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Chiari I malformation and altered cerebrospinal fluid dynamics—the highs and the lows

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Abstract

Purpose This paper reviews the altered cerebrospinal fluid dynamics that can be associated with paediatric Chiari I malformation and we present our own institutional experience.

Methods We conducted a thorough review of the literature and retrospectively analyzed all cases of operatively managed paediatric Chiari 1 malformation at our institution between February 2006 and February 2019.

Results Acquired Chiari malformation (ACM) can radiologically mimic Chiari I and has been associated with both intracranial hypotension (either secondary to lumboperitoneal shunting or spontaneous CSF hypotension) and idiopathic intracranial hypertension (IIH). At our institution, 61 paediatric cases (range, 2–15 years) underwent foramen magnum decompression (FMD) for Chiari I malformation. Whilst 80% (50/61) of cases underwent FMD with no preceding or post-operative problems of CSF dynamics, 8% (5/61) of cases had hydrocephalus at initial presentation requiring CSF diversion followed by FMD for persistent Chiari, and 10% (6/61) developed hydrocephalus following FMD and required long-term CSF diversion.

Conclusions In paediatric ACM, the management of intracranial hypotension involves thorough radiological assessment and inclusion/adjustment of a valve in the case of lumboperitoneal shunting or epidural blood patch or interventional techniques in the case of spontaneous CSF leak. Thereby, unwarranted posterior fossa decompression surgery is avoided. In the case of IIH and Chiari I malformation, children who have recurrent symptoms despite adequate posterior fossa decompression surgery (failed Chiari), there is a strong role for intracranial pressure monitoring as raised intracranial pressure may indicate long-term CSF diversion.

Keywords Chiari malformation · Intracranial hypotension · Lumboperitoneal shunt · Cerebrospinal fluid leak · Intracranial hypertension · Paediatrics

Introduction

Chiari I malformation is defined radiologically as a displacement of the cerebellar tonsils 5 mm or greater below a straight line drawn between the basion and opisthion (McRae line) at the foramen magnum [1]. Chari I malformation is often associated with bony abnormalities such as a small posterior fossa, and with characteristic clinical symptoms including cough-induced headache. Acquired Chiari malformation (ACM) can have a very similar radiological appearance with respect

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to cerebellar tonsillar descent below the foramen magnum, but is caused by intracranial hypotension, and as such, the radiological indicators of intracranial hypotension can help differentiate ACM from true Chiari I malformation. Intracranial hypotension itself may be secondary to lumbar spinal CSF diversion procedures (in particular, lumboperitoneal shunting) or more rarely spontaneous CSF leakage. Ideal therapy requires recognition of the acquired Chiari syndrome and treatment directed to the cause and/or site of the spinal CSF hypotension [2]. Chiari I malformation has also been associated with idiopathic intracranial hypertension [3], although the exact relationship is poorly understood. Appropriate management of each of these and thorough review of the literature, together with presentation of our own institutional experience, will be addressed in this paper.

Discussion

Chiari malformation and CSF diversion—lumboperitoneal shunting

Lumboperitoneal (LP) shunting's effectiveness necessitates free CSF communication between the ventricular system and the spinal subarachnoid pathways (i.e. the absence of obstructive hydrocephalus) and, although largely reserved for adults, represents a rare but nevertheless important treatment option for CSF diversion in children with hydrocephalus, idiopathic intracranial hypertension, and CSF leak. From the first attempted cases of LP shunting back in 1898 by Ferguson [4] through to the 1950s, there were relatively high rates of morbidity as well as mortality with the open technique and the use of somewhat fragile polyethylene tubing [5]. Since then, the operative technique has undergone significant refinements, including introduction of silicone tubing in the 1960s [6, 7], the percutaneous technique [6], the need for secure ties and anchors to prevent shunt migration [8], and insertion of valve mechanisms to reduce the risk of overdrainage problems including postural headache and acquired Chiari malformation [9].

The rarity of LP shunting in children is well known and well demonstrated by published data from our own institution, a large tertiary centre for paediatric neurosurgery. Over the last 10 years, of over 1000 CSF diversion procedures performed in children at our institution, only 16 patients (1.6%) were LP shunting procedures, and of these, only 6 were new shunt insertions mainly for IIH; the remaining 10 cases were revision operations [9]. The vast majority of CSF diversion procedures were VP shunts (85%) and endoscopic third ventriculostomies. There are several other reports of the rarity of paediatric LP shunting in the literature [10–13].

In assessing adequate LP shunt function, as well as clinical evaluation which is, of course, paramount and should include visual acuity, fields and funduscopic assessment for papilloedema, post-operative CT, or MRI can also be important. These imaging modalities can demonstrate normalization of ventricular size in patients operated for hydrocephalus with ventriculomegaly, although it should be pointed out that ventricular size and LP shunt function have been poorly correlated [13]. Considered more reliable in ruling out a blocked LP shunt and confirming adequate CSF drainage—especially in patients with IIH—cranial imaging may demonstrate attenuated or obliterated basal cisterns (the so-called absent cistern sign) (Fig. 1), which has a reported sensitivity of 75% and a specificity of 57% [13].

ACM following LP shunting has a varied reported incidence in the literature as high as 70% in some studies [10, 11, 14, 15] and therefore is not uncommon, although it is mostly asymptomatic. The rare instances of symptomatic cases (approximately 5% of cases) may manifest as persistent disabling low-pressure headache that does not resolve over time (especially in IIH patients), neck pain, and lower cranial

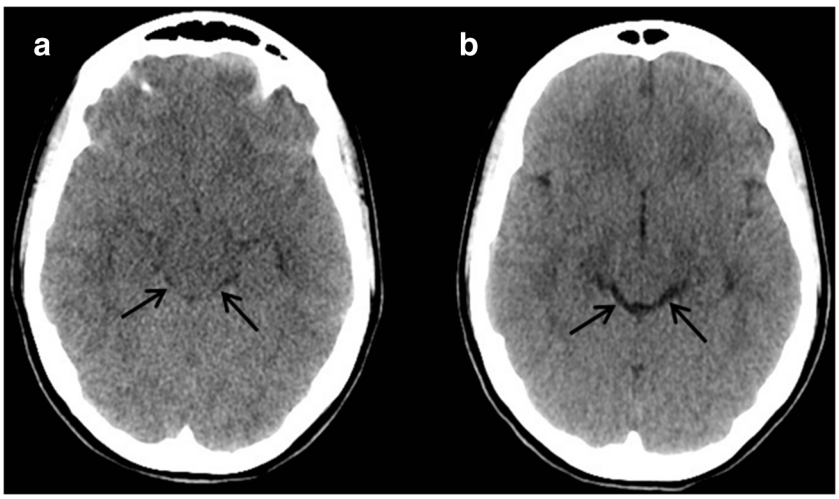

Fig. 1 'Absent cistern' sign following successful lumboperitoneal shunting in 15-year-old boy with IIH (**a**, black arrows) versus pre-operatively with present ambient cisterns (**b**, black arrows)

nerve palsies from the lower brainstem or upper cervical cord compression. It is in such symptomatic cases that revision surgery to insert a valve (if not previously used) or revise the valve setting if appropriate (if valve in situ) may be effective, and if not, then converting to a VP shunt should be considered. In support of this practice, the incidence of ACM has been reported to decrease significantly with the use of adjustable or programmable valves [16], and several studies show a reversal of ACM with the use of valve systems [17–20] (Fig. 2).

The study of myelomeningocoele and its repair may offer an in utero correlate of lumboperitoneal shunting and acquired Chiari malformation. Myelomeningocoele is the most severe form of spina bifida—the most common congenital malformation of the central nervous system—and is defined as the protrusion of the spinal cord and the meninges through a defect in the vertebral column. This defect can result in significant disability including lower limb paralysis and bladder and bowel dysfunction. The vast majority of neonates born with myelomeningocoele have the Chiari II malformation, which defines a range of anomalies including hindbrain herniation (downward displacement of the medulla, fourth ventricle, and cerebellum into the spinal canal), brainstem abnormalities, low-lying venous sinuses, and a small posterior fossa. It is also associated with hydrocephalus and developmental brain abnormalities. Previous cohort studies have suggested improved outcomes with prenatal surgery for myelomeningocoele [21–24]. However, comparisons between infants who were treated in utero and historical controls have understandably been heavily criticized for bias. Subsequently, an important randomized trial called the Management of Myelomeningocoele Study (MOMS) was undertaken and, although associated with greater obstetric complications, did demonstrate greater improvement in (and reduced rate of) hindbrain herniation and significantly reduced rate of CSF shunting procedures with antenatal repair of the myelomeningocoele (40% shunted) versus standard post-natal repair within 24–48 h of birth (82% shunted) [25]. It has been postulated that reduced hindbrain herniation may have led to improved flow of CSF and accounted for the reduced need for CSF shunting. The MOMS data provide an in utero correlate of improvement in Chiari and restoration of normal CSF dynamics when the primary spinal defect was repaired as early as possible (antenatally before 26-week gestation), suggesting that the open spinal defect is in effect a 'lumboperitoneal shunt in utero' [11]. Interestingly, patients with open spina bifida also often have tectal beaking and perimesencephalic changes and, as mentioned above, in lumboperitoneal shunt patients, the 'absent cistern sign' is used as an indicator of shunt functioning—further evidence of the anatomical distortion that is possible by draining CSF from the lumbar region.

Chiari malformation and spontaneous intracranial hypotension

Spontaneous intracranial hypotension (SIH) is extremely rare, with an estimated prevalence of one in 50,000 children [26]. The most consistent symptom is postural (orthostatic) headache but it is worth emphasizing that SIH is an extremely rare diagnosis for the cause of headache in children and adolescents. Other features of SIH may include symptoms of hindbrain herniation (low cranial nerve palsies, ataxia), cranial nerve dysfunction (such as abducens nerve palsy or less frequently oculomotor nerve or trochlear nerve palsy), and radicular arm pain [26, 27]. Whilst SIH is increasingly recognized in the adult population, only four case reports and one larger case series have been published on children and adolescents [28]. Schievink and his team, based in Los Angeles, USA, specialize in treating SIH, with numerous publications in the field, and has treated just over 20 children over an 11-year period [29]. Our own experience at a high volume tertiary paediatric centre in Leeds, UK, similarly demonstrates a low volume of cases, approximately one paediatric case per year at most.

SIH is due to spontaneous cerebrospinal fluid (CSF) leak. Spinal fistulae may particularly be identified in cases in which the leak involves the thoracic spine [29], with postulated

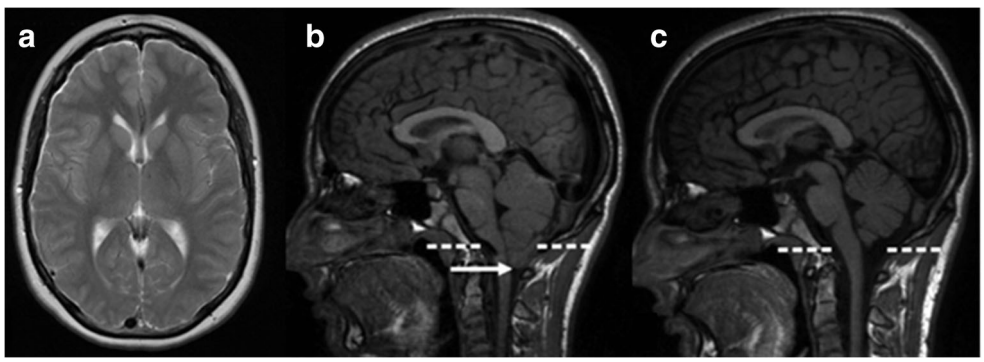

Fig. 2 Acquired Chiari malformation in a 13-year-old boy with a lumboperitoneal shunt for IIH. Magnetic resonance imaging shows bilateral thin convexity CSF subdural hygromas (**a**) and 11-mm tonsillar descent (white arrow) below McRae line (dotted lines) (**b**), with re-ascent of the cerebellar tonsils above the foramen magnum following insertion of a valve into the shunt system (**c**)

causes including minor trauma, weakness of the dural sac, or a combination of both [30–32]. Several studies have indicated an association with connective tissue disorders. Abnormalities in fibrillin-1 synthesis and deposition have been linked to the pathogenesis of SIH, but other extracellular matrix components may be involved [33, 34].

The clinical suspicion of SIH is mainly hinged on the presence of postural headache as this is the most consistent symptom (23 of 24 patients had postural headache in the only reported paediatric series of SIH) [28]. Evidence of low CSF pressure is then required to confirm the diagnosis. This is usually demonstrated by cranial computed tomography (CT) or cranial magnetic resonance imaging (MRI), with radiological features of intracranial hypotension including bilateral subdural hygromas, pachymeningeal enhancement, attenuated prepontine cisterns, narrow ventricles, sagging of the brain, flattening of the brainstem, downward herniation of the cerebellar tonsils (acquired Chiari I malformation), and enlarged cerebral veins or dural venous sinuses (Fig. 3) [2, 28, 35–37]. Around 80% of cases have one or more of these MRI appearances. Sagging of the brain and enhancement of the pachymeninges are the most frequent findings, whilst engorgement of venous structures is a much less consistent feature (around 5% of cases), in the paediatric age group based on the only paediatric SIH series by Schievink et al. [28]. CT myelography is the most reliable method to localize the CSF leak [36].

Chiari-like displacement of the cerebellar tonsils occurs in about 20% of adult patients [2] but may well be an even less consistent radiological feature of SIH in children. To the best of our knowledge, ACM with SIH has only been reported four times in the paediatric age group. The first case was a 12-year-old girl with postural headache, who underwent posterior fossa decompression [38]. Post-operatively, however, symptoms were worse. MR myelography indicated a relevant CSF leak at S1. After epidural blood patching, the headache resolved and cranial MRI demonstrated reversal of the cerebellar tonsil displacement. The second report was published on a 16-year-old boy, also suffering from postural headache [39]. Cranial MRI showed bilateral subdural hygromas, whilst spinal MRI with gadolinium-contrasted views did not reveal a CSF leak. Nevertheless, symptomatic relief was observed after epidural blood patching. The third case was a 15-year-old boy with sacral CSF leak and SIH with ACM and postural headache successfully treated by epidural blood patching. The fourth case was of a 13-year-old girl with postural headache and ACM secondary to SIH due to lumbar CSF leak verified on CT myelography [40]. Epidural blood patch successfully resolved her symptoms and reversed the Chiari.

Conservative management is seldom sufficient, and in the majority of cases, epidural blood patch and percutaneous injection of fibrin glue are the main and effective treatments [41], whilst surgical correction of the CSF leak is reserved for refractory cases [42]. Most of the patients published in the paediatric case series by Schievink et al. required repatching (specific rate unknown), fibrin glue injection (35%), or surgical repair (22%) [28]. Verification that the patch covers its target area may improve outcome especially in difficult-to-treat cases [43]. Following successful treatment, as well as symptomatic relief, resolution of the cranial MRI appearances is seen, including reversal of the acquired Chiari malformation to a normal-lying position of the cerebellar tonsils above the foramen magnum.

Although no foramen magnum decompression procedure has been performed for ACM secondary to CSF hypotension in our institution, the worldwide literature reports several patients with an ACM accompanied by low CSF pressure, and yet symptoms of headache have often been attributed to tonsillar herniation as the primary insult, with many of these patients having subsequently been treated with foramen magnum decompression surgery (with all its attendant risks) without benefit [31, 44, 45]. This highlights the clinical diagnostic difficulty faced by paediatric neurosurgeons and neurologists as well as radiologists, in that the radiological presence of tonsillar descent does not alone secure the primary diagnosis of Chiari 1 malformation, even in the presence of headache. Rather, the precise nature of the symptoms (including orthostatic headache of SIH versus the more classic Chiari cough-induced headache) should mandate a more systematic workup

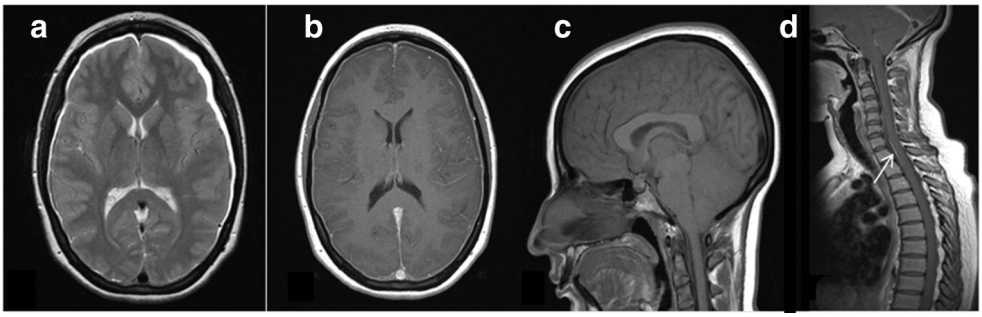

Fig. 3 Acquired Chiari malformation in an 11-year-old girl secondary to SIH. MRI demonstrates bilateral convexity CSF subdural hygromas (**a**), pachymeningeal enhancement (**b**), tonsillar descent of 7-mm below the foramen magnum, (**c**) and evidence of cervico-thoracic CSF leak (white arrow) on T1 post-gadolinium spinal MRI (**d**)

to confirm or rule out the presence of SIH and establish the site of CSF leak as appropriate. Cranial MRI should be undertaken and also used to evaluate Chiari malformation typical bony abnormalities, such as a small posterior fossa. We recommend MRI of the entire spine to reveal any associated spinal anomalies and hydrosyringomyelia [46]. If clinical and radiological suspicion of SIH remains, MR or CT myelography should be initiated. These modalities' sensitively revealed either CSF leaks (50%) or dural ectasia and meningeal diverticula (42%) in Schievink's paediatric series [28]. If, as in the first reported paediatric case of ACM in SIH outlined earlier [38], MR myelography had been part of the pre-operative investigations indicated given the patient's postural headache symptoms, one would unlikely have performed posterior fossa decompression. There remains the caveat however that in a large proportion of cases, the site of CSF leak may not be detected [47–50]. Only after thorough clinical and radiological investigation into ACM and SIH can appropriate treatment be offered, usually epidural blood patching or interventional techniques in the first instance.

Chiari malformation and idiopathic intracranial hypertension

Idiopathic intracranial hypertension (IIH) (previously known as 'pseudotumour cerebri') presents with clinical signs and symptoms of elevated intracranial pressure (ICP), most commonly headaches, and there is no ventricular enlargement or mass lesions on cranial imaging [51]. IIH is therefore diagnosed based on clinical signs and symptoms of elevated ICP, documented high ICP (for example, by lumbar puncture or ICP monitoring), and is commonly associated with papilloedema on formal ophthalmological testing. IIH predominantly affects young overweight women (body mass index > 25). Increased cerebral oedema, disrupted CSF absorption, and abnormal intracranial compliance have all been implicated in studies of IIH [51]. Treatment includes weight reduction, acetazolamide to reduce CSF production, venous sinus stenting to improve CSF absorption, and surgical intervention including CSF shunt surgery.

Tonsillar herniation (acquired Chiari) is frequently seen in the presence of active hydrocephalus—especially in the presence of a posterior fossa mass—and such an association is easily understood. However, there is evidence that Chiari I malformation and IIH may coexist, and the cause and effect in this situation is unclear. Notable historical studies include Banik et al. [3] and Johnston et al. [51], who demonstrated that 11% and 6% of IIH patients had concomitant radiological evidence of Chiari I (tonsillar ectopia ≥ 5 mm), respectively. This represented 16 times and 8 times the incidence of Chiari I in the general population, respectively. A more recent retrospective study by Aiken et al. demonstrated an even higher figure of 21% of IIH patients with radiological Chiari I malformation [52]. However, the precise relationship between Chiari I and IIH is unclear. Raised ICP and brain oedema in IIH may cause downward herniation of the intracranial contents through the foramen magnum corresponding to radiological Chiari I malformation. Alternatively, a subset of Chiari I patients may have abnormal CSF flow at the foramen magnum leading to raised intracranial hypertension, which may only become evident after Chiari decompression. In such cases, a pre-existing or peri-operative change to CSF dynamics may result in an IIH-like condition [51, 53–55]. In children who have undergone posterior fossa decompression surgery for Chiari and still have persistent symptoms (deemed 'failed Chiari' syndrome), measuring ICP may be useful to diagnose high intracranial pressure and suggest a possible therapeutic role of CSF diversion.

For example, a 'Chiari-pseudotumor cerebri syndrome' has been described in adults and children who have failed to improve after Chiari I decompression surgery and who are found to have elevated ICP with small ventricles [53–55]. In children who reported to have coexistence of Chiari I and IIH, recurrence of Chiari I symptoms after Chiari decompression (adequate on post-operative MRI) (failed Chiari) has been reported from 2 months to 3 years after surgery [55]. Case reports of acute presentation of increased intracranial pressure within a week of posterior fossa decompression have also been described, likely due to multifactorial causes of oedema and venous hypertension from surgery.

In Fagan et al.'s retrospective review, 42% (15 out of 36) of failed posterior fossa decompression patients (mostly children) for Chiari I malformation were diagnosed with IIH [55]. If there is adequate decompression as noted on MRI, then a lumbar puncture may be used to measure opening pressures, and also to perform diagnostic and therapeutic CSF drainage to determine whether drainage improves symptoms. Both ventriculoperitoneal and lumboperitoneal shunts are options in patients who show symptomatic relief from the diagnostic lumbar puncture and CSF drainage in this setting. CSF shunt placement generally relieves signs of high ICP such as optic nerve swelling and to a lesser extent headache. Fourteen of the 15 failed Chiari patients with IIH described by Fagan et al. [55] underwent shunt placement (nine children and five adults). Of the 9 children, 7 experienced symptomatic improvement and 2 had moderate improvement. The 5 adults all showed a decrease in optic nerve swelling with no improvement in the headaches. Bejjani et al. [54] reported symptomatic relief in 6 similar adult patients who underwent shunting.

Our own clinical experience supports the literature on altered CSF dynamics with Chiari malformation and following Chiari decompression surgery. At our institution, over the last 13 years (from February 2006 to February 2019), 61 paediatric patients (range, 2–15 years) underwent foramen

magnum decompression (FMD) for Chiari I malformation. Of these 61 cases, five (8%) had hydrocephalus at initial presentation but with ventricles considered not to be large enough for safe endoscopic navigation for third ventriculostomy (ETV, our preferred first choice). These five cases underwent CSF shunting (3 ventriculoperitoneal shunts; 2 ventriculo-atrial shunts indicated for hostile abdomen) with improvement in the hydrocephalus but persistent Chiari, and all five patients subsequently underwent FMD. Fifty cases (82%) underwent FMD with no preceding or post-operative problems with CSF dynamics and who did not require any further procedures. Six cases of Chiari (10%) did require CSF diversion following FMD: three cases of post-operative hydrocephalus, for which one underwent ventriculoperitoneal shunting 8 days after FMD, whilst two underwent ETV both more than 4 years after FMD; one case was complicated by CSF leak and required a lumbar drain for temporary CSF diversion; finally, there were two cases of persistent headache with small ventricles on CT and MRI, but with raised pressure on formal intracranial pressure monitoring and lumbar puncture measurements and who underwent ventriculoperitoneal shunting at 12 months following the original FMD surgery with good symptomatic relief. Our data therefore corroborate wider reports of a complex and not yet fully understood relationship between Chari malformation together with surgical decompression and altered CSF dynamics.

Conclusion

Chiari I malformation is defined radiologically as a displacement of the cerebellar tonsils 5 mm or greater below the basion-opisthion line at the foramen magnum. Acquired Chiari malformation (ACM) can have a very similar radiological appearance with respect to cerebellar tonsillar descent below the foramen magnum, and has been associated with both intracranial hypotension (either secondary to over-drainage of lumboperitoneal shunting or spontaneous CSF hypotension) and idiopathic intracranial hypertension (IIH). In the case of intracranial hypotension, careful clinical assessment, ascertaining postural headache, and thorough radiological assessment for intracranial CSF hypotension can help differentiate ACM from true Chiari I malformation and guide appropriate treatment, usually involving epidural blood patch or interventional techniques, and avoid unwarranted surgery. In the case of IIH and Chiari I malformation, the relationship is complex and not well understood, but certainly, in children who have recurrent symptoms despite adequate posterior fossa decompression surgery (failed Chiari), there is a strong role for ICP monitoring as raised ICP may suggest long-term CSF diversion as a potential therapeutic option.

Compliance with ethical standards

Conflict of interest On behalf of all authors, the corresponding author states that there is no conflict of interest.

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