defined as clinical indication for imaging was to investigate a diagnosis of CM1), presenting signs and symptoms, syrinx, surgical approaches, postoperative complications, and follow-up length. The outcomes of PFD patients were compared to a similar cohort of patients that underwent PFDD to determine if a subset of PFD patients was more likely to manifest as a surgical failure. For this study, clinical parameters of interest included Chiari headache (defined as occipital headache and neck pain worsened by Valsalva maneuver, coughing, or sneezing), and neurological symptoms (defined as paresthesia, motor deficits, nystagmus, hyperreflexia of the lower extremities, or gait disturbances). Patients reported their symptoms based on the state of those clinical symptoms at the time of the follow-up appointment, compared to prior to the surgery, on a scale ranging from "better," "stable," or "worse." The radiological parameter of this study was syringomyelia, which was evaluated by periodic MRI's. This was objectively assessed based on the syrinx grade (defined as the ratio between the syrinx maximum anterior-posterior (AP) diameter and the diameter of spinal cord at the level of the maximum AP syrinx diameter (grade I <25%, grade II 26–50%, grade III 51–75%, grade IV >76%)), and its extent of involvement along the length of the spinal cord (cervical, thoracic, cervicothoracic, and holocord syrinx). The syrinx was labeled as "better," "stable," or "worse" based on the syrinx grade and length of spinal cord involvement at the most recent MRI compared to the imaging obtained prior to the surgery. Failure was defined as the need for reoperation to address residual or recurrent clinical symptoms and/or syringomyelia.

Statistical analysis

Demographic and outcome descriptive statistics are reported for the overall sample. The mean with corresponding range is presented for select parametric continuous variables. Categorical variables are presented as frequency (percentage). Clinical and radiologic outcomes were compared by extradural and intradural approaches. Cohorts were compared using independent samples *t*-test and ANOVA for parametric outcomes, while chi-square and Fisher's exact test were used for adequate cell-count categorical outcomes and low cell-count categorical outcomes (≥25% of cells with expected cell-count ≤5), respectively. All statistical analysis was performed using SAS Version 9.4 (SAS Institute Inc., Cary, NC). A $p < 0.05$ was statistically significant.

Results

Study population

A total of 281 patients ≤18-years-old were identified, of which 221 were excluded due to <5 years of follow-up. A total of 60 patients met the inclusion criteria of which 28 were female (46.67%) and 32 were male (53.33%). The mean age at time of surgery was 7.41 years (range 5 months to 18 years). Thirty-seven patients (61.67%) had a Chiari headache, 25 patients (41.67%) had neurological symptoms, and 43 patients (71.67%) had syringomyelia. CM1

Table 1 Patient characteristics

Characteristic variable	Entire cohort ($n = 60$)	PFD cohort ($n = 25$)	PFDD cohort ($n = 35$)	p value
Mean age at time of surgery (years)	7.41 (range 0.4–18 years)	7.63 (range 1–15.8)	7.34 (range 0.4–18)	0.76
Sex Female Male	28 (46.67) 32 (53.33)	11 (44.0) 14 (56.0)	17 (48.57) 18 (51.43)	0.80
Presentation type Incidental Non-incidental	16 (26.67) 44 (73.33)	5 (20.0) 20 (80.0)	11 (31.43) 24 (68.57)	0.52
Chiari headache	37 (61.67)	15 (60.0)	22 (62.86)	0.82
Neurological symptoms	25 (41.67)	6 (24.0)	19 (54.29)	0.02*
Syringomyelia	43 (71.67)	17 (68.0)	26 (74.29)	0.60
Follow-up length (years)	8.23 (range 5.0–21.5 years)	8.81 (range 5.0–21.5)	7.88 (range 5.0–19.82)	0.43

Notation in the parentheses indicate the percentage for sex, presentation type, Chiari headache, neurological symptoms, syrinx, and type of surgery

PFD osseous-only posterior fossa decompression, *PFDD* posterior fossa decompression with duraplasty

*Statistical significance

was discovered incidentally in 16 patients (26.67%) who had imaging as part of a work-up for an unrelated diagnosis. The most common indications for imaging in this group included seizures (18.75%), trauma (12.50%), and non-specific headaches (12.50%). Surgical indication in the incidental cohort was due to de novo development of syringomyelia in 8 patients (50%), both syringomyelia and new-onset Chiari headache in 3 patients (18.75%), and development of neurological complaints in 5 patients (31.25%). For the entire cohort, mean time from diagnosis to surgery was 157.20 days (range 1 day to 3.80 years). The PFD cohort had 25 patients (25/60, 41.67%), while there were 35 (35/60, 58.33%) in the PFDD cohort. Both cohorts had similar rates of Chiari headache and syringomyelia (Table 1). However, neurological symptoms were significantly higher in the PFDD cohort ($p = 0.02$). Mean follow-up length from time of surgery to the last follow-up appointment was 8.23 years (range 5.0 to 21.5 years) for the entire cohort. PFD and PFDD groups had similar mean length of follow up (8.81 vs 7.88 years, $p = 0.43$) (Table 1).

Surgery types and outcomes

There was no statistical difference between the PFD and PFDD cohorts with regard to improvement in Chiari headache (73.33% vs 81.82%, $p = 0.31$), and neurological symptoms (66.67% vs 78.95%, $p = 0.73$) (Table 2). There was also no difference in failure rate based on the degree of tonsillar ectopia ($p = 0.86$). However, PFDD was associated with significantly greater syrinx improvement relative to PFD (80.77% vs 47.06%, $p = 0.03$) (Table 2). Nevertheless, there was no statistical difference with regards to outcomes of *non-holocord syringes* (cervical syrinx, thoracic syrinx, and cervicothoracic syrinx) as improvement rates were similar between PFD and PFDD cohorts (63.63% vs 72.22%, $p = 0.61$). However, there was statistically significant higher

Table 2 Comparison of extradural and intradural surgical approaches

Outcome	PFD ($n = 25$)	PFDD ($n = 35$)	p value
Chiari headache	15 (60.0)	22 (62.86)	0.82
Improved	11 (73.33)	18 (81.82)	0.31
Stable	2 (13.33)	-	
Worse	2 (13.33)	4 (18.18)	
Neurological symptoms	6 (24.0)	19 (54.28)	0.02*
Improved	4 (66.67)	15 (78.95)	0.74
Stable	1 (16.67)	3 (15.79)	
Worse	1 (16.67)	1 (5.26)	
Syrinx	17 (68.0)	26 (74.29)	0.59
Improved	8 (47.06)	21 (80.77)	0.03*
Stable	6 (35.29)	1 (3.85)	
Worse	3 (17.65)	4 (15.38)	

Notations in the parentheses indicate the percentage

*Statistical significance

improvement of *holocord syringes* in the PFDD group compared to PFD (75.0% vs 33.33%, $p = 0.03$). With regard to syrinx grades, there was no statistical difference with respect to improvement of grade I syrinx ($p = 0.56$), grade II syrinx ($p = 0.82$), grade III syrinx ($p = 0.26$), or grade IV syrinx ($p = 0.15$) between the two groups.

Over the follow-up period, 13.33% (2/15) of PFD patients with Chiari headache and 16.66% (1/6) of PFD patients with neurological symptoms reported an unchanged (stable) status of their clinical symptoms. During follow-up appointments, these patients reported either an "improved" or "stable" status of their clinical symptoms, never labeling them as "worse." Therefore, a shared clinical decision between the attending neurosurgeon and legal guardian was made to conservatively manage rather than proceed with PFDD for a second operation. Six out of seventeen (35.3%) PFD patients with syringes who underwent recurring MRI evaluations had objectively stable radiographic findings. The decision to conservatively manage or re-operate on these patients was based on correlating the radiographic findings with their clinical presentation (Chiari headache and neurological symptoms).

Complications

Complications amongst the two cohorts were divided to major and minor ones. Minor complications included superficial wound dehiscence and pseudomeningoceles managed conservatively. Major complications included CSF leaks, infections requiring antibiotics treatment, and pseudomeningoceles requiring surgical management. There were 14 (14/60, 23.33%) complications (major 12% and minor 12%) in the entire cohort. There was one superficial wound dehiscence in the PFD cohort (1/35, 2.86%), and six conservatively managed pseudomeningoceles in the PFDD cohort (6/35, 17.14%). There was one major (1/25, 4%) complication in the PFD group, which was an infection requiring antibiotics. In the PFDD cohort, there were 6 major complications (6/35, 17.14%), which included 2 CSF leaks, 1 infection treated with antibiotics, and 3 pseudomeningoceles requiring surgical management (Table 3). Major complication rate was significantly higher in the PFDD cohort (17.14% vs 4.0%, $p = 0.04$).

Failures

There were 6 failures (requiring repeat surgery) in this study, all of which were patients that had syringomyelia. Patients #1 and #3 also had Chiari headaches, and patient #2 had neurological symptoms as indications for second surgery in addition to syringes (Table 4). Of the failures, 5 of them received a PFD for index decompression. By comparison, there was one failure due to syringomyelia in the PFDD cohort resulting in significantly worse failure rate in the PFD cohort (20% vs 2.85%, $p = 0.03$). Following index decompressive surgery, patients #1, #2, and #3 initially showed improvement in their clinical symptoms, with stable syringes. However, after 12 months, 22 months, and 10 months, respectively, (mean 14.67 months), they displayed return of their clinical symptoms, and progression of their syringes. Patients #4 and #5 never saw improvement of their syringes following the index surgery, and thus underwent second decompression at 26 months, and 9 months after their initial operations. Patient #6 initially had radiologic improvement/stability of her holocord syrinx. However, 5 years after the initial operation, there was progression of the syrinx, eventually requiring a second operation. A duraplasty was performed at all repeat surgeries and a fourth ventricle stent was placed in 5 out of the 6 patients, due to significant adhesions at the site of CSF outflow. Patient #3 did not have a fourth ventricle stent placed at the second surgery and required a third surgery 1.7 years later for stent placement due to a progressive grade IV cervical syrinx, which subsequently remained stable. The placement of a fourth ventricular stent has been supported in cases of adhesions at the site of CSF outflow and considerable syrinx involvement. All six failure patients in this series had extensive syringes and significant adhesions at the site of CSF outflow, thus favoring the placement of a stent to achieve adequate surgical intervention. From our institution's 20-year experience and Boston case series of 100 Chiari decompressions, a fourth ventricular stent placement leads to favorable outcomes [12, 18]. Mean length of follow-up after the second surgery was 4.62 years (range 1.80 to 7.42 years) (Table 4). The mean time to clinical and radiologic improvement was 2.3 and 8.5 months, respectively.

Table 3 Comparison of major complications amongst PFD vs PFDD cohorts

Major complication	PFD ($n = 25$)	PFDD ($n = 35$)	p-value
Infection requiring antibiotics	1 (4.0%)	1 (2.86%)	-
CSF leak	-	2 (5.71%)	-
Pseudomeningocele requiring surgical management	-	3 (8.57%)	-
Total major complications	1/25 (4.0%)	6/35 (17.14%)	0.04*

PFD osseous-only posterior fossa decompression, *PFDD* posterior fossa decompression with duraplasty

*Statistical significance

Discussion

In this study, the long-term surgical outcomes in pediatric CM1 patients that underwent a PFD were analyzed and compared to those who underwent a PFDD to determine

Table 4 Failures that required second operation

Patient number	Sex/age (years)	Indications for index surgery	Index surgery	Time interval between surgeries (years)	Indications for second surgery	Second surgery	Postoperative complications	Length of follow up after second surgery (years)	Outcome
1	F/14	Chiari headache, neurological symptoms, grade IV holocord syrinx	PFDD	1.61	Chiari headache, neurological symptoms and progressive grade IV holocord syrinx	PFDD with fourth ventricular stent placement	Hydrocephalus requiring CSF diversion	3.94	Improved clinical symptoms and syrinx
2	M/12	Chiari headache, neurological symptoms, grade IV holocord syrinx	PFD	2.32	Neurological symptoms and progressive grade IV holocord syrinx	PFDD with fourth ventricular stent placement	None	3.61	Improved syrinx, stable neurological symptoms
3	M/3	Chiari headache, neurological symptoms, grade IV cervical syrinx	PFD	1.13	Chiari headache and progressive grade IV cervical syrinx	PFDD	None	7.42	Required a 3rd surgery with fourth ventricular stent 1.7 years later due to a progressive cervical syrinx which subsequently remained stable
4	M/11	Grade III holocord syrinx	PFD	2.20	Stable grade III holocord syrinx	PFDD with fourth ventricular stent placement	Pseudo-meningocele	5.93	Improved syrinx
5	M/7	Grade IV holocord syrinx	PFD	0.80	Stable grade IV holocord syrinx	PFDD with fourth ventricular stent placement	CSF leak	5.02	Improved syrinx
6	F/6	Grade IV holocord syrinx	PFD	7.0	Progressive grade IV holocord syrinx	PFDD with fourth ventricular stent placement	None	1.80	Improved syrinx

PFD osseous-only posterior fossa decompression, *PFDD* posterior fossa decompression with duraplasty

if there was any difference between cohorts in risk of failure resulting in reoperation. Defining "failure" in CM1 patients can be a difficult task given the variable presentation of patients. In this series, "failure" was defined as any patient that required a second surgery due to new onset symptoms, progressive symptoms, or significant residual syringomyelia.

The most common presenting symptom in this series was Chiari headache, which is consistent with previous findings in the literature [7, 11, 19]. For the entire cohort, clinical improvement was seen in a large majority of patients with Chiari headache, neurological symptoms, and syringes (Table 2), which is in concordance with previous findings that surgical management of CM1 leads to good clinical outcomes [7, 20].

In the literature, there is a trend over the past decade to utilize PFD for treating CM1 [9, 11, 12, 21]. This is due to similar clinical outcomes between PFD and PFDD [9, 15, 22]. However, there are studies that have demonstrated more favorable clinical outcomes with PFDD, particularly in patients with syringomyelia [14, 15]. In this series, there was similar clinical improvement in patients with respect to Chiari headache and neurological symptoms between PFD and PFDD cohorts (Table 2). In addition, PFD and PFDD had similar improvement rates when comparing non-holocord syringes ($p = 0.61$). However, there was statistically significant improvement of holocord syringes in the PFDD group compared to PFD ($p = 0.03$). The greater radiological syrinx improvement among PFDD patients may serve as an indicator of the adequacy of decompression as well as addressing any 4th ventricular outflow obstruction (i.e., adhesions). However, we did not detect a superiority of PFDD with respect to headache and neurological improvement or in other smaller syringes, which may suggest that such improvement requires less decompression than that needed to treat syringomyelia.

There are divergent reports on re-operation rates between PFD and PFDD, which has contributed to the ongoing debate in finding the superior approach. Durham et al. [22] reported significantly lower reoperation rates in the PFDD group. Similarly, Mutchnick et al. [23] reported a reoperation rate of 12.5% in the PFD group versus 3.1% in the PFDD group. However, there are other reports that have presented similar reoperation rates between the two approaches [14, 24]. In majority of these studies, the indications for reoperation rates were reported as persistent/progressive syringomyelia; however, a major limitation of such studies was the lack of differentiation between small and moderate syringes, compared to holocord and grade IV syringes as indications for reoperation [10, 13, 23, 25]. In our series, PFD was significantly associated with higher reoperation rates when compared to the PFDD approach in the setting of holocord and grade IV syringes as the indication for reoperation ($p = 0.03$). However, PFD and PFDD had similar improvement rates with respect to non-holocord syringes ($p = 0.61$), demonstrating that PFD is as effective as PFDD when treating small to moderate syringes. Our study is unique in that it investigated the nuances of indications for reoperation by differentiating between the degree and extensiveness of syringes in failures. Given the clinical and radiologic improvement in all failure patients following an intradural approach for their second surgery, it may be advisable to consider an intradural approach as the index surgery in patients with holocord syringes, in order to ensure a higher initial success rate.

Whether managed conservatively or surgically, studies have demonstrated the importance of long-term follow-up in CM1 patients, particularly in those with syringes [12, 26, 27]. In this series, the importance of long-term follow-up was highlighted, particularly in setting of failure patients. A total of 3/6 failure patients (patients #1, #2, #3) initially showed clinical and radiological improvement after the index surgery; however, they displayed clinical and radiological progression with mean of 14.67 months after the surgery. Patient #6 initially had stability of her holocord syrinx; however, progression was noted about 5 years later, eventually requiring a second decompression. These cases highlight the importance of long-term follow-up as there can be progression of clinical symptoms and radiologic findings years after the first operation. Thus, we recommend patients continuing long-term follow-up with periodic imaging, as patients may require a second decompression, even after initial improvement following the index surgery.

There are several potential mechanisms for failure. If the dura is not opened, there is a relative limitation to the amount of decompression that may be achieved, which in some cases may be insufficient to re-establish enough room for CSF flow to improve symptoms and syringomyelia [14]. Failure to open the dura may also fail to recognize the presence of subarachnoid adhesions obstructing CSF outflow from the 4th ventricle. Delayed failures can also result from re-ossification of the occipital bone or C1 lamina [7, 12]. In some cases, epidural scarring can recreate a constriction of the dura at the cervicomedullary junction that is effectively similar to the transverse fibrous bands that are opened during the initial decompression [7, 12]. When the dura is opened, failure may occur as a result of an inadequately sized duraplasty, unaddressed arachnoid bands, or new scarring and adhesions at the foramen of Magendie or obex [7, 12], especially after manipulation of the cerebellar tonsils.

PFD is associated with a lower rate of post-operative complications compared to PFDD [10, 13, 28]. In this study, the aggregate complication rate (major and minor complications) was 23.33%, similar to range of 0–33% in

previous reports [22, 23]. The PFDD group was associated with a significantly higher rate of major post-operative complications (Table 3). The most common major complication in the PFDD group was a pseudomeningocele. Although not statistically analyzed in this study, other factors including operative time and hospital length of stay may differ drastically between the two surgical approaches. Studies have reported longer operation time and hospital length of stay in PFDD group compared to PFD patients [9, 13]. Our own institution's 17-year experience of surgically treating 125 PFDD patients and 70 PFD patients demonstrated a significantly longer hospital length of stay in the PFDD group (4.6 vs 2.9 days, $p < 0.01$) [12]. Such findings underscore a well-demonstrated increased risk of complications and costs in PFDD patients, which may be avoided by using an extradural approach with reported similar clinical outcomes [11, 12, 15, 22].

Study limitations

As a single-center evaluation, there were only 60 patients included after 221 patients were excluded due to a follow-up length of less than 5 years, thus reducing the statistical power of the study. Although a statistically significant relationship was not detected between the degree of tonsillar ectopia and surgical failure rate ($p = 0.86$), this may have been due to the small sample size, as this relationship has been previously demonstrated in larger studies [29]. The demographic variables included age, sex, presenting signs and symptoms, and length of follow-up. Possible confounding variables such as concurrent Ehlers-Danlos syndrome and craniocervical anomalies were not controlled. In addition, there was a non-random assignment of patients to each surgical approach, which was performed at the discretion of the individual surgeon. It is possible that some patients had additional follow-up and surgery elsewhere; however, there is no way for us to know if any of the patients lost to follow-up ultimately underwent a second Chiari decompression for a surgical failure. Lastly, clinical improvement of Chiari headache and neurological symptoms was subjectively reported by patients and tracked simply as better, stable, or worse. There was a lack of validated clinical assessment, which would have provided more of an objective evaluation of clinical outcomes in our patients [9, 11, 13].

Conclusion

Short-term analysis revealed lower post-operative complication rates in PFD compared to PFDD. Long-term analysis showed the PFD approach provides a safer treatment option with similar neurological and radiological improvements to PFDD, including patients with non-holocord syringomyelia. However, surgical failure can be minimized by utilizing a PFDD in patients with a holocord syrinx due the significantly higher rate of reoperation for persistent syringomyelia in patients who initially underwent a PFD.

Funding The authors received financial support from the American Syringomyelia and Chiari Alliance Project (ASAP) for the above clinical research at Children's National Hospital.

Availability of data and material The datasets generated and analyzed during the current study are not publicly available due to Health Insurance Portability and Accountability Act (HIPAA), but are available from corresponding author on reasonable request.

Declarations

Ethics approval This prospective and retrospective chart review study involving human participants was in accordance with the ethical standards of the institutional and national research committee and with the 1964 Helsinki Declaration and its later amendments or comparable ethical standards. The Institutional review board approved this study (Pro00013560).

Consent to participate Informed consent was obtained from legal guardians

Consent for publication Informed consent was obtained from legal guardians regarding publishing patients' data and photographs.

Conflict of interest The authors declare no competing interests.

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