Hindbrain-Related Syringomyelia

10

Jörg Klekamp

Contents

10.1 Anatomical Abnormalities

About one in every two patients with syringomyelia demonstrates pathology at the craniocervical junction. Any pathology in this region, which compromises the passage of cerebrospinal fluid (CSF), may lead to syringomyelia. The most common is the Chiari I malformation. Less common entities are basilar invagination, Chiari II malformation and foramen magnum arachnoiditis.

Hans Chiari described four varieties of malformations in his monograph (Chiari 1896). The most common is the type I, which is characterised by herniation of cerebellar tonsils into the spinal canal. In type II, the brainstem, the cerebellar tonsils and part of the vermis are displaced into the spinal canal. In type III, the features of type II are combined with an occipital meningoencephalocoele. Type IV is characterised by hypoplasia of the vermis. Chiari considered these abnormalities to be causally related to hydrocephalus. Modern imaging techniques and experimental studies, however, disclose a different aetiology.

10.1.1 Chiari I Malformation

It has been shown, in the majority of patients, that Chiari I malformation is a disorder related to a small posterior fossa volume, forcing the tonsils into the spinal canal (Stovner et al. 1993; Badie et al. 1995; Trigylidas et al. 2008; Nyland and Krogness 1978; Nishikawa et al. 1997; Boyles et al. 2006; Milhorat et al. 1999)

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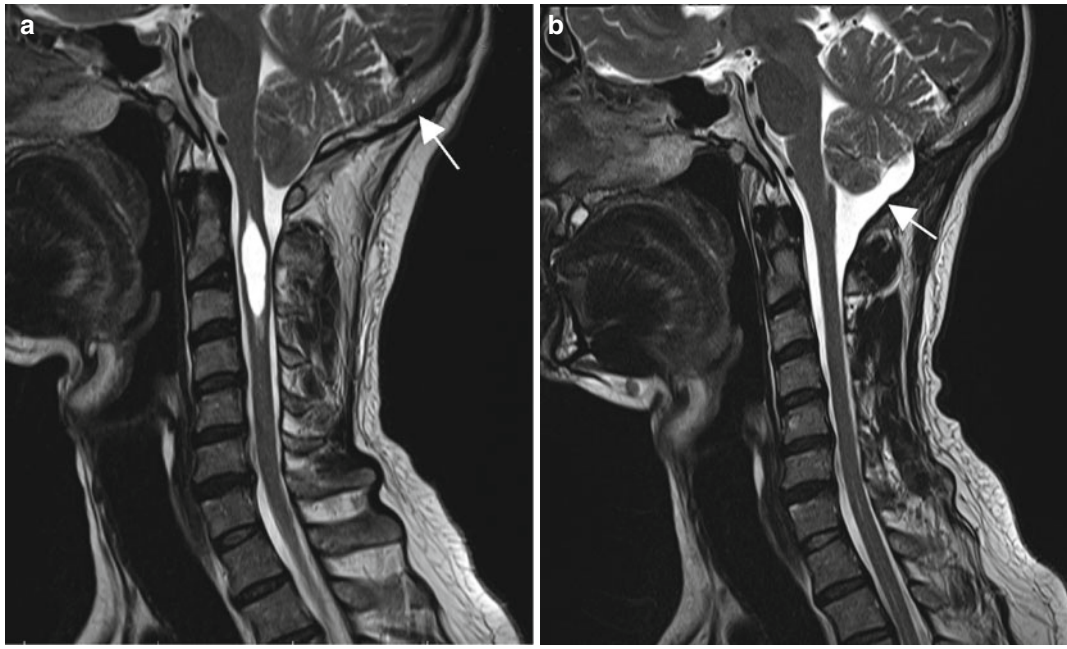

Fig. 10.1 (**a**) Preoperative T2-weighted MRI of a patient with Chiari I malformation. The image demonstrates a small posterior fossa, a slight caudal displacement of the tentorial insertion (*arrow*) and a syringomyelia at C2–C3. (**b**) The postoperative scan shows the decompression at the foramen magnum (*arrow*) with resolution of the syrinx

(Fig. 10.1). Marin-Padilla could demonstrate this effect in hamsters more than 30 years ago (Marin-Padilla and Marin-Padilla 1981). There may be a genetic disposition in some patients (Boyles et al. 2006; Milhorat et al. 1999; Tubbs et al. 2011). Not all Chiari I malformations are caused by a small posterior fossa, and they may develop after lumboperitoneal shunting (Payner et al. 1994; Chumas et al. 1993), birth trauma (Aghakhani et al. 1999; Hida et al. 1994; Williams 1977), arachnoid pathologies at the craniocervical junction (Aghakhani et al. 1999), posterior fossa arachnoid cysts (Galarza et al. 2010) or solid tumours in the posterior fossa (Klekamp et al. 1995).

Apart from a small posterior fossa volume, additional bony anomalies are common in Chiari I malformation and may involve the articulations at the craniocervical junction. Assimilations of the atlas to the occiput, basilar invaginations or Klippel-Feil syndromes may also be encountered (Kagawa et al. 2006; Tubbs et al. 2011; Smith et al. 2010). It is important to recognise that the compression of neural structures and CSF flow obstruction are localised at the foramen magnum in all variants of Chiari I malformation although these may not be the only mechanisms responsible for the patients' symptoms. Instabilities of the craniocervical junction or upper cervical spine are important features to recognise in a significant proportion of Chiari I patients.

10.1.2 Basilar Invagination

Basilar invagination is defined as a protrusion of the odontoid peg into the foramen magnum (Fig. 10.2). A line between the posterior rim of the foramen and the hard palate constitutes Chamberlain's line. If the odontoid crosses this line for more than 2.5 mm, this is considered pathological. Basilar invagination may be associated with osteogenesis imperfecta, Hajdu-Cheney syndrome, Paget's disease (Menezes 2008b), Marfan's syndrome (Hobbs et al. 1997), Down's syndrome (Menezes 2008a) or rheumatoid arthritis (Krauss et al. 2010). The congenital form is caused by bony anomalies of the

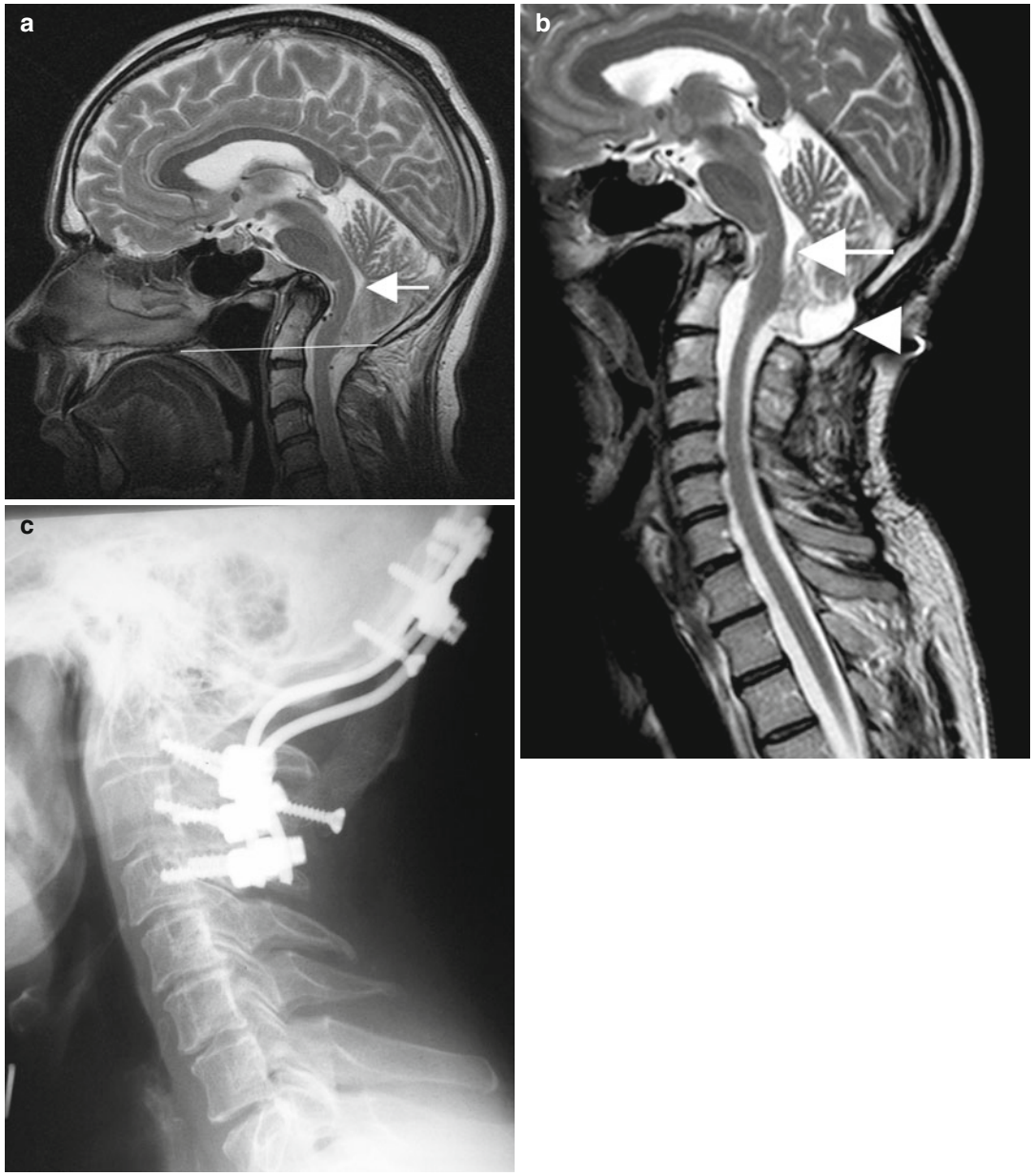

Fig. 10.2 (**a**) The preoperative T2-weighted MRI shows a Chiari I malformation associated with a profound basilar invagination and compression of the brainstem (*arrow*). The odontoid extends far above Chamberlain's line (*white horizontal line*). (**b**) The postoperative MRI demonstrates the result of a combined decompression with transoral resection of the dens (*arrow*) and posterior decompression (*arrowhead*) and fusion. (**c**) The postoperative lateral radiograph demonstrates the position of all implants and a good sagittal profile of the cervical spine

clivus, occipital bone, atlas and upper cervical vertebrae. The result of this altered anatomy is a gradual upward shifting of the upper cervical vertebrae towards the foramen magnum. The C1/2 intervertebral joints appear to play a major role for this effect as distraction of these joints may reverse the ventral compression by the odontoid peg (Jian et al. 2010; Goel 2004).

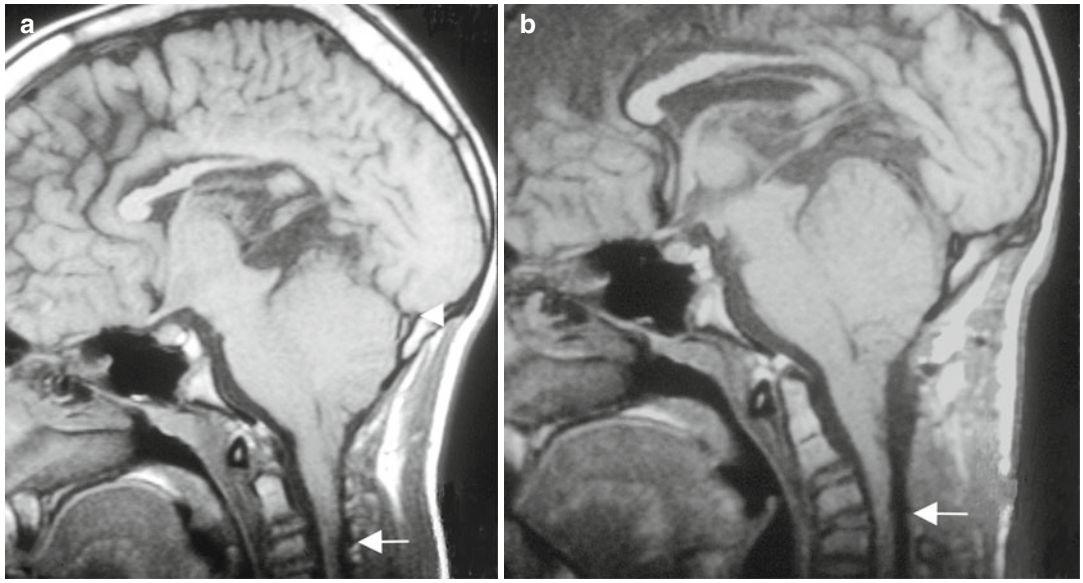

Fig. 10.3 (**a**) Preoperative T1-weighted MRI of a 14-year-old boy with a Chiari II malformation, demonstrating the enlarged foramen magnum with cerebellar tonsillar herniation to C3 (*arrow*). The tentorium inserts close to the posterior margin of the foramen magnum (*arrowhead*). (**b**) The postoperative image demonstrates the decompression of the upper cervical spine and discloses a kyphotic angulation at C3/4 (*arrow*)

10.1.3 Chiari II Malformation

In Chiari II malformation, the compression and cerebrospinal fluid flow obstruction occur in the upper spinal canal and not at the level of the foramen magnum. In contrast with Chiari I, the foramen magnum is enlarged in Chiari II, and the tonsils, the vermis and the brainstem are all herniated into the cervical canal (Fig. 10.3). Almost all patients with this malformation will also have a spinal myelomeningocoele. The pathophysiology of this malformation has been elegantly described by McLone and Knepper. Due to the spinal myelomeningocoele, CSF drains in utero into the amniotic fluid, resulting in a low intracranial pressure, which then inhibits the formation of a normally sized posterior fossa. The growth of the brain finally leads to herniation of cerebellar tonsils, vermis and brainstem into the spinal canal (McLone and Knepper 1989). As in the majority of Chiari I patients, the size of the skull forces the brain to grow towards the spinal canal in Chiari II. The major difference is the timing: in Chiari I, this effect takes place after birth, whereas in Chiari II the major pathological changes occur before birth with much graver consequences. Support for this hypothesis comes from results of intrauterine operations on myelomeningoceles before the 26th week of gestation. If the spinal dysraphism could be closed successfully, then no Chiari II malformation developed (Danzer et al. 2011; Tulipan et al. 1999).

10.1.4 Foramen Magnum Arachnoiditis

Foramen magnum arachnoiditis is the only pathology at the craniocervical junction associated with syringomyelia without there being additional compression of brainstem or spinal cord (Fig. 10.4). Arachnoiditis at this level may be related to a previous episode of meningitis or trauma or other causes of haemorrhage (Klekamp et al. 2002; Appleby et al. 1969).

10.2 Neuroradiology

The diagnosis of Chiari I malformation should be straightforward these days, given the wide accessibility of magnetic resonance imaging

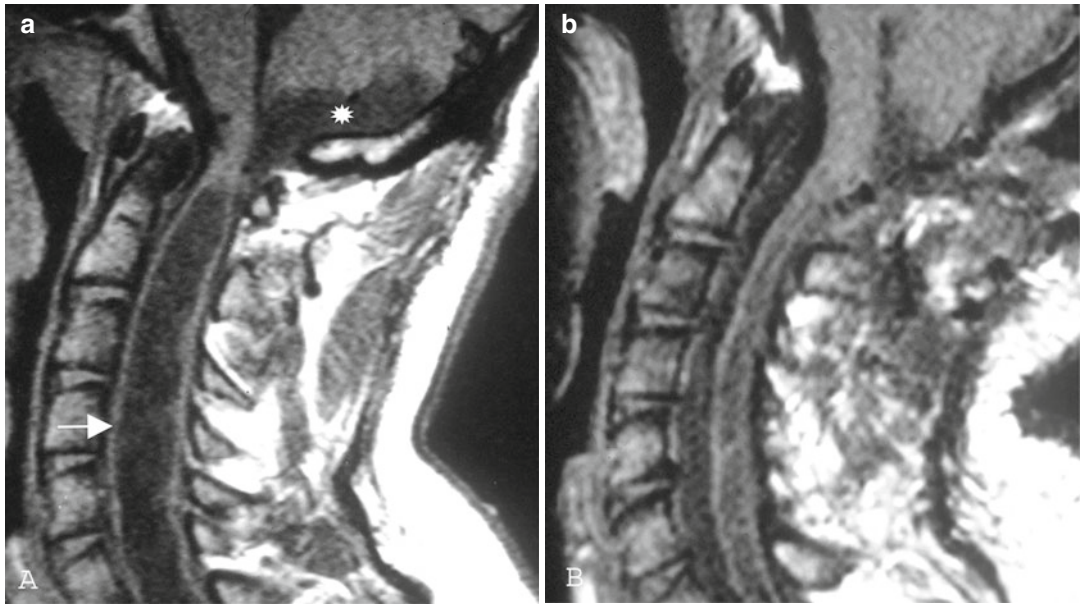

Fig. 10.4 (**a**) Preoperative T1-weighted MRI indicates foramen magnum arachnoiditis related to birth injury in a 29-year-old man with progressive tetraparesis. There is not a cerebellar tonsillar herniation, but there is an arachnoid pouch close to the foramen magnum (*) and a significant syrinx (*arrow*). (**b**) The postoperative MRI shows a collapse of the arachnoid pouch and the syrinx

(MRI) in western countries. To what extent tonsillar herniation may sometimes be seen as physiological and how neuroradiological criteria should be defined are, however, still a matter of controversy. A tonsillar herniation of more than 5 mm is widely considered pathological in adults (Aboulezz et al. 1985), but in young children, cerebellar growth causes a physiological herniation of the cerebellar tonsils. Conversely, in old age, atrophy of the brain may lead to tonsillar ascent (Mikulis et al. 1992). Tonsillar descent of less than 5 mm does not exclude the diagnosis of a Chiari I malformation (Milhorat et al. 1999), and in doubtful cases cardiac-gated cine MRI is very helpful to demonstrate a CSF flow obstruction and a clinically relevant herniation (Haughton et al. 2003; Panigrahi et al. 2004; Ellenbogen et al. 2000; Tubbs et al. 2007; Milhorat et al. 1999; Hofkes et al. 2007). Likewise neurophysiological examinations (Henriques Filho and Pratesi 2006) and neuro-otological evaluations (Kumar et al. 2002) have been proposed, as means of providing evidence of compression of the medulla oblongata or spinal cord.

In patients with Chiari I malformation, the radiological examination should include more than simply defining how far the tonsils are descended into the spinal canal. It is also important to consider the bony anatomy of the craniocervical junction. For example, is there evidence of anterior compression by the odontoid peg, i.e. basilar invagination (Fig. 10.2)? Is the atlas assimilated to the occiput? Are cervical segments fused, i.e. Klippel-Feil syndrome? If these anomalies are present, then instability of the craniocervical junction must be ruled out with flexion and extension studies, using either conventional radiography or CT imaging. The latter has the advantage of being able to visualise each of the different joints, in multiple planes and in both flexion and extension. In addition, sagittal and coronal reconstructions are particularly useful.

For all craniocervical pathologies, ventricular sizes should be evaluated. In Chiari I malformation, overt hydrocephalus is rare, but some degree of ventricular enlargement is not uncommon and was observed in 9 % of the author's series. With Chiari II malformation, on the other hand, hydrocephalus is almost ubiquitous.

In fact, it is uncommon for the neurosurgeon to encounter such a patient who has not already been managed with a ventricular shunt. If a Chiari II patient is evaluated because of new neurological symptoms, the first priority must be to assess the function of a previously implanted ventricular shunt. This requires a comparison of recent and old CT or MRI images, to look for changes of ventricular size that may indicate under- or over-drainage.

One important aspect for surgical planning in Chiari patients is the position of the tentorium, in relation to foramen magnum and the external occipital protuberance. Normally, the tentorium will insert at the level of this protuberance, and this indicates the position of the large intradural sinuses, such as the transverse sinus and the confluence of sinuses. In Chiari I patients, this tentorial insertion may be shifted by a centimetre or more towards the foramen magnum. When planning an occipital craniectomy, this must be taken into account and is even more important with regard to the dural incision. In Chiari II malformation, the foramen magnum is widened, and the tentorium usually inserts at the level of or very close to the foramen magnum (Fig. 10.3). For this reason, the suboccipital dura should not be incised in patients with a Chiari II malformation.

In patients with primary foramen magnum arachnoiditis, bony anomalies will not be apparent. The diagnosis is made by MRI and, in the absence of tonsillar herniation and brainstem or cervical cord compression, will require a cine MRI to demonstrate a CSF flow obstruction in the foramen magnum area. Such obstruction often involves the fourth ventricle exit foramina as well, and ventriculomegaly is common, being observed in half of the author's series of foramen magnum arachnoiditis. Another common feature of primary foramen magnum arachnoiditis is the formation of arachnoid cysts or pouches in the posterior fossa (Fig. 10.4).

Finally, the presence or absence of an associated syringomyelia or syringobulbia should be demonstrated as well as the entire extension of the cavity, to rule out additional abnormalities such as a tethered cord.

10.3 Clinical Presentation

With all craniocervical pathologies, clinical symptoms may evolve from different pathophysiological components. These may include hydrocephalus, compression of the brainstem and spinal cord, craniocervical instability, disturbances of brainstem or spinal cord blood flow, tethering mechanisms related to chronic arachnoiditis and CSF flow obstruction leading to syringomyelia. Clinical and radiological examinations therefore need to be analysed carefully, to identify the appropriate targets for treatment.

Considering all Chiari patients, it is interesting to note that the presentation is very much dependent on the age of the patient. In early childhood below age 2 years, signs of brainstem compression predominate, with apnoeic spells, cyanosis attacks and swallowing problems, whereas in later childhood scoliosis becomes the most common presenting sign. What are regarded as the more typical clinical features of a Chiari I malformation – occipital headaches, gait ataxia, sensory disturbances and motor weakness – are uncommon in children and are observed predominantly in adults (Rauzzino and Oakes 1995; Menezes et al. 2005) (Table 10.1). This age-related clinical profile can be explained by the postnatal growth of the cerebellum. At birth, most parts of the brain have reached about a third of their adult volume, but the cerebellum is the smallest part of the central nervous system, with just 15 % of its adult volume at this time; presumably, this serves to protect the brainstem during delivery. The adult volume of the cerebellum is reached late in the second year of life, indicating that the cerebellar volume increases by a factor of seven in that period (Klekamp et al. 1989). Therefore, if a Chiari malformation does become symptomatic before 2 years of age, dramatic presentations with respiratory problems are likely to be observed, something unknown in adult patients. Once the cerebellum is fully grown, the clinical course tends to be less dramatic and is characterised by slow progression.

As with every rule, however, there are exceptions. Minor traumas may cause acute symptoms in formerly asymptomatic patients with Chiari I

Table 10.1 Preoperative neurological symptoms (author's series)

Group	Occipital pain (%)	Neuropathic pain (%)	Hyperaesthesia (%)	Gait (%)	Motor power (%)	Sphincter function (%)	Swallowing difficulties (%)
Chiari I	79	50	71	62	40	16	20
Chiari II	25	19	64	100	67	92	22
FMA	74	39	83	78	83	48	15

The percentages given in this table represent the total number of patients presenting with a particular symptom
FMA foramen magnum arachnoiditis

malformation (Yarbrough et al. 2011; Murano and Rella 2006). In extremely rare cases, even sudden deaths have been reported (Wolf et al. 1998; Stephany et al. 2008; Agrawal 2008; Yoshikawa 2003). This raises questions as to whether asymptomatic children with Chiari I malformations should be allowed to participate in sport activities and whether prophylactic surgery for such patients is warranted. Chiari decompressions certainly cannot be considered to be no-risk procedures and are associated with a mortality of about 1 %. Given the fact that severe neurological deficits after minor trauma are extremely rare – no such instance was reported by a single patient in the author's series of more than 600 patients – it appears reasonable to leave decisions regarding timing of surgery and which sport activities can be pursued to the patients and their parents, without pressing them one way or the other.

In the author's series, patients with Chiari I presented with an average age of 43 ± 16 years, while those with Chiari II tended to be younger, at 17 ± 13 years. Compared to patients with a Chiari I malformation, patients with foramen magnum arachnoiditis presented at a similar age (37 ± 10 years) but with more severe neurological deficits (Table 10.1). Interestingly, this appeared not to hold for swallowing dysfunctions, which were more common with Chiari malformations, despite the significant scarring at the medulla oblongata level, which was present in all patients with foramen magnum arachnoiditis. The length of history was significantly longer for patients with arachnoiditis compared to Chiari patients (101 ± 96 months, compared to 71 ± 106 months for Chiari I and 26 ± 39 months for Chiari II).

10.4 Management

10.4.1 Chiari I Malformation

For all Chiari I patients, treatment of symptomatic hydrocephalus should be prioritised. Endoscopic third ventriculostomy is now preferred to traditional ventriculoperitoneal shunting as the first option for CSF diversion whenever patients present with clinical signs of raised intracranial pressure (Massimi et al. 2011).

Management options for arachnoid cysts include resection or fenestration (Fig. 10.5). Insertion of cyst shunts is not considered by the author due to their high failure rate. Treatment for Chiari I malformations due to solid masses requires tumour resection with foramen magnum duraplasty (Fig. 10.6).

For Chiari I hindbrain hernias, there is general agreement that surgical treatment should be reserved for symptomatic patients. In children, however, it may be unclear which complaints are linked to the malformation. It can also be a challenge to differentiate between physiological and pathological tonsillar descent, as revealed by MRI. In the absence of neurological symptoms or progressive scoliosis, the author does not recommend decompression for children with a Chiari I malformation, unless they demonstrate the typical occipital headache provoked by Valsalva-like manoeuvres, such as sneezing or coughing.

With all patients, it should be borne in mind that not every headache is due to tonsillar herniation, even when such an abnormality is present. Nor does the presence of syringomyelia constitute an indication for surgery in its own right, in children or adults alike. When neurological symptoms are present, however, surgery should

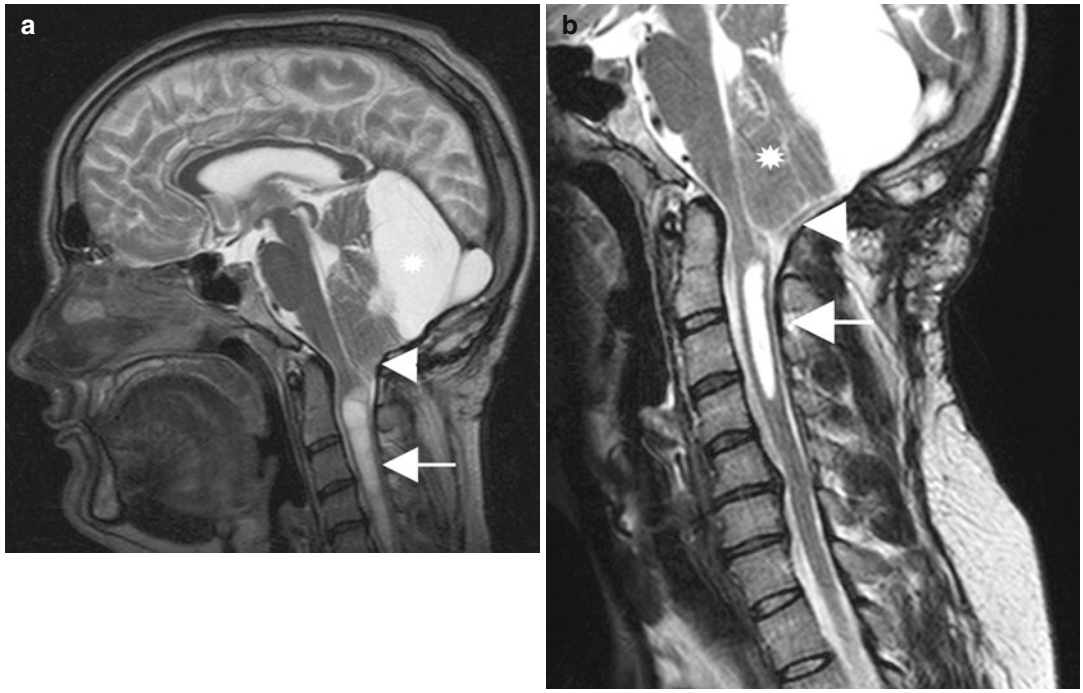

Fig. 10.5 (**a**) Preoperative T2-weighted MRI of a patient with syringomyelia (*arrow*) and a cerebellar tonsillar herniation (*arrowhead*) secondary to a large retrocerebellar arachnoid cyst (*). (**b**) The postoperative scan demonstrates a decrease in syrinx size (*arrow*) and a decompression at the foramen magnum (*arrowhead*). Despite large fenestration of the arachnoid cyst, the vermis has not changed its shape (*)

be recommended. As a general rule, progression of neurological symptoms occurs more rapidly than does enlargement of an underlying syrinx. The author has not observed enlargement of a syrinx in an asymptomatic patient with a Chiari I malformation.

Even though foramen magnum decompression is widely accepted as the treatment of choice for Chiari I malformation, there is no general agreement on how this operation should be performed (Schijman and Steinbok 2004; Haroun et al. 2000). Gardner's original operation consisted of a wide craniectomy of the posterior fossa and opening of the 4th ventricle in order to plug the obex with a piece of muscle. The dura was left open. He reported 5 mortalities after 74 such procedures (Gardner 1965). Similar mortality rates, as well as significant morbidity caused by manipulations at the obex (Williams 1978), led other surgeons to modify this operation. Guided by modern imaging techniques such as MRI in the 1980s and modern CT scanners, the anatomy of patients with Chiari I malformation could be studied in much more detail than was previously possible. This led to less invasive procedures, such as leaving the arachnoid intact after dural opening (Logue and Edwards 1981), incising only the outer dural layer (Gambardella et al. 1998) or even restricting the operation to a purely bony decompression (James and Brant 2002). It should, of course, be the intention of every neurosurgeon to restrict any operation to its essential requirements in order to limit surgical morbidity, complications and discomfort for the patient but, at the same time, to do so without compromising the beneficial effects of the procedure.

The following account of a surgical technique describes the author's preferred method, as do subsequent accounts of operative techniques in this chapter. It should be noted, however, that there are many variations, and it is true to say that the only manoeuvre that all methods have in common is removal of bone from the occipital squama.

Surgery is performed in prone position. The decompression is limited to a foramen magnum decompression of 3–4 cm, together with removal

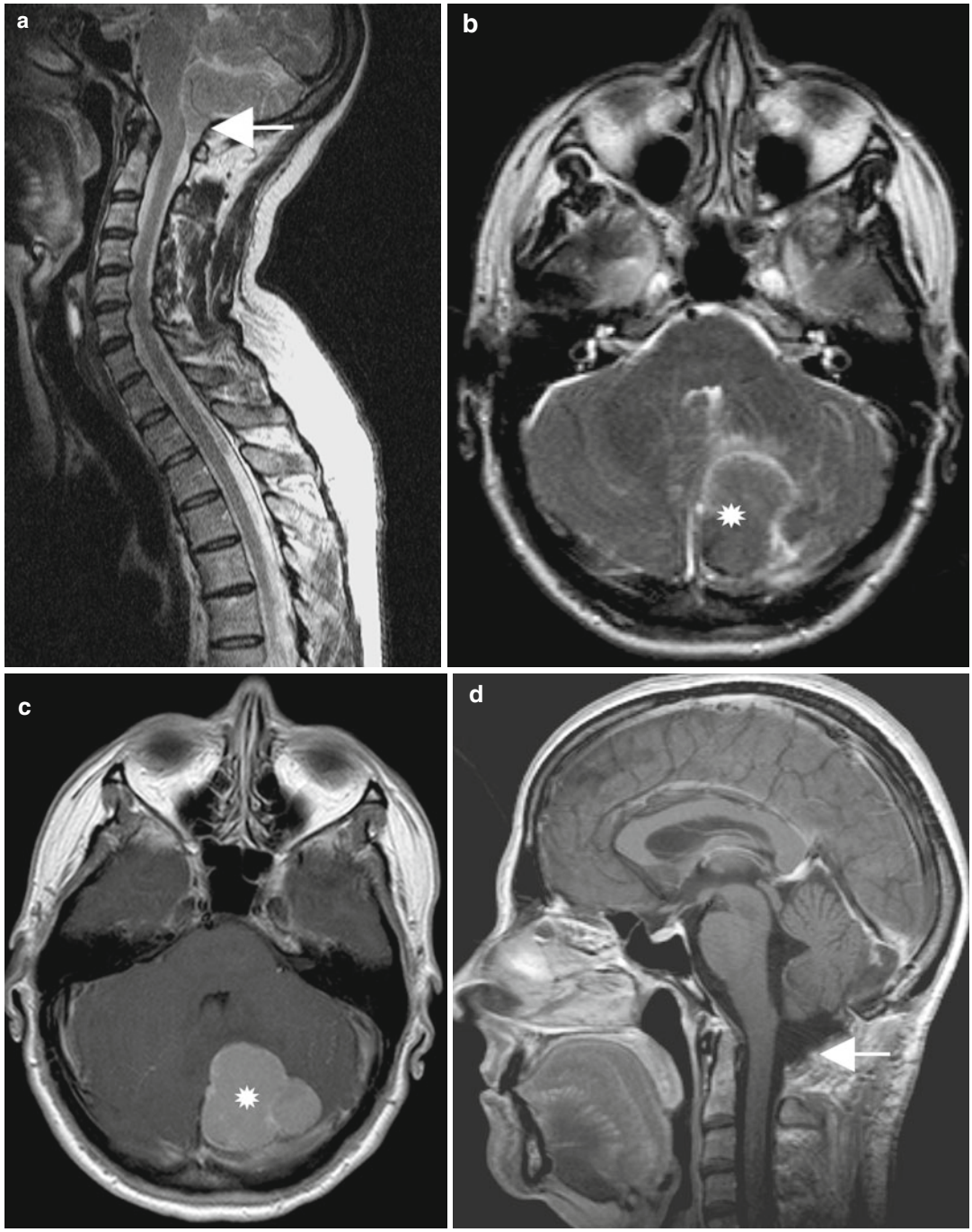

Fig. 10.6 (**a**) This preoperative T2-weighted MRI seems to demonstrate a typical Chiari I malformation (*arrow*). (**b**) The axial T2-weighted image indicates a lesion underneath the tentorium on the left side (*). (**c**) This turned out to be a large meningioma in T1 after contrast (*). (**d**) The postoperative T1-weighted scan with gadolinium shows a complete tumour removal and a decompressed foramen magnum (*arrow*)

of the posterior arch of the atlas. The atlanto-occipital membrane is coagulated and dissected off the dura, which is then incised in a Y-shape, under the microscope, and held open by sutures. Care should be taken to leave the arachnoid intact in order not to pull on and tear underlying bridging

veins or small blood vessels supplying the spinal cord, brainstem or cerebellum. Leaving the arachnoid intact at this stage also avoids contamination of the subarachnoid cisterns with blood. Venous sinuses may be encountered upon opening the dura, in the midline or at the foramen magnum, and these will require suturing. The arachnoid should be examined for evidence of scar formation or adhesions to the cerebellum, brainstem or spinal cord. The arachnoid is then incised, starting below the cerebellar tonsils and continuing to an extent that allows these structures to be spread apart for inspection of the foramen of Magendie. For this purpose, the cerebellar tonsils are coagulated at their tips and medially. Resection of tonsils is advised against as this may risk injury to important blood vessels such as the posterior inferior cerebellar artery (PICA). If Magendie is patent and no arachnoid adhesions are detectable elsewhere, then no further intradural dissections need be performed. If the foramen is obstructed, then it should be opened by sharp dissection. In patients with severe arachnoid scarring, dissection will need to create at least a communication between the cranial and the spinal subarachnoid channels. In such instances, it may not always be possible to open Magendie without risking injury to important structures, such as the PICA. The dissection should not, in these circumstances, be carried out laterally, to avoid injury to perforating vessels of brainstem or spinal cord. A duraplasty is then inserted using alloplastic material. To avoid formation of adhesions between nervous tissue and the duraplasty or suture line, the graft is lifted off the cord by tenting sutures, which are fixed to muscle attachments laterally. Finally, the wound is closed, paying particular attention to the muscular layer in order to avoid CSF fistulas. Postoperatively, all patients should be supervised on the intensive care unit for at least 24 h before returning to the normal ward.

10.4.2 Basilar Invagination

In patients with additional basilar invagination, a combination of ventral and dorsal compression may be associated with instability of the craniocervical junction. These pathophysiological components may not, however, be relevant in all affected patients. Of a group of 53 patients with basilar invagination in the author's series, 35 were managed surgically. In 16 patients, there was neither a ventral compression by the odontoid nor craniocervical instability. These patients were managed with foramen magnum decompression for their Chiari malformation as the only procedure. In another 10 of the 35 patients, who demonstrated no clinical signs of ventral compression by the odontoid such as caudal cranial nerve deficits but either radiological evidence of craniocervical instability or assimilation of the atlas to the occiput, foramen magnum decompression was combined with craniocervical stabilisation. In the remaining 9 patients, ventral compression of the medulla oblongata had caused caudal cranial nerve dysfunctions. These patients underwent transoral resection of the odontoid, followed by posterior decompression and craniocervical fusion (Fig. 10.2) (Klekamp and Samii 2001).

Whenever the position of the odontoid leads to brainstem compression, the key elements of surgical treatment are distraction of the C1/2 intervertebral joints and C1/2 fusion. This distraction may reverse the ventral compression to a degree that no additional transoral resection of the odontoid is required. In the author's series, the decision for a transoral decompression was based on clinical signs of caudal cranial nerve deficits in the presence of compression of the medulla by the odontoid. Whether a transoral resection of the odontoid is obsolete (Goel and Shah 2009) or still required for patients with substantial and irreducible ventral compression (Smith et al. 2010) remains a controversial issue.

For craniocervical stabilisations, the implants need to be adjusted carefully to the abnormal anatomy. Precise planning is required to allow their safe fixation at the occipital bone as well as the allocation of the bone graft despite of the craniectomy required for foramen magnum decompression. All implants need to be covered completely by the muscular layer during closure, without too much strain being placed on soft tissues; otherwise, local discomfort or even CSF fistulas may result.

10.4.3 Chiari II Malformation

If clinical signs of hydrocephalus are present in Chiari II patients, treatment of the increased intracranial pressure has always the first priority. Endoscopic third ventriculostomy is again an optional alternative to ventriculoperitoneal shunts (Elgamal et al. 2011). In the literature, it has been stated in two large series that if the hydrocephalus has been managed successfully, then only a small minority of Chiari II patients require a decompression (Talamonti and Zella 2011; Rauzzino and Oakes 1995).

If one considers the pathophysiological considerations put forward by McLone (McLone and Knepper 1989) and the positive results obtained following intrauterine operations on foetuses (Danzer et al. 2011; Tulipan et al. 1999), Chiari II malformations are potentially preventable or reversible, if the decompression is carried out early enough in symptomatic patients with a sufficiently treated hydrocephalus. A number of studies have reported a benefit if decompression for Chiari II is performed as soon as neurological symptoms begin (Rauzzino and Oakes 1995; Teo et al. 1997; Pollack et al. 1992, 1996). Yet, there appears to be very little scientific evidence to support routine use of this approach for symptomatic infants (Tubbs and Oakes 2004). This may reflect the fact that, once severe brainstem dysfunctions are present, a decompression may not reverse established neurological deficits (Kirsch et al. 1968).

In the author's series, 42 patients presented with a Chiari II malformation. Surgical management was only recommended in 13 of these patients, when there was no evidence for ventricular shunt malfunction but a clear history of progressive brainstem or cervical cord dysfunction (Charney et al. 1987; Kirsch et al. 1968). Six were under 2 years of age, presenting with signs of central dysregulation. Three patients presented between 5 and 14 years of age (Fig. 10.3) and the remainder in adulthood, with progressive upper extremity dysfunction.

When operating for Chiari II malformation, the decompression must be undertaken at the spinal levels corresponding to the tonsillar descent, rather than at the foramen magnum, which is enlarged in this entity. Patients are operated in prone position. The exposure extends from the foramen magnum to the lowest lamina covering the herniated tonsils. After laminectomy of these segments, the atlanto-occipital membrane should be coagulated and dissected off the dura. The dural incision then starts at the level of the foramen magnum and extends over all levels involved. Then the arachnoid is opened. Dissection should be limited strictly to the midline in order to avoid injury to perforating vessels or caudal cranial nerves. It should be borne in mind that the brainstem is displaced caudally in these patients, taking with it important structures such as the PICA or caudal cranial nerves. As outlined for Chiari I operations, it is also desirable to open the foramen of Magendie in Chiari II patients, although this should be undertaken only if it can be performed safely. Coagulation of tonsils, which is a safe technique in Chiari I to gain access to Magendie, is not recommended in Chiari II as these are very often tightly adhered to the underlying brainstem and any such manoeuvre carries considerable risks. Arachnoid dissection should concentrate on creating a passage between the intracranial and spinal subarachnoid channels. Once that is achieved, a duraplasty should be inserted. As described for Chiari I surgery (above), the duraplasty is lifted off the cord by tenting sutures.

Laminectomies in this patient group carry a significant risk of producing postoperative kyphosis or swan-neck deformities (Lam et al. 2009) (Fig. 10.3). They should therefore be combined with posterior fusion, which can be achieved elegantly with lateral mass screws (Fig. 10.7).

10.4.4 Foramen Magnum Arachnoiditis

In the author's series, ventriculomegaly was more common in foramen magnum arachnoiditis compared to Chiari I malformations (52 % vs. 8.6 %). This implies that if a borderline tonsillar herniation is associated with ventricular dilatation, then arachnoiditis at the foramen magnum may well

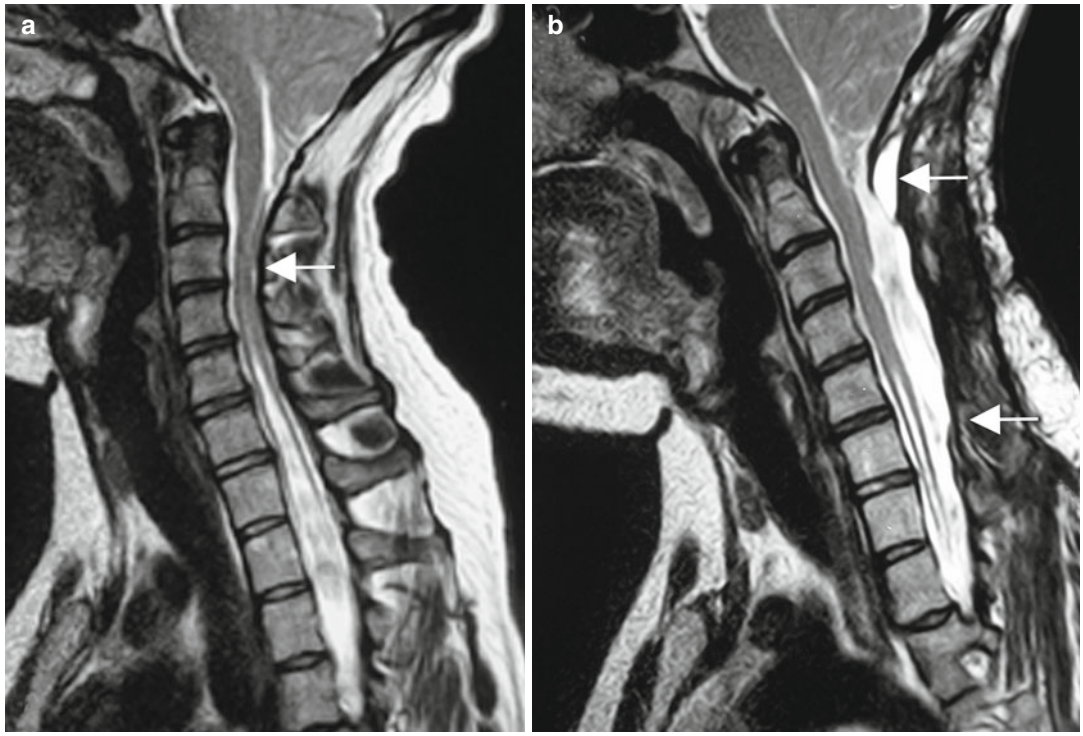

Fig. 10.7 (**a**) Preoperative T2-weighted MRI of an adult patient with Chiari II malformation presenting with progressive weakness of both hands. The image shows the tonsillar descent to C3 (*arrow*) with osteochondrosis and stenosis of the cervical spine at that level. (**b**) After bony decompression and duraplasty from C1 down to C5 (*arrows*), the enlarged subarachnoid space and decrease of the syrinx are apparent. To prevent a kyphotic deformity, a stabilisation was added with lateral mass screws. Postoperatively, she regained function in her hands

be present. Depending on the extent and severity of the arachnoiditis, surgical management may require CSF diversion, in addition to foramen magnum decompression.

When foramen magnum arachnoiditis is not diagnosed preoperatively, its presence, extent and severity must be determined after dural opening. Dural opening has to respect the arachnoid layer in such cases in order to avoid surgical morbidity related to vascular injuries in particular. Thereafter, the aim of surgery is to create a free CSF passage between the intracranial and spinal subarachnoid spaces. It is not advisable to dissect all arachnoid scarring off the spinal cord, medulla oblongata and cerebellar tonsils. On the contrary, such attempts are risky and simply lead to new adhesions and scar tissue formation. In foramen magnum arachnoiditis, the foramen of Magendie is always obstructed. A decision must therefore be made, intraoperatively, as to whether or not it can be opened safely. If important structures such as the PICAs are embedded in arachnoid scar tissue, then the risk may be too high. Some authors recommend placement of small catheters to provide an outflow for the 4th ventricle (Abe et al. 1995). However, even with ultrasound guidance, this remains a very risky manoeuvre, and the author has encountered patients with severe neurological deficits as a consequence of malpositioned catheters in this region. A safer strategy is to leave the foramen Magendie closed and place a supraventricular shunt. Indeed, because of such concerns, some surgeons have previously recommended limiting the management of foramen magnum arachnoiditis to ventricular shunting (Appleby et al. 1969).

10.5 Surgical Results and Complications

10.5.1 Chiari I Malformation

Two published analyses of foramen magnum decompressions gave extensive overviews on complications encountered during and immediately after surgery as well as delayed postoperative problems but did not provide any data (Menezes 1991; Mazzola and Fried 2003). When it comes to quantifying such complications, analysis of the literature shows enormous variations in reported figures. Not all studies seem to use the same standards when complications are analysed; how else can one explain figures of 2.4 % (Tubbs et al. 2011) and 37 % (Zerah 1999), both from series of more than 100 children, undergoing the same decompression procedure, carried out in respected institutions?

Tables 10.2 and 10.3 provide a literature overview, comparing complication rates, syrinx reduction rates and the frequency of surgical revisions, for different surgical decompression techniques for Chiari I. These include decompressions involving only bone removal, those with incision of the outer dural layer, those opening the dura completely and those where additional arachnoid dissection was performed.

The most common complication in the author's series was a CSF fistula, occurring in 6 % of cases overall but more often after revision surgeries than first operations (9 and 5.5 %, respectively). Use of autologous material for duraplasty was not associated with a lower rate for CSF fistulas compared to artificial materials although this is in contrast with the findings of other studies (Vanaclocha and Saiz-Sapena 1997). In order to limit the risk of a fistula, it is important to close the duraplasty with a tight running suture as well as ensure a good closure of the muscular layer, which appears to be the most effective barrier to CSF leakage. For that reason, the author does not use monopolar electrocautery for soft tissue dissection as this may cause significant damage to the muscular layers in particular. Leaving the arachnoid intact does not exclude fistulas because small lacerations and tears in this thin membrane are very common after dural opening. Nor does the additional use of tissue sealants appear to lower rates for fistulas (Parker et al. 2011).

Aseptic meningitis does seem to be related to the type of material used for duraplasty and was seen exclusively when lyophilised dura, fascia lata or galea had been used. To avoid any postoperative problems related to duraplasties, Bernard

Table 10.2 Literature review of the results of decompressions for Chiari I malformations, without dural opening

Authors	Group	*N*	Follow-up period	Peri- and postoperative complications	Syrinx size reduced	Recurrences/ deaths
Bony decompression only						
James and Brant (2002)	C	4	Not reported	None	Not reported	None
Hayhurst et al. (2008)	A, C	16	43 months[a]	27 %[a]	87 %[a]	25 %/none
McGirt et al. (2008b)	C	116	25 months	1 %	Not reported	7.8 %/none
Mutchnik et al. (2010)	A, C	56	Not reported	None	Not reported	12.5 %/none
Yilmaz et al. (2011)	A	24	Not reported	8.3 %	91.1 %	9.5 %/not reported
Bony decompression with outer dural decompression						
Gambardella et al. (1998)	A	8	Not reported	Not reported	88 %	12.5 %/none
Munshi et al. (2000)	A	11	Not reported	10 %	50 %	18.2 %/none
Navarro et al. (2004)	C	71	28 months[a]	5.6 %	65.7 %[a]	10.8 %/none
Limonadi and Selden (2004)	C	12	15.7 months	Not reported	No syrinx cases	Not reported
Caldarelli et al. (2007)	C	30	55 months	Not reported	50 %	6.7 %/none
Chauvet et al. (2009)	A	11	18 months	9.1 %	80 %	None

Abbreviations: *N* number of patients, *A* adults, *C* children
[a]Pooled data for different subgroups

Table 10.3 Literature review of the results of decompressions for Chiari I malformations, with dural opening

Authors	Group	*N*	Follow-up period	Peri- and postoperative complications	Syrinx size reduced	Recurrences/deaths
Bony decompression with arachnoid left intact and dura left open						
Di Lorenzo et al. (1995)	A	20	29 months	Not reported	100 %	15 %/none
Zerah (1999)	C	79	Not reported	37 %[a]	69 %[a]	1.6 %[a]/none
Bony decompression with arachnoid left intact and duraplasty						
Guyotat et al. (1998)	A, C	42	39 months	Not reported	58 %	50 %/4.7 %
Zerah (1999)	C	79	Not reported	37 %[a]	69 %[a]	1.6 %[a]/none
Munshi et al. (2000)	A, C	34	Not reported	42 %	100 %	None/none
Limonadi and Selden (2004)	C	12	14.8 months	8.3 %	100 %	Not reported
Navarro et al. (2004)	C	24	28 months[a]	42.1 %	65.7 %[a]	4.2 %/none
Galarza et al. (2007)	C	20	21 months[a]	8.3 %[a]	64[a]	Not reported
McGirt et al. (2008b)	C	140	29 months	3 %	Not reported	7.1 %/None
Wetjen et al. (2008)	A	29	36 months	Not reported	100 %	Not reported
Hoffman and Souweidane (2008)	A, C	40	11.4 months	CSF related 2.5 %	Not reported	5 %/None
Attenello et al. (2008)	C	49	41 months	10 %[a]	55 %[a]	10.2 %[a]/none
Sindou et al. (2002); Sindou and Gimbert (2009)	A	44	48 months	20.5 %	60 %	Not reported/none
Attenello et al. (2009)	C	27	Not reported	4 %	80 %	None Gore-Tex
Attenello et al. (2009)	C	40	Not reported	10 %	52 %	None Galea
Mutchnik et al. (2010)	A, C	64	Not reported	4.6 %	Not reported	3.1 %/none
Spena et al. (2010)	A	36	40 months	8.1 %	80.5 %	5.5 %/none
Yilmaz et al. (2011)	A	58	Not reported	12.1 %	84.2 %	Not reported
Valentini et al. (2011)	C	80	Not reported	6.3 %	91.5 %	6.3 %/none
Mottolese et al. (2011)	C	82	Not reported	18 %	Not reported	Not reported
Bony decompression with arachnoid opened and duraplasty						
Fischer (1995)	C	19	Not reported	26.3 %	93 %	Not reported
Vanaclocha and Saiz-Sapena (1997)	A	26	27 months	42.3 %	No syrinx cases	Not reported
Guyotat et al. (1998)	A, C	8	28 months	None	100 %	None
Aghakhani et al. (1999)	A	214	79 months	24 %	95 %	12.4 %/0.7 %
Zerah (1999)	C	105	Not reported	37 %[a]	69 %[a]	1.6 %[a]/none
Tubbs et al. (2003)	C	130	50 months	2.3 %	Not reported	6.9 %/none
Navarro et al. (2004)	C	14	28 months[a]	50 %	65.7 %[a]	28.6 %/none
Guo et al. (2007)	A, C	115	36 months	CSF 9.82–18.75 %	82–88 %	Not reported
Galarza et al. (2007)	C	40	21 months[a]	8.3 %[a]	64.3[a]	Not reported
Zhang et al. (2008)	A, C	234	Not reported	15.8 %[a]	66.5 %	Not reported/1.3 %[a]
Kumar et al. (2008)	A, C	87	34 months	17.2 %	Not reported	10.3 %/none
Aghakhani et al. (2009)	A	157	88 months	9.5 %	75.64 %	3.8 %/0.63 %
Zhang et al. (2011)	A, C	132	27 months	28 %	81.8 %	Not reported
Author's series[b]	A, C	203	52 months	19.2 %	87.9 %	5.9 %/1 %

Abbreviations: *N* number of patients, *A* adults, *C* children

[a]Pooled data for different subgroups

[b]Data for first decompressions with arachnoid opening and alloplastic material for duraplasty

Williams recommended against duraplasty. He favoured to suture the dura into the muscle and to close all soft tissues with tight sutures (Williams 1994). However, it has been claimed that omission of a duraplasty after arachnoid opening and dissection may predispose to severe arachnoiditis and recurrent CSF flow obstruction (Munshi et al. 2000).

Hydrocephalus is an important postoperative complication (Tubbs et al. 2003). In the author's series of 371 decompressions, it occurred after 3 % of decompressions within 30 days of surgery. An additional 2 of the 371 patients developed hydrocephalus months after the operation. An analysis of complications after posterior fossa surgery in general determined a rate of 4.6 % for postoperative hydrocephalus after 500 operations (Dubey et al. 2009). Most authors relate this problem to formation of subdural hygromas in the posterior fossa (Filis et al. 2009; Suzuki et al. 2011; Bahl et al. 2011; Marshman et al. 2005; Elton et al. 2002), but not all patients with postoperative hydrocephalus in the author's series demonstrated such collections. Whether or not postoperative ventriculomegaly needs surgical treatment should be based on the clinical course of the patient. The problem may resolve spontaneously (Marshman et al. 2005) or may require an intervention. Treatment options are either a ventriculoperitoneal shunt or a third ventriculostomy (Kandasamy et al. 2008).

Swallowing dysfunctions after craniovertebral decompression are related to surgical manipulations at or close to the obex and were observed early in 2.5 % of cases in the author's series, but this complication was not observed following application of the microsurgical dissection technique outlined above, which leaves the area of the obex and the fourth ventricular floor untouched.

The author's overall complication rate of 22 % is in line with those of several other studies (Zhang et al. 2011; Aghakhani et al. 1999; Guo et al. 2007; Fischer 1995; Kumar et al. 2008; Navarro et al. 2004; Zerah 1999; Aghakhani et al. 2009; Parker et al. 2011; Mottolese et al. 2011). This is a considerably higher complication rate than after bony decompression alone or following a decompression with incision of the outer dura alone (McGirt et al. 2008b; Mutchnick et al. 2010; Navarro et al. 2004; Yilmaz et al. 2011; Chauvet et al. 2009; Limonadi and Selden 2004) (Table 10.2). As to whether leaving the arachnoid intact lowers the complication rate, the author's experience suggests that this is not the case. Furthermore, the same range of complication rates are reported in studies involving duraplasties, with or without arachnoid dissection (Guo et al. 2007; Tubbs et al. 2003, 2011; Zerah 1999; Zhang et al. 2008, 2011; Aghakhani et al. 1999; Sindou et al. 2002; McGirt et al. 2008b; Munshi et al. 2000; Sindou and Gimbert 2009). Yet other publications report considerably lower complication rates than the author's experience but, once again, independent of whether or not the arachnoid was opened (Mutchnick et al. 2010; McGirt et al. 2008b; Attenello et al. 2009; Spena et al. 2010; Yilmaz et al. 2011; Valentini et al. 2011; Tubbs et al. 2003; Aghakhani et al. 2009) (Table 10.3).

Decrease in syrinx size was observed in 81 % of 281 craniovertebral decompressions in the author's series. No change occurred in 15 and 3.7 % demonstrated a further expansion. The number of postoperative syrinx reductions was significantly greater after primary decompressions, as compared to secondary operations (85 and 72 %, respectively).

In the first postoperative year, improvements can be anticipated for all symptoms. The most profound effect will be seen with occipital pain, which almost always improves after surgery. In the author's series, there were no differences in short-term results between adults and children. Patients undergoing a revision usually had more severe preoperative motor deficits and gait disturbances, and these deficits improved only marginally with a secondary decompression. For all foramen magnum decompressions combined, 3 out of 4 of patients in the author's series considered their condition to have improved 3 months after surgery, while 1 in 5 reported no change, and 1 in 20 experienced a worsening of symptoms (Table 10.4).

Long-term results in the author's series were analysed with Kaplan-Meier statistics, to determine the rates at which patients experienced

Table 10.4 Postoperative results after 3 months for Chiari malformations and foramen magnum arachnoiditis (author's series)

Type	Chiari I first surgery (%)	Chiari I revision surgery (%)	Basilar invagination (%)	Chiari I all surgery (%)	Chiari II (%)	Foramen magnum arachnoiditis (%)
Improved	76.3	65.1	85	73.6	56	45
Unchanged	19.7	23.3	15	21.0	38	36
Worse	4.0	11.6	–	5.5	6	18

Table 10.5 Neurological recurrence rates for Chiari I malformations: Kaplan-Meier statistics

Group	5 years (%)	10 years (%)	*P*-value
First surgery	13.1	19.1	0.007
Revision surgery	34	34	
Adults	14.4	15.6	0.66
Children	11.8	–	
Arachnoid opened	13.5	14.7	0.023
Arachnoid not opened	34.1	–	
Artificial graft	10	11.5	0.022
Autologous graft	31.8	31.8	
Syringomyelia	13.8	15.2	0.61
No syringomyelia	15.5	15.5	
All	14.3	15.4	
Optimal first surgery	7	8.7	

neurological deterioration, compared with those maintaining a stable neurological status (Table 10.5). Overall, recurrences amounted to 14 % within 5 years and 15 % within 10 years. No significant differences were seen between adults and children or between patients with and without syringomyelia. Recurrence rates were, however, significantly lower for primary compared to revision operations. It is also interesting to note that recurrence rates were significantly higher after decompressions with duraplasties using autologous material, as compared with those using alloplastic materials.

When comparing results in the literature for different types of foramen magnum decompression, there is a lower frequency of complications in all types of procedures that leave the inner layer of the dura intact. This, however, is counterbalanced by a lower frequency of syrinx collapses and a higher frequency of patients experiencing a recurrence of symptoms and/or requiring revision surgery – if these data are reported (Tables 10.2 and 10.3). The two largest series where decompression was restricted to a bony removal did not provide such numbers (McGirt et al. 2008b; Mutchnick et al. 2010). The largest series reporting on decompression with incisions of the outer dural layer reported rates for syrinx decrease of between 50 and 66 % (Caldarelli et al. 2007; Munshi et al. 2000; Navarro et al. 2004).

When it comes to opening the dura fully, series recommending a duraplasty but leaving the arachnoid intact once again often do not provide data for syrinx reductions (Mottolese et al. 2011; McGirt et al. 2008b; Mutchnick et al. 2010; Hoffman and Souweidane 2008). Studies do report syrinx reduction rates in the range of 50–60 % (Guyotat et al. 1997; Sindou et al. 2002; Attenello et al. 2009), although some smaller series observed a syrinx reduction in all their patients (Limonadi and Selden 2004; Wetjen et al. 2008; Munshi et al. 2000). Higher rates for postoperative improvements and syrinx reductions were found in 34 patients receiving duraplasties, as compared to 11 who underwent incisions of the outer dura (Munshi et al. 2000). A study comparing decompressions involving tonsillar coagulation and duraplasties, with decompressions confined to incision of the outer dura alone, found better results in the former group after a mean follow-up of 21 months, both in terms of clinical improvements and syrinx reductions (Galarza et al. 2007). This overview suggests but does not prove that postoperative syrinx reduction rates are lower after decompressions that do not open the arachnoid (Tables 10.2 and 10.3).

Looking on rates for postoperative revisions and recurrences, a study comparing results for 56 patients undergoing a pure bony decompression with 64 patients receiving duraplasty but without arachnoid opening concluded that the revision rate was higher in the former group (12.5 compared to 3.1 %), although the paper did not

state the follow-up time (Mutchnick et al. 2010). A similar study on 82 patients reported a higher rate of revision after decompressions without duraplasty, compared to those with duraplasty – 9.5 and 3.6 %, respectively (Yilmaz et al. 2011). A meta-analysis from the literature of 582 patients came to the same conclusion (Durham and Fjeld-Olenec 2008). A study of results in children, however, concluded that decompression with incision of the outer dura gave comparable results to more invasive forms of decompression and avoided higher complication rates (Navarro et al. 2004). A report comparing results for 16 bony decompressions with 80 decompressions involving arachnoid dissection noted a lower recurrence rate after dura and arachnoid opening – 7.5 compared to 25 % after bony decompressions only (Hayhurst et al. 2008).

Some investigators have tried to tailor surgical steps for individual patients according to intraoperative ultrasound findings. A study to evaluate the effect of bony decompressions in children noted tonsillar pulsations and a sufficient subarachnoid space in 116 operations, so the dura was not opened. A duraplasty was performed without opening the arachnoid in a further 140 instances. Following this policy and at an average follow-up of 27 months, 15 % of children had a mild or moderate recurrence of symptoms, and another 7 % had severe symptom recurrence, requiring a revision surgery. Kaplan-Meier analyses showed a recurrence rate for headache of more than 40 % and a recurrence rate for brainstem and cranial nerve symptoms of more than 20 % within 80 months after surgery. A syrinx decrease was detected in 62 % of patients (McGirt et al. 2008b). Another group also modified their operative steps according to intraoperative ultrasonic CSF flow measurements and found arachnoid dissection necessary in the overwhelming majority of patients (Milhorat and Bolognese 2003). Others have also used ultrasound to establish an adequate decompression after bony removal but then restricted the operation to splitting the outer dural layer in only a minority of patients, who did not have a syrinx and where the tonsillar herniation was mild, not reaching C1. The majority of the 363 paediatric patients underwent dural opening, arachnoid dissection, opening of the foramen of Magendie and duraplasty (Menezes et al. 2005). Neither set of authors gave any information on postoperative outcomes for their patients.

In summary, the outcome data as published in the literature for syrinx reduction and revision rates are considerably worse for limited decompressions when compared to those with arachnoid opening and duraplasty, in the author's and other similar series (Tables 10.2 and 10.3). Given the low morbidity rates of the technique described in this chapter and the much better long-term results for decompressions that involve at least a duraplasty, the author does not support methods of decompression that do not involve duraplasties.

This leaves the question of whether the arachnoid should be opened or not. In the author's series, the arachnoid was not opened in a subgroup of 24 Chiari I patients without syringomyelia because it appeared entirely normal after dural opening. In the long term, however, a significantly higher neurological recurrence rate[1] was observed for these patients compared to the other decompressions with arachnoid dissection (Table 10.5). So far, a few studies have analysed the influence of arachnoid changes on clinical symptoms and outcomes in Chiari I malformation. Some authors comment that severe arachnoid scarring is a sign of unfavourable long-term prognosis (Sakamoto et al. 1999; Aghakhani et al. 2009). However, there are little data on the frequency of arachnoid changes in Chiari I and even less information on their significance. One study found arachnoid scarring in 6 of 14 patients without and 19 of 51 patients with syringomyelia but made no statements as to whether this influenced the clinical outcome (Ellenbogen et al. 2000). Another found that patients with arachnoiditis showed more severe neurological signs before surgery and had a worse postoperative outcome than patients without arachnoid pathology (Aghakhani et al. 2009). In the author's series, severe arachnoid scarring was detected in 48 of

[1] The reappearance or progression of preoperative symptoms, or development of new neurological symptoms, related to the Chiari or the syrinx

371 decompressions (13 %). Severe scarring was significantly more common in secondary operations as compared to primary procedures (63 and 6 %, respectively). More data are available concerning patency of the foramen of Magendie. The foramen was obstructed in 12 % of 500 paediatric patients that were surgically managed for Chiari I (Tubbs et al. 2011). This group also emphasised in an earlier paper that such obstructions cannot be detected without arachnoid dissection (Tubbs et al. 2003). The foramen was found to be partially or completely obstructed in another study in 14 out of 105 children (Zerah 1999). In a third study, the foramen was obstructed in 7.4 %, with some arachnoid scarring at this level in 17 % of patients (Menezes et al. 2005). In the author's series, Magendie was obstructed in 33 % of patients. Again, this rate was significantly higher in revision as against primary surgeries (65 and 28 %, respectively). Again in the author's series, severe arachnoid scarring was associated with more severe preoperative neurological deficits, a lower frequency for syrinx reduction postoperatively and a higher frequency of long-term neurological deterioration.

These observations suggest that the arachnoid does play a role in the pathophysiology of Chiari I malformations at least for the formation and postoperative resolution of syringomyelia but also for the function of brainstem and spinal cord irrespective of a syrinx. Decompression techniques that do not address the arachnoid neglect a component that is relevant for a significant number of patients. Techniques that treat the arachnoid pathology can be expected to be associated with improved long-term results (Table 10.3). To settle this issue, however, a prospective study comparing postoperative results for decompressions with and without arachnoid dissection will be required.

10.5.2 Basilar Invagination

In the author's series of 35 operated patients, 30 patients with basilar invagination considered their condition improved following surgery, and 5 were unchanged 3 months postoperatively (Table 10.4). Four required further procedures subsequently. Patients with basilar invagination treated by posterior decompression only require postoperative monitoring for evidence of instability. In one of the author's patients without preoperative evidence of craniocervical instability, postoperative MR scans demonstrated pannus formation around the odontoid peg and instability on functional X-rays about 6 months after decompression. A posterior fusion was necessary, which then led to neurological improvement. Among 9 patients treated by ventral and dorsal decompression and fusion, two revisions for late implant failures were required. Another patient in this group with a stable postoperative neurological status underwent a revision at another institution resulting in neurological worsening. Apart from these 4 patients, no subsequent operations were required or clinical recurrences detected in the group of patients with basilar invagination.

10.5.3 Chiari II Malformation

In the literature, good results have been reported for infants undergoing decompression before severe brainstem dysfunctions, such as bilateral vocal cord palsies, had developed, with success rates in the order of 80 % (Zerah 1999; Vandertop et al. 1992; Ishak et al. 1980; Teo et al. 1997; Pollack et al. 1996; Talamonti and Zella 2011). The author's series of six children under 2 years of age included four who presented as neonates with severe respiratory problems and two 1-year-old children who had cyanotic attacks when stressed or upon coughing. Following decompression, all infants showed improvement of respiratory functions, although one child still died, 17 months after surgery, due to recurrent respiratory problems. In children, radiological evidence of instability is common after decompressions and was found in 5 of 9 patients in one study addressing this problem, specifically. However, this appeared not to be clinically relevant as no clinical symptoms were related to it (Lam et al. 2009). Whether this conclusion still holds once these children have reached adulthood remains to be seen.

For adult patients with Chiari II malformation, few data exist in the literature. One study recommends a combination of decompression

and stabilisation if surgery is required in this age group and reported good outcomes in 4 patients so treated (Rahman et al. 2009). Looking at short-term results for 7 older patients in the author's series undergoing 10 decompressions in total, 3 operations were followed by improvements, and 6 left the patients unchanged, while 1 patient considered his condition worsened within 3 months after decompression. In the longer term, the neurological status was stabilised in 4 of these 7 patients, requiring a total of 6 decompressions. The other 3 progressed despite 4 decompressions presumably related to compromised cervical stability. Therefore, a decompression for Chiari II malformation should be combined with a posterior fusion to prevent neurological deteriorations related to post-laminectomy deformities.

All patients with Chiari II malformations require a lifelong medical surveillance by neurologists, orthopaedic surgeons and neurosurgeons, to maintain as much quality of life and autonomy as possible.

10.5.4 Foramen Magnum Arachnoiditis

In the author's series, 10 patients with foramen magnum arachnoiditis underwent decompression at the foramen magnum. Five considered their condition improved 3 months after decompression, 3 were unchanged and 2 worsened (Table 10.4). In the long term, 6 clinical recurrences were detected. In this series, arachnoiditis had the worst prognosis among all pathologies at the foramen magnum, with only 33 % achieving a stable neurological status according to a Kaplan-Meier analysis. In the literature, no outcome data for a patient series treated by decompression exists.

10.6 Recurrences

This section will deal with surgical revisions for Chiari I malformations only. For Chiari II or foramen magnum arachnoiditis, the number requiring revision surgery was low in the author's series, and cases in the literature were discussed above.

10.6.1 Assessment

Once hydrocephalus is ruled out, the further evaluation of patients representing after a foramen magnum decompression must start with the clinical history. What symptoms were apparent before the previous decompression, and how did these symptoms respond to surgery? Was the neurology unchanged or improved, or did symptoms progress further without an interval of stable neurology? In the author's series of 107 patients presenting after a Chiari I foramen magnum decompression, 56 did not go on to a further surgical procedure, mainly because their neurological status either was stable or was considered unlikely to be stabilised by another intervention. Most of these patients not undergoing a revision presented because they were disappointed by the result of their initial decompression. Although many symptoms had improved and the syrinx had decreased in size, burning dysaesthesias persisted and proved extremely difficult to treat with analgesics in the great majority. It is certainly important to inform any patient, before any surgery, that dysaesthesias may not respond to an otherwise successful decompression. Indeed, such pains may even worsen following collapse of a syrinx (Milhorat et al. 1996). This phenomenon, however, was not observed in the author's series.

Progression of symptoms without an interval of clinical stability suggests an insufficient operation. In most instances, this will be related to an insufficient decompression; untreated features of an associated basilar invagination, such as anterior compression by the odontoid peg; or the result of craniocervical instability. In the majority of patients, however, the clinical history reveals a stable interval after foramen magnum decompression with or without improvement of preoperative symptoms. It should then be noted how and when the deterioration started. In the author's experience, the longer the interval of clinical stability before the deterioration began, the less likely the cause was related to the foramen magnum. The only clinical symptom, which always suggested a foramen magnum problem in these patients, was a recurrence of occipital headaches or swallowing dysfunctions.

As the next step, careful neuroradiological assessment is essential for these patients. First of all, the area of the previous operation needs to be evaluated, comparing pre- and postoperative MRI scans. There might be evidence of insufficient decompression or recurrent compression. It has been reported that new bone formation may cause recurrent compression (Zerah 1999; Aoki et al. 1995; Hudgins and Boydston 1995). An oversized craniectomy may result in cerebellar ptosis and medullary compression (Holly and Batzdorf 2001). The newly created artificial cisterna magna may not be of sufficient size. There may be a pseudomeningocoele pushing the dura anteriorly onto the cervicomedullary junction. MR imaging will also reveal whether there is basilar invagination with persistent anterior compression of the odontoid or if there is a precursor of craniocervical instability, such as an assimilated atlas, a Klippel-Feil syndrome of the upper cervical spine or pannus formation around the odontoid (Smith et al. 2010). Functional X-rays or CT scans should be part of the diagnostic workup in such patients to evaluate craniocervical stability.

Another important consideration is the postoperative course of a syrinx. If the syrinx is seen, radiologically, to have decreased after surgery and to have remained so, it was unlikely that any new symptoms were related to the foramen magnum, with the one exception that craniocervical instability still had to be ruled out. Next, a cardiac-gated cine MRI should be performed, to evaluate the CSF passage at the foramen magnum. This modality is the most sensitive method to detect or exclude arachnoid scarring and postoperative adhesions (Armonda et al. 1994; Bhadelia et al. 1995; Hofkes et al. 2007; McGirt et al. 2008a). If such a study demonstrates adequate CSF flow at the foramen magnum and the neuroradiological evaluation had excluded all the other above-mentioned possibilities, then the clinical deterioration was due to a mechanism or cause unrelated to the previous decompression.

In patients with syrinx shunts, the shunt catheter may have caused tethering of nerve roots or spinal cord leading to radicular or myelopathic symptoms, which are often provoked by neck or arm movements (Batzdorf et al. 1998). The MRI in these patients will show adherence of the cord to the dura at the level of the shunt.

If this, in turn, has been excluded, degenerative changes of the cervical spine should be evaluated. Many patients with a well-treated Chiari malformation and a collapsed syrinx demonstrate considerable spinal cord atrophy. MRI scans may then give the impression that a slight or moderate degree of cervical stenosis is not clinically relevant, but this is a dangerous assumption. Such patients have little functional reserve in their spinal cord, and any additional physical insult, even a minor one, may be enough to cause significant new deficits. It has even been suggested that Chiari patients may be particularly prone to the effects of degenerative problems of the cervical spine (Takeuchi et al. 2007). In particular, signs of hypermobility of cervical segments should be looked for by radiographs in ante- and retroflexion. Cervical fusion should be offered to patients with neurological deterioration despite a sufficiently treated Chiari I malformation but with signs of hypermobility in the cervical spine in functional studies. In the author's experience, such fusions may stabilise the neurological course of these patients.

10.6.2 Revision Surgery

Indications for revision surgery at the craniovertebral junction include untreated or new instability of the craniocervical junction, insufficient primary decompression or an obstruction of CSF flow, the latter related to arachnoid scarring or compression of the cisterna magna by a pseudomeningocoele (Fig. 10.8). In view of the poor response of Chiari-related syringomyelia to direct shunting, the author would not choose this treatment for patients after a failed primary decompression (Klekamp et al. 1996). Instead, if a syrinx reappears or does not regress in the first place, the reason for this must be established and treated at the level of the foramen magnum. This will require a revision with opening of the dura or duraplasty and then careful arachnoid dissection, to establish free outflow of CSF from the foramen

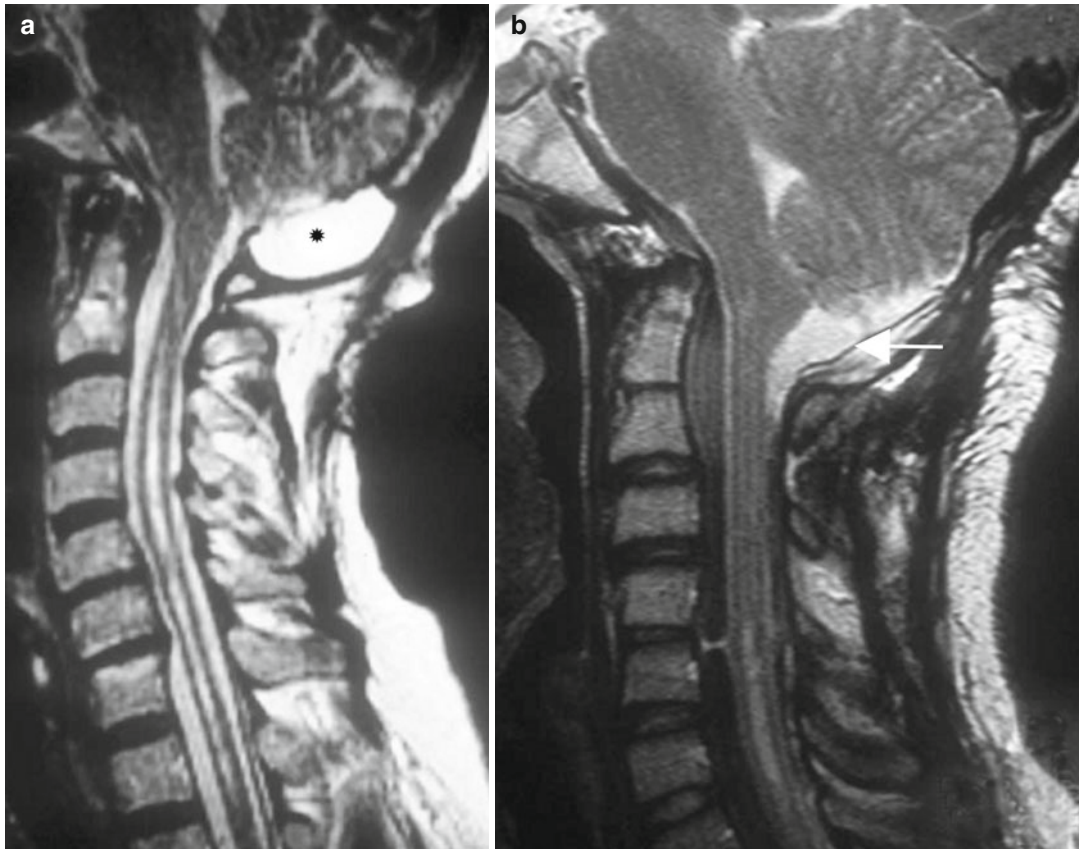

Fig. 10.8 (**a**) This preoperative T2-weighted MRI shows a situation after foramen magnum decompression for Chiari I malformation. C1 was not removed. A pseudomeningocoele developed (*), pushing the duraplasty anteriorly and resulting in CSF flow obstruction. Even though the syrinx had not enlarged, the patient complained of local discomfort and deteriorating gait. (**b**) After foramen magnum revision, with removal of C1 and exchange of the duraplasty, CSF flow is restored (*arrow*) and the patient improved

of Magendie. Several authors have mentioned the importance of opening Magendie during foramen magnum decompressions (Zerah 1999; Menezes et al. 2005; Tubbs et al. 2011) and particularly so during revisions (Sacco and Scott 2003; Tubbs et al. 2011). In some cases, such revisions may need to be combined with craniocervical fusion and even transoral resection of the odontoid. Severe arachnoid scarring related to surgical manoeuvres or postoperative meningitis is the most common feature in patients demonstrating a CSF flow obstruction, usually in the form of adhesions between dural graft and cerebellum or the spinal cord, although other factors may sometimes operate (Table 10.6) (Fig. 10.8) (Klekamp et al. 1996; Sakamoto et al. 1999; Ellenbogen et al. 2000; Mazzola and Fried 2003; Rosen et al. 2003; Yanni et al. 2010; Pare and Batzdorf 1998). Leaving the arachnoid intact during the first decompression may also result in insufficient CSF flow postoperatively leading to a subsequent revision.

Table 10.6 Causes of postoperative cicatrix

Pseudomeningocoele pushing the graft anteriorly
Use of autologous graft material
Insufficient arachnoid dissection at primary surgery
Plugging the obex
History of meningitis

The more extensive and dense the arachnoid pathology, the less likely it is that a revision will

produce a lasting benefit. Furthermore, the risk of secondary surgery is certainly higher than with primary decompressions. It is, therefore, very difficult to predict the outcome for a patient before revision surgery. Unless there is a history of meningitis or a clear description of severe arachnoid changes in the original operation notes, it is impossible to foresee exactly what will be discovered after reopening of the dura. A surgical strategy has to be adopted intraoperatively which improves CSF flow but minimises the risk of recurrent postoperative arachnoid scarring. Limiting the arachnoid dissection to the midline, with sharp transection of arachnoid adhesions obstructing the foramen of Magendie and the posterior spinal subarachnoid space, is all that is required. Blunt dissection or tackling arachnoid adhesions laterally carries the risk of damage to small perforating arteries and caudal cranial nerves and should be avoided. Finally, a spacious dura graft, using artificial material, provides reasonable protection against postoperative arachnoid scarring, which may otherwise cause another clinical recurrence. The realistic outlook for patients undergoing a foramen magnum revision is clinical stabilisation of the previously progressive course. Long-term results in the author's series, determined by Kaplan-Meier statistics, revealed a further recurrence rate of 1 in 3 within 5 years after foramen magnum revision (Table 10.5) (Klekamp et al. 2002).

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