Management of hydrocephalus and subdural hygromas in pediatric patients after decompression of Chiari malformation type I: case series and review of the literature

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OBJECTIVE Hydrocephalus associated with subdural hygromas is a rare complication after decompression of Chiari malformation type I (CM-I). There is no consensus for management of this complication. The authors present a series of 5 pediatric patients who underwent CM-I decompression with placement of a dural graft complicated by posterior fossa hygromas and hydrocephalus that were successfully managed nonoperatively.

METHODS A retrospective review over the last 5 years of patients who presented with hydrocephalus and subdural hygromas following foramen magnum decompression with placement of a dural graft for CM-I was conducted at 2 pediatric institutions. Their preoperative presentation, perioperative hospital course, and postoperative re-presentation are discussed with attention to their treatment regimen and ultimate outcome. In addition to reporting these cases, the authors discuss all similar cases found in their literature review.

RESULTS Over the last 5 years, the authors have encountered 194 pediatric cases of CM-I decompression with duraplasty equally distributed at the 2 institutions. Of those cases, 5 pediatric patients with a delayed postoperative complication involving hydrocephalus and subdural hygromas were identified. The 5 patients were managed nonoperatively with acetazolamide and high-dose dexamethasone; dosages of both drugs were adjusted to the age and weight of each patient. All patients were symptom free at follow-up and exhibited resolution of their pathology on imaging. Thirteen similar pediatric cases and 17 adult cases were identified in the literature review. Most reported cases were treated with CSF diversion or reoperation. There were a total of 4 cases previously reported with successful nonoperative management. Of these cases, only 1 case was reported in the pediatric population.

CONCLUSIONS De novo hydrocephalus, in association with subdural hygromas following CM-I decompression, is rare. This presentation suggests that these complications after posterior fossa decompression with duraplasty can be treated with nonoperative medical management, therefore obviating the need for CSF diversion or reoperation.

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KEYWORDS Chiari I malformation; complication; hydrocephalus; posterior fossa decompression; subdural hygromas

DESCENT of the cerebellar tonsils greater than 5 mm below the level of the foramen magnum, known as Chiari malformation type I (CM-I), is a well-recognized entity in both adult and pediatric patients.^{17,22} It is generally accepted that suboccipital craniectomy with a C1 laminectomy, with or without dural expansion, is the

standard of care for patients with symptomatic primary CM-I.^{1,8,10,14,30} This is especially true with an associated syrinx, which may serve as an indicator of abnormal CSF dynamics.^{2,18} While techniques for the decompression may vary from surgeon to surgeon, the creation of space at the cervicomedullary junction, with improvement of CSF flow

ABBREVIATIONS CM-I = Chiari malformation type I; ETV = endoscopic third ventriculostomy; FMD = foramen magnum decompression.

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obstruction and brainstem compression, is the accepted general goal of surgery. Common complications following CM-I decompression include CSF leakage, pseudomeningocele, meningitis (bacterial or chemical), postoperative hemorrhage, and cerebellar ptosis or slump.¹² Less common complications include vascular injury and stroke.²⁰ Subdural hygromas and postoperative hydrocephalus after posterior fossa decompression are rare but recognized complications without a consensus for management.^{4–6,9,11,19,23,24,26,27,31} A variety of treatments have been described, including temporary CSF diversion, permanent CSF diversion, endoscopic third ventriculostomy (ETV), reoperation to widen the arachnoidal opening, and evacuation of subdural hygromas.^{24,27,31} There are rare reports of good outcomes after nonoperative management.^{9,11,19} We present our experience with 5 pediatric patients at 2 institutions and discuss nonoperative strategies for treatment. Additionally, we present a review of pediatric and adult cases of CM-I complicated by hydrocephalus with subdural hygromas that have been reported in the literature and discuss the possible pathogenesis and treatment of this rare complication.

Methods

At both pediatric institutions, a retrospective review of the last 5 years was done in order to identify operative cases of CM-I decompression with duraplasty. Data were obtained identifying patients who presented with hydrocephalus and subdural hygromas. For the purposes of this study, we defined CM-I as tonsillar herniation greater than 5 mm below the foramen magnum with presenting symptoms attributable to the malformation. Five pediatric patients (age 1.5–17 years, average 8.9 years) were identified from the database. IRB approval was obtained for purposes of this review. The patients were those who underwent foramen magnum decompression (FMD) for CM-I, and whose postoperative courses were complicated by delayed postoperative subdural fluid collections (either cerebellar or cerebral) and evidence of intracranial hypertension and enlarged ventricles. Preoperative, operative, and postoperative records for these cases were reviewed. Asymptomatic patients and patients with CM-I attributable to other causes, such as mass lesion, hydrocephalus, spinal cord tethering, and other secondary type I Chiari malformations (e.g., from lumboperitoneal shunting or intracranial hypotension), were excluded. Table 1 contains the summary for our patients' data.

Operative Technique

Our cohort of pediatric patients underwent surgery performed by 4 pediatric neurosurgeons at 2 pediatric neurosurgical centers. The operations were performed with the patient in the prone position. A midline cervical dissection was used to expose the occiput and high cervical spine. Suboccipital craniectomy with C1 laminectomy was performed to decompress the foramen magnum. The dura and arachnoid mater were then opened. In 4 of our patients, the cerebellar tonsils were cauterized to shrink and elevate them. In all cases, a thorough arachnoidolysis was done, with specific attention to remove any veil or tissue webs

that could potentially obstruct the CSF pathway around the outlet of the fourth ventricle. Careful arachnoidolysis was also done laterally toward the gutters around the tonsils in order to release any potential arachnoid pockets. An autograft dural graft (pericranium) was used in 3 cases, whereas allograft dural graft was used in the remaining 2 cases. The graft was sutured into place in order to enlarge the intradural diameter at the craniovertebral junction (CVJ) and prevent further compression of the intradural contents at the CVJ. The patched dura and subsequently the overlying fascia were both closed in a watertight fashion. Operative goals and immediate postoperative results were equivalent despite minor technical differences due to surgeon preferences. Intraoperative ultrasound was not routinely used.

Literature Review

MEDLINE and PubMed searches were performed using the keywords “Chiari malformation,” “foramen magnum decompression,” “hydrocephalus,” “subdural hematoma,” “subdural hygroma,” “cerebellar ptosis,” “cerebellar sag,” and “cerebellar slump.” All articles discussing postoperative intracranial hypertension with subdural fluid collections (either cerebellar or cerebral) as a complication following posterior fossa decompression, along with respondent commentaries regarding these articles, were included. We extracted pediatric case data from the published case reports and case series for comparison and analysis. Adult data are presented for a thorough and complete analysis but are not considered in our analysis, as this report focuses on a pediatric population.

Case Presentations

Table 1 contains a summary of our patients' data.

Case 1

This 14-year-old girl with CM-I and syringomyelia presented with a 3-month history of pain and numbness moving up the left upper limb to the left back, chest, and neck in a hemicape-like distribution. Neurological examination revealed hypesthesia to temperature and pin sensation in the left upper extremity down to approximately the T4 level (slightly beyond midline) as well as to the posterior neck and head. All other examination findings were normal. MRI revealed a CM-I with cerebellar tonsils terminating 6 mm below the foramen magnum, as well as a large cervicothoracic syrinx extending from C1 to T8 and deviating eccentrically to the left side. Of note, no hydrocephalus or ventriculomegaly was evident at admission (Fig. 1A).

The patient underwent a standard posterior fossa decompression as previously described. The arachnoid was widely opened, and the obex was visualized without any signs of obstructing tissue or web. The tonsils were not cauterized. Blood loss was estimated to be 75 ml. An allograft dural graft was utilized for the duraplasty, which was closed in a watertight fashion. The patient was discharged home in good condition after an uneventful 3-day postoperative course. Fourteen days after surgery, the patient developed headache, malaise, nausea, vomit-

TABLE 1. Summary of our patients

Case No.	Age, Sex*	Presenting Sx	Tonsillar Herniation (mm), Syrinx?	Surgical Technique	Time Btwn Op & Re-Presentation (days)	Postop Sx	Postop Radiographic Findings		Treatment	Outcome
1	14, F	Pain & numbness of upper extremity	6, syrinx	SOC, DP, AO	14	HA, malaise, N, V, & dizziness	Ventriculomegaly, bilat IT hygromas, pseudomeningocele, decrease in syrinx size		Acetazolamide, steroids	Gradual improvement w/ complete resolution in 5 mos
2	4, M	HAS	15, no syrinx	SOC, DP, AO, TC	14	HA, altered mental status	Ventriculomegaly, IT hygromas		Acetazolamide, steroids, head-down position	Gradual improvement, complete resolution in 30 days
3	15 mos, F	Swallowing, sleep apnea, frequent irritability, & episodes of discomfort	6, syrinx	SOC, DP, AO, TC	32†	Fussiness, vomiting	Slight ventriculomegaly, ST & IT hygromas		Acetazolamide, steroids, head-down position	Gradually improved, resolution in 52 days
4	17, M	Occipital/nuchal pain	23, syrinx	SOC, DP, AO, TC	10	Episodes of blurred vision	Ventriculomegaly, ST & IT hygromas		Acetazolamide, steroids	Gradually improved, complete resolution after 4 wks
5	8, F	HAS, itching of the hands & feet, sleep apnea	17, syrinx	SOC, DP, AO, TC	14	Activity-induced headaches, neck pain, itching, coughing, & a change in voice	IT hygromas		Acetazolamide, steroids	ASx at 3 mos

AO = arachnoid opening; ASx = asymptomatic; DP = duraplasty; HA = headache; IT = infratentorial; N = nausea; SOC = suboccipital craniectomy; ST = supratentorial; Sx = symptoms; TC = tonsil cauterization; V = vomiting.

* Age is in years unless stated otherwise.

† This patient was seen 14 days after surgery at an outside facility, but only a CT scan was done. She was discharged home on steroid treatment for 12 days.

FIG. 1. Case 1. Axial, sagittal, and coronal T2-weighted MRI scans obtained pre-FMD (A), and 14 days (B), 21 days (C), and 5 months (D) post-FMD, showing the progressive appearance and decrease of bilateral infratentorial subdural fluid collections in addition to normalization of hydrocephalus.

ing, and dizziness on cervical rotation. She presented to a local emergency department a day after onset of symptoms where a lumbar puncture was performed to rule out meningitis. CSF results were without signs for suspected infectious process, and results of the final culture were negative. Her symptoms improved after the lumbar puncture. The patient was transferred to our facility for further evaluation. On examination, there was no evidence of CSF leak, wound discharge, or papilledema. Her neurological examination findings were normal aside from residual left upper extremity hypesthesia. MRI of the brain revealed ventriculomegaly consistent with hydrocephalus, bilateral infratentorial extraaxial fluid collections, and pseudomeningocele (Fig. 1B). There appeared to be a slight decrease in the size of the syrinx. A conservative treatment regimen (high-dose dexamethasone for 2 weeks, acetazolamide for 1 month) was initiated, with gradual alleviation of the patient's symptoms. She was monitored in the hospital for 7 days with repeat imaging that demonstrated mild improvement (Fig. 1C). Complete resolution of ventriculomegaly was noted 5 months later, with continued improvement of left posterior head numbness and residual syrinx (Fig. 1D).

Case 2

This 4-year-old boy was complaining at presentation of headaches several times a week for several months. His caregivers described him holding his neck and the back of his head complaining of pain. He also had episodes of irritability and temper outbursts but at other times would be quite agreeable and complacent. Staring events and stiffening of limbs during sleep were reported, yet 48-hour electroencephalography demonstrated no epileptic activity. He also had mild speech delay.

MRI was performed, which demonstrated a CM-I with 15 mm of tonsillar herniation below the foramen magnum and an impacted foramen magnum. There was no

evidence of ventriculomegaly or syrinx on preoperative imaging (Fig. 2A).

The patient underwent posterior fossa decompression with a laminectomy of C1. The fourth ventricle and the region of the obex were inspected, and extensive arachnoidolysis was performed. There was no scarring or arachnoidal veil visible. The tonsils were very large, extending to C2, and they were cauterized. Blood loss was minimal and estimated around 15 ml. A pericranial graft was utilized for the duraplasty, which was closed in a watertight fashion. The muscle, fascia, and subcutaneous tissues were also tightly closed. The patient did well postoperatively and returned home 3 days after surgery. Fourteen days after surgery, he presented to a local hospital with a complaint of severe headache that woke him from sleep in the early morning hours. He had headaches for a few days before presentation, but they were not severe enough for his family to seek medical attention. The patient was then transferred to our facility. He was sleepy and complained of severe headache (9 of 10 on a numeric rating scale). His symptoms were worse while in a sitting position. He had no signs of infection. Of note, he had sustained a minor fall from his bike the day before admission. CT scans obtained at the local facility after the fall showed ventriculomegaly of the supratentorial ventricles and a normal-sized fourth ventricle as well as posterior fossa subdural hygromas. MRI performed at our institution better delineated these findings (Fig. 2B). The ophthalmology examination results were normal, without signs of papilledema.

The child was then placed in a head-down position and experienced immediate relief of his symptoms. He was started on acetazolamide and high-dose dexamethasone. He was able to sit up for periods of time on day 3 of treatment, with a significant decrease in headache symptoms. MRI done on day 2 after admission showed a mild decrease in ventricular size, as well as a decrease in the posterior fossa subdural fluid. By day 6 the patient was

FIG. 2. Case 2. Sagittal, coronal, and axial T1- and T2-weighted MRI scans obtained pre-FMD (A), and 14 days (B), 21 days (C), and 1.5 months (D) post-FMD, showing the progressive appearance and decrease of bilateral infratentorial subdural fluid collections in addition to normalization of hydrocephalus.

ambulating and playing with no headaches. MRI done on day 7 showed a significant decrease in ventricular size and the subdural hygromas (Fig. 2C). A dexamethasone wean was started on day 6 and completed over a week. Acetazolamide continued for a total of 4 weeks with a gradual wean starting week 2. The patient and his imaging continued to improve, and he returned to baseline 30 days after his presentation (Fig. 2D). His preoperative symptoms resolved, and his family felt he was more active.

Case 3

This 15-month-old girl presented with problems swallowing, sleep apnea, and frequent irritability and episodes of discomfort. MRI was done and showed a significant CM-I with about 6 mm of tonsillar descent below the foramen magnum. Preoperative MRI showed no signs of ventriculomegaly or syrinx (Fig. 3A). She underwent a posterior fossa decompression with laminectomy of C1, extensive arachnoidolysis, and cauterization of the cerebellar tonsils. Inspection of the obex confirmed no arachnoid veil or tissue obstructing the CSF flow from the fourth ventricle. A pericranial dural graft was utilized in order to close the dural opening in a watertight fashion. Blood loss was 10 ml. The patient did well postoperatively and was discharged home on postoperative day 2. Fourteen days after surgery, she was admitted to a regional facility. She presented with fussiness and had 1 episode of vomiting. Otherwise, she had no other complaints. The patient was

started on dexamethasone for 12 days, and 4 days after weaning off the dexamethasone she was admitted again to our emergency department with complaints of irritability for several days, and again with 1 episode of emesis. There were no signs of infection or other viral illness. On examination, she had no focal neurological deficit and no sign of a pseudomeningocele.

The patient's MRI scan (32 days after surgery) showed extraaxial fluid collections around both cerebellar and cerebral convexities and along the interhemispheric fissure (Fig. 3B). There was a slight increase in the ventricle size. In comparison to her CT scan from 2 weeks prior, as reported by the other facility, MRI demonstrated an increase in the size of the extraaxial fluid supratentorially and infratentorially. She was started on acetazolamide, intravenous fluids, and high-dose dexamethasone and was positioned in bed with her head down. Three days after admission she felt much better and started to ambulate and play. On day 6 after presentation the repeat MRI showed almost complete resolution of the extraaxial fluid around the cerebrum and cerebellum (Fig. 3C). She clinically felt much better and was discharged home. When seen in follow-up her scanning results had returned to normal (Fig. 3D).

Case 4

This 17-year-old boy with developmental delay and known scoliosis was diagnosed with holocord syringomy-

FIG. 3. Case 3. Sagittal, coronal, and axial T1- and T2-weighted MRI scans obtained pre-FMD (A), and 32 days (B), 38 days (C), and 52 days (D) post-FMD, showing the progressive appearance and decrease of bilateral infratentorial and supratentorial subdural fluid collections with gradual normalization.

elia and 23 mm of tonsillar ectopia while being assessed for his scoliosis with spinal imaging (Fig. 4A). On preoperative MRI there was no evidence of ventriculomegaly. A decision was made to take the patient to surgery for a posterior fossa decompression, laminectomy of C1, and duraplasty. Due to the low position of the tonsils, their fairly large size, and the anatomy at the foramen magnum, both cerebellar tonsils were cauterized. Thorough arachnoidolysis was done between the tonsils and lateral to the gutters around them. An inspection was done, revealing there was neither an arachnoid veil nor tissue obstructing the outlet of the fourth ventricle. Blood loss was 75 ml. A pericranial dural graft was utilized and installed in a watertight fashion.

Postoperatively the patient was treated with dexamethasone for 5 days. He experienced a few episodes of low oxygen saturation, which required administration of oxygen. No reason was found for the desaturation episodes. The patient did well and was discharged home 3 days after surgery. Ten days after surgery he presented to the emergency department with 2 episodes of binocular vision loss as reported by ophthalmology, without loss of consciousness, lethargy, headaches, or vomiting. An MRI of the brain demonstrated cerebellar hygromas and a slight increase

in ventricle size with the formation of supratentorial and infratentorial hygromas as well (Fig. 4B). There was no apparent change in the position of the cerebellar tonsils. The patient had no evidence of papilledema on ophthalmological examination. He was started on acetazolamide and high-dose dexamethasone. Two days after admission and medical treatment, his episodes of blurred vision stopped, and his MRI findings improved with a minimal decrease in ventriculomegaly and a partial resolution of the subdural hygromas (Fig. 4C). The patient was discharged home on the medications and plans for further ambulatory follow-up. He was seen 2 weeks after discharge and had no new clinical symptoms. His imaging showed continued resolution of the hydrocephalus as well as complete resolution of the hygromas (Fig. 4D). He resumed his regular activities and was weaned from both dexamethasone and acetazolamide over the next 2 weeks.

Case 5

This 8-year-old girl had a known history of CM-I and syringomyelia of C4–T4. The patient had tonsillar decent of 17 mm below the foramen magnum. She had undergone 2 surgeries for foramen magnum decompression 5 years and 2 years prior to the current surgery. Her initial

FIG. 4. Case 4. Sagittal, coronal, and axial T1- and T2-weighted MRI scans obtained pre-FMD (A), and 10 days (B), 12 days (C), and 28 days (D) post-FMD, showing the progressive appearance and decrease of bilateral infratentorial subdural fluid collections in addition to normalization of hydrocephalus.

presentation 5 years earlier was activity-induced occipital headaches and progressive itching of her hands and feet. MRI demonstrated the CM-I with an associated syrinx, and without evidence of ventriculomegaly. Foramen magnum decompression with duraplasty was performed. After the first surgery, she had resolution of her symptoms and remained asymptomatic for 3 years. She again developed activity-induced headaches, neck pain exacerbated by movement, itching in her legs, intermittent coughing, and a change in her voice. The patient underwent a second foramen magnum decompression surgery but with a smaller duraplasty this time. She was well for 2 years after that surgery when she again presented with similar symptoms, but this time with an additional diagnosis of central sleep

apnea. She underwent a posterior fossa decompression, opening of scar tissue, extensive arachnoidolysis, cauterization of the tonsils, and a duraplasty using an allograft dural graft. Inspection of the obex revealed that there was no arachnoid veil or tissue obstructing the CSF pathways. Blood loss was estimated at 25 ml. After surgery, the patient initially did well but then developed a pseudomeningocele after severe coughing, which occurred 3 months after the decompression surgery. She presented with slightly enlarged ventricles (Fig. 5A) and eventually required exploratory surgery to repair the pseudomeningocele 5 months after the operation. Fourteen days after this surgery, the patient presented with headaches and an MRI demonstrated infratentorial hygromas and associated

FIG. 5. Case 5. Sagittal, coronal, and axial T1- and T2-weighted MRI scans obtained post-FMD in the past and pre-fixation of pseudomeningocele (A), and 14 days after surgery with the formation of intratentorial hygromas (B), and 3 months after surgery with partial resolution in ventricular size and partial decrease of bilateral infratentorial subdural fluid collections (C).

mass effect (Fig. 5B). She was treated for several weeks with high-dose dexamethasone and a 2-week course of acetazolamide. She had slight resolution of the hygromas and significant improvement in her symptoms after 3 months (Fig. 5C).

Literature Review

The results of the literature search are shown in Tables 2 and 3. Table 2 includes all 13 published pediatric cases from 2001 to 2014. Zakaria et al. published the largest series of patients, which included 4 pediatric patients and

TABLE 2. Summary of published pediatric cases

Authors & Year	Age (yrs), Sex	Presenting Sx	Tonsillar Herniation (mm), Syrinx?	Surgical Technique	Time Btwn Op & Re-Presentation (days)	Postop Sx	Postop Radiographic Findings	Treatment	Outcome
Bosma et al., 2002	8, F	Scoliosis, diplopia, HA	Unk, syrinx	SOC, unk	7	HA	HCP, ST hygroma	EVD, VPS	VPS, ASx at 16 mos
Elton et al., 2002	10, M	Scoliosis, HA	Unk, no syrinx	SOC, DP, unk	10	HA, V, bradycardia	HCP, IT hygroma, pseudomeningocele	EVD x2	ASx at 6 mos
	14, F	Colitis, depression, HA	Unk, syrinx	SOC, DP, unk	5	HA, N, ataxia	HCP, IT hygroma	EVD	ASx at 6 mos
	14, F	Nocturnal interscapular pain	Unk, syrinx	SOC, DP, unk	25	HA, N, V	HCP, IT hygroma, pseudomeningocele	EVD	Delayed LPS for subsequently diagnosed PTC, ASx at 3 mos
	17, F	NF1, HA	Unk, syrinx	SOC, DP, AO	6	HA, V, syncope	HCP, ST & IT hygroma	EVD, VPS	VPS, ASx at 3 mos
Marshman et al., 2005	2, F	Cyclical vomiting, HA	Unk, syrinx	SOC, durotomy w/o DP	10	HA, N, V	HCP, ST & IT hygroma	Acetazolamide	ASx at 1 mo
Bahuleyan et al., 2011	18, F	Unk	Unk, syrinx	SOC, DP, arachnoidotomy	7	HA, V, papilledema, bradycardia, decerebrate posturing	ST & IT hygroma	VPS	VPS, ASx at 6 mos
Suzuki et al., 2011	15, F	Neck & shoulder pain	Unk, syrinx	SOC, DP, arachnoidotomy	11	HA	HCP, ST & IT hygroma	Reop w/ resection of arachnoid	ASx at 24 mos
Zakaria et al., 2012	1, M	Unk	Unk, no syrinx	SOC, DP, AO	9	CSF leak	HCP	EVD, VPS	VPS, ASx at 12 mos
	2, M	Unk	Unk, no syrinx	SOC, DP, AO	8	CSF leak	HCP, hygroma	EVD	ASx at 13 mos
	7, M	Unk	Unk, syrinx	SOC, DP, AO	8	CSF leak	None	EVD, VPS	ASx at 39 mos
	9, M	Unk	Unk, syrinx	SOC, DP, AO	42	HA, V	HCP	Evacuation of hygroma	ASx at 15 mos
Pereira et al., 2014	10, F	Occipital/nuchal pain	Unk, syrinx	SOC, DP, arachnoidotomy	6	HA, N, V, CSF leak, papilledema, CN VI palsy (over a period of 3 hospital admissions)	IT hygromas	Evacuation of hygroma x2, wound exploration & closure, acetazolamide, LP, lumbar drain	ASx at 3 mos

CN = cranial nerve; EVD = external ventricular drain; HCP = hydrocephalus; LP = lumbar puncture; LPS = lumboperitoneal shunt; NF1 = neurofibromatosis type 1; PTC = pseudotumor cerebri; unk = unknown; VPS = ventriculoperitoneal shunt.

TABLE 3. Summary of published adult patients

Authors & Year	Age (yrs), Sex	Tonsillar Herniation (mm), Syrinx?	Presenting Sx	Surgical Technique	Time Btwn Op & Re-Presentation (days)	Postop Sx	Postop Radiographic Findings	Treatment	Outcome
Ranjan & Cast, 1996	55, F	Unk, no syrinx	RLE weakness, numbness	SOC, DP, arachnoidotomy	14	HA, V, drowsiness	HCP, ST & IT hygroma	Conservative	ASx at 3 mos
Marshman et al., 2005	59, F	Unk, no syrinx	HA	SOC, DP, arachnoidotomy	5	HA, N, V	HCP, IT hygroma	Conservative	ASx at 3 mos
Perrini et al., 2008	37, M	Unk, syrinx	Klippel-Feil, HA, BUE dysesthesias	SOC, arachnoid opened, no dural closure	20	Dizziness, RUE weakness	ST & IT hygroma	Burr hole evacuation of hygroma, VPS	VPS, baseline at 4 mos
Bahuleyan et al., 2011	38, M	Unk, unk	Unk	SOC, DP, arachnoidotomy	15	HA, N, V, vertigo, papilledema	ST & IT hygroma	Evacuation of hygroma	ASx at 8 mos
	19, M	Unk, no syrinx	Unk	SOC, DP, AO	42	HA, V	HCP	VPS	Persistent HA at 6 mos, pending valve change
	31, M	Unk, no syrinx	Unk	SOC, DP, AO	8	CSF leak	None	EVD, VPS	Awaiting OC fusion
	33, F	Unk, syrinx	Unk	SOC, DP, AO	15	CSF leak	None	LPS, VPS	ASx at 42 mos, valve changed due to low-pressure HA
Zakaria et al., 2012	35, F	Unk, syrinx	Unk	SOC, DP, AO	11	HA, V	HCP, hygroma	EVD, ETV, VPS	ASx at 27 mos
	41, F	Unk, syrinx	Unk	SOC, DP, AO	5	HA, CSF leak	None	VPS	ASx at 64 mos
	42, F	Unk, no syrinx	Unk	SOC, DP	30	HA, papilledema	HCP, hygroma	Evacuation of hygroma, VPS	Persistent HA at 51 mos, normal ICP
	46, F	Unk, syrinx	Unk	SOC, DP, AO	210	HA, ataxia	HCP	VPS	ASx at 63 mos
	61, F	Unk, no syrinx	Unk	SOC, DP, AO	31	HA	HCP, hygroma	Conservative	ASx at 36 mos
	44, F	Unk, unk	HA, unsteadiness, falls	SOC, DP, no open arachnoid, dura sutured to muscle	21	HA, unsteadiness, falls	Hygroma, posterior fossa mass effect	Reopening extending arachnoid opening	ASx at 12 mos
Bahl et al., 2011	41, F	5-6, none	HA, shooting pain lt arm, lt arm weakness	SOC, DP, no open arachnoid, dura sutured to muscle	9	HA, V, gait ataxia	HCP, hygroma	Reopening extending arachnoid opening	ASx at 18 mos
	27, M	Unk, syrinx	Lancinating pain to rt arm w/ numbness	SOC, DP, no open arachnoid, dura sutured to muscle	12	Severe HA	HCP, hygroma	Reopening extending arachnoid opening	At 36 mos still some sensory deficit in the rt arm, syrinx improved considerably
Thakar et al., 2014	21, M	Unk, syrinx	Cough-induced HA & paresis in all limbs	SOC, DP (technique is not fully described)	7	HA, tightness in all limbs, difficulty in swallowing	HCP, hygroma	1st treated conservatively w/ acetazolamide w/o significant improvement & then treated w/ VPS	At 6 mos still w/ preop Sx but w/ much improvement
Munakomi et al., 2016	30, F	Unk, syrinx	Chronic dull neck pain, cape-like dissociative sensory loss C2-T4	SOC, DP (technique is not fully described)	21	HA, dizziness	HCP, hygroma	1st treated conservatively w/ acetazolamide on day 6 when acute HCP developed, treated w/ VPS	At 24 mos remnants of sensory deficits w/ much improvement

BUE = bilateral upper extremities; ICP = intracranial pressure; OC = occipital cervical; RLE = right lower extremity.

8 adult patients.³¹ The largest group of pediatric patients examined at one institution was by Elton et al., who reviewed 200 consecutive pediatric patients undergoing posterior fossa decompression and as such were able to publish information on the incidence of this complication.⁹ The age ranged from 1 to 18 years with a mean of 9.7 years. We were unable to determine the average length of tonsillar herniation based on published data, since different publications did not include the specific radiological description of every patient. Almost all pediatric patients in the published literature underwent suboccipital craniectomy and C1 laminectomy in combination with duraplasty and arachnoidal opening. On average, most of the patients presented again 11.8 days after surgery with a median of 8 days (range 5–42 days). In our series, patient age ranged from 15 months to 17 years with a mean of 8.8 years. The mean and median times to presentation were 13.25 and 14 days after surgery, respectively (range 10–32 days). Most patients reported on in the literature presented with headache, nausea, and vomiting, although 5 patients presented with more concerning symptoms of bradycardia or cranial neuropathy.⁵ One of these patients exhibited decerebrate posturing. While nonoperative management has been described by Filis et al.,¹¹ the other 12 patients were managed by CSF diversion (either temporarily via EVD or permanently via shunting) or operative reexploration. In the literature, the average length of follow-up was 11.3 months, with all patients being asymptomatic at last follow-up.

Discussion

Symptomatic primary CM-I is typically treated by posterior fossa decompression with the addition of a C1 laminectomy.^{4,5,10,17} There is much debate regarding the surgery for CM-I decompression, such as whether to perform bony decompression alone or to include additional steps, such as duraplasty, opening the arachnoid, or tonsillar cauterization.^{2,8,10,18,27} Delayed postoperative subdural hygromas and acute hydrocephalus after CM-I decompression are rare complications but have been previously discussed in the literature. Most of the known literature focuses on the adult population. In the largest published series of pediatric CM-I decompressions, by Tubbs et al., 4 of 500 patients (0.8%) developed this complication.²⁹ Despite its rarity, most high-volume pediatric neurosurgical centers will likely encounter this complication, given the prevalence of symptomatic CM-I in the pediatric population.

Various mechanisms have been proposed to explain the pathophysiology of this complication. Some authors have theorized that small openings in the arachnoid at the time of surgery (either purposefully or incidentally) result in the formation of a 1-way slit-valve mechanism that allows CSF egress from the subarachnoid space into the subdural space.^{9,26} Some have suggested that the accumulated subdural fluid around the foramen magnum can potentially migrate with brain pulsation to the cisterna magna, which then compresses the cerebellar hemisphere either down toward the foramen magnum or anteriorly, compressing the aqueduct and fourth ventricle.⁵ Closure of the aqueduct (so-called “torqueing of the aqueduct”) or closure of the foramina of Luschka and Magendie potentially can lead to

obstructive hydrocephalus either above or below the fourth ventricle, with resulting clinical sequelae. Another potential explanation is related to cerebellar slump or sag, which is caused by the downward migration of the hindbrain after decompression of the foramen magnum. This descent of the cerebellum downward toward the foramen magnum in an already small posterior fossa in the pediatric population can potentially cause the partial obstruction of CSF pathways around the foramen magnum.^{3,7,15,16,25} In reviewing our cases, neither our radiologists nor we recognized any findings such as inflammation, the presence of hemorrhage, fourth ventricle obstruction, or diffuse edema that could potentially explain the findings of hydrocephalus or subdural hygromas.

In 2016, Guan and colleagues published an assessment of clinical risk factors for Chiari-related hydrocephalus in a cohort of 297 consecutive patients.¹³ They concluded that age younger than 6 years, a large amount of intraoperative blood loss, and the presence of fourth ventricular outlet obstruction are all associated with a greater chance for requiring long-term CSF diversion. In our cases, blood loss was minimal for those younger than 12 years and 75 ml or less for older children. As mentioned in the description of the cases, in each case a thorough inspection for an arachnoid veil or web was done, and extensive arachnoidolysis was performed in order to prevent potential arachnoidal pockets or webs from obstructing the CSF pathways.

To date, there is no consensus regarding the best method to treat postoperative hydrocephalus associated with CM-I decompression. Several publications, describing either adult or pediatric patients, stress that an effort should be made to prevent this complication by both avoiding durotomy and arachnoidolysis whenever possible or, conversely, opening the arachnoid widely. Some authors have even suggested reoperation for closing the tiny hole in the arachnoid or opening the arachnoid widely and suturing it to the dura.^{9,26,31} However, in all of our cases, a wide arachnoidal opening was achieved but hydrocephalus was still observed, which leads to the possible conclusion that there are other alterations in CSF dynamics that may result in this pathology.

Currently, possible treatment options for post-CM-I decompression-associated hydrocephalus include conservative treatment, ventriculostomy, shunting, lumbar puncture, ETV, wound exploration for further decompression, and evacuation of hygromas. The overall prognosis, regardless of treatment modality, is excellent, with all reported patients achieving resolution of symptoms and radiographic abnormalities by 6 months (Tables 1–3). The vast majority of pediatric patients reported with this complication were treated via temporary or permanent CSF diversion (9 of 13, or 69%).⁹ Of the 4 patients managed without CSF diversion, 2 were managed by reoperation to widen the arachnoidal opening, and 1 was managed by evacuation of the subdural hygromas. Only 1 patient was managed nonoperatively. Among adult patients, most were treated with the use of CSF diversion (10 of 17, 58.9%). Five patients were treated initially nonoperatively and 2 of them eventually did not improve and required CSF diversion (Table 3).

In review of our data, we identified an incidence of

2.6% (5/194 patients) for this condition. We add 5 pediatric cases to the literature that were managed nonoperatively and suggest that in patients without a precipitous decline in neurological function, a trial of observation and medical therapy is reasonable. All 6 pediatric patients managed nonoperatively (5 from our series and 1 reported by Filis et al.)¹¹ had excellent long-term outcomes and did not ultimately require CSF diversion. While this sample set is small, it appears clear that the postoperative hydrocephalus experienced by these patients is distinct from intrinsic increased intracranial hypertension that must be ruled out in the setting of CM-I prior to decompression (i.e., secondary Chiari malformation). We believe that the hydrocephalus is likely a result of altered CSF pathways by mass effect or inflammation around the surgical bed causing transient swelling of the tissue and obstructing CSF flow, eventually leading to the accumulation of cerebellar subdural hygromas. If the patient's neurological condition is stable, a trial of conservative, nonoperative treatment with acetazolamide and steroids, prescribed at the appropriate dose for weight, may eliminate the need for a more invasive therapy while natural CSF pathways are restored. Since most reported patients presented with classic symptoms of hydrocephalus, including headache, nausea, and vomiting, and only a minority of them presented with a deteriorating level of consciousness and symptoms of impending herniation (lethargy and bradycardia), we believe that the more aggressive treatment of CSF diversion solutions can be withheld in most of the cases while treating the patient conservatively.

Conclusions

De novo hydrocephalus associated with subdural hygromas is a known, albeit rare, complication after CM-I decompression with duraplasty. In carefully selected patients who are without signs or symptoms of impending herniation, it is reasonable to attempt nonoperative management.

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Disclosures

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

Author Contributions

Conception and design: Carey, Vivas, Shimony, Jallo. Acquisition of data: Vivas, Shimony, Jackson, Xu. Analysis and interpretation of data: Vivas, Shimony. Drafting the article: Carey, Vivas, Shimony, Xu, Jallo. Critically revising the article: Carey, Vivas, Shimony, Jackson, Jallo. Reviewed submitted version of manuscript: Vivas, Shimony, Jackson, Jallo, Rodriguez, Tuite. Approved the final version of the manuscript on behalf of all authors: Carey. Administrative/technical/material support: Vivas, Shimony. Study supervision: Carey, Vivas, Shimony, Jallo.

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