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Chiari 1 malformation and raised intracranial pressure

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Abstract

Background The pathophysiology of Chiari 1 malformation (CM1) is inextricably related to intracranial pressure (ICP). The characteristic cerebellar tonsil herniation at the foramen magnum may either cause raised ICP by disturbing CSF flow (as observed in idiopathic CM1) or may itself be the effect of raised ICP (as observed in acquired CM1). Distinguishing between these two phenomena, therefore, is of paramount importance in successfully alleviating the symptoms of the condition and preventing serious complications.

Objectives In this article, we discuss the pathophysiology of raised ICP in CM1 and review the current evidence for its investigation and treatment. We also share our own clinical experience which investigates the utility of ICP monitoring in a series of 26 children with CM1.

Keywords Chiari 1 malformation · Hydrocephalus · Intracranial pressure

Background

Chiari 1 malformation (CM1) is a structural abnormality of the hindbrain and is characterized by the herniation of the cerebellar tonsils through the foramen magnum [1]. CM1 has an estimated prevalence of 1% in the pediatric population [2] and patients typically present in adolescence or early adulthood. Despite the condition being eponymously named more than 100 years ago by Professor Hans Chiari, CM1 remains poorly understood and a condition of contention in many respects. In this article, we aim to provoke debate regarding the convoluted relationship between CM1 and intracranial pressure. We will review the pathogenesis, investigation, and management of raised ICP and hydrocephalus in the context of CM1. We firstly present data from our pediatric neurosurgery center on our experience of ICP monitoring in children with CM1.

Methods

We searched all available records from the Department of Paediatric Neurosurgery at the John Radcliffe Hospital (Oxford, UK) for children who underwent ICP monitoring to inform the management of their CM1. Patients included had clinical features suspicious of raised ICP [1] but either no or subtle ventriculomegaly. We disregarded patients with CM1 who had ICP monitoring for the purpose of determining shunt failure and also children who had ICP monitoring for the purpose of managing craniosynostosis in the presence of a CM1 considered incidental. ICP was measured continuously, typically for 24 h, using either a *Codman* Microsensor or a *Raumedic* intraparenchymal pressure monitor inserted into the frontal lobe. The ICP waveform was inspected manually and the mean ICP was measured computationally. As per a prior review from our unit, in children with a closed skull, we considered raised ICP to be present when there was a consistently raised baseline of more than 15 mmHg or when there were more than three B-waves observed within 24 h [3].

Results

We identified 26 patients between the ages of 2 to 18 years who had CM1 and underwent ICP monitoring between January 2009 and December 2018. Nineteen children had not or not yet undergone foramen magnum decompression ("pre-FMD") (Table 1) and eight children had ICPM after FMD ("post-FMD"). One child underwent ICP monitoring both before and after FMD and is therefore included in both groups. Eight children had craniosynostosis.

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In the pre-FMD group, 2/19 patients were found to have raised ICP. The first child was a 2-year-old boy with bilambdoid synostosis who had a tight foramen magnum, dysmorphic ventricles, and a CM1. He underwent total calvarial remodeling and evaded the need for either cerebrospinal fluid (CSF) diversion or FMD. The other child was a 2-year-old girl with a history of neonatal hypoxic-ischemic encephalopathy who was found to have a baseline pressure of 19-21 mmHg on monitoring. While the rationale is not clear on retrospective review, the clinicians at that time first trialed a course of acetazolamide. When there was no symptom benefit, the patient then underwent an uncomplicated FMD.

A particular case of interest in the pre-FMD group was a 3-year-old boy with a significant CM1 who had a history of sagittal and left unicoronal synostosis that was previously treated with calvarial remodeling. He later developed headaches suspicious of raised ICP and underwent monitoring revealing a mean ICP value of 10 mmHg. He then proceeded to have an FMD, but the postoperative course was complicated by a pseudomeningocele, CSF leak, requirement for lumbar drainage, and then meningitis. Repeat ICP monitoring was performed on his recovery and showed a baseline of 24 mmHg, so further calvarial expansion was performed which improved his symptoms.

Other than the 3-year-old boy aforementioned, none of the other eight children in the post-FMD group were found to have raised ICP and their symptoms were managed conservatively.

Discussion

Chiari 1 malformation—a cause or effect of raised pressure?

While the pathogenesis of CM1 remains poorly understood, we do understand that CM1 is a heterogeneous condition. We consider that most patients have an idiopathic CM1, meaning that there is no other causative pathology causing cerebellar herniation. The typical appearance of a congested, small posterior fossa floor supports a theory that CM1 is the result of a mesodermal defect causing an under-developed and insufficient cranium [4]. This theory supersedes Hans Chiari's original theory that CM1 is the result of resolved fetal

Table 1 Clinical details of nineteen children who underwent ICP monitoring who had not or not yet undergone foramen magnum decompression

Age (yrs)	Cranio-synostosis	Ventriculomegaly	ICP	Management	Surgical complications
4	No	No	Normal	Conservative	
15	No	No	Normal	Conservative	
13	No	No	Normal	Conservative	
14	No	No	Normal	Conservative	
12	No	No	Normal	Conservative	
18	Yes	No	Normal	Conservative	
6	No	No	Normal	FMD	None
4	No	No	Normal	FMD	None
9	Yes	No	Normal	FMD	None
16	No	No	Normal	FMD	None
12	Yes	No	Normal	FMD	None
14	Yes	No	Normal	FMD	None
11	No	No	Normal	FMD	Postoperative hydrocephalus treated conservatively with acetazolamide and bed-rest.
16	No	No	Normal	FMD	No complications
3	No	No	Normal	FMD	Small pseudomeningocele, managed conservatively.
9	No	Subtle	Normal	FMD	No complications
3	Yes	No	Normal	FMD	Pseudomeningocele and wound leak, treated with lumbar drain. Then developed meningitis. Repeat ICP monitoring showed raised ICP (mean 24 mmHg). Then treated with repeat calvarial remodeling.
2	Yes	Dysmorphic	High	Total calvarial expansion	None
2	No	No	High	Trial of acetazolamide, followed by FMD	None

FMD foramen magnum decompression

hydrocephalus. Congestion at the foramen magnum and posterior fossa causes a disturbance of CSF circulation and outflow from the fourth ventricle. Hydrocephalus is, therefore, a logical complication of CM1 but, according to the literature, occurs only in around 6.5–9.6% of cases [5, 6]. Syringomyelia, present in 45–75% of patients [7], is also a downstream consequence of a disturbance of CSF flow and pressure differential at the craniocervical junction. As such, in these cases of idiopathic CM1, FMD is therefore as an intervention to treat the symptoms by relieving congestion and normalizing the circulation and hydrodynamics of CSF.

On the other hand, there are cases of CM1 considered to be acquired due to a comorbid and aggravating pathology. These cases have been reported in association with craniosynostosis [8, 9], cranial space-occupying-lesions [10], tethered cord [11], idiopathic intracranial hypertension (IIH) [12], and spinal CSF diversion or leakage [13–15]. While in some cases causation is tentative, there are some examples of more convincing paradigms. For example, Payner et al. demonstrated 10 children with prior normal hindbrain anatomy who developed tonsillar descent following lumbar shunting [16]. In terms of pressure, raised ICP in craniosynostosis, brain tumors, and IIH will increase the craniospinal pressure gradient and cause a "push-down" effect, whereas spinal CSF diversion will cause a "pull-down" effect on the cerebellar tonsils. Tethered cord has a more complex relationship with CM1 that is not well understood. In these acquired cases, where CM1 may be the effect or consequence of raised pressure, FMD alone is less likely to significantly change the cranio-spinal gradient, alter the anatomy of the malformation, or alleviate its symptoms.

While this dichotomy of CM1 into idiopathic or acquired cases is arbitrary and somewhat contentious within the literature, what is imperative, in our opinion, is to carefully consider the relationship between the CSF physiology and the hindbrain anatomy before choosing how to manage the CM1 on any one child. However, determining this pivotal distinction is not always straight forward.

Management of pre-FMD raised ICP

There are currently no guidelines to bring consensus to the investigation or treatment of CM1 [17, 18] and surgical practice varies widely [19]. There appears to be an agreement in the literature that patients with CM1 with overt hydrocephalus or objectively proven raised ICP should have CSF diversion prior to consideration of FMD (Fig. 1) [6, 20]. A study of 936 children undergoing FMD for CM1 by Greenberg et al. showed that comorbid hydrocephalus was independently associated with an increased rate of "surgical complications" occurring within 90 days [5]. Similarly, Vedantam et al. noted an association with an increased risk of complications in children with pre-FMD hydrocephalus [21], but as per the Greenberg study, the individual complications accredited to hydrocephalus were not specified. Furthermore, there is data to suggest that CSF diversion prior to FMD for CM1 associated with hydrocephalus will reduce the requirement for later FMD [20, 22] and this reflects our anecdotal experience also. For example, Hayhurst et al. evaluated the results of endoscopic third ventriculostomy (ETV) in 16 patients (children and adults) with CM1 in one center [22]. They showed that performing ETV first evades the requirement of a shunt in 15/16 patients and the need for FMD in 10/16 patients.

Although there is a distinct lack of data on the management of acquired cases of CM1, we would support that a pathology clearly causing CM1 by imposing intracranial hypertension should be dealt with prior to the consideration of FMD. For example, our pediatric neurosurgical practice collaborates with a craniofacial service in which we observe the well-recognized association between CM1 and both syndromic and non-syndromic craniosynostosis [9]. Perhaps related to early intervention (typically calvarial expansion), we see very few children with craniosynostosis later requiring FMD for comorbid CM1, but further data is required in this field.

The cases that pose a greater diagnostic and therapeutic challenge are patients with CM1 with clinical features of raised ICP, but without hydrocephalus or an obvious pathological cause. For example, cases of IIH can be difficult mimics of idiopathic CM1 due to normal or small ventricular size in association with an acquired CM1 [12]. Zakaria et al. from Liverpool have suggested that in the absence of overt hydrocephalus, but in the presence of pressure-like symptoms, ICP monitoring should precede FMD [20]. So far, this has also been the practice in our center.

The decision to invasively measure ICP, however, can understandably be a daunting proposition to patients or to parents. Recent data from our institution shows that ICP monitoring is relatively safe, but not risk-free. In 385 children undergoing ICP monitoring over 6 years, there were no cases of mortality or lasting morbidity [23]. There was, however, an overall complication rate of 8.3% in which the most frequent complications were hardware failure (4.2%), CSF leakage (3.6%), hemorrhage (0.5%), and infection (0.3%). Similar figures have been reported by another UK center, but they reported also a 0.9% incidence of device misplacement and 0.3% incidence of postoperative seizures [24]. The risks of ICP monitoring, therefore, need to be considered and balanced against the aforementioned risks of performing FMD in patients who potentially have raised ICP.

Management of post-FMD raised ICP

Postoperative hydrocephalus following FMD is a recognized phenomenon, however statistical reporting of this complication in published studies is variable (Table 2). A large, retrospective, US, multi-center study of 2649 children undergoing

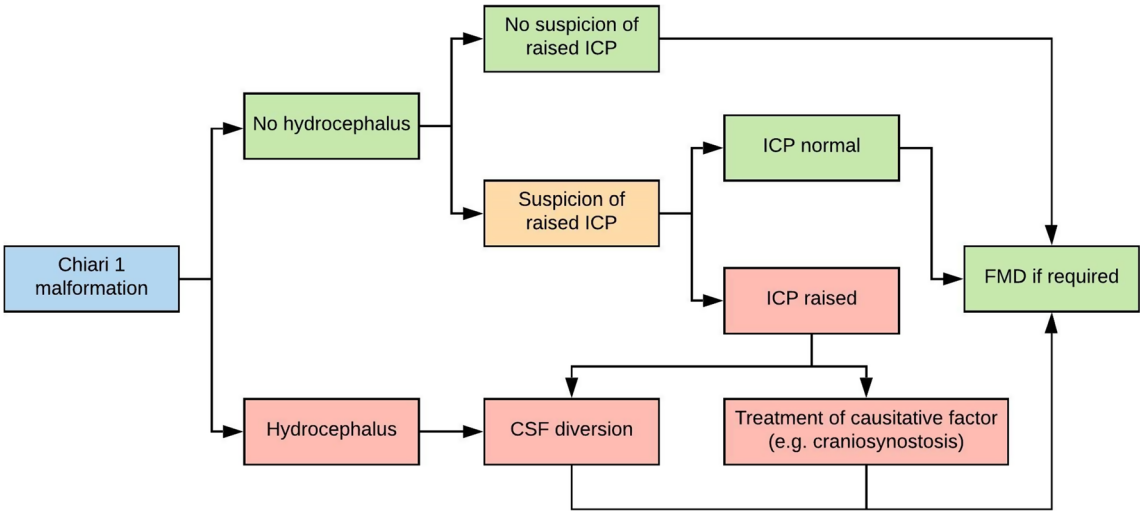

Fig. 1 Proposed management algorithm for the surgical management of CM1

FMD by Shweikeh et al. showed that the rate of hydrocephalus following FMD was 5.9% [26]. Interestingly, the same study showed that although pseudomeningocele rates were significantly higher in cases undergoing FMD with durotomy and duraplasty rather than those undergoing a "bone-only" FMD, there was no significant difference in rates of post-FMD hydrocephalus between these groups. A recent systematic review has supported that FMD with duraplasty causes more "CSF-related complications" than bone-only FMD, but does not specify these further [27]. Guan et al. showed that in 297 children undergoing FMD for CM1, the factors associated with hydrocephalus within 12 months after FMD were age (higher risk in those aged less than 6 years), high intraoperative blood loss, and the finding of a fourth ventricular web [25]. Based on the data available, it would seem necessary to include CSF diversion as a potential complication following FMD when consenting patients for the procedure.

The requirement for permanent CSF diversion post-FMD has been reported between 1 and 7.4% in pediatric studies [20, 21, 25]. There are some children, however, who develop post-FMD hydrocephalus or raised ICP who can evade this requirement for permanent CSF diversion by a temporary measure. For example, a study of 138 children and adults with CM1 showed that 12 (8.7%) developed post-FMD hydrocephalus [20], of which 9/12 required permanent CSF diversion and 3/12 evaded permanent CSF diversion. This mirrors our anecdotal experience, such that not all children with postoperative hydrocephalus require shunting and that strict, flat bed rest and acetazolamide for a number of days can restore CSF circulation. An interesting but small study ($n = 11$) by Fric et al. showed that FMD with expansive duraplasty did not convincingly lead to normalization of either pulsatile or static ICP in the first three postoperative days [28]. The authors suggested that CSF circulation does not normalize in the immediate postoperative phase and this may be in some way related to the pathophysiology of the post-FMD hydrocephalus phenomenon. Further research on the subject of postoperative hydrocephalus is required and a question remains as to

Table 2 A sample of studies investigating CSF complications following FMD for CM1 in children

First author	No.	Age range (years)	Post-FMD CSF complications				
			Hydrocephalus	CSF leak	Hygroma	Pseudomeningocele	Permanent CSF diversion
Greenberg [5]	936	</= 18	NR	7.4%*	NR	7.4%*	4.0%**
Guan [25]	297	0–18+	7.4%*	5.7%	NR	11.1%	7.4%*
Shweikeh [26]	2649	0–20	5.9%	NR	NR	3.9%	NR
Tubbs [6]	500	2–20	NR	0.2%	0.8%	NR	NR
Vedantam [21]	1459	0–18	NR	1.7%*	NR	1.7%*	1%
Zakaria [20]	138	0–61	8.7%	4.3%	3.6%	NR	6.5%

NR not reported

*Outcomes reported together

**Data combined with shunt revision or exploration

whether temporary diversion techniques (lumbar puncture or drainage) could defer or eliminate the need for permanent diversion.

Raised ICP after FMD may cause patients typical pressure symptoms and overt ventriculomegaly on imaging, but in some cases raised ICP may alternatively manifest as CSF-leak through the wound (0.2–7.4%) [5, 6, 20, 21, 25], pseudomeningocele (3.9–11.1%) [5, 21, 25, 26], or subdural hygromas (1–7.4%) [20, 21, 25]. These CSF complications due to raised pressure, even in the absence of ventriculomegaly, are best treated with CSF diversion [29], however the method of diversion remains disputed. While the conventional methods would be ventriculoperitoneal shunting or even ETV, some have attempted cysto-peritoneal shunting or subdural-peritoneal shunting.

In our practice, we have observed in some cases the unsatisfactory post-FMD phenomenon of "slumping" or "ptosis" of the cerebellum, as described in 1978 by Williams [30]. This complication causes deranged CSF flow and can cause hydrocephalus. Slumping and consequent disturbed CSF flow may be prevented by limiting the size of the suboccipital craniectomy (dubbed "less is more" by Abd-El-Barr and Groff [31]). In cases of slumping, we would advocate strict bed rest for 2–4 days with a Trendelenburg position and acetazolamide. While we find that, in most cases, further surgery is not required, others have advocated for posterior fossa reconstruction using bone grafts [32] or synthetic plates [33].

Limitations

As per Zakaria et al. [20], our approach has also been to use ICP monitoring to exclude raised ICP in the presence of raised pressure symptoms but without overt hydrocephalus. This small, minority of cases that we have examined, in both the pre- and post-FMD cohort, are those in which there is diagnostic uncertainty and we are therefore bound to have a low pickup rate of raised ICP. We recognize the bias in this data since patients with overt hydrocephalus or high clinical suspicion of raised pressure may bypass ICP monitoring and continue with CSF diversion. Furthermore, the outcome data from such a small sample reported in our case series cannot inform us of the benefit of ICP monitoring in these situations.

Although we found that only 2/19 children had raised ICP on monitoring prior to FMD, we showed that in 9/11 children with reassuring ICP results who then went on to have FMD had no CSF complications. Furthermore, objective monitoring proving normal ICP may have prevented unnecessary CSF diversion. In the case of the child who had severe CSF complications and repeat monitoring revealing raised pressure, it is impossible in retrospect to know if the first ICP monitoring was inaccurate and missed raised ICP caused by craniosynostosis or whether the raised ICP was due to complications from the FMD procedure.

This is the second case series, to our knowledge, looking specifically at ICP monitoring in CM1. The other study, reported by Dyson et al., was a series of 16 patients, but the full data was not published [34]. There is therefore a distinct lack of data that can give evidence to the utility of ICP monitoring in CM1 and the benefits versus the risks cannot yet be weighed. Considering that it took 10 years to measure 26 patients in one center, it is clear that a collaborative and multi-center approach to answering these questions in CM1 is required [35].

Beyond "raised ICP"

Most children with CM1 will not have hydrocephalus or proven raised ICP baseline, but abnormal ICP may still be the cause of their symptoms. Indeed, the Valsalva maneuver (such as coughing or straining) causes a transient rise in ICP, increases the craniospinal pressure gradient, and exacerbates herniation of the tonsils. There is a growing idea that an impairment in intracranial compliance and a disorder of CSF pulsatility is a key pathophysiological concept in CM1 [29, 36]. A study by Frič et al. analyzed simultaneous intracranial and lumbar pressure in 26 patients and found that most patients had an abnormal pulsatile ICP and pulsatile pressure gradient [29]. The study by Dyson et al. found that 14/16 patients with CM1 without hydrocephalus undergoing ICP monitoring had abnormal pulsatility (defined > 4 mmHg median pulse amplitude) but all patients had a median ICP of < 10 mmHg [34]. Six of the patients underwent ventriculoperitoneal shunting as a primary procedure and experienced symptomatic improvement at two months, leading the authors to conclude that ventriculoperitoneal shunting is an effective alternative to FMD for CM1 in the absence of hydrocephalus. The study requires corroboration, but the question of whether parameters other than static ICP should influence decision making in CM1 is certainly interesting.

Conclusions

Based on our clinical experience and literature review, we iterate the following suggestions for practice:

1. Consider that a CM1 may be either an idiopathic or acquired malformation.
2. CM1 may either be the cause or the effect of raised ICP and/or hydrocephalus and the correct management of the CM1 must take this into account.
3. The literature suggests that comorbid hydrocephalus is better dealt with by supratentorial CSF diversion prior to FMD.
4. Further evidence is required to determine the utility of ICP monitoring in CM1 in children with symptoms suspicious

of raised ICP but without hydrocephalus. Larger, collaborative, and multi-center studies are required.
5. Postoperative hydrocephalus is a recognized complication following FMD. It should be discussed with patients consenting for FMD along with the risk of requiring either temporary or permanent CSF diversion. Further research is required to establish which patients are susceptible to postoperative hydrocephalus and how it should be best treated.
6. CM1 physiology is more complex than what can be interpreted by static ICP data and further research is required to determine how more advanced CSF hydrodynamic analysis will influence practice.

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Compliance with ethical standards

Conflict of interest None to declare.

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