Paediatric Perspectives

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Contents

With Shane Tubbs and Melandee Brown

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13.1 Introduction

In the paediatric population, the diagnosis of syringomyelia is most commonly associated with hindbrain hernias (Chiari malformations). In this chapter, syringomyelia that accompanies the pathology of Chiari I and Chiari II malformations will be discussed. Syringomyelia may also be encountered in cases without frank tonsillar ectopia but with noted crowding of the cisterna magna. This so-called Chiari 0 will also be reviewed. The relationship between syringomyelia, hindbrain hernia and scoliosis will also be addressed.

Hindbrain malformations were first studied in detail in the 1890s, by Hans Chiari. He described three types of herniation, as well as cases of hypoplasia or complete aplasia of the cerebellum and tentorium cerebelli – the Chiari IV malformation (Soleau et al. 2008).

The three types of hindbrain herniation share a common pathophysiology that involves the loss of free movement of cerebrospinal fluid (CSF) out of the fourth ventricle and into the cervicomedullary subarachnoid cisterns. An understanding of the pressure gradients that develop between the intraventricular and largely intracranial space and the subarachnoid channels surrounding the cervicomedullary junction allows for an appreciation of the developmental differences between the Chiari I and Chiari II malformations. It is this pressure gradient that is also thought to be responsible for the development of the syringomyelia that frequently accompanies these conditions (Attenello et al. 2008; Ball and Dayan 1972).

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13.2 Chiari I Malformation and Syringomyelia

The more common Chiari I malformation is classically defined as caudal displacement of the cerebellar tonsils 5 mm or more below the foramen magnum. Affected individuals are distinguished from Chiari II patients by the normal location and appearance of the brainstem and caudal displacement of the cerebellar tonsils, rather than the vermis. The true incidence of tonsillar herniation is not known, but a review of radiological images of over 22,000 patients reported it to be 1 in 130 (Meadows et al. 2000). A more recent series, based on a review of MRI images of 5,248 patients under the age of 20, found the incidence of Chiari I malformation to be 1 in 102 (Tubbs et al. 2011)

13.2.1 Presentation

Although Chiari I malformation more often presents symptomatically in early adult life, the widespread use of magnetic resonance imaging (MRI) has led to increased recognition of this disorder in children (Meadows et al. 2000). At the same time, given this increasing frequency of radiological diagnosis of Chiari I malformation, care must be taken to differentiate between patients with recognisable symptoms and individuals with symptoms that are unrelated to the finding of tonsillar herniation on imaging. Patients with Chiari I malformation may present with a variety of complaints, ranging from headache and neck pain to symptoms of myelopathy and brainstem compression. The most common presentation is one of pain in the occipital region or upper cervical spine that is exacerbated by Valsalva or tussive manoeuvres. In infants and non-verbal children, headache may be signalled by generalised irritability and neck grabbing. With headaches that are not Valsalva-related and pain that is remote from the occipital-cervical junction, it is unlikely that the symptoms are attributable to the presence of a Chiari I malformation (Soleau et al. 2008).

More unusual presentations of Chiari I malformations include features of brainstem compression, cranial nerve dysfunction or cerebellar signs. Symptoms of brainstem dysfunction include tongue atrophy, down-beat nystagmus and isolated abducens nerve palsies. Symptomatic lower cranial nerve dysfunction has been reported to occur in 12–25 % of patients, the most common symptoms including dysphagia, gagging or chronic emesis and sleep apnoea (Soleau et al. 2008). In a recent series, however, fewer than 10 % of patients presented with cranial nerve deficits and 5 % presented with central apnoea, confirmed by a sleep study (Tubbs et al. 2011). Rarely, patients may present with an acute crisis, and in another series of 500 patients, three presented with symptoms of dysphagia, anisocoria and hemiparesis that progressed rapidly over 2–3 days (Tubbs et al. 2011). Three other reported cases presented with abrupt onset of severe symptoms, two following mild head injury (Massimi et al. 2011).

Clinical myelopathy can result from direct compression of the upper cord, by the herniated cerebellar tonsils, or can be due to the presence of a syrinx. Loss of pain and temperature sensation in the upper extremities, with preservation of light touch and proprioception, in a Chiari I patient, should alert the examiner to the presence of an accompanying syringomyelia. Scoliosis in a Chiari I patient may also signal the presence of a syrinx. Patients with a left convex thoracic curve, abnormal or absent abdominal reflexes or complaints of non-dermatomal pain should also raise the suspicion of an underlying syrinx (Soleau et al. 2008). Syringomyelia has been described as affecting between 35 and 75 % of patients with Chiari I malformation (Attenello et al. 2008). In one series of 500 patients, who underwent surgery for Chiari I malformation, 57 % were found to have syringomyelia. Syrinx cavities were most commonly located in the cervicothoracic cord, followed by the cervical cord, lumbar cord and brainstem. A surprising 39 % of patients had holocord syringes (Tubbs et al. 2011).

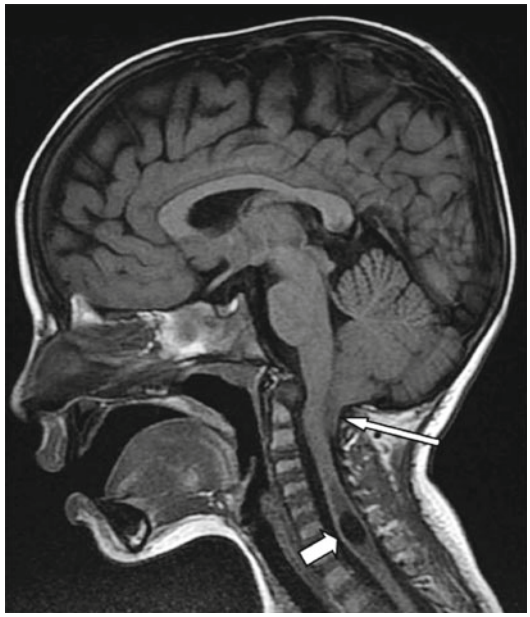

Fig. 13.1 Sagittal T1-weighted MRI. Note the Chiari I malformation (*long arrow*) and cervical syrinx (*short arrow*)

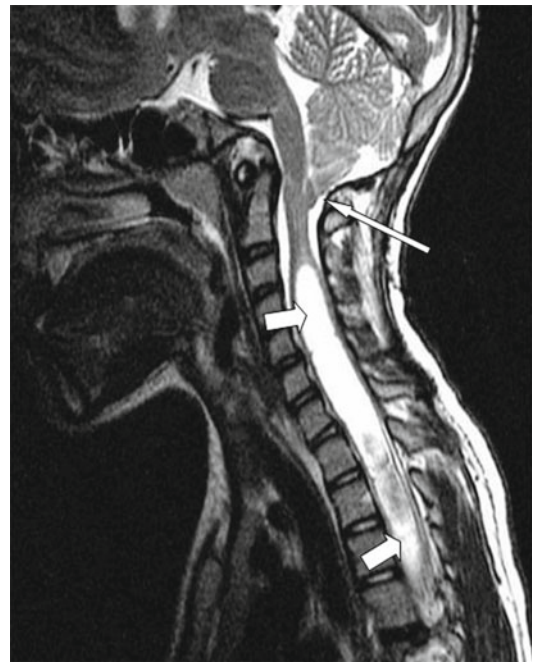

Fig. 13.2 Sagittal T2-weighted MRI. Note the Chiari I malformation (*long arrow*) and holocord syrinx (*short arrows*)

13.2.2 Imaging

Conventional computerised tomography (CT) is sometimes the imaging modality that leads to the initial diagnosis of Chiari I malformation. The radiologist may identify characteristic crowding of the foramen magnum, although the axial plane of the standard head CT makes it difficult to appreciate fully the extent of the hindbrain herniation. Sagittal reconstructions of the source data may be helpful, but CT imaging alone is inadequate in evaluating a patient for the presence of a syrinx or Chiari malformation.

On magnetic resonance imaging (MRI), Chiari I malformation is characterised by significant herniation of the cerebellar tonsils through the foramen magnum. An associated cervical, cervical thoracic or holocord syrinx may also be found (Figs. 13.1 and 13.2). The herniated cerebellar tonsils also often lose their rounded appearance and become peglike (Fig. 13.3). They frequently lead to obliteration of the retrocerebellar subarachnoid space (Aitken et al. 2009). Although extension of one or both tonsils 5 mm

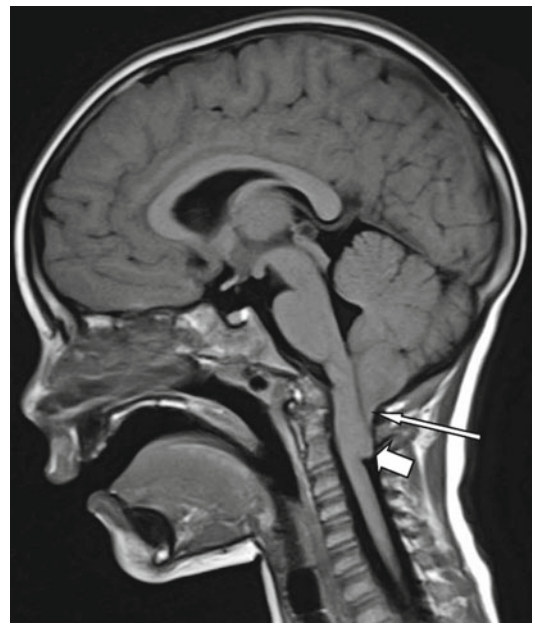

Fig. 13.3 Sagittal T1-weighted MRI. Note the pointed cerebellar tips (*long arrow*) often seen in the Chiari I malformation and elongation of the brainstem (*short arrow*)

below the foramen magnum is sufficient for a radiological diagnosis, these findings should be considered in conjunction with the patient's symptoms. Patients that do not have symptoms or syringomyelia, even when the MR imaging is impressive, can be managed conservatively with observation (Soleau et al. 2008). The presence of syringomyelia will, however, lead the practitioner towards surgical intervention even in the presence of minimal symptoms (Oakes 1991).

Patients suspected of having Chiari I malformation should also undergo imaging of the brain, as a small percentage of patients will be found to have associated hydrocephalus, nearly one in ten cases in one large series. Those with untreated hydrocephalus should have this addressed, prior to any decompressive procedure at the craniovertebral junction, and those patients with an already shunted hydrocephalus should have the functioning of the shunt confirmed.

Other radiographic abnormalities are seen in association with the Chiari I malformation, including retroverted odontoid process which was found in 24 % of operative Chiari I patients in one series. Other reported associations include scoliosis (18 %), atlanto-occipital fusion (8 %), Klippel-Feil anomaly (3 %) and basilar invagination (3 %) (Tubbs et al. 2011). Plain flexion-extension radiographs of the cervical spine are helpful in resolving the question of spinal instability and can further delineate the extent of the bony abnormalities. The presence of significant basilar invagination should alert the practitioner to the possible need for an anterior decompression, prior to a posterior decompression (Oakes 1991).

Cine-MRI may be used to assess flow across the foramen magnum. Some authors find it a valuable tool in decision making, whilst others find that its usefulness is dependent upon the software used to process the acquired sequences (Soleau et al. 2008; Ventureyra et al. 2003).

13.2.3 Surgical Intervention

Surgical manoeuvres in the paediatric Chiari I patient include bony decompression of the posterior fossa, usually with exploration and decompression of the fourth ventricular outlet. In most cases this necessitates removal of the posterior arch of C1 as well, with excision of the subadjacent fibrous band and duraplasty. Coagulation or partial resection of the herniated tonsils may be necessary, if they obstruct fourth ventricular outflow or in the event that a re-operation is needed due to failure of the syrinx to collapse after the initial procedure.

Controversy exists regarding the necessity for duraplasty in the treatment of Chiari I malformation. A small retrospective study of 11 adult patients, who underwent bone only decompression, with removal of the posterior arch of C1, found that eight of the patients had improvement in their symptoms. Seven of these patients also had syringomyelia, and three had radiological decrease in the size of their cavities and improvement in their symptoms. All three of these patients also had increased posterior fossa volume on post-operative imaging (Munshi et al. 2000). A study of brainstem auditory evoked potentials in 11 paediatric patients, before and after duraplasty, found that no additional improvement was noted following this manoeuvre (Anderson et al. 2003). Intraoperative ultrasound has been employed in an effort to identify patients who might benefit from duraplasty, in addition to bony decompression (McGirt et al. 2008; Yeh et al. 2006). A retrospective study of 256 paediatric patients stratified the success rates of bone only decompression versus duraplasty, according to the extent of tonsillar herniation. In children with tonsillar herniation caudal to C1, suboccipital decompression alone was associated with a twofold increase in the risk of symptom recurrence, when compared with those children who also underwent duraplasty. Children who had tonsillar herniation rostral to C1 had equivalent outcomes when undergoing duraplasty or bone only decompression (McGirt et al. 2008).

A large retrospective review of 500 surgically treated paediatric Chiari I patients, of whom 285 also had syringomyelia, found that 12 % of this latter group had an arachnoid veil occluding the fourth ventricular outlet. Fifteen patients in total required re-operation, 13 for persistent syringes

and two for persistent Valsalva-related headache. One of the latter, two improved following re-operation with duraplasty. Of the 13 patients with persistent syrinx, 11 had resolution of their hydromyelia with re-operation and tonsillar coagulation. The two patients that did not improve following re-operation enjoyed some decrease in their syrinx size following subsequent placement of syringopleural shunts (Tubbs et al. 2011).

Some authors have noted an association between duraplasty and a higher complication rates (Navarro et al. 2004). Others have noted a small but significant correlation between duraplasty and longer hospitalisations (Yeh et al. 2006). Other authors report low complication rates but including transient acute hydrocephalus and extra-axial subdural collections (Elton et al. 2002). Other complications include direct vascular or neural injury, bleeding from dural venous sinuses, CSF leak, meningitis and pseudomeningocoele formation. Sagging or slumping of the cerebellum may also be encountered, in the setting of overly generous craniectomy and dural grafting. In this case, a partial cranioplasty, with revisional duraplasty, to create a cerebellar 'sling', can be effective in supporting the herniated cerebellum (Holly and Batzdorf 2001). Bone regrowth at the foramen magnum and at the site of the C1 bone removal has been reported in young children and infants (Aoki et al. 1995).

13.2.4 Response to Cervicomedullary Decompression

A review of 49 paediatric patients operated on for Chiari I and syringomyelia observed that just over half had radiological improvement in their syringomyelia during a mean follow-up period of 41 months. The median time until radiological improvement was 14 months following surgery. The same proportion of children enjoyed symptomatic improvement with a median time to improvement of 4 months (Attenello et al. 2008).

A series of 500 operated paediatric patients, of which 57 % had syringomyelia, included a re-operation rate of 3 %, mostly for persistent syrinx. Posterior fossa decompression was sufficient in relieving symptoms in 80 % of patients at the time of the first operation and in 95 % of patients following a second operation. In the 13 patients with persistent syringomyelia, all but two improved with re-exploration alone. The two patients who did not improve following re-operation underwent placement of syringopleural shunts, but given the success rates of re-operation, the authors have almost completely abandoned the practice of syrinx shunting (Tubbs et al. 2011). In this same series, an arachnoid veil was found to be obstructing the outlet of the fourth ventricle in 12 % of patients with syringomyelia. Thirty patients in the series underwent placement of stents, passing from the fourth ventricle into the subarachnoid space. These patients were less likely to have resolution of their syringomyelia, although this may be because patients selected for stent placement had unfavourable anatomical features.

13.3 Chiari II Malformation and Syringomyelia

The Chiari II malformation is characterised by caudal displacement of the cerebellar vermis, brainstem and fourth ventricle below the level of the foramen magnum (Fig. 13.4). It occurs exclusively in association with myelodysplasia. In the United States, the historical prevalence of myelomeningocoele is cited as one to two per 1,000 live births, but the advent of prenatal screening and increased awareness of the need for maternal folic acid supplementation has decreased the prevalence to between 2 and 4.6 cases per 10,000 births (Honein et al. 2001; Piatt 2010), although regional variations in prevalence do exist (Shurtleff 2004). The frequency of syringomyelia in Chiari II patients varies between different reports, from 40 to 80 %, and was documented in 20 out of 26 (3 in 4) autopsy cases of young children born with myelomeningocoele (Piatt 2004). The true incidence may, in fact, be underestimated because some pre-existing but undiagnosed cavities may collapse, following shunting of hydrocephalus in infancy.

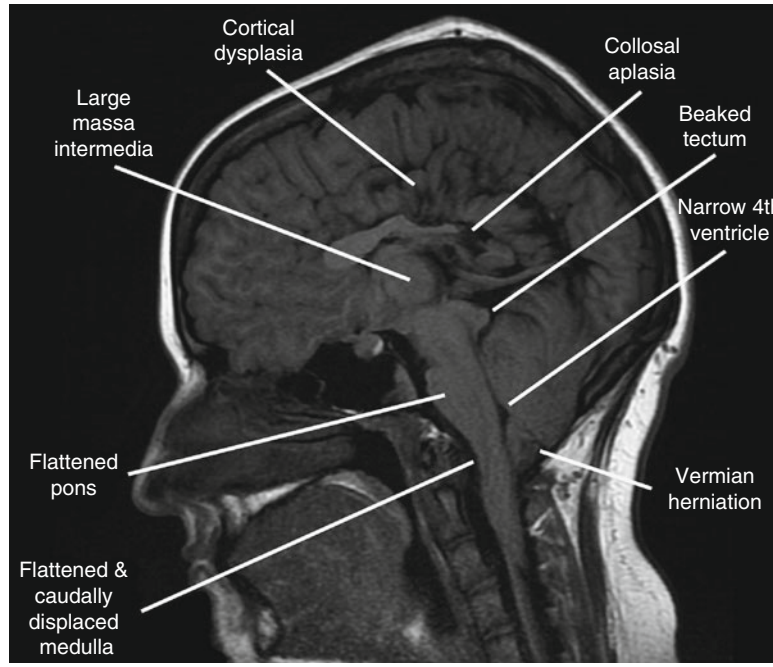

Fig. 13.4 Sagittal T1-weighted MRI. An adult patient with Chiari II malformation

Syringomyelia cavities are often found in the lower cervical and upper thoracic cord and may be missed on routine cranial imaging. Low lumbar syrinx cavities are rare but are frequently associated with rapidly progressive scoliosis and are less likely to respond to ventricular shunting (Piatt 2004). Spinal arachnoid cysts may also be found in association with Chiari II malformation.

Biopsy specimens of the cyst walls, when analysed by electron microscopy, have demonstrated the presence of axons which suggests that some of these cysts are actually formed by dorsal rupture of eccentrically located syringes (Heinz et al. 1992).

There also appears to be an association between untreated or partially treated hydrocephalus and syringomyelia. A study of 18 Chiari II patients with progressive myelopathy, who underwent radionucleotide ventriculography, demonstrated the accumulation of tracer in the syrinx cavities of the 14 patients with untreated hydrocephalus (Batnitzky et al. 1976). These observations underscore the need to confirm the presence of a functioning shunt in Chiari II patients with shunted hydrocephalus that present with syringes.

13.3.1 Presentation of Chiari II

Patients with Chiari II malformations present at birth with myelomeningocoele. The outlook for this population is somewhat restricted, and historically, approximately one third of the neonates with symptoms of brainstem dysfunction did not survive beyond infancy. Survival has, however, increased with prompt ventricular shunting, shunt revision when needed and posterior fossa decompression when appropriate (Talamonti et al. 2007). Overall, some 20–30 % of Chiari II patients will become symptomatic from their hindbrain herniation at some point in their lifetime.

Clinical presentation varies in different age groups. Infants and neonates can present with apnoea, inspiratory stridor, dysphagia, bradycardia and opisthotonus. The dysphagia may be severe enough to necessitate placement of a gastrostomy tube. When present at birth, such symptoms suggest hypoplasia or aplasia of brainstem nuclei and often predict a poor prognosis. More commonly, symptoms begin within the first few months of life and when they are seen in a previously well infant attention should

first be given to normalisation of the intracranial pressure through shunt placement or shunt revision if necessary (Holinger et al. 1978; Soleau et al. 2008). Feeding and swallowing difficulties are the most common presenting symptoms for infants at risk and were found in 59–71 % of symptomatic Chiari II patients in one series (Pollack et al. 1992b). Infants may present with weak suck and prolonged feeding times, and they may demonstrate weight loss and failure to thrive, as their physical growth outpaces their ability to meet their nutritional needs. Breathing difficulties are the most dangerous symptom in Chiari patients and have been reported to occur in between 29 and 76 % of patients (Rauzzino and Oakes 1995). Inspiratory stridor is the result of bilateral tenth nerve paresis, with weakness of the vocal cord abductors. It may be caused by direct traction on the tenth nerve or by damage to the dorsal motor nuclei, secondary to microhaemorrhage or compressive ischemia (Benjamin et al. 2009; Linder and Lindholm 1997; Rauzzino and Oakes 1995). One series observed that all infants began with normal swallowing and feeding that progressively deteriorated (Pollack et al. 1992b). In these cases, they found that feeding problems preceded respiratory difficulties and were a recognisable warning sign. In another publication, the same group found that all neonates that underwent surgical decompression, prior to the development of bilateral vocal cord paralysis, demonstrated some improvement in neurological function following surgery (Pollack et al. 1992a).

Older children more commonly have symptoms of spinal cord compromise, and the syringomyelia associated with Chiari II malformation behaves in a manner similar to that seen in Chiari I patients. Classic presentation is with lower motor neuron features in the arms and upper motor neuron findings in the legs. These include atrophy of the small muscles of the hands, together with disassociated sensory loss. Progressive enlargement of the syrinx cavity leads to loss of the ability to perform fine motor tasks, and fasciculation and loss of tendon reflexes may be evident in the upper limbs. Further expansion of the syrinx may impede descending corticospinal tracts, such that patients who were previously ambulatory may report a history of increasing falls, coming on over months or years.

Syringobulbia, or expansion of the syrinx into the brainstem, will manifest itself as dysfunction in multiple brainstem nuclei, which may wrongly be attributed to the underlying Chiari II malformation. Neurogenic athropathies may also be present. Back pain and scoliosis (see below) may also be the initial presenting complaints.

Changes in bowel or bladder function, however, should prompt imaging to exclude a tethered cord (Rauzzino and Oakes 1995). Headache and posterior cervical neck pain are also common and may be due to the distortion of the descending fibres of the spinal trigeminal tract or the upper cervical roots.

13.3.2 Imaging Chiari II

Radiographic assessment of patients with Chiari II malformation can begin with plain films of the cervical spine. Flexion and extension views can be used to assess for cervical instability. The posterior arch of C1 is incomplete in up to 70 % of cases and may be replaced by a compressive band of periosteal tissue. Full spine radiographs permit assessment of scoliosis, segmentation errors and other occult dysraphisms. Rarely, split cord malformation, with a bony median septum, may be identified on plain films (Rauzzino and Oakes 1995).

CT images are inadequate for studying the pathology of hindbrain herniation, but head CT imaging is helpful in evaluating ventricular size and can provide an objective measurement of the adequacy of ventricular shunting, although surgical exploration remains the ‘gold standard’ for the determination of the adequacy of shunt function.

MRI is the diagnostic tool of choice in evaluating patients with suspected Chiari malformations. The hallmark of the Chiari II malformation is the elongation and caudal displacement of the cerebellar vermis, medulla and lower brainstem, below the foramen magnum. In up to 70 % of

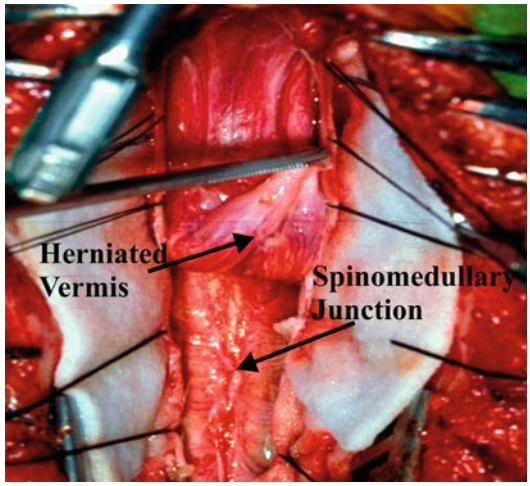

Fig. 13.5 Operative view of the Chiari II malformation

Chiari II patients, a medullary kink may be identified. Elongation of the fourth ventricle into the upper cervical cord is also a common finding. The cerebellum of Chiari II patients is smaller than normal and is contained in a hypoplastic posterior fossa with obliterated retrocerebellar CSF spaces (Fig. 13.5). The tentorium cerebelli has a low insertion and can also be hypoplastic, allowing the cerebellum to tower above it. Other frequently observed cerebellar findings include persistent foetal location with extension around the midbrain into the cerebellopontine cisterns and cerebellar dysplasias and heterotopias (Rauzzino and Oakes 1995).

13.3.3 Surgery for Chiari II Malformations

The surgical management of Chiari II malformation generally consists of bony decompression followed by some form of dural expansion manoeuvre. Prior to any posterior fossa decompression, however, patients with questionable shunt function should first undergo shunt revision. This approach can sometimes avoid the need for posterior fossa surgery because reducing the downward pressure on the cerebellum may lead to resolution of symptoms. Obtaining effective ventricular drainage may also lead to collapse of the syrinx, without recourse to major posterior fossa surgery. On the other hand, ignoring an active hydrocephalus and performing a craniovertebral decompression may sometimes lead to a potentially dangerous exacerbation of a hydrocephalus that was previously in a state of hydrodynamic equilibrium.

Reference to pre-operative MRI assists in the localisation of the vermian herniation, a medullary kink, the fourth ventricle and a low-lying torcular Herophilus and transverse sinus. For patients with a large foramen magnum, occipital craniectomy may not be required. If the torcular is low-lying, care must be taken to avoid inadvertent entry into the major venous sinuses, which could lead to dangerous blood loss. Care must be taken to distinguish the medullary kink from the cerebellar vermis, and intraoperative ultrasonography may be useful in this regard (Tubbs and Oakes 2004). In Chiari II patients, the choroid plexus usually retains its embryonic extraventricular location and marks the caudal end of the fourth ventricle.

Unlike with Chiari I malformations, adequate decompression of the Chiari II hindbrain hernia requires that the decompression should extend to the level of the displaced posterior fossa content. The herniated brainstem, unlike the cerebellar tonsils, cannot be delivered upwards or coagulated (Fig. 13.6). The fourth ventricular outlet should be identified and widely opened, and it is often necessary to develop several tissue planes, before the floor of the fourth ventricle can be clearly identified. Misidentification of anatomical landmarks or excessive tissue manipulation can result in damage to at-risk parts of the medulla or lower pons. Dense adhesions and hypervascularity may be encountered at points of compression or traction, making identification and dissection of these structures more difficult.

13.3.4 Surveillance

Early surgical therapy may prove to be life saving for symptomatic Chiari II patients, when the symptoms are attributable to brainstem

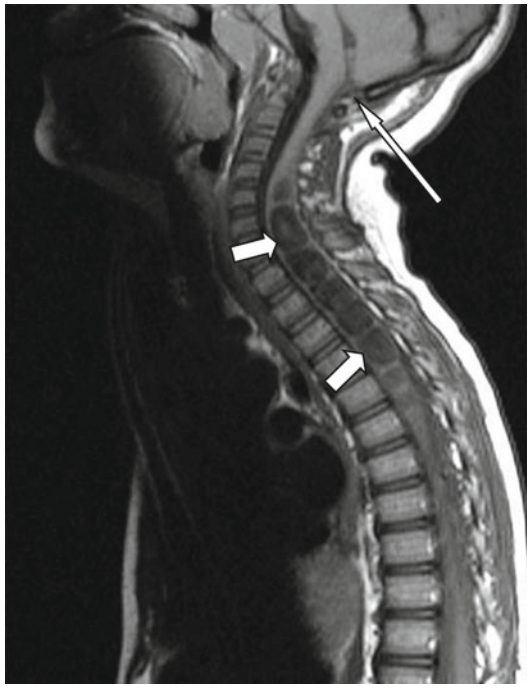

Fig. 13.6 Sagittal T1-weighted MRI of the Chiari 0 malformation. Note the absence of cerebellar tonsillar herniation (*long arrow*) and large cervicothoracic syrinx (*short arrows*) that resolved following posterior fossa decompression

dysfunction. For infants and neonates who present with respiratory distress and stridor, the timing of decompression is important. Beyond this age group, approximately 10 % of patients who survive will have late symptoms of brainstem dysfunction (Talamonti et al. 2007). Continued well-child surveillance and family education can therefore help to ensure that late presentations of brainstem compression do not go unrecognised. The importance of having a functioning shunt in this patient population should again be stressed. Obstructive apnoea, which can have a mortality rate as high as 60 %, can be reversed by an optimally functioning shunt (Cochrane et al. 1991; Hesz and Wolraich 1985; Soleau et al. 2008).

The extensive laminectomy required to decompress a Chiari II malformation increases the risk of delayed cervical instability, and the patients should undergo post-operative cervical spine evaluation (Oakes 1991).

13.4 Chiari 0

The term Chiari 0 malformation has been used to describe a subset of patients with syringomyelia and without cerebellar tonsillar ectopia but with observed 'crowding' of the posterior fossa (Tubbs et al. 2001; Weprin and Oakes 2001). These patients do not have caudal displacement of their cerebellar tonsils, beyond that which is considered to be within the normal anatomical range (Soleau et al. 2008). Analysis of the posterior fossa in these patients has, however, demonstrated that they have caudal displacement of their brainstems, with a low-lying obex and a hypoplastic posterior fossa (Weprin and Oakes 2001). These patients are considered to have impaired circulation of CSF at the outlet of the fourth ventricle and across the craniovertebral junction. In addition, those that come to surgery are often found to have barriers to free egress of CSF. The first published series described five patients with syringomyelia but without hindbrain herniation. They all underwent posterior fossa decompression that entailed a suboccipital craniectomy, removal of the posterior arch of C1 and duraplasty. Additionally, the patients underwent lysis of adhesions, opening of the fourth ventricular veil (if present) and partial resection of one of the cerebellar tonsils. Four patients had resolution of their symptoms post-operatively, and all went on to display a diminution in the size of their syringes (Iskandar et al. 1998).

13.5 Scoliosis

There is a particularly marked association between scoliosis and syringomyelia in children. One review identified scoliosis in 4 out of 5 syringomyelia patients under 20 years of age, compared with 1 in 6 older patients (Isu et al. 1990). Scoliosis is the presenting feature of syringomyelia in as many as two thirds of children (Isu et al. 1992). Features that alert an orthopaedic surgeon to the possibility of such underling cord pathology include male sex and curves that are convex to the left; idiopathic curves are more commonly convex to the right and have a predilection for girls. The presence of neurological symptoms and

signs as well as rapid progression of a curve also suggest a neurogenic aetiology. Extensive cord cavitations, comprising 50 % or more of the cross-sectional diameter of the cord, are more likely to experience progression of their scoliosis.

Our understanding of the pathophysiological mechanisms underlying the association between syringomyelia and scoliosis is limited. A syrinx cavity is rarely symmetrical in its transverse dimensions, so there is usually asymmetrical impairment of function in the anterior horn cells. The assumption is that the resultant imbalance in the innervation to the axial musculature results in progressive spinal curvature. Alternatively, abnormal intramedullary pressure may interfere with postural tonic reflexes (Eule et al. 2002). It is generally accepted in veterinary medicine that scoliosis occurs secondary to dorsal grey column damage and disruption of proprioceptive information coming from Golgi tendon organs (see Chap. 14). In animal models, sectioning dorsal roots can be followed by development of scoliosis (Alexander et al. 1972; Liszka 1961; MacEwen 1973), although paraspinal muscle dissection and laminectomy may play a role in the development of scoliosis in these animals. It is important to bear in mind that syrinx cavities may have diverse aetiologies (e.g. post infectious, and post traumatic) and in some instances neurological damage and spinal innervation may be compromised by mechanisms other than, or in addition to the syrinx. Additionally, in the paediatric population congenital vertebral malformations are common and may co-exist with syrinx cavities however the causal relationship between these two entities is by no means clear. That there is a causal relationship between syringomyelia and scoliosis is now generally accepted and supported by many studies (Bertran et al. 1989; Muhonen et al. 1992), however, it is not always clear whether a syrinx is the cause of or a consequence of a scoliosis. Greitz argued that the spinal subarachnoid pressure is lower adjacent to a syrinx, as a result of the Bernoulli principle (Greitz 2006). This states that an increase in fluid velocity, as might occur where the subarachnoid space is narrowed (as for example at the apex of a scoliotic curve), is accompanied by a decrease in the pressure exerted by that fluid. This drop in CSF pressure increases the pressure gradient between the spinal cord parenchyma and subarachnoid space, with the result that fluid is drawn from the cord capillary bed into the parenchyma, initiating formation of a syrinx. If we apply this reasoning to a case of scoliosis, then it is possible that the spinal curvature might compromise the streamline flow of CSF in the subarachnoid channels and, thereby, encourage the accumulation of fluid within the substance of the cord. Furthermore, an idiopathic syrinx can also occur in the absence of scoliosis, the finding of syringomyelia in association with a scoliosis may be simply coincidence.

Table 13.1 Clinical associations between Chiari malformation, syringomyelia and scoliosis

Chiari with syrinx and scoliosis
Idiopathic syrinx with scoliosis
Chiari with scoliosis but no syrinx

The issue of the Chiari I malformation adds a further dimension, and trying to unravel the interaction between hindbrain hernia, syringomyelia and scoliosis is challenging, particularly as regards which is the primary anomaly and which component of this triad should receive our initial attention. Three scenarios that are commonly encountered in paediatric neurosurgical practice (Table 13.1).

13.5.1 Scoliosis and Chiari-Related Syringomyelia

The increasing trend for all paediatric scoliosis patients to be evaluated with MRI has resulted in the identification of a variety of intraspinal anomalies, of which syringomyelia is the most common (Diab et al. 2011; Lewonowski et al. 1992). Further, the term syrinx is frequently used to cover a spectrum of appearances, ranging from short segments of hydromyelia, that may be little more than focal dilatations of the central canal, through to larger intramedullary fluid collections, more consistent with classical syringomyelia. Chiari-related syringomyelia is the most commonly seen association with scoliosis in the paediatric population. A series of 500 paediatric Chiari I malformations reported a syrinx to be present in 57 % and scoliosis in 18 % of cases (Tubbs et al. 2011)

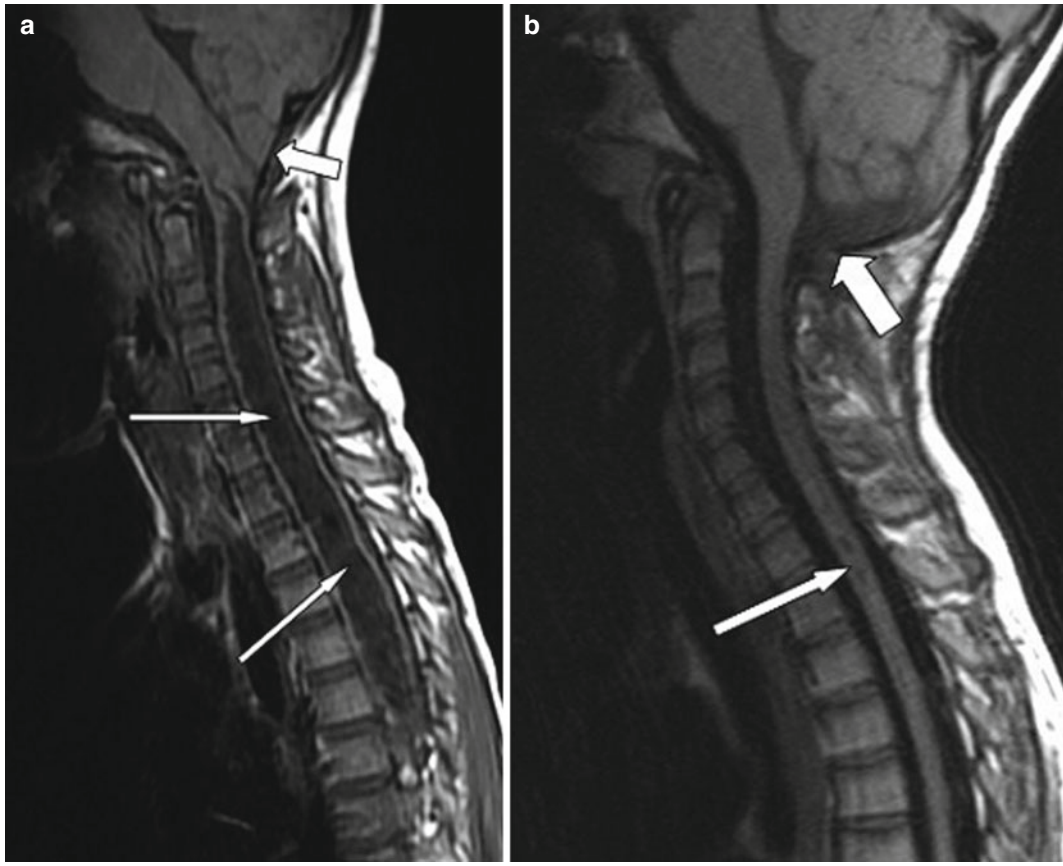

Fig. 13.7 Sagittal T1-weighted MRI scans, before and after craniovertebral decompression. (**a**) Showing syringomyelia (*narrow arrows*) and a Chiari I malformation (*broad arrow*) in a child presenting with scoliosis. (**b**) Following foramen magnum decompression and duraplasty (*broad arrow*), there has been resolution of the syrinx (*narrow arrow*). There has also been some improvement in the sagittal alignment of the spinal column. This is revealed by scrutiny of the lower part of the images, where the midline of the lower vertebral bodies lies outside the plane of the image before surgery, whereas they lie in the midsagittal plane after surgery

Abnormal CSF flow in the region of the foramen magnum can also lead to syringomyelia formation, in the absence of Chiari malformation – the Chiari 0 condition (Klekamp et al. 2002; Kyoshima et al. 2002). As with syringomyelia associated with Chiari I and II malformations, scoliosis may coexist. In some of these cases, foramen magnum decompression and intradural exploration can result in improvement in the syrinx (Fig. 13.7) (Goel and Desai 2000; Iskandar et al. 1998). The scoliosis may improve as well, and in one series of 15 children with Chiari 0, eight had scoliosis and foramen magnum decompression led to the curve improving in four cases and stabilising in another three (Chern et al. 2011).

13.5.2 Scoliosis and Idiopathic Syringomyelia

The natural history of idiopathic syringomyelia in the paediatric population seems to be benign. In a study of 48 children with an idiopathic syrinx, followed over a 2½ year period, 9 out of 10 either improved or remained clinically stable and asymptomatic. Almost all of these cavities also remained stable radiologically or reduced in size (Magge et al. 2011). In those children who had a scoliosis as well, neither the site nor size of the syrinx correlated with the severity or progression of the scoliosis. In another study of children with apparently idiopathic scoliosis, neither the presence of syringomyelia nor Chiari

had any bearing on the eventual outcome of the scoliosis surgery (Diab et al. 2011). Orthopaedic surgeons are, however, likely to continue screening scoliosis children because associated bony malformations, such as hemivertebrae or other structural anomalies increase the chance of scoliosis surgery being required (Magge et al. 2011).

13.5.3 Scoliosis and Chiari Without Syringomyelia

Occasionally, in the investigation of scoliosis, MRI will reveal a Chiari I malformation but no demonstrable syrinx. This was the case in 2 of 22 patients in one study (Brockmeyer et al. 2003). It has been suggested that direct compression on the cervicomedullary junction, by the impacted cerebellar tonsils, might interfere with spinal cord function, providing the impetus to initiate and then propagate the scoliosis (Brockmeyer 2011).

13.5.4 Treatment

A number of reasons have been put forward to justify treating syringomyelia in the child presenting with scoliosis (Table 13.2). It is important to emphasise that the following discussion pertains to the effect of syrinx treatment on the scoliosis. Clearly, there may be concomitant neurological indications to treat a syrinx, and neurological features may improve or stabilise after syrinx drainage procedures, despite there being no demonstrable benefit in terms of scoliosis progression (Farley et al. 1995).

Numerous articles support the assertion that scoliosis can be improved or at least stabilised by treating the underlying syringomyelia (Brockmeyer et al. 2003; Eule et al. 2002; Isu et al. 1992; Krieger et al. 2011; Muhonen et al. 1992; Nohria and Oakes 1990; (Ozerdemoglu et al. 2003). This approach, however, needs qualification. Age, for example, seems to be a pertinent factor, and scoliosis is more likely to improve after syrinx treatment in young children compared with adolescents and young adults (Brockmeyer et al. 2003; Ozerdemoglu et al. 2003). The severity of the deformity may also influence the outcome, with curves less than 40° often being halted by early posterior fossa decompression, whereas more severe curves are less likely to improve (Eule et al. 2002). There may also be other aetiological factors influencing outcome, besides the syrinx itself. Congenital scoliosis and scoliosis associated with myelomeningocoele will, for example, have a much lower chance of improvement following treatment of an accompanying syrinx (Ozerdemoglu et al. 2003). The surgical procedure used to treat the syrinx is also likely to be important. After the foramen magnum decompression, the proportion of patients in whom scoliosis improves or is stabilised ranges widely, from 25 % to more than 80 % (Brockmeyer et al. 2003; Eule et al. 2002; Muhonen et al. 1992; Sengupta et al. 2000). The success rate of direct syrinx drainage procedures on the evolution of scoliosis is considerably less. In one group of four patients, progression of the curve was seen in all, in spite of syringe-arachnoid shunting (Phillips et al. 1990). Such variable results may, of course, reflect differences in the severity of the scoliosis pre-operatively, as well as the child's age at the time of surgery.

Another commonly quoted argument in favour of treatment of a syrinx, prior to correction of a scoliosis, is that its decompression makes subsequent correction of the scoliosis safer. The presence of syringomyelia could increase the risk of neurological complications of scoliosis surgery in various ways. The syrinx might make the spinal cord more vulnerable to direct injury, the spinal cord vasculature might be compromised or adhesions around the spinal cord might make it vulnerable to traction forces (Phillips et al. 1990). Evidence to support such assertions is, however, weak and largely stems from case reports and anecdotal experi-

Table 13.2 Suggested reasons for treating syringomyelia associated with scoliosis

Favourably influences the natural history of the scoliosis?
Safer subsequent correction of the scoliosis?
Tonsillar herniation represents a risk under anaesthesia?

ence (Huebert and MacKinnon 1969; Noordeen et al. 1994; Nordwall and Wikkelso 1979). In one study neurological complications were seen in 3 out of 38 patients whose scoliosis was treated without prior syrinx treatment, compared with 1 out of 37 patients where there had been prior intervention to treat syrinx (Ozerdemoglu et al. 2003). In a large multicentre study of idiopathic adolescent scoliosis, 1 in 10 patients who were screened with MRI were found to have an abnormality of the neuraxis. Anomalies included Chiari malformation in 1 in 3 cases and syrinx in 2 out of 3 cases (Diab et al. 2011). None of these patients received neurosurgical intervention prior to scoliosis surgery, and there was no increased morbidity observed in these children. This suggests that, in the absence of symptoms, syringomyelia cavities do not require treatment as a prelude to treating the scoliosis. More recent and larger studies have also shown how benign idiopathic syringomyelia can be and have questioned the dogma that syrinx treatment is essential, to reduce the risk of spinal orthopaedic surgery (Diab et al. 2011 ; Magge et al. 2011). This being so, we may also conclude that there is no role for surgical treatment of Chiari malformation in scoliosis patients when there is no associated syrinx or when there are no symptoms arising from the Chiari itself. When a syrinx is detected, then, unless it is unduly large or there are associated clinical symptoms or signs in addition to the scoliosis, then surgical treatment of the syrinx may, once again, not be needed.

With the scoliosis child facing up to corrective surgery, questions may be raised as to the safety of anaesthesia in the presence of a Chiari malformation. For the otherwise asymptomatic patient, there is no clear evidence to support the assertion that compression of the brainstem at the foramen magnum, by the herniated cerebellar tonsils, represents a risk under anaesthesia. Chiari I malformations can, however, be associated bony anomalies at the craniovertebral junction and can then, occasionally, cause bulbar dysfunction. This may lead to problems such as central respiratory disturbance, vocal cord paresis and reduced pharyngeal sensation. Whilst these disturbances may well be of anaesthetic relevance, it is difficult, in the absence of clear clinical symptoms of bulbar compromise, to identify an evidence base to support posterior fossa decompressive surgery, solely to ameliorate an anaesthetic risk.

13.6 Summary

Chiari malformations share the common pathology of impaired cerebrospinal fluid circulation through the foramen magnum. Children with symptomatic Chiari malformations and accompanying syringomyelia should be managed by first treating elevated intracranial pressure if present, followed by adequate bony decompression of the cerebellum and brainstem with intradural exploration and duraplasty. With this approach, the vast majority of patients will have resolution of their pre-operative symptoms, diminution of their syrinx and sustained clinical stability.

The interplay between syrinx, Chiari and scoliosis is complex, and the evidence base to guide neurosurgical management in these cases is incomplete and sometimes conflicting. It is clear that there are some patients in whom neurosurgical intervention to decompress a Chiari malformation and improve a syrinx will beneficially affect the natural history of scoliosis, particularly when that intervention is performed at young age and at an early stage in the evolution of the spinal deformity. There are, however, many instances in which a Chiari or a syrinx is identified, simply as a result of screening with MRI and here the need to intervene with a neurosurgical procedure should be evaluated critically. The risks of Chiari or syrinx surgery are not insignificant, and any surgical risk needs particular justification in the case of a neurologically asymptomatic patient. On the basis of current evidence, it is not possible to define rigorous criteria to guide patient selection for surgery. Perhaps more appropriate is a recommendation that scoliosis patients harbouring an abnormality of the neuraxis receive a comprehensive, individualised evaluation, by a neurosurgeon, looking carefully for additional symptoms or signs, which together strengthen the argument for a neurosurgical procedure.

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