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ORIGINAL ARTICLE

Posterior fossa decompression for children with Chiari I malformation and hydrocephalus

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Abstract

Introduction Chiari I malformation (CMI) and hydrocephalus often coexist, with no clear understanding of the cause-and-effect relationship. In the absence of other associated etiologies, the traditional teaching has been to treat the hydrocephalus first, partly to minimize the risk of cerebrospinal fluid (CSF) leak from CMI decompression in the setting of elevated ICP. We describe a series of consecutive pediatric patients with CMI and hydrocephalus, the majority of whom were managed with posterior fossa decompression.

Methods A retrospective review was conducted on consecutive children who presented to the senior author with both hydrocephalus and CMI, with emphasis on rationale for and outcomes of surgical intervention, including the need for additional surgery.

Results There were 14 patients aged 2 weeks to 16 years (median 2 years) who presented with Chiari I and hydrocephalus. Four of these underwent posterior fossa decompression without duraplasty (PFD) as first-line therapy (one of whom eventually required duraplasty), 7 had PFD with duraplasty (PFDD), 1 received a ventriculoperitoneal shunt (VPS), and two had endoscopic third ventriculostomy (ETV). Of the 11 who had PFD/D, 9 (90%) had significant symptom improvement/resolution, 7 (55%) showed decrease in ventricle size, and 1 (10%) required VPS placement for persistent hydrocephalus. Both ETV patients improved clinically, and 1 showed decrease in ventricle size. There were no pseudomeningoceles, infections, or neurological deficits. One CSF leak occurred after an ETV and was successfully treated with wound revision.

Conclusion In patients with both CMI and hydrocephalus, treating the CMI first in an effort to avoid a shunt can be safe and effective. In this series, PFDD in the setting of hydrocephalus did not result in CSF leak or pseudomeningocele. While limited by a small sample size, these data support a causative relationship between CMI and hydrocephalus.

Keywords VP shunts · Posterior fossa decompression with duraplasty · Shunt placement · Pseudomeningocele · CSF leak

Introduction

Chiari I malformation (CMI) and hydrocephalus can be congenital or acquired. Causes of acquired CMI include disorders that increase intracranial pressure (ICP), such as hydrocephalus or intracranial masses, or low ICP, such as chronic lumboperitoneal shunting. On the other hand, causes of acquired hydrocephalus include intraventricular hemorrhage, infection, trauma, obstruction (tumor, cyst), and CMI. CMI is associated with hydrocephalus in 5.9–9.6% of cases depending on the study [1, 2]. Initially attributed to hydrostatic forces created by the overproduction of cerebrospinal fluid (CSF) driving cerebellar contents downward [3], the exact pathogenesis underlying the relationship between CMI and hydrocephalus remains elusive [4–8].

Identifying the causal relationship between CMI and hydrocephalus is of both diagnostic and therapeutic importance. Traditionally, in the absence of other etiologies, such as congenital or acquired craniosynostosis, jugular venous stenosis, and others [9], neurosurgeons were trained to treat coexistent CMI/hydrocephalus with a ventriculoperitoneal shunt (VPS), partly based on the

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theoretical concern that a CMI decompression in the face of elevated intracranial pressure may result in a pseudomeningocele and/or CSF leak [10–12]. Yet, primary treatment of the hydrocephalus may in some situations require subsequent foramen magnum decompression for persistent CMI symptoms [10, 12], suggesting that, in these patients, either the CMI is the cause of the hydrocephalus, or, less likely, the CMI and hydrocephalus are causally independent. We describe our institutional experience in managing patients with both CMI and hydrocephalus, with special emphasis on those (the majority) treated with posterior fossa decompression. Our results indicate that posterior fossa decompression in the setting of hydrocephalus is (1) safe with no increased risk of pseudomeningocele or CSF leak and (2) within the constraints of a small series, effective in resolving hydrocephalus as well as CMI symptoms in the majority. While this suggests the possibility of CMI as a primary cause of hydrocephalus, experienced neurosurgeons recognize that CSF hydrodynamics are complex, with feedback loops between hydrocephalus and tonsillar herniation leading to a causality dilemma.

Methods

Patient data

After approval by the University of Wisconsin-Madison Institutional Review Board, a retrospective analysis was completed on children (ages 17 or less) who presented with both hydrocephalus and CMI between 1999 and 2017. Data collected included age, gender, presenting signs and symptoms, surgical approach, and requirement for additional surgery. Complete brain and spinal MRI were obtained in all patients preoperatively. Brain and targeted spinal MRI scans were obtained in a delayed fashion (at 3 months and/or 1 year) postoperatively except for one patient from South America who underwent immediate postoperative imaging only, after which she returned to her home country with phone and photo follow-up. The diagnosis of hydrocephalus was made based on the presence of ventriculomegaly on MR imaging, in association with clinical suspicion of increased intracranial pressure in most patients. The latter included progressive macrocephaly, irritability, developmental delay, and papilledema (documented by an ophthalmologist). The MRI diagnosis of CMI was made when the tip of one or both cerebellar tonsils was positioned at least 5 mm below the foramen magnum, often with accompanying syringomyelia and appropriate clinical signs and symptoms, such as Valsalva-induced headaches.

Posterior fossa decompression surgery with or without duraplasty

Posterior fossa decompression (PFD/D) was performed via a suboccipital craniectomy with a C1 laminectomy (and C2 laminectomy when indicated) with duraplasty (PFDD) or without (PFD). Those who did not have duraplasty underwent splitting of the outer layer of the dura; tonsillar resection was performed on one patient early in the senior author's practice. PFDD is the standard CMI decompression procedure in the senior surgeon's practice. However, occasional PFD is done based on family preference and intraoperative ultrasound evidence of pulsating tonsils.

Results

Patient demographics and clinical presentation

Between 1997 and 2019, 14 patients (8 male and 6 female) presented with hydrocephalus and a CMI. Ages at presentation ranged from 2 weeks to 16 years (median 1.96 years). The most frequent presenting symptoms were headache ($n=7$, 50%), irritability ($n=5$, 36%), nausea and/or vomiting ($n=3$, 21%), and developmental delay ($n=6$, 43%). Additional symptoms included diplopia ($n=1$) and dysphagia ($n=2$). The most common presenting exam signs were macrocephaly ($n=8$, 53%) and bulging anterior fontanelle ($n=3$, 21%). Additional presenting signs included papilledema ($n=3$), scoliosis ($n=1$), and syringomyelia ($n=3$). Average pre-op FOHR 0.4 (range 0.3–0.53). Clinical characteristics of the cohort are described in Table 1. Postoperative follow-up ranged from 0.58 to 15 years with an average of 5 years and a median of 3.4 years.

Illustrative case (Patient 1 in Table 1 and Fig. 1g)

This otherwise healthy 11-year-old girl presented with progressive headaches, which were more severe in the morning, and associated with nausea, vomiting, and horizontal diplopia. Physical examination revealed severe papilledema. A brain MRI (Fig. 1g) showed both hydrocephalus and a CMI malformation. CINE images (not shown) showed absence of dorsal CSF flow at the foramen magnum with abnormal bidirectional flow ventrally and elevated flow velocities especially with neck extension, as previously described [13]. Given an extensive family history of CMI (both the father and brother) without hydrocephalus, we presumed that the primary diagnosis in our patient was familial CMI, and that hydrocephalus occurred as a result of the foramen magnum obstruction. She underwent PFD with expansile duraplasty using bovine pericardium. Her symptoms and papilledema resolved, and she remains symptom-free after 14 years of follow-up.

Table 1 Patient demographics including pre- and post-operative clinical presentation; initial surgical approach; cerebellar ectopia, presence of syrinx, and need for additional surgery. Note that some symptoms could have been due to either hydrocephalus or CMI and were accordingly displayed in a shared cell. Abbreviations: BF, bulging fontanelle; DD, developmental delay; ETV, endoscopic third ventriculostomy. HA, headache; HC, hydrocephalus; MC, macrocephaly; M-CM, macrocephaly-capillary malformation syndrome; NA, not applicable; N/V, nausea and vomiting; PFD, posterior fossa decompression with stripping of outer layer of dura; PFDD, posterior fossa decompression with duraplasty; VM, Valsalva maneuver; VPS, ventriculoperitoneal shunt placement

ID	Age (years)	Tonsil position	Initial surgery	Clinical presentation		Clinical improvement		Radiographic improvement		Additional surgery
				HC	Chiari I	HC	Chiari I	Ventricle size	Syrinx	
1	11	18 mm	PFDD, tonsillar resection	N/V, morning frontal radiation	HA with occipital	Symptom resolution		Decreased	NA	None
2	13	17 mm	PFD	Papilledema	HA with Valsalva, dysphagia, sleep apnea	Symptom resolution	Symptom resolution	Unchanged	NA	PFDD after 2.5 years
3	2	8 mm	PFDD	MC Hypotonia, speech and motor delay	Dysphagia	Resolution of symptoms, improved speech, motor function, and hypotonia		Decreased	NA	None
4	2	7 mm	PFD	MC Speech and motor delay		Improved speech and motor function		Decreased	NA	None
5	1	6.5 mm	PFD	MC, BF, irritability Speech and motor delay		Symptom resolution		Decreased	NA	None
6	16	9 mm	PFDD	NA	Occipital HA, severe scoliosis, syrinx	NA	Headache resolution	Unchanged	Decreased	Spinal fusion with syrinx shunt
7	1	5 mm	PFDD	MC, BF (carries diagnosis of M-CM) Hypotonia, speech and motor delay	Syrinx	Improved speech, and motor function, improved hypotonia		Decreased	Decreased	None
8	12	28 mm	PFDD	Morning N/V	Occipital HA with temporal radiation	Symptom resolution	Symptom resolution	Decreased	NA	None
9	1	8 mm	PFDD	MC, irritability Speech and motor delay		No change	NA	Unchanged	NA	VPS after 17 months
10	9	12 mm	PFDD and shunt removal after 8 years of shunting for hydrocephalus	NA (VPS present, original pre-shunt presentation of elevated intracranial pressure)	Syrinx	Remains asymptomatic		NA	Decreased	None
11	8 months	13 mm	PFD	MC, irritability, motor delay HA		Symptom resolution		Decreased	None	None

Table 1 (continued)

ID	Age (years)	Tonsil position	Initial surgery	Clinical presentation		Clinical improvement		Radiographic improvement		Additional surgery
				HC	Chiari I	HC	Chiari I	Ventricle size	Syrinx	
12	1	9.3 mm	VPS	Irritability vomiting HA		Symptom resolution		Decreased	None	None
13	2 weeks	7 mm	ETV	MC Poor feeding		Symptom resolution		Unchanged	None	None
14	14	6.2 mm	ETV	Papilledema, bifrontal HA	NA	Symptom resolution	NA	Decreased	None	None

Success of primary procedure

PFD/D as primary procedure

Seven patients underwent posterior fossa decompression with duraplasty (PFDD) and four underwent bone-only decompression (PFD). One of the PFD patients required reoperation for duraplasty 2.5 years postoperatively for severe central apnea (90 events per hour), with complete resolution of the apnea. The ventricles decreased in size in 8 of 11 patients (73%) (Fig. 1) and remained unchanged in the other 3 (27%) (Fig. 2). Average FOHR was 0.4 (range 0.3 to 0.53) preoperatively and 0.38 (range 0.28 to 0.45) postoperatively. However, in some patients, although the FOHR was within normal limits, other ventricular abnormalities suggested the presence of hydrocephalus, namely, dilatation of the temporal horns, cerebral aqueduct, and/or fourth ventricle. Ten patients (including the reoperated patient) showed significant clinical improvement or complete resolution of symptoms and signs without the need for further intervention. One PFDD patient had ongoing ventriculomegaly and elevated ICP (20–30 s mmHg) on 48-h ICP monitoring; he received a ventriculoperitoneal shunt (VPS) 17 months post-operatively, with improvement in both neurological development and ventricle size (Fig. 3).

VPS as primary procedure

One patient received a VPS as primary surgery, leading to improvement in symptoms and ventricle size.

ETV as primary procedure

Two patients underwent successful endoscopic third ventriculostomy (ETV) and remain both shunt- and symptom-free, with decrease in ventricle size in one of them.

Complications

None of the PFD/D and VPS patients had postoperative complications, including CSF leak, pseudomeningocele, infection, and new neurological deficit. One of the ETV patients had a CSF leak from the burr hole site, which was successfully treated with skin staples.

Discussion

The association between CMI and hydrocephalus continues to be of great clinical interest, as the pathogenesis and management implications remain unclear [14]. In this study, we show that PFD/D is a safe approach to

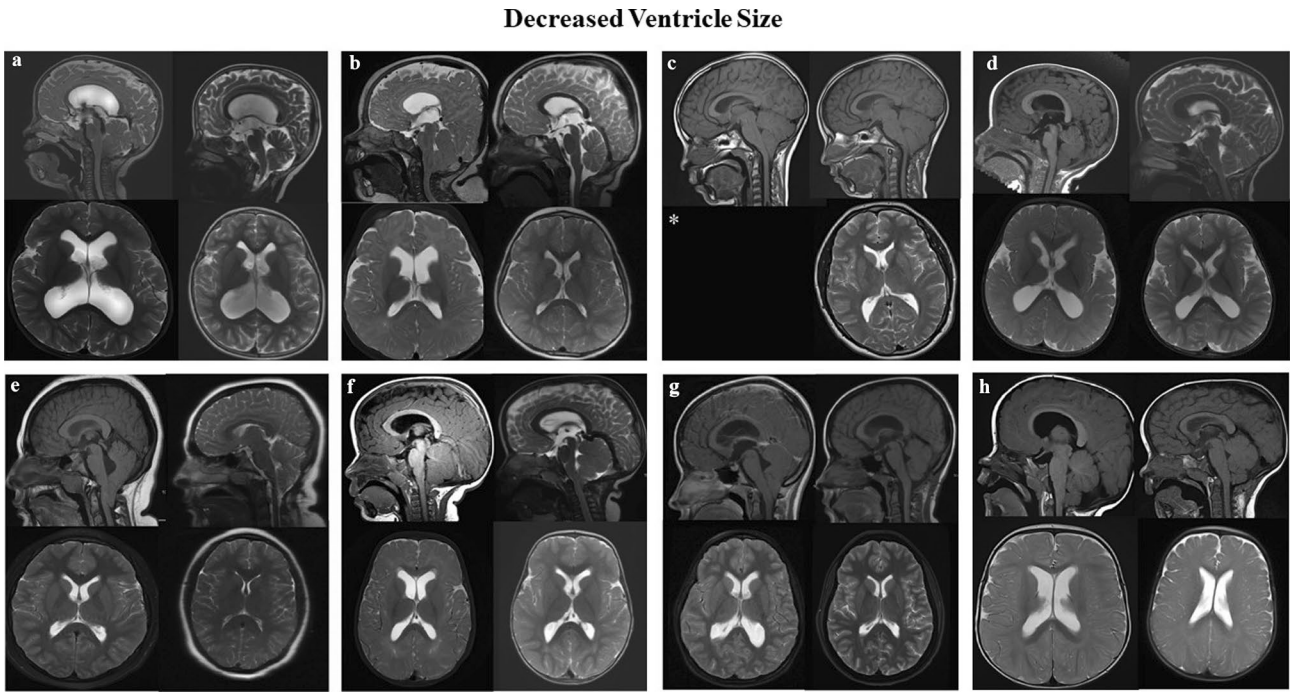

Fig. 1 PFD/D patients whose ventricles decreased in size postoperatively. Sagittal and axial MRI images before (left panel) and after (right panel) PFD/D, showing decrease in ventricle size. Postoperative sagittal images were obtained at 3 months to 1 year to show absence of pseudomeningocele. Postoperative axial images represent the most recent scan. **e** *This is a patient who presented after having been shunted for hydrocephalus at an outside institution. His original (pre-shunt) images were unavailable to us. When he presented with shunt malfunction, he received a PFDD, after which the shunt was removed successfully (**a** patient 3, **b** patient 4, **c** patient 10, **d** patient 7, **e** patient 8, **f** patient 11, **g** patient 1, **h** patient 5)

these patients with little if any risk of CSF leak. Notably, even the patient whose hydrocephalus persisted after PFDD eventually requiring shunting showed no evidence of pseudomeningocele or leak. In addition, within the constraints of our small series, PFD/D is effective in the majority of patients.

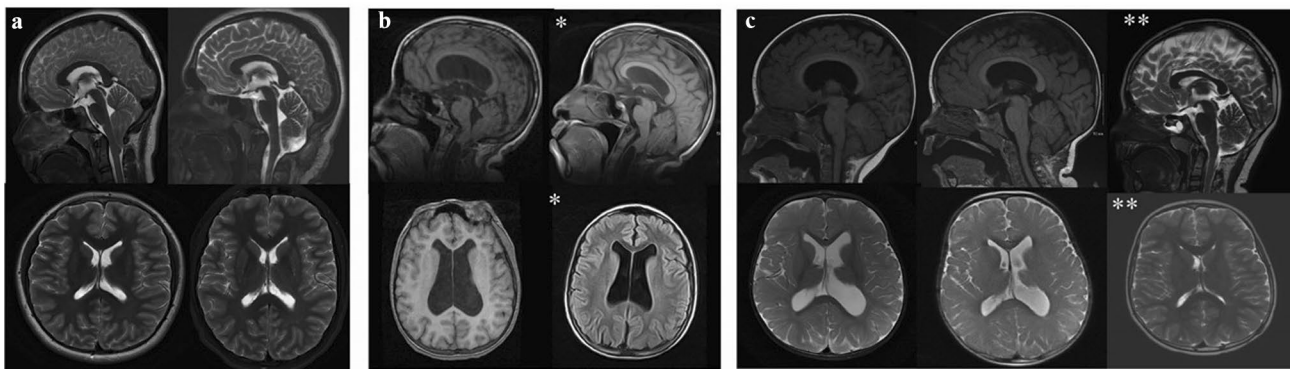

Fig. 2 PFD/D patients whose ventricles did not change in size postoperatively. Sagittal and axial MRI images before (left panel) and after (right panel in **a**, **b**; middle panel in **c**) PFD/D, showing no change in ventricle size. Postoperative sagittal images were obtained at 3 months to 1 year to show absence of pseudomeningocele. Postoperative axial images represent the most recent scan (**a** patient 2, **b** patient 6 *immediate post-op imaging). The patient had complete resolution of symptoms but did not have delayed images (3 months or longer postoperatively) because she returned to her home country; **c** patient 9. **The patient failed PFDD requiring shunt placement (right panel shows post-shunt scan)

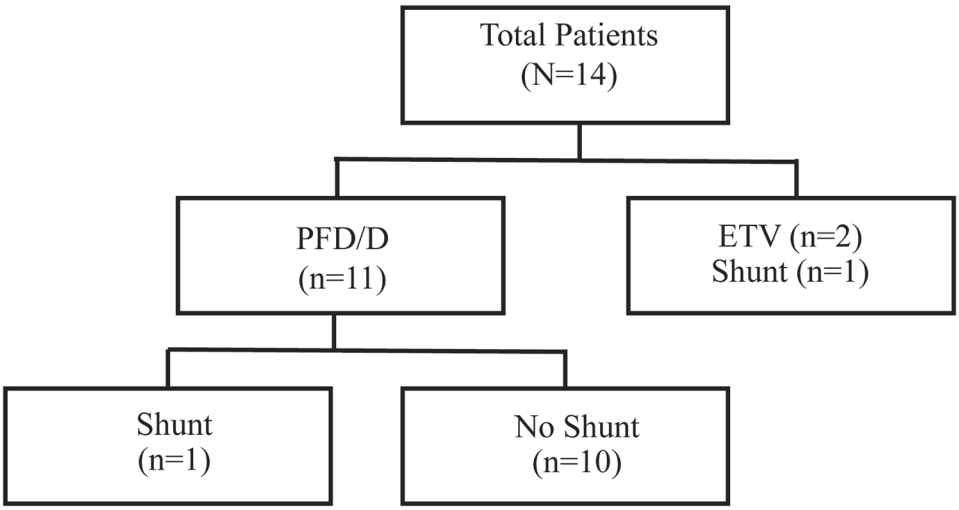

Fig 3 Flowchart depicts the initial surgical approaches with outcome. ETV, endoscopic third ventriculostomy; PFD/D, posterior fossa decompression with or without duraplasty; shunt/no shunt, the patient received/did not receive a VP shunt

Simple vs. complex CMI

Di Rocco et al. [9] argue that patients with coexistent CMI and hydrocephalus may be classified based on simple and complex pathophysiological hypotheses: the "complex" hypothesis includes complicating etiological factors such as cephalocranial disproportion and jugular venous stenosis (e.g., patients with bone dysplasias). Treating such patients depends on the underlying problem. The "simple" hypothesis indicates coexistence of CMI and hydrocephalus with no other complicating factor [9]. In our series, only one patient was considered "complex" and carried a diagnosis of macrocephaly-capillary malformation (M-CM) syndrome. He responded to PFDD with resolution of the hydrocephalus and syringomyelia.

The literature

The most common surgical technique used to treat the CMI/hydrocephalus association in the absence of other etiologies is CSF diversion via either VPS or more recently, ETV [10, 12, 14–22]. CSF diversion leads to resolution of symptoms of elevated ICP in virtually all patients, and a decrease in ventricle size in most shunted patients and a subset of ETV patients [23, 24], consistent with literature on treatment of hydrocephalus from other etiologies [25, 26]. However, the success of CSF diversion in improving CMI symptomatology is variable. Specifically, Haurust et al. [10] reported that, of 16 patients with CMI and hydrocephalus who underwent ETV, 1 required a shunt, and 6 (37.5%) others required foramen magnum decompression. Similarly, in 2018, Wu et al. [12] reported that, of 10 patients treated with ETV, 2 failed, one of whom underwent PFD. On the other hand, Massimi et al. [14] reported treating 15 patients with ETV with 0 failure and none requiring subsequent PFD at a mean follow-up period of 35 months. The literature also includes 13 case reports of successful ETV (Table 2).

Hydrocephalus-CMI cause-and-effect: A "chicken and egg" paradox?

Importantly, if future prospective studies confirm our data, this would suggest that CMI may be a primary cause of obstructive hydrocephalus (instead of the other way around: hydrocephalus as the cause of the CMI). Interestingly, one of the patients in this series had a sibling with CMI and a father with CMI and syringomyelia, but no family members with hydrocephalus. In this patient, the ventriculomegaly, symptoms, and papilledema all resolved after PFDD. The familial CMI and lack of CSF leak complications suggest, at least in this family, that the primary defect is the CMI, with hydrocephalus being secondary. Notwithstanding, experienced neurosurgeons recognize the complex interrelatedness of conditions of abnormal CSF hydrodynamics. For example, if a CMI patient develops hydrocephalus secondarily, the hydrocephalus can theoretically, in turn, cause worsening of the tonsillar herniation. Conversely, if a patient with untreated hydrocephalus shows secondary tonsillar herniation, the resulting foramen magnum obstruction could presumably lead to further ventricular dilatation. Only large studies and registries can more specifically scrutinize this "chicken and egg" paradox representing the complex relationship between these conditions. Additional compelling questions emerge in light of ETV success in relieving hydrocephalus associated with CMI, namely, whether tonsillar crowding of the foramen magnum represents the obstructive process that led to ventriculomegaly. It is notable that the ETV literature pertaining to CMI-associated hydrocephalus does not discern other obstructive lesions, such as aqueductal stenosis.

Table 2 Literature of ETV used to treat hydrocephalus associated with Chiari I malformation. Summary of adult and pediatric case series in the literature. In addition, our review of the literature includes at least a total of 13 individual case reports not listed here

ETV study	Number of patients	Outcomes			Additional surgery	
		High ICP symptoms *N* (%)	Ventricle size *N* (%)	Chiari I symptoms *N* (%)	PFD for persistent Chiari I symptoms *N* (%)	VP shunt for persistent hydrocephalus *N* (%)
Hayrust et al. [10]	16	Resolved in 15 (94%)	Reduced in 5 (31%)	Persistent in 6 (37.5%)	6 (37.5%)	1 (6%)
Massimi et al. [14]	15	Resolved in 15 (100%)	Resolved in 6 (40%) Reduced in 9 (60%)	Resolved in 6/10 (60%) Improved in 4/10 (40%)	0 (0%)	0 (0%)
Wu et al. [12]	10	Improved in 6 (60%) Resolved in 1 (10%) Stable in 1 (10%) Deteriorated in 1 (10%)	Data not available	Persisted in 1 (10%)	1 (10%)	0 (0%)

Children vs. adults

Our data in children confirm those in an adult study by Deng et al. [27] who show that all 38 patients with CMI and ventriculomegaly who underwent PFDD had successful resolution of the CMI symptoms and no complications. Interestingly, none of the reported adult patients presented with signs or symptoms of elevated ICP that would suggest the need for CSF diversion, and one had a change in ventricle size postoperatively [27]. These observations suggest that the ventriculomegaly in the Deng series may not represent hydrocephalus, in contrast to our series, in which hydrocephalus was suspected clinically and/or radiographically in the majority of patients.

Treating the hydrocephalus vs. the CMI

If both ETV and PFD/D prove to be successful options for the treatment of the CMI/hydrocephalus association, the choice between the 2 procedures rests on their respective risks. Advantages of ETV over PFD/D include shorter procedure time, smaller skull opening, and absence of muscle dissection leading to less postoperative pain. While, theoretically, ETV carries less risk of a CSF leak or pseudomeningocele than a PFDD, the only patient in our series who suffered a leak was an ETV patient. Conversely, advantages of PFD/D over ETV include lack of brain entry or proximity to the basilar artery and hypothalamus.

Incidental low tonsillar position and/or ventriculomegaly

Finally, we caution that (1) tonsil position below the foramen magnum has a high prevalence in the normal population [28], (2) hydrocephalus is a disorder with diverse etiologies, and (3) asymptomatic ventriculomegaly may not require treatment. This means that co-occurrence of low tonsils and hydrocephalus/ventriculomegaly may in some cases be coincidental. Similarly, as stated previously, CMI can be associated with other etiologies such as congenital and acquired craniosynostosis. Therefore, as previously recommended [9], a thorough evaluation for other causes of hydrocephalus and CMI would be wise before committing the patient to surgical intervention. In addition, only one patient in this series was considered complex, and most patients had clinical symptomatology and/or imaging criteria that indicated that the low tonsillar position and the ventriculomegaly were unlikely to be incidental.

Conclusion

We describe a cohort of children treated for CMI and concomitant hydrocephalus, the majority of whom underwent PFD/D with 90% success and no complications. When CMI and hydrocephalus coexist in the absence of other congenital or acquired anomalies, PFD/D can be safe and effective, obviating the need for long-term shunting. While this is a small retrospective series, which prevents us from making definitive statements regarding best therapy, it represents the entire population of children evaluated by the senior author with this specific combination of malformations. This arguably compels us to consider tonsillar crowding as a principal obstructive cause of hydrocephalus in these children, or at least, as a safe point of intervention for a complex hydrodynamic pathology. Notably, both ETV and PFD/D may be sensible treatment options, the choice of

which should be considered on a case-by-case basis and motivated by surgeon-family discussion.

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Declarations

Conflict of interest The authors have no conflict of interest with any part of this manuscript.

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