# Chiari I Malformation and Associated Syringomyelia

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## Introduction

There are few conditions in the entire practice of pediatric neurosurgery that are surrounded by such controversy and that evoke such diversity of opinion than is the case for CIM. This controversy extends through terminology, pathophysiology, indications for treatment and surgical technique. CIM is a common reason for pediatric neurosurgical referral and the challenge for the pediatric neurosurgeon is to try and identify those patients who may benefit from neurosurgical intervention as there are many cases that are asymptomatic, incidentally found, or have symptoms unrelated to the radiological finding. Moreover, once a child is deemed to be symptomatic it is then incumbent upon the neurosurgeon to identify the underlying pathophysiological mechanism that has led to the CIM so that an appropriate intervention can be offered. While foramen magnum decompression has been and remains the mainstay of surgical treatment for CIM, this is not appropriate for all patients and indeed can be deleterious in some.

The aim of this chapter is to try and present a contemporary view of the entity of Chiari I malformation highlighting the areas of ongoing controversy and to try and provide an evidence-based approach to the evaluation of patients and selection of the most appropriate surgical intervention.

## Terminology

The original monograph of Hans Chiari concerns "cerebellar changes caused by hydrocephalus of the cerebrum." The entities he described led to the four tier classification that bear his name, these are summarized in Table 1. A major shortcoming of this nomenclature is the implication that these four entities are part of a spectrum of increasing anatomical and clinical severity, whereas in actuality they are distinct pathological conditions. CIM is typically a focal abnormality comprising prolapse of the cerebellar tonsils, sometimes with distortion of the brainstem at the level of the foramen magnum. By contrast, the other Chiari anomalies are not only associated with more extensive changes in

**Table 1** The Chiari anomalies, diagnostic criteria, and associated malformations

| Chiari type | Essential features | Typical associations |
|---|---|---|
| Chiari I | Cerebellar tonsillar herniation through the foramen magnum. | None |
| Chiari II | A pan cerebral malformation that includes brainstem dysplasia with downward descent of the cerebellum, medulla, pons, and fourth ventricle. | Myelomeningocele |
| Chiari III | Hindbrain malformation with herniation or displacement of the posterior fossa contents (cerebellum, medulla, fourth ventricle). | Occipital or occipitocervical encephalocele |
| Chiari IV | Severe cerebellar hypoplasia, posterior fossa under development and abnormal venous sinuses. | Occipital encephalocele |

the rest of the rhombencephalic derivatives (e.g., descent of the pons, cervicomedullary kinking, and small fourth ventricle in Chiari II) but throughout the entire central nervous system as well. For example, the supratentorial changes that are seen in the context of Chiari II malformation include fusion of the massa intermedia, interdigitation of cortical gyri, and "beaking" of the tectum. The use of terms such as Chiari 0 and Chiari 1.5 while referring to recognizable radiological entities is apt to confuse, as the introduction of these additional "grades" of Chiari tends to propagate the incorrect notion that the Chiari series is a spectrum of anomaly.

Indeed it is perhaps regrettable that the condition we know as CIM has been a part of Chiari's classification. While Chiari types II, III, and IV are indeed true congenital malformations of the central nervous system, the same cannot be said of CIM. Some authors have used the terms "hindbrain hernia" or "cerebellar tonsillar ectopia" in an attempt to emphasize that CIM should be more appropriately considered an acquired *deformation* of the normal hindbrain rather than a true *malformation*. There are a number of lines of argument that can be cited to support this assertion. Firstly, CIM is rarely seen in the newborn; when it does appear in early childhood, this is most likely due to a discrepancy between postnatal growth of the cerebellum and that of the posterior cranial fossa. Secondly, histological analysis of the cerebellar tonsils in CIM is usually normal, perhaps with areas of pressure necrosis or gliosis rather than the neuronal and nuclear disorganization that have been described, for example, in Chiari II malformation. Thirdly, the morphology of the hindbrain and tonsil in CIM will often normalize following foramen magnum decompression, and there are also numerous examples in the literature that demonstrate the reversibility of cerebellar tonsillar ectopia following treatment of the underlying cause.

While the term CIM will continue to be used in this chapter, the reader should perhaps consider that CIM is generally an acquired and potentially reversible anomaly while the other Chiari malformations are congenital and irreversible.

## Defining Chiari I Malformation

There is no agreed definition of CIM. The most commonly cited definition is decent of the cerebellar tonsils of at least 5 mm below the foramen magnum, though definitions range from 0 mm (tonsils at the level of the foramen magnum) to 1.5 cm. Defining CIM by measurement is not only arbitrary but potentially misleading with a risk of both overdiagnosis and underdiagnosis. The reasons for this are as follows: the plane of the foramen magnum is not flat but ellipsoid or "saddle shaped" thus making it difficult to establish an appropriate point of reference. Additionally, since the tonsils are para median structures and yet measurements are usually taken from a midline sagittal MRI sequences, this measure will not necessarily reflect the true extent of tonsillar descent. A further complicating factor is the variability in the level of the cerebellar tonsil found in the normal population. Barkovitch et al. found that in 14% of normals the tonsils were at, or below the level of the foramen magnum (Barkovich et al. 1986). Finally, there is no clear correlation between the extent of tonsillar herniation and the likelihood of symptoms or syrinx formation, indeed the term 'Chiari 0' was introduced to account for those patients with syrinx but with cerebellar tonsils at or above the foramen magnum whose syrinx improved after a Chiari operation (Chern et al. 2011).

Therefore, including a numerical measure in the definition of CIM is not only imprecise, but it lacks clinical relevance and thus should be abandoned. A more useful definition of CIM is that of a functional obstruction at the foramen magnum by the inferior cerebellum, one that may result in neuraxis compression and/or obstruction to CSF flow across the craniovertebral junction. Such a definition obviates the need for a measurement and reflects the fact that there are other mechanisms that might result in compromise of this region, for example, local scarring of the subarachnoid spaces or bony malformation at the craniovertebral junction. Thus Chiari I, Chiari 0, and Chiari 1.5 would all be included under such a scheme, each representing some degree of acquired obstruction at the level of the foramen magnum.

## Pathophysiology

### Craniocerebral Disproportion

The traditional view has been that CIM occurs as a result of disproportionate growth between the posterior cranial vault and its contents, such that the cerebellum exceeds the confines of the posterior fossa and herniates into the upper cervical spinal canal (Fig. 1). A number of morphometric studies have suggested a correlation between small posterior fossa volumes and CIM in children. Moreover, experimentally inducing a small posterior fossa by administering vitamin A to hamsters has been shown to result in hindbrain herniation (Marin-Padilla and Marin-Padilla 1981); this is commonly cited as experimental evidence to support the craniocerebral disproportion hypothesis. The association between growth hormone deficiency and CIM (Tubbs et al. 2003) and between rickets and CIM (Tubbs et al. 2004) provides further clinical evidence corroborating the hypothesis that reduced cranial growth might be an important etiological factor in pathogenesis of hindbrain herniation (Fig. 2).

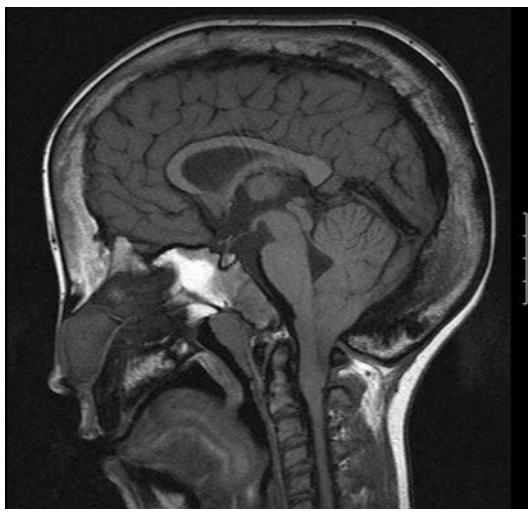

**Fig. 1** Sagittal T1 weighted MRI scan demonstrating hindbrain herniation secondary to craniocerebral disproportion in a case of craniometaphyseal dysplasia. There is extensive thickening of the cranial vault compromising the size of the posterior fossa

However, a number of studies have failed to demonstrate a consistent association between PFV and CIM. Sgorous et al. found no difference in PFV between cases of isolated CIM compared with control though they did find a reduced PFV in cases of CIM associated with syringomyelia, the implication being that these two clinical scenarios (CIM with and without syrinx) may stem from different underlying pathophysiological mechanisms (Sgouros et al. 2006). Some caution needs to be exercised in interpreting volumetric data since the calculation of PFV is not straightforward, the majority of these studies have relied on estimates extrapolated from linear measurements, such linear measures have subsequently been found to correlate poorly with PFV (Bagci et al. 2013).

What now appears to be clear is that there is no simple correlation between PFV and tonsillar descent. While in many cases the traditional treatment practice of enlarging the posterior fossa through foramen magnum decompression might be appropriate, it is essential that other pathophysiological mechanisms should be borne in mind that might be better addressed through other means.

### Raised Intracranial Pressure

Chiari's original monograph describing the malformations that bear his name is entitled "On Cerebellar Changes Caused by Hydrocephalus of the Cerebrum." This clearly indicates that he believed that the pressure generated by hydrocephalus was an important etiological factor in the cases that he had encountered and indeed hydrocephalus is reported to be present in approximately 10% of pediatric patients with CIM. However, it requires a detailed clinical history, sometimes supplemented by fundoscopy and intracranial pressure monitoring in order to differentiate between active hydrocephalus and stable, normotensive ventriculomegaly. Of course the majority of cases of hydrocephalus do not lead to hindbrain herniation and, even where "active" hydrocephalus is deemed to be present it can be unclear as to whether the hydrocephalus is primary, resulting in cerebellar tonsillar descent

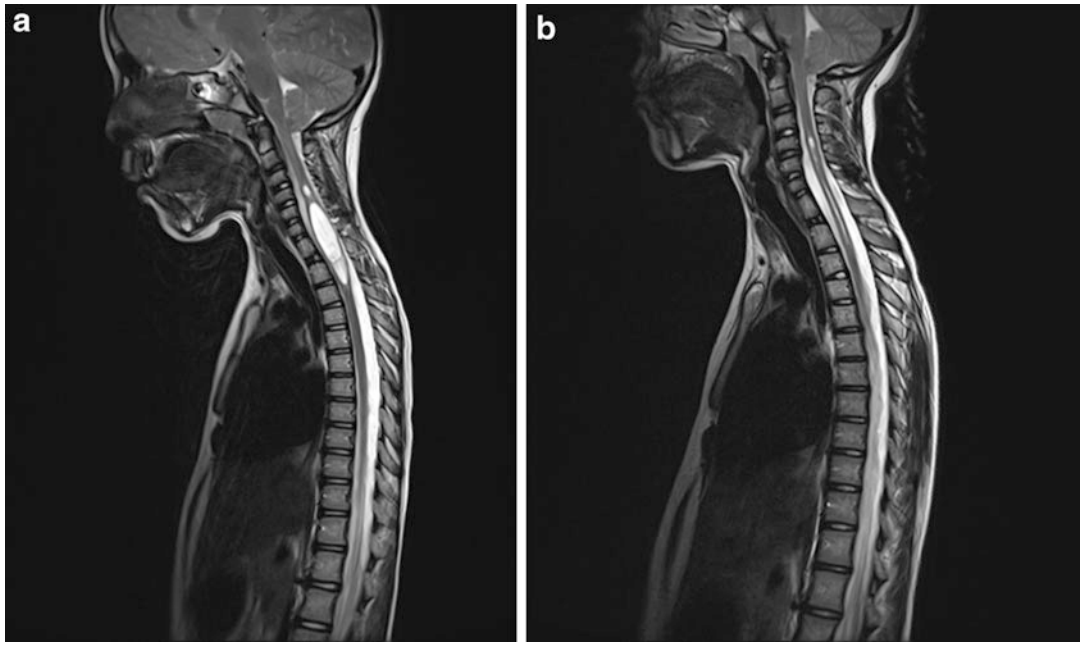


**Fig. 2** Sagittal T2 weighted spinal MRI scans showing CIM and syringomyelia in a girl with vitamin D deficiency before (**a**) and after (**b**) vitamin D replacement

through hydrostatic pressure or whether it is in fact secondary, due to obstruction of CSF outflow from the fourth ventricle. Most neurosurgeons would concur that while the interplay between ventriculomegaly and CIM is complex, where there is evidence of hydrocephalus this should be treated in the first instance (Di Rocco et al. 2011). Whether the hydrocephalus is the cause or consequence of the CIM, treatment by means of endoscopic third ventriculostomy is gaining popularity as the preferred modality of hydrocephalus management in this context. In addition to controlling the symptoms and signs of raised ICP, reversal of the tonsillar herniation and associated syringomyelia is frequently seen following endoscopic third ventriculostomy, thus obviating the need for foramen magnum decompression (Hayhurst et al. 2008) (Fig. 3).

It is now well established that it is not only hydrocephalus but also many other causes of raised ICP that may result in hindbrain herniation. In vein of Galen malformation for example, the engorgement of the brain parenchyma due to raised venous pressure results in extrusion of the cerebellar tonsils through the foramen magnum; moreover, this phenomenon typically reverses following successful treatment of the vascular anomaly (Girard et al. 1994). In cases of idiopathic intracranial hypertension, prolapse of the cerebellar tonsils (sometimes with classical "pointed" appearance) occurs in as many as 20% of cases (Aiken et al. 2012). Fagan et al. found that in a retrospective study of 192 patients who had undergone posterior fossa decompression for CIM, 36 did not improve; of these 41.6% were found to have a pseudotumor syndrome that responded well to CSF diversion (Fagan et al. 2006).

The instances described above support the hypothesis that, at least in some instances a positive pressure differential across the foramen magnum can result in cerebellar tonsillar descent, thus supporting a role for raised ICP in the etiology of CIM. By contrast, it is also recognized that inducing a pressure differential, by reducing intraspinal pressure can also lead to the same effect, as has

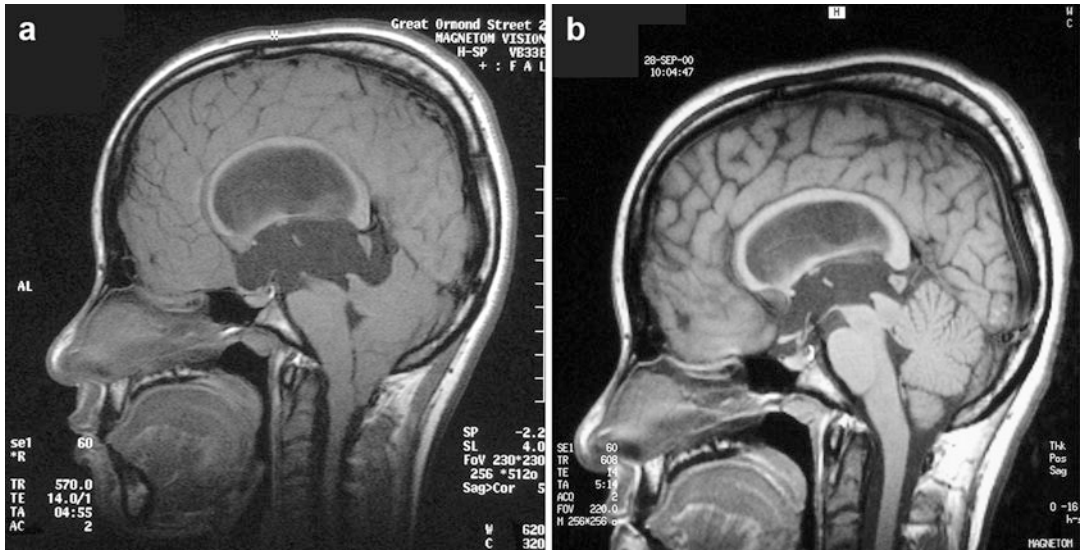


**Fig. 3** Sagittal T1 MRI scan demonstrating CIM secondary to hydrocephalus (**a**). Following endoscopic third ventriculostomy there is improvement of the CIM (**b**)

been demonstrated following lumboperitoneal shunt placement. Reports of tonsillar herniation secondary to spontaneous spinal CSF leak, for example, in association with dural ectasia and Marfan syndrome most likely represent the same mechanism (Puget et al. 2007).

## Craniovertebral Malformation

Until relatively recently, attention had focused on reduced posterior fossa volume as a principle mechanical factor in the etiology of CIM (see above). However, in the past 20 years or so there has been a shift of emphasis toward the role of craniovertebral junction malformation and instability. Not infrequently, anomalies of segmentation and bone maturation will compromise not only the space available for the posterior fossa contents, leading to herniation of the hindbrain structures, but may also cause the craniovertebral axis to become more angulated resulting in basilar invagination, which, in itself may cause ventral compression of those same structures. Furthermore, instability of the craniovertebral junction, either secondary to bone malformation or as a result of connective tissue disorders such as Ehlers Danlos syndrome, is now well documented in cases of CIM providing an additional mechanism for the development of symptoms and neurological deterioration. The true incidence of clinically significant basilar invagination and instability in pediatric CIM patients is difficult to ascertain since the diagnostic criteria for these phenomena are imprecise; many published series inevitably have an inherent bias due to the subspecialty experience of the author. In one of the largest pediatric CIM series to be published only 4 of 500 procedures (0.8%) went on to require ventral decompression and occipitocervical stabilization (Tubbs et al. 2011), whereas in another series of 101 cases, occipitocervical fixation was required in 19% and transoral decompression in 3%. In this later series, failure to recognize the significance of craniovertebral anomalies was cited as a major cause of treatment failure following simple posterior fossa decompression (Bollo et al. 2012).

Assimilation of the atlas is the commonest single bony anomaly to occur in the context of CIM and is associated with reduced height of the posterior fossa and reduced posterior fossa volume. Menezes identified atlas assimilation in more than 550 patients with craniovertebral junction anomalies, and in this group the incidence of hindbrain herniation was 38% (Menezes 2008).

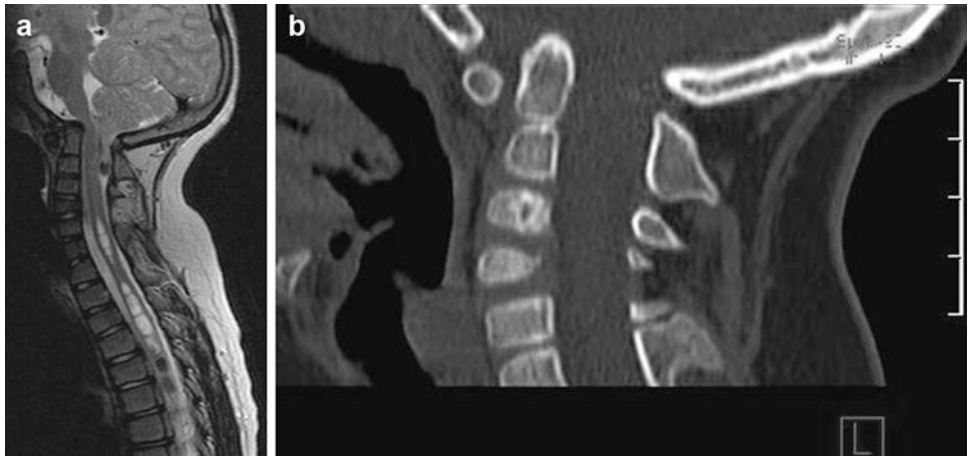


**Fig. 4** A case of CIM and syringomyelia secondary to craniocervical junction segmentation anomaly. The sagittal T2 MRI scan of the spine (**a**) shows the CIM and syrinx. Sagittal reformatted CT scan (**b**) of the craniocervical junction illustrates the associated bony malformation with translocation of the odontoid peg and cervical segmental fusions

Congenital fusions of the upper cervical vertebrae (particularly between C2 and C3) frequently coexist with occipito-atlantal assimilation, a situation that is apt to result in atlanto-axial instability since the atlanto-axial joint in such circumstances becomes the sole motion segment of the craniovertebral articulation. With growth of the child there is a slow upward migration of the axis and flattening of the clivus leading to basilar invagination and ventral compression of the neuraxis. Menezes suggested that early in the evolution of this process the deformity is often reducible and can be successfully treated by traction, posterior decompression, and stabilization, while in older children and adults the deformity becomes irreducible, increasing the likelihood of ventral decompression being required (Fig. 4).

Where CIM exists alongside craniovertebral anomaly, it can be difficult to ascertain whether simple posterior decompression may suffice, or whether this may need to be supplemented by occipitocervical fixation or even ventral decompression. A number of studies have tried to identify objective and reproducible measures of ventral compression and basilar invagination that might aid in the decision-making process. Smoker described the clivus-canal angle, the angle formed between the slope of the clivus and the long axis of the odontoid. The normal value lies between 150 and 180 (Smoker 1994). A value $<150$ deg implies pathological, kyphotic deformity of the craniovertebral junction and has been used as a measure of basilar invagination. The clivus-canal angle is however somewhat position dependent and therefore has poor reproducibility; furthermore, it relies purely on bone morphometry and therefore fails to take account of the effect that this angulation may have on the neuraxis. In an attempt to overcome these shortfalls, Grabb et al. described the "pB-C2 line" as a more useful discriminator of the likely need for ventral decompression surgery (Grabb et al. 1999). This line is constructed as follows (Fig. 5): a line B-C2 is drawn from the basion to the infero-posterior aspect of the C2 vertebral body, a second line pB-C2 is then drawn perpendicular to B-C2 to the odontoid tip, where this meets the dura. Where pB-C2 is less than 9 mm ventral decompression is not warranted. The Grabb line however was specifically used to describe the extent and therapeutic implications of ventral compression; it was not intended as a measure of the need for posterior stabilization. In children, even where there is clinically significant ventral compression, this can usually be addressed by traction, either preoperatively or intraoperatively, followed by posterior decompression and occipitocervical fixation without recourse to a ventral decompression

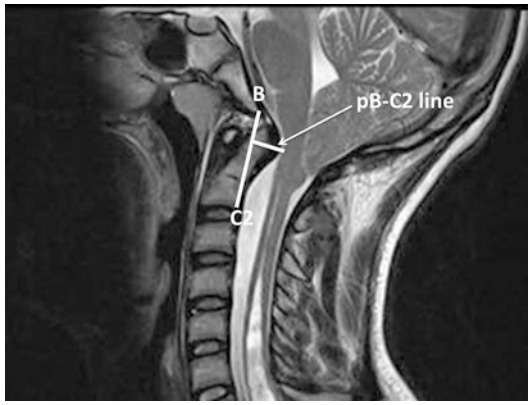


**Fig. 5** Sagittal T2 weighted MRI scan showing a case of "Complex CIM" with retroversion of odontoid peg. Calculation of the Grabb line (pB-C2) is demonstrated

procedure. In fact, it is important that patients are not subjected to anterior decompression until irreducibility has been verified.

The clinical presentation of CIM patients who ultimately require craniovertebral fixation shows some differences when compared with more "typical" CIM patients. Those CIM patients requiring fixation have an increased incidence of impaired gag reflex, downbeat nystagmus, and upper extremity sensory changes or motor weakness (Fenoy et al. 2008). In a more recent study, Bollo et al. sought to identify what combination of radiological factors might predict which patients with CIM would likely require occipitocervical fixation. Factors that they found to be significant, after univariate analysis were "complex CIM," i.e., with descent of the obex as well as the tonsils below the plane of the foramen magnum (referred to as Chiari 1.5 by some), basilar invagination, and clivo-canal angle $<$125 deg (Bollo et al. 2012).

## Craniosynostosis

The occurrence of hindbrain herniation in the context of syndromic craniosynostosis has been recognized for many years. This concurrence represents a particularly complex form of CIM in which there are multiple, interdependent pathogenetic mechanisms involved; these include skull base deformity, hydrocephalus, and venous hypertension (Cinalli et al. 1995). The relative contribution of each of these mechanisms may vary between syndromes and indeed may change during the child's life as a consequence of growth and surgical interventions (Thompson et al. 1997).

The incidence of CIM among the craniofacial syndromes is by no means evenly distributed. CIM is almost ubiquitous in Kleeblatschadel (cloverleaf skull) and extremely common in Crouzon syndrome (70%) and Pfeiffer syndrome (50%) but distinctly unusual in Apert syndrome ($<$2%). These differences are attributed to the pattern of closure of the skull base sutures, in particular the lambdoid sutures (Fig. 6). In Crouzon syndrome, these sutures tend to close in infancy resulting in a small posterior fossa and distorted basiocciput at a time of significant postnatal cerebellar maturation. This leads to a situation of cranial-cerebral disproportion and thus hindbrain herniation. By contrast, in Apert syndrome these basal sutures remain patent for longer; as a result the basioccipital region and posterior fossa are normal (or sometimes larger than normal) and thus do not typically constrain growth of the posterior fossa contents (Cinalli et al. 2005).

In addition to its effect on skull base growth, closure of the basal sutures also leads to anomalies of cranial venous drainage. In one angiographic study, 18/23 patients with craniosynostosis had either complete or more than 50% occlusion of one or both sigmoid/jugular sinuses. The intracranial venous hypertension induced by these changes, and frequently exacerbated by chronic $CO_2$ retention due to upper airways obstruction, leads to a state of brain "turgor." The consequences of this will depend upon whether or not the skull can expand. Raised venous pressure impairs CSF reabsorption, and where skull growth is constrained by fusion of the calvarial sutures the state of high brain turgor persists with normal ventricular size, a situation that has been likened to idiopathic intracranial hypertension. However, where skull expansion can occur (if the calvarial sutures and fontanelles are patent, or following cranial vault expansion) the restriction to ventricular enlargement is not present and progressive ventriculomegaly (hydrocephalus) as

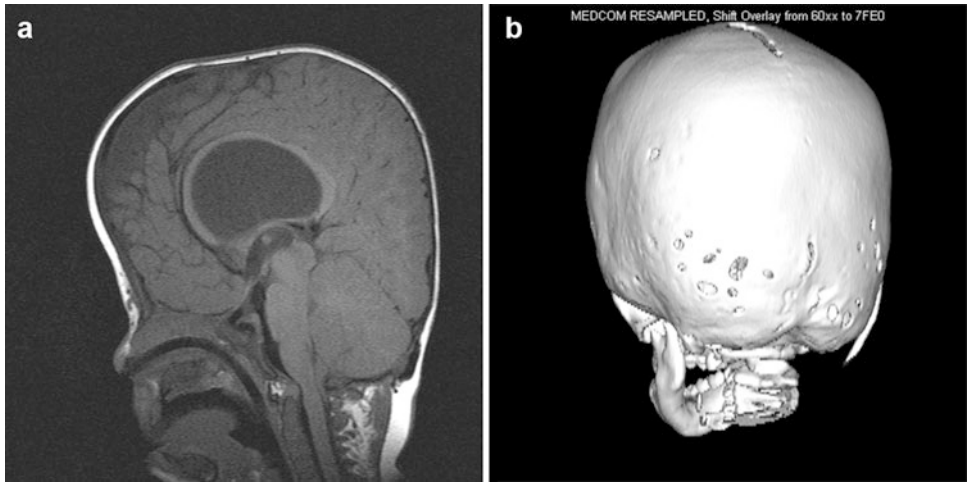


**Fig. 6** Sagittal T1 weighted MRI scan showing CIM in a case of Pfeiffer syndrome (**a**). The 3D CT scan (**b**) confirms closure of the posterior cranial vault sutures

well as progressive hindbrain herniation can ensue (Thompson et al. 1997). The development of collateral venous drainage may eventually compensate and restore the *status quo*, though throughout early childhood, managing this precarious interaction of ICP, hydrocephalus, cranial vault deformity, and hindbrain herniation can be exceedingly problematic. This underscores the importance of experienced neurosurgical input to the multidisciplinary care of these cases.

While the association between CIM and craniosynostosis is most commonly seen and best documented for syndromic cases, there is an increasing recognition of an association between nonsyndromic or monosutural synostosis and CIM (Fig. 7). Leikola et al. reported an incidence of 5.6% of C1M in a series of 124 patients with nonsyndromic craniosynostosis (Leikola et al. 2010) though none of these were associated with Chiari symptoms. Strahle described similar incidence in sagittal (2.8%) and coronal (6.3%) synostosis but, in keeping with the experience from syndromic cases, a much higher incidence in isolated lambdoid (55.6%) and multisuture (35%) synostosis (Strahle et al. 2011a). In this latter series, calvarial vault remodeling was associated with subsequent reduction in the extent of tonsillar herniation in six out of seven cases. This observation has been previously documented and has led to the suggestion that, in selected cases, posterior cranial vault expansion might be the initial procedure of choice for treatment of CIM, in preference to foramen magnum decompression.

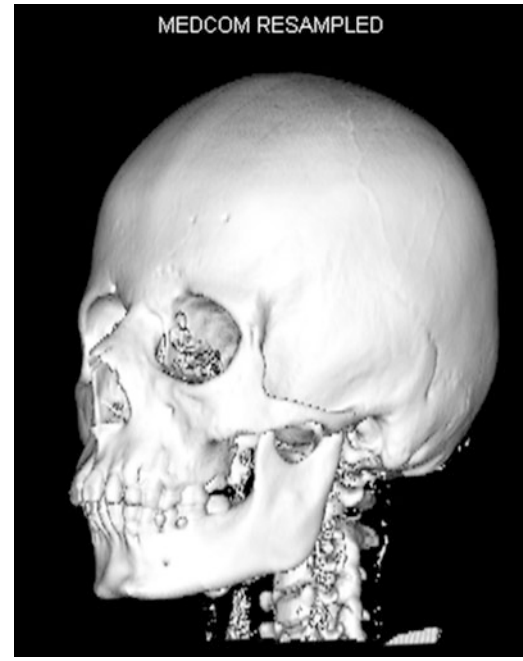


**Fig. 7** 3D CT scan showing pan synostosis in a child with nonsyndromic craniosynostosis discovered during the evaluation of CIM

## Chiari Associated Syringomyelia and Scoliosis

### Syringomyelia

Although as many as two-thirds of children with CIM have a syrinx, it is unclear why some children with CIM develop a syrinx while others do not. Age may be a factor as young children, less than 5 years, are much less likely to have syringomyelia than older. The extent of tonsillar descent, as a predictor of syringomyelia has been variably reported. However, in a recent large study that included over 500 children with CIM, there was a strong correlation between level of the tonsils and the presence of a syrinx (Strahle et al. 2011b). Moreover, in this study pegged (pointed) tonsils were more likely than rounded tonsils to be associated with syrinx, and the combination of low, pegged tonsils was associated with more severe alterations of CSF flow at the foramen magnum.

The pathophysiological basis of syringomyelia secondary to CIM has been extensively debated. The most commonly accepted model is that proposed over 20 years ago by Oldfield (Oldfield et al. 1994), he combined observations from intraoperative ultrasonography and phase contrast MRI to explore CSF flow and tonsillar motion at the foramen magnum. The functional obstruction to CSF flow at the foramen magnum, caused by the tonsillar ectopia in this scenario though could equally apply to scarring, tumor, or other obstructing mechanism, means that the normal physiological pressure waves that are generated along the spinal CSF pathways (due to Valsalva, coughing, etc.) cannot be dissipated into the cranial compartment; the result is that CSF is forced along the perivascular spaces into the spinal cord. Ultimately, this fluid results in cavitation within the parenchyma of the spinal cord, which, in the presence of repeated pressure waves through the CSF compartment propagates the syrinx.

Decompression at the foramen magnum equilibrates the pressure gradient across cervicomedullary junction, thus disabling the filling mechanism for the syrinx (Fig. 8). In this model, the syrinx fluid is therefore derived from without, rather than from within the spinal cord as had been predicted by previous theories. This explanation is not universally accepted; it has also been proposed that the pressure gradient between the spinal cord and the subarachnoid space is in fact in the opposite direction, the pressure wave originating within the spinal cord, resulting in distension of the spinal

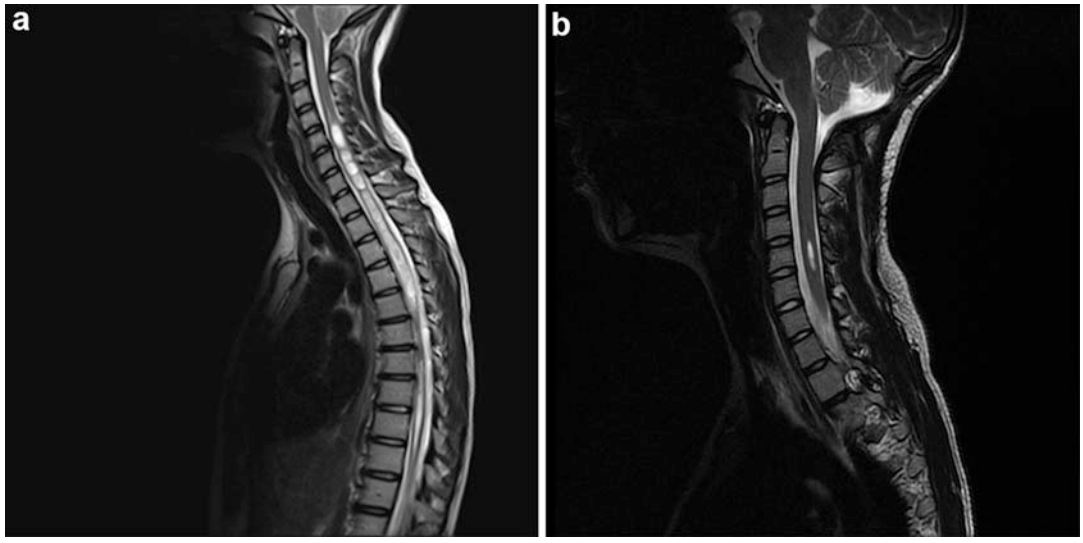


**Fig. 8** Sagittal T2 weighted MRI scans of CIM associated with syringomyelia (**a**). Following foramen magnum decompression with duraplasty there is evidence of improved CSF around the cervicomedullary junction with resolution of syringomyelia (**b**)

cord and an egress of fluid from the microvascular compartment into the spinal cord parenchyma (Greitz 2006). Greitz invokes the Bernoulli principle to explain why the spinal subarachnoid pressure is in fact lower adjacent to a spinal cord syrinx. This principle states that an increase in fluid velocity (as might occur where the subarachnoid space is narrowed by cerebellar tonsils, scarring, etc.) is accompanied by a decrease in the pressure exerted by that fluid. The transient drop in CSF pressure creates a pressure gradient from spinal cord parenchyma to the subarachnoid space with the result that fluid is drawn from the capillary bed into the parenchyma initiating the syrinx.

### Scoliosis

Scoliosis secondary to CIM associated syringomyelia is particularly common in children compared with adults, and this is likely to reflect the vulnerability of the growing spine to asymmetric neuromuscular influences. Although the precise pathophysiology of syrinx associated scoliosis is incompletely understood, it is claimed that the syrinx cavity (which is rarely symmetrical in the axial dimension), interferes with the function of the anterior horn cells of the spinal cord; this results in an imbalance of the innervation to the axial musculature and thus postural change. In experimental animal models, sectioning the dorsal roots can be shown to cause scoliosis, a finding that lends some credence to this hypothesis, although the effect of paraspinal muscle dissection and laminectomy on the subsequent development of scoliosis in these animal models are confounding factors in such studies (Alexander et al. 1972).

While clinical experience supports the causal sequence of CIM leading to syrinx and thence scoliosis, a not infrequent clinical scenario is CIM with scoliosis in the absence of syringomyelia. Here, the etiological role of the CIM in the genesis of the scoliosis is much less clear, it has been suggested that asymmetric pressure on the spinal cord at the level of the foramen magnum might be a potential mechanism, though this would seem unlikely. In a recent large study, designed to address this question, Strahle et al. concluded that Chiari I was not independently associated with scoliosis (Strahle et al. 2015). Currently, there is no evidence to support preemptive neurosurgical intervention in this situation.

## Clinical Presentation

The list of symptoms and signs that have been attributed to CIM is extensive; moreover, many of these are common neurological complaints, and it requires clinical judgment and experience, frequently supplemented by further investigations to ascertain whether or not such symptoms are indeed referable to the radiological finding of cerebellar tonsillar ectopia. Additionally, it is important to appreciate that age has a significant impact on the clinical presentation of CIM; infants, school aged children, and adolescents each having somewhat differing patterns of symptoms (Table 2).

Clinical presentation can be considered under the following headings: classical, atypical, and incidental.

## Classical

There are, in essence, two modes of "classical" presentation of CIM in childhood; these can be

**Table 2** Symptoms, signs, and associated disorders associated with Chiari I according to age at presentation

| 0–3 years | 4–10 years | >10 years |
|---|---|---|
| Oropharyngeal dysfunction | Headache/neck pain | Headache (Valsalva induced) |
| Vomiting/gastro-esophogeal reflux | Ataxia | Scoliosis and syrinx |
| Stridor, sleep disordered breathing | Craniocervical junction anomaly | Sensorimotor disturbance |
| Craniosynostosis | Vomiting | Extraoccular movement disorder |
| Irritability | Reduced gag reflex | |
| Hydrocephalus | | |

linked to the underlying pathophysiological basis of the condition, namely local compression and CSF obstruction, of course aspects of both may present in the same individual.

## Local Compression

Headache is perhaps the most consistent presenting symptom of CIM; it is present in up to 70% of cases and is thought to be due to impaction of the cerebellar tonsils within the foramen magnum. Typical CIM headache characteristics are a posterior occipital or suboccipital location, with exacerbation on coughing, sneezing, or any Valsalva type maneuver, for example, playing wind instruments or weight training. The headaches tend to occur in paroxysms, of relatively short duration, lasting less than 5 min (Headache Classification Committee of the International Headache Society (IHS) 2013). By contrast, chronic headache or headache localized to the frontal or temporal regions is much less likely to be due to tonsillar ectopia, and other headache etiologies should be considered, such as tension headache and migraine.

Local compression may also result in bulbar symptoms including ocular movement disorders such as nystagmus, swallowing difficulties with recurrent aspiration or choking, and sleep apnea. Sleep apnea syndrome is uncommon in childhood, with a prevalence of 1–3%; however, it is found in as many as 60% of children with CIM (Dauvilliers et al. 2007). Sleep disordered breathing may take the form of hypoventilation, central or obstructive breathing patterns. Central breathing disturbance is thought to be due to direct medullary compression, whereas the obstructive component is considered to be due to impaired vocal cord movement or poor pharyngeal muscle tone secondary to lower cranial nerve impairment.

Young children have a greater propensity to have a bulbar presentation; in one study, 77% of children with CIM who were under 2 years at presentation presented with oropharyngeal dysfunction, comprising such symptoms as reflux, vomiting, and choking (Albert et al. 2010). CIM occurring in the context of craniovertebral malformation, where there is commonly ventral compression from a retroflexed dens or basilar invagination, is also more likely to present with bulbar symptoms, in particular lower cranial nerve palsies, including hearing loss. Cerebellar symptoms and signs, other than nystagmus, are surprisingly unusual in CIM.

## CSF Obstruction

It is obstruction to CSF flow at the foramen magnum that appears to be the initial event in the development of Chiari associated syringomyelia. The clinical presentation of CIM in cases with syringomyelia will accordingly be modified, with a propensity to symptoms of spinal cord dysfunction. In one study, specifically looking at children with CIM and syrinx the presentation included skeletal deformity, mainly scoliosis (71%), sensory disturbance (24%), and motor weakness (6%) (Isu et al. 1990).

The combination of CIM, syringomyelia, and scoliosis is particularly well recognized in pediatric practice. In a series of 500 case of pediatric CIM reported by Tubbs et al., a syrinx was present in 57% and scoliosis in 18% (Tubbs et al. 2011). The incidence of scoliosis in syringomyelia patients is particularly high in children. In a review of 28 patients, Isu et al. identified scoliosis in 82% of patients aged less than 20 years of age and only in 16% of older patients with syringomyelia (Isu et al. 1990). Particular features that alert the orthopedic surgeon to the potential for underlying spinal cord pathology in children with scoliosis include male sex and left thoracic curves (idiopathic scoliosis is more commonly right sided and has a predilection for girls). The presence of neurological symptoms and signs and rapid progression of a curve are also features suggestive of a neurogenic etiology. Spinal MRI is therefore routinely indicated in the evaluation of the child with scoliosis.

## Atypical

Rare and atypical presentations of CIM include hiccups, drop attacks, and ataxia. Recurrent vomiting and dysarthric speech may also be seen. It is exceptional however for these atypical symptoms to be the sole presenting features of CIM. Before attributing these atypical symptoms

to a finding of Chiari additional investigations (see below) should be considered in an attempt to localize such symptoms to the lower brainstem. Scoliosis occurring in the presence of CIM but without syrinx should also be considered an atypical presentation, the evidence to treat the CIM in this scenario is much less than when syringomyelia is also present.

### Incidental

The widespread use of MRI in the evaluation of diverse neurological symptomatology has resulted in increased diagnosis of CIM; this is particularly so in the pediatric population where the prevalence of low cerebellar tonsils is greater compared with adults. In the majority of these cases, the finding of CIM is likely to be incidental and unrelated to the reason for the MRI. In a study of over 14,000 MRI scans in children, Strahle et al. identified CIM (tonsils >5 mm below the basion-opisthion line) in 509 cases (3.4%). Of those with CIM 70% were deemed to be asymptomatic (Strahle et al. 2011b). This not infrequently results in unnecessary anxiety for patients and parents and an increased workload for the pediatric neurosurgeon who has to council these victims of modern imaging technology (VOMIT).

## Investigation of CIM

### MRI

Multiplanar MRI with T1 and T2 weighted sequences is the imaging modality of choice in the evaluation of the child with CIM. In addition to assessment of the craniovertebral region, it is essential to look for associated cranial, cerebral, and spinal abnormalities and therefore the entire neuraxis should be imaged. In the majority of cases conventional MRI will be sufficient to establish the diagnosis and form a basis for surgical management; however, given the poor correlation between radiological findings and clinical symptoms of CIM, there has been a trend toward the use of more sophisticated imaging techniques in an attempt to better characterize the local mechanical and hydrodynamic consequences of low lying cerebellar tonsils.

### Advanced MRI Techniques

MRI-based studies of CSF flow, tonsillar motion, and even axoplasmic flow along local white matter tracts are among the advanced MRI techniques that some centers are now incorporating into their CIM evaluation protocol.

#### Phase Contrast MRI

Phase contrast MRI is a flow sensitive sequence that measures the changing signal intensity from moving protons (water in CSF). During cardiac systole, the delivery of the extra volume of blood to the intracranial compartment causes a rise in intracranial pressure (ICP); this is responsible for the initial peak or percussion wave (P1) in the normal ICP waveform. Under normal circumstances this wave is dampened, due to bulk flow of CSF from the intracranial compartment into the spine across the foramen magnum (in keeping with the Monroe Kellie doctrine). In phase contrast MRI, this CSF motion is "encoded" as changes in MR signal intensity such that downward flow of CSF gives rise to the bright signal of CSF motion. During diastole, the flow is in the reverse direction, encoded as dark signal on cine MRI. By selecting an area of interest the signal changes can be "decoded" to provide a quantitative measure of CSF flow velocity.

When there is obstruction at the level of the foramen magnum, the velocity, amplitude, and pulsitility of the CSF flow are altered. There are differing patterns of altered CSF flow that are observed in CIM. In some patients, velocities are increased and in others, nonuniform or even reduced velocities are seen. Moreover, changes in flow are frequently different anterior to the cervicomedullary junction compared with posterior. That improvements in MRI flow studies can be demonstrated following surgery, and that these can be retrospectively correlated with clinical outcome has been demonstrated in a number of

studies (McGirt et al. 2006). It should be mentioned however that the preoperative interpretation of the flow changes demonstrated by phase contrast imaging and extrapolating these findings to surgical decision making is not always straightforward and thus far there has been no claim that this studies can predict which patients are most at risk for future deterioration. Perhaps more importantly is the potential for such studies to identify those patients, with ostensibly normal or satisfactory CSF flows who are not likely to benefit from surgery and who can be safely observed, and also in investigating those patients who have early or late failure from surgery who are being evaluated for second look surgery.

### Cine MRI

Fast imaging employing steady state acquisition (FIESTA) is a technique in which the MRI signal acquisition is synchronized or "gated" to the cardiac cycle and or respiratory cycle. FIESTA has been used to study movement of the hindbrain structures, specifically the cerebral tonsils and medulla. Oscillation of the tonsils has been shown to be somewhat greater in CIM patients compared with controls (Cousins and Haughton 2009) although the sensitivity of this technique as a means of patient selection for surgery has yet to be proven.

### Diffusion Tensor Imaging (DTI)

A more recent application of advanced MRI techniques in the evaluation of CIM patients has been DTI. This quantitative technique measures directional diffusion of water through white matter tracts (fractional anisotropy). Axial diffusivity through the middle cerebellar peduncle was shown, in one study to be significantly different between CIM patients and controls (Eshetu et al. 2014), furthermore improvements in brainstem diffusivity have recently been demonstrated following surgical decompression (Krishna et al. 2016).

## Respiratory Sleep Studies

Given the high prevalence of sleep disordered breathing in children with CIM and, in recognition of the potential long-term morbidity associated with this condition, respiratory sleep studies (nocturnal polysomnography recordings) have found an increasing role in the investigation of children with CIM (Dhamija et al. 2013). Improvement in sleep apnea and disordered breathing following surgical decompression is now recognized in the pediatric population, and this presentation of CIM should be considered as an indication for treatment.

## Evaluation of Bulbar Function

Assessment of ocular movements and fundoscopy should be part of the initial outpatient assessment of the child with CIM; this may need to be supplemented by formal ophthalmological assessment. On rare occasions, other means of investigating potential bulbar symptoms due to Chiari may be indicated, for example, bulbar EMG and video swallow.

## Intracranial Pressure Monitoring

Before embarking upon posterior fossa decompression, it is important to establish that there is no associated intracranial hypertension that might be precipitating the CIM. Hydrocephalus will usually be evident from the brain imaging; however, tonsillar herniation, secondary to raised intracranial pressure with normal sized ventricles may occur in idiopathic intracranial hypertension and also in craniosynostosis. A finding of raised ICP will mean that alternative treatment strategies should be considered since posterior fossa decompression in such circumstances can be deleterious. Where there is doubt a period of intracranial pressure monitoring should be considered.

## Treatment

Once a diagnosis of CIM has been made, it is exceedingly likely that this will prompt neurosurgical referral. Since up to two-thirds of childhood cases of radiologically diagnosed CIM are asymptomatic, the neurosurgeon is then faced with a dilemma, whether to proceed with prophylactic surgery or to pursue a nonoperative course of action.

## Nonoperative Management

During the past two decades there have been a number of surveys of neurosurgical practice in respect of surgical attitudes to the prophylactic treatment of CIM and syringomyelia. Pediatric neurosurgeons have been presented with hypothetical scenarios, and their opinions to observe or treat have been sought. In a survey from 2000, only 9% of the respondents favored prophylactic surgery for CIM and syringomyelia, preferring instead a period of observation and radiological surveillance (Haroun et al. 2000). In 2004, another survey found that while only 8% of respondents would offer surgery for an asymptomatic patient with CIM only, 75% would offer surgery to an asymptomatic child with CIM and syrinx (Schijman and Steinbok 2004). A more recent survey, also with a scenario of CIM and syrinx, reported that 85% of respondents would advocate prophylactic surgery (Rocque et al. 2011). While such surveys have inherent methodological limitations and comparisons between them should be made with caution, the implication is that neurosurgeons perceive the natural history of CIM with syrinx to be worse than without. Furthermore, it appears that surgical attitudes to the asymptomatic patient have become more aggressive over time.

Interestingly, over the same time period of these surveys there have been a number of observational studies that shed light on the natural history of CIM with and without syringomyelia and the findings do not support the opinions expressed in the surveys. Whitson reported the natural history of 52 asymptomatic children with CIM followed for up to 7 years. None developed neurological findings during follow-up, and the radiological appearance of the tonsils improved in 50%, stabilized in 38%, and progressed in only 12% (Whitson et al. 2015). In another study comprising 124 cases of nonoperated Chiari patients (81 symptomatic and 43 asymptomatic), Benglis et al. recorded no new deficits in any patients and indeed spontaneous improvement in 6 of 14 patients with typical Chiari symptoms who had declined surgery (Benglis et al. 2011). Pomeraniec et al. also described 70 children with Chiari (48 symptomatic, 22 asymptomatic) followed for a mean of 5 years again no new neurological findings occurred and 42% of the symptomatic patients improved (Fig. 9) (Pomeraniec et al. 2016).

With respect to the syrinx, although studies are generally smaller, they suggest a similarly benign natural history. A study of nine adult patients with CIM and syrinx with 10 years follow-up revealed that the majority remained clinically and radiologically stable over time, with only one patient showing neurological progression (Nishizawa et al. 2001). This finding has been replicated in pediatric patients, in a series of 17 children with syringomyelia, who had mild or no symptoms the syrinx was unchanged (7) or reduced (8) in 15 (88%) (Singhal et al. 2011). Among a large series of pediatric CIM patients followed without surgery Strahle et al. reported 13 cases with associated syrinx, of these the syrinx was either unchanged (6) or improved (5) in 11 (85%). In the remaining two cases the syrinx progressed (Strahle et al. 2011c).

In summary, on the basis of best available current data it is reasonable to conclude that the natural history of asymptomatic CIM, with or without syrinx is more favorable than generally perceived, with only a minority of children demonstrating clinical or radiological deterioration. Therefore nonoperative management with observational follow-up is to be favored over surgery in this setting.

## Surgical Management

### Foramen Magnum Decompression

There is ample evidence that attests to the efficacy of foramen magnum decompression in appropriately selected symptomatic pediatric patients, with improvements in symptoms (chiefly headache) and syrinx decompression observed in 70% or more of cases. While most neurosurgeons agree that foramen magnum decompression is the mainstay of surgical treatment, there is considerable diversity of opinion regarding the extent of such surgery and a wide range of permutations of operative technique are described (Table 3). Perhaps the most widely debated aspect of surgical

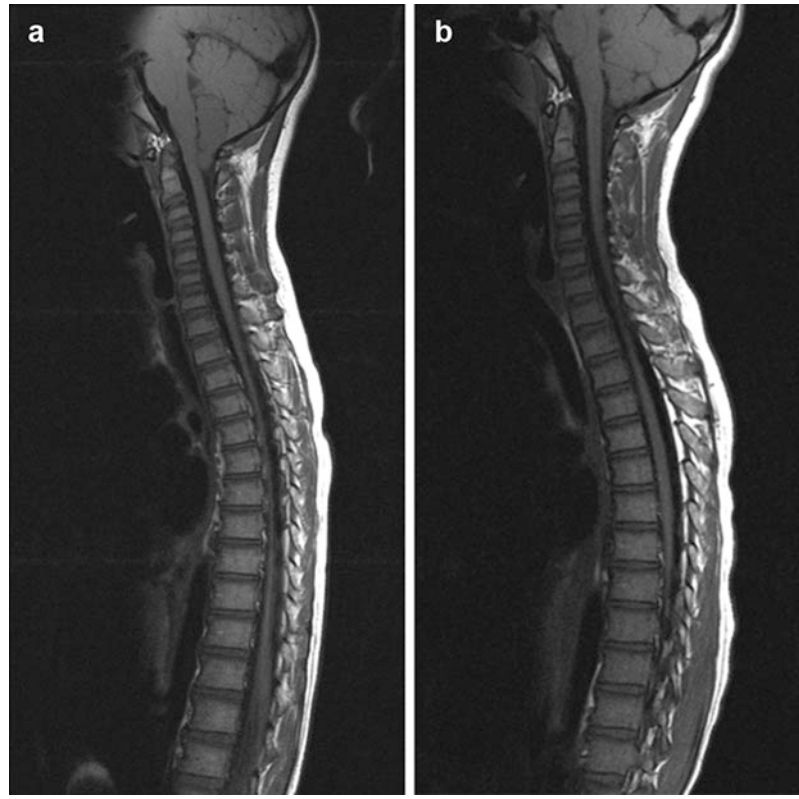


**Fig. 9** Serial T1 weighted MRI scans in a child with CIM demonstrating spontaneous improvement over time

**Table 3** The spectrum of operative permutations described for foramen magnum decompression for CIM

| Bone | Dura | Arachnoid | Parenchymal | Additional |
|---|---|---|---|---|
| Foramen magnum | Dura open no duraplasty | Arachnoid preservation | Tonsil resection | Obex plugging |
| Foramen magnum + C1 | Dura open with duraplasty | Arachnoid opening | Tonsil shrinkage (coagulation) | Fourth ventricle stent |
| Posterior fossa augmentation | – autograft<br>– allograft<br>– synthetic | | | |

technique is whether satisfactory decompression can be achieved by posterior fossa bone decompression alone (PFD) or whether the dura needs to be opened and augmented with duraplasty PFDD. The advantages cited in favor of PFD include shorter procedure time, reduced hospital stay, and reduced complication rate. Proponents of PFDD argue that a fundamental requirement of the surgery is the creation of a capacious CSF cistern at the foramen magnum and that this is better achieved with dural opening (Fig. 10). Moreover advocates of PFDD suggest that the results of PFD are unpredictable, pertinent intradural pathology such as arachnoidal veils and scarring will be missed and re-operation rates are higher if the dura is not opened.

Two recent meta-analyses have sought to address this question (Durham and Fjeld-Olenec 2008; Hankinson et al. 2011), and although the data was insufficient to make clear recommendations, there are noteworthy observations. Comparable rates of clinical improvements are claimed for both procedures 78.8% for PFDD versus 64.6% for PFD. With respect to the effect on

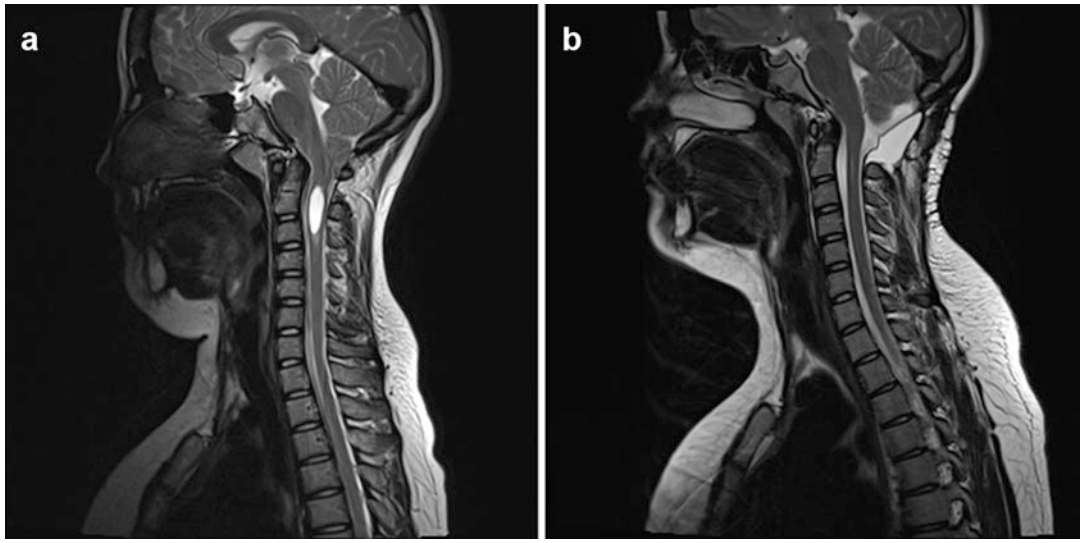


**Fig. 10** Pre- and postoperative T2 weighted MRI scans demonstrating the creation of a capacious CSF cistern at the site of the previous CIM following foramen magnum decompression with duraplasty

syringomyelia, it is a commonly held view that syrinx resolution is more frequently observed following PFDD compared with PFD, and while Durham et al. observed that syrinx decompression seemed more likely in PFDD (87%) compared with PF (56.3%), the effect was nonsignificant due to the small numbers evaluated. They were however able to confirm that re-operation rate following PFDD (2.1%) was less than following PFD (12.6%) although the rate of CSF related complications was higher for PFDD (18.5%) than PFD (1.8%) (Durham and Fjeld-Olenec 2008).

A variety of dural substitutes have been used following foramen magnum decompression, the most common being autologous pericranium or nonautologous collagen matrix (Fig. 11). Pericranium is nonimmunogenic and suggested to have a lower rate of aseptic meningitis and wound complications such as infection and pseudomeningocele; however, it requires the skin incision to be extended, and in children pericranium may be thin and difficult to harvest and late ossification of the graft is also reported. Collagen matrix such as DuraGen (Integer Neuroscience Plainsboro NJ) is a commonly preferred alternative that can either be used as an onlay graft or sutured into the dural defect. A recent literature review concluded that there was no clear evidence to favor autologous or nonautologous but suggested that where feasible pericranium offered advantages (Abla et al. 2010).

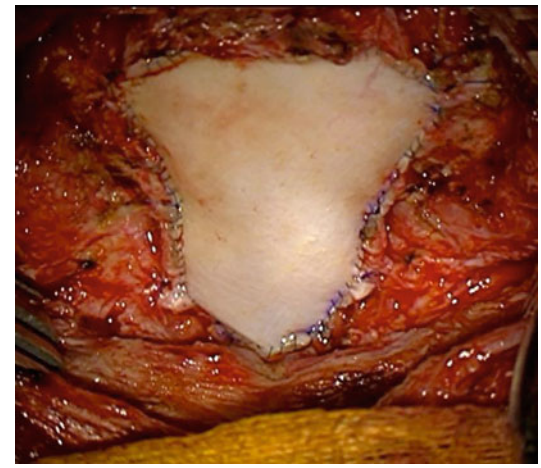

**Fig. 11** Intraoperative photograph of allograft (bovine pericardium) duraplasty

Some surgeons advocate dura opening but without duraplasty, simply closing the muscle layers over the open dura. It is important to appreciate that the role of duraplasty is not only to facilitate creation of a cisterna magna and prevent CSF leak but also to reduce the risk of blood

products and serous fluid contaminating the CSF pathways, and so the technique of leaving dura open my increase the risk of complications such as 'aseptic' meningitis, hydrocephalus and wound related complications.

It is clear that the number of permutations of technique for foramen magnum decompression is extensive, and there is no conclusive evidence base to support any one technique. The more invasive procedures including intra-arachnoidal dissection, tonsillar shrinkage/resection, and placement of fourth ventricular stent are likely to increase the risk of surgery, and there is little evidence to support the routine use of such measures in primary procedures. Some surgeons advocate keeping the arachnoid layer intact in an attempt to gain the supposed advantages of dural opening while avoiding risks associated with intra-arachnoidal manipulations (Fig. 12). A number of authors have also advocated the use of intraoperative ultrasound as a means of tailoring the operative procedure according to the CSF flow and tonsil pulsations. Ultrasound assessment is however qualitative and precise criteria to ascertain the adequacy of decompression are yet to be defined.

Since it is not possible to provide unequivocal guidance on surgical technique it is recommended that individual neurosurgeons and departments who undertake this surgery engage in prospective audit and critical evaluations of their own CIM practice to ensure that whatever their chosen technique this achieves an appropriate balance between clinical efficacy and low operative morbidity, such that their outcomes are comparable with best published data.

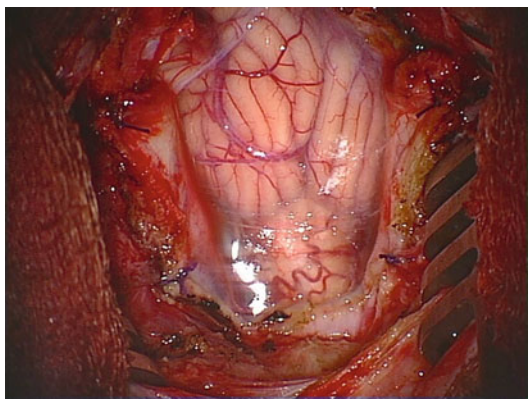

**Fig. 12** Intraoperative photograph of foramen magnum decompression with dural opening and preservation of arachnoid (prior to duraplasty)

## Cranial Vault Expansion

Where possible any underlying predisposition to cerebellar tonsillar herniation should be accounted for in selecting the appropriate surgical treatment. Foramen magnum decompression in the presence of raised intracranial pressure is associated with increased risk of complications such as neurological deterioration, pseudomeningocele, persisting or worsening syringomyelia, and cerebellar "slump." Any associated hydrocephalus should be treated in the first instance either by ventricular shunt or endoscopic third venticulostomy and resolution of syringomyelia as well as improvement in hydrocephalus can be anticipated in the majority of patients treated in this way.

The situation is somewhat more complicated in the context of CIM and craniosynostosis with raised ICP. Particularly in young children, placement of shunt may further compromise cephalocranial disproportion leading to worsening of the hindbrain herniation and again some would advocate the use of ETV over shunting in this scenario. An alternative approach is posterior cranial vault expansion. This procedure is well established in the early management of craniosynostosis as a means of addressing raised ICP; however, it has also been observed that, following expansion of the supratentorial compartment, the position of the cerebellar tonsils may also improve, particularly when the procedure is carried out in young children (Leikola et al. 2012).

## Craniovertebral Fixation

In the same way that raised ICP should modify the surgical strategy for CIM, the presence of structural abnormalities at the craniovertebral junction

should alert the neurosurgeon that the child may not be appropriate for "standard" foramen magnum decompression, and indeed may be at significant risk for treatment failure or worsening neurology following such an intervention. Anomalies that raise concern and constitute what has been termed "complex Chiari" include retroflexed odontoid (Fig. 5), herniation of the brainstem, as well as cerebellar tonsils and basilar invagination with reduced clivus-canal angle (Bollo et al. 2012).

The principles of treatment of "complex Chiari" are identical to those that govern the management any other craniovertebral malformation namely, reduction, decompression, and stabilization. Aggressive attempts to reduce the bone deformity should be pursued before consideration is given to decompression. Either on table reduction, with intraoperative neurophysiological monitoring and fluoroscopic screening, or preoperative halo traction can be used. Successful reduction may not only preclude the need for complex decompression (in particular transoral decompression) but can also result in improvement of the hindbrain hernia, obviating the need for formal foramen decompression and duraplasty. It may be difficult to decide whether decompression and expansion duraplasty are required in addition to craniovertebral fixation, in such cases intraoperative ultrasound may be of help; however, if there is any doubt, the decompression should be performed as redo surgery after craniovertebral fixation presents a major challenge.

A recent study of 65 patients (pediatric and adult) with hindbrain hernia, including 55 with syringomyelia, reported clinical improvement in over 90% of cases following atlantoaxial arthrodesis alone (Goel 2015); the authors controversially suggesting that CIM, with or without basilar invagination was always associated with atlantoaxial instability and moreover that fixation to the occiput was unnecessary in these cases.

Where arthrodesis is indicated in the management of complex Chiari malformations most pediatric neurosurgeons would advocate an occipitocervical construct as the skull base anomaly is, in most cases, an integral component of the deformity that needs to be addressed (Fig. 13). It is rarely necessary to extend the fixation beyond the level of C2.

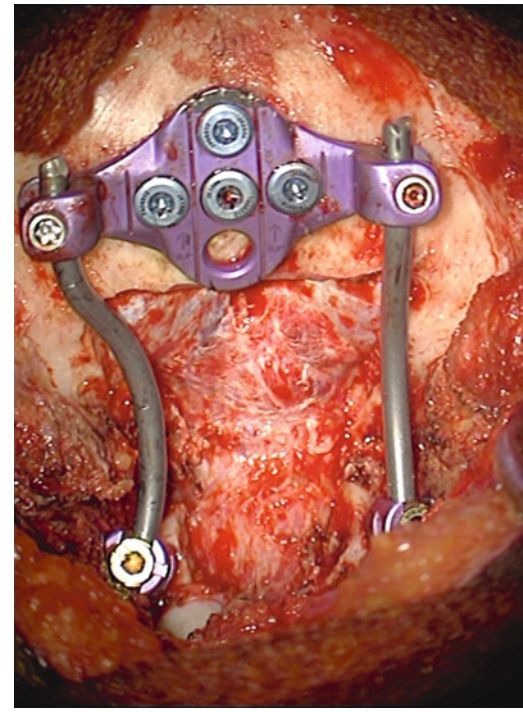

**Fig. 13** Occipitocervical fixation with foramen magnum decompression in a child with "complex CIM." The occipital plate is connected via rods to C2 pedicle screws

## Summary of Operative Technique Might Be Indicated

In summary, the investigation and management of CIM needs to reflect not only the apparently benign natural history in the asymptomatic child but also the multifactorial etiology of this condition in pediatric practice (Fig. 14). With respect to the procedure of foramen magnum decompression within this scheme, currently there is no clear evidence to dictate the optimal surgical strategy nor indeed whether the extent of surgery can be tailored to specific situations, for example, which child might benefit from dural opening and intra-arachnoidal dissection and which might be better served by a more conservative bone only decompression.

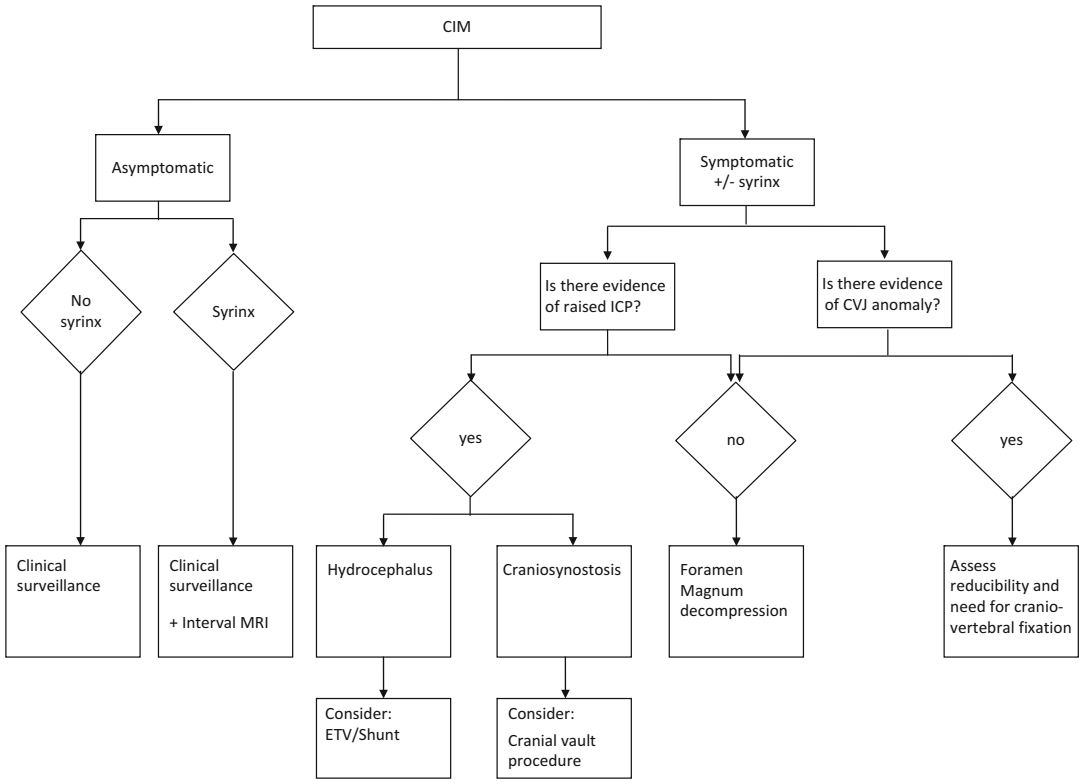


**Fig. 14** Algorithm of management for pediatric CIM

## Outcomes for CIM

### Outcomes Scales

The heterogeneity in the underlying etiology of CIM combined with the variable symptom profile and a diverse array of surgical techniques for treatment make the evaluation of outcome for CIM exceedingly difficult. Yarbrough et al. have critically reviewed currently used outcome measures for CIM surgery highlighting the limitations of current reporting and emphasizing the potential utility of general quality of life and disability scales from other disciplines where patient defined outcomes take precedence over the simplified "gestalt" evaluations (such as same, improved, worse) that predominate in the literature (Yarbrough et al. 2015).

Many of the scoring systems are not appropriate for children (e.g., Asgari score) or have a bias toward specific clinical issues such as pain or physical disability (Japanese Orthopaedic Association score for cervical myelopathy). There are however scales that have been devised specifically to evaluate CIM and or syringomyelia, these include the Chiari Symptom Profile, a patient questionnaire comprising 57 questions covering four domains of physical, functional, psychological, and social (Mueller and Oro 2013). This tool has the disadvantage that it is time consuming to apply and has yet to be validated on a wider scale. A more simplified outcome measure is the Chicago Chiari Outcome Scale (CCOS), again this attempts to be a multifaceted assessment covering four areas, pain, nonpain (e.g., bulbar, ataxia, sensory), functionality, and complications (Aliaga et al. 2012). While easier to apply than the Chiari Symptom Profile, some domains of the CCOS are vulnerable to variable and subjective interpretation, and in the original description of the CCOS, good outcomes were disproportionately represented thus compromising

the discriminatory ability of the scale. In spite of the inherent limitations of clinical outcome scales, such attempts to provide some degree of uniformity and subjectivity into the evaluation of CIM surgery are not only laudable but essential if we are to see any possibility of resolving many of the continuing controversies surrounding patient selection and operative technique in the future.

A major obstacle in defining outcomes for CIM is how to reflect both clinical and radiological outcome criteria since these two measures may not complement each other and indeed may even be contradictory; an excellent radiological result in a patient with persisting symptoms or vice versa. In an extensive review of the literature (adult and pediatric), Sindou et al. attempted to evaluate the relative efficacy of a variety of surgical techniques to treat CIM associated syringomyelia, techniques ranged from simple bone decompression through to dural opening with various intradural procedures (Sindou 2002). While the more extensive procedures were associated with good rates of syrinx improvement, the rates of surgical complications and clinical aggravation tended to be higher in such procedures. They concluded that the optimal balance between high surgical efficacy and low rate of aggravation was achieved by the combination of bone decompression with duraplasty, but preservation of the arachnoid layer, by contrast the least efficacious technique was bone decompression with dural and arachnoid opening but no duraplasty.

## Clinical Outcomes

In a large review of the published literature, improved headache was reported in 88% of pediatric cases (Arnautovic et al. 2015). In that study headache outcomes were statistically better for children compared with adults. The importance of accurate diagnosis of CIM associated headache, differentiating this from other varieties of childhood headache is likely to be pivotal in achieving good outcomes.

Syrinx improvement should be anticipated in at least 75% of patients following surgery. Failure to demonstrate improvement on follow-up MRI scan after at least 6 months following surgery warrants further investigation. Factors associated with failure of syrinx improvement include inadequate primary decompression with persisting obstruction to CSF flow at the cervicomedullary junction, arachnoid webs or veils at the fourth ventricular outflow, subclinical raised intracranial pressure, or craniovertebral malformation. Syrinx shunting should not be considered until potential causes for persisting syrinx have been fully investigated.

The effect of foramen magnum decompression on associated scoliosis is less predictable as this depends on the severity of scoliosis at the time of surgery and the age of the patient. Children who have scoliosis measuring less than 30 deg and who are less than 10 years old have a much greater chance of curve improvement and reduced need for spinal fusion compared with older children with more severe curves (Mackel et al. 2016).

## Surgical Complications

The operative morbidity and mortality from CIM surgery is commonly underemphasized and underreported. A recent multicenter study of 30 day outcomes following CIM surgery reported a complication rate of 5.3%, mostly comprising wound related morbidity and in this series 3.4% of patients required an unplanned re-operation (Vedantam et al. 2016). A similar finding was revealed in an extensive review of CIM literature (from 1965–2013) where the rate of reported complications was 3.5% (IQR 2.3–14.3%); however, less than half of the studies in this review reported their serious complications (Arnautovic et al. 2015). The most frequently reported complications of foramen magnum decompression included pseudomeningocele (5–47%), aseptic meningitis (1–60%), CSF leak (0.2–21%), and wound infection (1–9%). The reported rate of neurological deficit following surgery was 1–4% (Arnautovic et al. 2015). This latter review additionally identified a mortality rate of 3% in the pediatric age group.

The incidence of hydrocephalus following CIM surgery varies from 0.9% to as high as 18% (Duddy et al. 2014) in contemporary series. This

risk is not restricted to those with ventriculomegaly or raised ICP preoperatively but does seem rare following bone only decompression suggesting that aspects operative technique might be relevant.

## Conclusions

The term Chiari I malformation is a misnomer, the condition that we encounter in current clinical practice is only occasionally secondary to hydrocephalus (as initially reported by Chiari), and furthermore, there is no evidence that CIM (in contrast to the other conditions described by Chiari) is a true CNS malformation. The implications of this conclusion are far more than just semantic, CIM should be considered as a deformation of the hindbrain in response to local mechanical and hydrodynamic factors, and it is beholden to the pediatric neurosurgeon to investigate and critically evaluate these factors for each patient before embarking upon treatment. It follows that the identification of CIM should be considered a starting point for investigation not a final diagnosis.

In pediatric practice in particular, management strategies for CIM should reflect the etiological heterogeneity of this condition and the (often benign) natural history of this condition in asymptomatic or minimally symptomatic patients. Pediatric neurosurgeons should be cognizant of the potential for predisposing anomalies such as raised intracranial pressure (with or without hydrocephalus), craniosynostosis, and craniovertebral anomaly. While foramen magnum decompression currently remains the mainstay of treatment, it should only be offered to patients with symptoms or evidence of radiological progression and only after thorough investigation. Moreover as there is no consensus as to the optimal surgical technique for foramen magnum decompression, it is beholden to surgeons to critically audit their own practice to ensure that whatever their preferred technique, their surgical complications and long-term outcomes are commensurate with best international practice.

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