190 Chiari Malformations

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The Chiari malformations are a collection of hindbrain abnormalities ranging from simple herniation of the cerebellar tonsils through the foramen magnum to complete agenesis of the cerebellum. It is unlikely that a unifying pathophysiology underlies all of these findings, which, compared with the past, are frequently recognized on magnetic resonance imaging (MRI). The focus of treatment for symptomatic patients with Chiari malformations mainly consists of restoring normal cerebrospinal fluid (CSF) dynamics across the craniocervical junction. There is great variability in the clinical presentation,[1,2] imaging findings, and technical aspects of decompression for each of the types of Chiari malformation. As such, careful patient selection is perhaps most important to achieve successful outcomes for this population. This chapter reviews the Chiari malformations, with special emphasis on these anatomic derailments in children.

HISTORY

In the early 1890s, Dr. Hans Chiari (1851-1916), professor of pathologic anatomy at the German University in Prague, used autopsy specimens to describe four congenital anomalies later termed the *Chiari malformations* (types I to IV) (Table 190-1). Chiari was not the first to observe and report the type II malformations. In *Observationes Medicae*, written by the Dutch physician and anatomist Nicholas Tulp (1593-1674), reference is made to hindbrain herniation in a myelodysplastic individual.[3] John Cleland (1835-1925) of Scotland reported a single myelodysplastic patient with hindbrain herniation and hydrocephalus in 1883. Contemporarily, Julius Arnold (1835-1915), professor of anatomy at Heidelberg, described a single myelodysplastic patient with hindbrain herniation and no hydrocephalus. Although the term *Arnold-Chiari malformation* has been used specifically in reference to hindbrain herniation in myelodysplastic patients, it was Chiari who described and attempted to delineate the pathophysiology of these posterior fossa abnormalities. As such, it is most appropriate to refer to this abnormality as the *Chiari II malformation*.[4]

TERMINOLOGY

Chiari I Malformation

The Chiari I malformation (CIM) consists of a 5-mm or more caudal displacement of the cerebellar tonsils inferior to the foramen magnum (Fig. 190-1). The most commonly associated findings are cervical syringomyelia and, on occasion, hydrocephalus (<10%). Multiple associations have been cited in the medical literature regarding this malformation. The foramen magnum potentially compresses the herniated cerebellar tissue and restricts normal CSF flow across the craniocervical junction. Some have referred to pathologic descent of the cerebellar tonsils due to raised intracranial pressure (e.g., tumor) as CIM. However, this chapter does not use such application of this term and will refer to only the congenital forms of the herniation.

Chiari II Malformation

The Chiari II malformation (CIIM) is seen almost exclusively in the setting of myelodysplasia and hydrocephalus. In type II malformations, the structures herniating through the foramen magnum include the cerebellar vermis, brainstem, and fourth ventricle (Fig. 190-2). In addition to these neural structures, the accompanying choroid plexus and the associated basilar artery and posterior inferior cerebellar arteries may also be caudally displaced. The posterior fossa is often small and the foramen magnum larger than normal, and syringomyelia is seen in many of these patients.

Chiari III Malformation

The Chiari III malformation is the rarest of the Chiari malformations, with herniation of cerebellum and brainstem into a posterior encephalocele and other intracranial anomalies that are seen in CIIM (Fig. 190-3). This is the most severe form of hindbrain herniation, and its management is often problematic from both a technical and an ethical point of view. Lesions that prominently involve the posterior fossa contents must be distinguished from high cervical myelomeningoceles, which may look the same superficially but carry a more favorable prognosis.[5,6] Patients with a Chiari III malformation generally have a poor prognosis. Severe neurological, developmental, and cranial nerve defects, in conjunction with seizures and respiratory insufficiency, are common. Surgical planning follows the same basic principles of any encephalocele closure.

Chiari IV Malformation

Chiari expanded his initial classification in a follow-up study published in 1896, in which he added a fourth subgroup consisting of cerebellar hypoplasia or aplasia (i.e., the Chiari IV malformation). In these patients, the posterior fossa is relatively normal in size, and there is an absence of any hindbrain herniation. Although this was included in Chiari's classification of rhombencephalic malformations, it is more appropriate to include this in the category of posterior fossa cysts. Therefore, details of this type of malformation will not be expanded on in this chapter.

Chiari 0 Malformation

The Chiari 0 malformation is defined as syringomyelia without tonsillar herniation that responds to posterior fossa decompression. Iskandar and associates[7] identified five patients with syringomyelia and no evidence of tonsillar herniation. MRI of the entire neuraxis ruled out other causes of a syrinx. Ultimately, abnormal CSF flow at the posterior fossa or foramen magnum was the suspected cause. All five patients underwent a posterior fossa decompression and duraplasty without direct fenestration or management of the syrinx. Significant syrinx and symptom resolution were observed in all patients. Their response to surgery suggests that "Chiari-like" pathophysiology may be present in the absence of tonsillar herniation, which may possibly be intermittent. This contention is supported by the identification of a crowded foramen magnum in two patients, multiple arachnoid adhesions in two others, and a fourth ventricular arachnoid veil in one. Each of these findings can alter CSF flow, although at present the pathophysiology resulting in syrinx formation in this group of patients is poorly understood. As such, a thorough evaluation to exclude other causes of syrinx is necessary before consideration of posterior fossa decompression. We stress that this group makes up only an extremely small portion of our practice and should be considered rare.

TABLE 190-1 The Chiari Malformations

Chiari Type	Features
I	Tonsillar herniation >5 mm inferior to the plane of the foramen magnum (basion-opisthion line) No associated brainstem herniation or supratentorial anomalies Hydrocephalus uncommon Hydrosyringomyelia common
II	Herniation of the cerebellar vermis, brainstem, and fourth ventricle through the foramen magnum Associated with myelomeningocele and multiple brain anomalies Hydrocephalus and syringomyelia very common
III	High cervical or occipital encephalocele containing herniated cerebellar and brainstem tissue
IV	Hypoplasia or aplasia of the cerebellum and tentorium cerebelli

Figure 190-1. Sagittal magnetic resonance image of child with Chiari I malformation. Note the caudalmost extent of the cerebellar tonsils displaced to the lower border of C2.

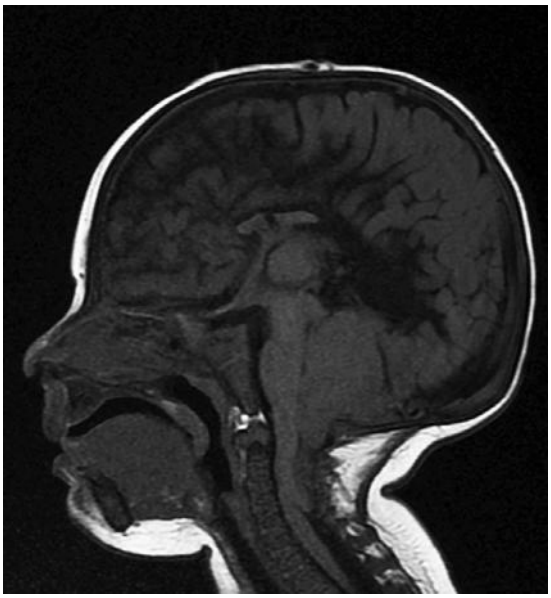

Figure 190-2. Sagittal magnetic resonance image demonstrating the Chiari II malformation. Note the hindbrain hernia into the neck and small cerebellum. Also note the absence of significant portions of the corpus callosum, a nearly vertical straight sinus, and an enlarged massa intermedia.

Figure 190-3. Child with the Chiari III malformation. The low occipital/high cervical sac containing herniated contents of the posterior fossa has ruptured, with the contents exposed.

Chiari 1.5 Malformation

Implying that only cerebellar tonsillar tissue is herniated through the foramen magnum, the CIM does not fully encompass all varieties of hindbrain herniation that also include descent of the brainstem. From our series of 130 surgical pediatric patients with CIM, we found that 17% had an additional component of brainstem descent.[8,9]

Because most cases of hindbrain herniation that come to medical attention are Chiari types I and II, the remainder of this chapter focuses on the presentation and management of these patients.

SIGNS AND SYMPTOMS

Chiari I Malformation

Patients with CIM may present with a variety of symptoms and signs ranging from headache to severe myelopathy and brainstem compromise (Box 190-1). The most common presenting symptom is pain (60% to 70%),[10-12] usually occipital and upper cervical in location,[13] and often induced by Valsalva maneuvers such as laughing, sneezing, and coughing. The typical pain is described as immediately following Valsalva and resolving shortly after the Valsalva maneuver is complete. In infants and children who are unable to communicate verbally, headaches may manifest simply as crying and irritability. Other common symptoms include weakness or numbness, loss of temperature sensation, and unsteadiness. Scoliosis due to an underlying syrinx is a common presentation. In our series of 500 surgical patients, this was found in 18%. Careful investigation reveals that many patients have some type of ophthalmologic or otologic disturbance at diagnosis. Ophthalmologic symptoms include blurry vision, nystagmus, extraocular muscle palsies, diplopia, and visual field deficits.[14-17] Otologic complaints consist of tinnitus, fluctuating hearing loss, vertigo, and nausea. Signs at presentation have included weakness, atrophy, hyperreflexia, cape-like sensory loss, ataxia,

BOX 190-1 Clinical Presentation of the Chiari I Malformation

SYMPTOMS

Occipitocervical headaches, dysesthesia
Nonradicular pain in the back, shoulders, and limbs
Motor and sensory symptoms
Clumsiness
Dysphagia
Dysarthria

CEREBELLAR SYNDROME

Truncal and appendicular ataxia

BRAINSTEM SYNDROME

Respiratory irregularities
Nystagmus
Lower cranial nerve dysfunction, including otologic disturbances
Recurrent aspiration
Glossal atrophy
Facial sensory loss
Trigeminal or glossopharyngeal neuralgia

SPINAL CORD SYNDROME

Motor and sensory losses, especially in the hands
Hyporeflexia
Hyperreflexia
Babinski response

OTHER SIGNS

Oscillopsia
Esotropia
Sinus bradycardia
Hoarseness
Hiccups
Urinary incontinence
Drop attacks
Scoliosis

BOX 190-2 Clinical Presentation of the Chiari II Malformation

NEWBORNS

(Usually asymptomatic)

INFANTS

(Signs of brainstem compression)

Stridor secondary to vocal cord paralysis
Central and obstructive apnea
Aspiration secondary to dysphagia with potential pneumonia
Failure to thrive secondary to dysphagia
Breath-holding spells with possible loss of consciousness
Hypotonia and quadriparesis
Irritability

OLDER CHILDREN AND YOUNG ADULTS

(Spinal, cerebellar, and ophthalmologic signs)

Occipitocervical pain
Hand weakness and loss of muscle bulk
Myelopathy
Ataxia
Strabismus
Nystagmus
Defects of pursuit movements and convergence
Defects of optokinetic movements
Scoliosis
Dysarthria

NEUROLOGICAL EMERGENCY

(Usually <2 years of age, commonly by 3 months of age)

Progressive neck pain
Apnea
Dysphagia
Stridor
Opisthotonos
Nystagmus
Progressive brainstem dysfunction

and lower cranial nerve dysfunction. By stretching the centrally located ventral white commissure through a syrinx, pain and temperature signals cannot cross to the contralateral spinothalamic tract. The loss of pain and temperature sensation occurs only at the levels that are served by stretched spinothalamic fibers. Down-beat nystagmus is reported to be specific for lesions involving the cervicomedullary junction. Abnormal abdominal reflexes are often seen in patients with associated syringomyelia. Children younger than 3 years are more likely to present with lower cranial nerve dysfunction.[18] This can manifest as poor feeding, failure to thrive, recurrent aspiration pneumonia, dysphagia, choking, or stridor. A diminished gag reflex is common. Vocal cord paralysis with stridor or hoarseness may be present. An inability to maintain airway patency with lower cranial nerve dysfunction may promote sleep apnea, which may be a cause for sudden death in this group.

Spinal cord dysfunction is the result of direct cord compression or syringomyelia. Based on our experience and that of others, the incidence of syringomyelia varies between 30% and 70% in CIM. Syrinx location is most often cervical, followed by cervicothoracic. Left untreated, permanent spinal cord damage can result. Progressive scoliosis is a relatively common manifestation of CIMs when there is coexistent syringomyelia.[12,19] Clinical and radiologic signs that raise the suspicion of an underlying neurological defect in a patient with scoliosis include a convexity to the left, leg or foot asymmetry, male gender or prepubescent female, and obviously, any neurological deficit.[5,6,19-23] The mechanism by which a syrinx causes scoliosis is not fully understood.

Chiari II Malformations

The CIIM occurs in most (>95%) patients with myelomeningocele and is the leading cause of death in treated myelodysplastic patients today. About one third of these patients develop brainstem symptoms by 5 years of age, and in excess of one third of those die, usually of respiratory failure (Box 190-2). In fact, as many as 20% of patients with symptomatic CIIM may present as a neurological emergency. When presenting acutely, there is dysfunction of the 9th and 10th cranial nerves, affecting respiration, swallowing, and vocal cord function; this is often accompanied by stridor, opisthotonos, and nystagmus. Symptomatic deterioration with progressive brainstem dysfunction may be irreversible and lead to death, irrespective of the type of therapy given or the speed with which treatment is instituted.[24-27] This potentially catastrophic syndrome occurs most frequently in infants younger than 2 years, particularly younger than 3 months, and has been shown to be unrelated to intracranial pressure and the size or extent of symptoms of the neural tube defect.[25] Patients with symptoms who survive the high-risk period (2 to 3 months of age) may improve and become clinically stable. Especially difficult to treat are neonates who fail to initiate adequate ventilation from birth. Although suspected of having inadequate respiratory drive centers, and therefore little or no potential for sustained independent ventilation, their management is associated with a poor outcome and difficult ethical decisions.

Unlike patients with CIM, there is a strong relationship between the type of symptoms and the age of onset. Newborns

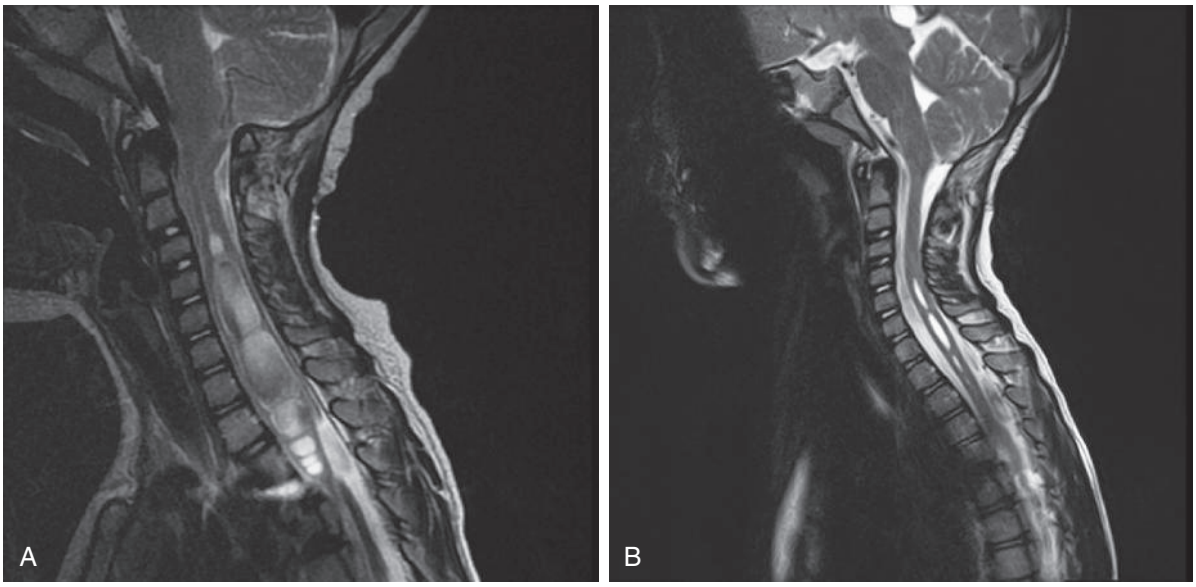

Figure 190-4. A, Child with Chiari I malformation and preoperative syrinx. **B,** Postoperative magnetic resonance image showing improvement of this patient's syrinx.

usually have no symptoms. Older children and young adults most commonly display symptoms and signs of spinal cord and cerebellar dysfunction.[4-6,28] A multitude of other symptoms and signs may occur in older patients. Common among these are ophthalmologic findings, which include strabismus, horizontal nystagmus (especially when looking upward), abnormalities of pursuit movements and convergence, and defects of optokinetic movements.[29]

DIAGNOSTIC STUDIES

Imaging

Chiari I Malformations

The diagnosis of CIM should include the absence of an intracranial mass lesion, Dandy-Walker malformation, or hydrocephalus—all of which may cause tonsillar displacement secondary to raised intracranial pressure.[30] The imaging method of choice is MRI, although some CIM cases have been diagnosed on computed tomography (CT). The true incidence of CIM is not known. However, Meadows and colleagues[31] found that of 22,591 patients who underwent MRI of the head, 175 (0.775%) were found to have tonsillar herniation extending more than 5 mm below the foramen magnum.

The position of the cerebellar tonsils in the normal population was studied by Aboulezz and colleagues.[32] In a review of 800 MRI examinations, the authors noted that normal or asymptomatic patients may have tonsils that extend 3 mm below the foramen magnum. The tonsillar herniation was noted to be clearly pathologic when it exceeded 5 mm and borderline between 3 and 5 mm. Similarly, Barkovich and colleagues[33] studied 200 normal patients and 25 patients with a "firm" diagnosis of CIM. The authors concluded that, in the absence of syringomyelia, 2 mm of tonsillar ectopia was of minimal clinical significance. Mikulis and associates[34] determined that age affects the normal position of the cerebellar tonsils, with ascent of the tonsils with increasing age.

Other associated radiologic anomalies occur infrequently and include most commonly atlanto-occipital assimilation, basilar invagination, and fused cervical vertebrae.[30,35] Despite the less frequent occurrence, these changes should be sought because they can lead to cervical instability.

In addition to the absolute position of the tonsillar tips, the configuration of the tonsil is important. The tonsillar tip may be pointed and drawn out, which may carry more pathologic significance, or it may be blunt and rounded and therefore less concerning. In general, the degree of herniation does not correlate with symptoms. Syringomyelia occurs commonly with CIM and can be seen in 30% to 70% of patients (Fig. 190-4A and B). Holocord syrinxes may present with no symptoms and only scoliosis.

Chiari II Malformations

CIIM is characterized by the elongation and caudal displacement of the cerebellar vermis and brainstem, the presence of a myelomeningocele in virtually all cases, and hydrocephalus in most patients. Syringomyelia is common in these patients and occurs in 40% to 95%.[36-42]

Associated neurological anomalies include tectal beaking, secondary to partial or complete fusion of the colliculi into a single backward pointed peak, and kinking at the level of the cervicomedullary junction (see Fig. 190-2). The latter anomaly is caused by caudal displacement of a portion of the medulla in conjunction with a spinal cord that is held in relative immobility by the denticulate ligaments. The cerebellum is usually smaller than usual, and upward herniation of the cerebellum may be evident. The choroid plexus usually maintains its extraventricular position (Fig. 190-5). Finally, callosal agenesis or dysgenesis, as well as abnormalities of the cerebral cortical pattern termed *polygyria* (not to be confused with the abnormal four-layered cortex seen in polymicrogyria), have frequently been described in Chiari II patients.[39]

In addition to hydrocephalus, the ventricular system displays multiple abnormalities. The third ventricle may be only mildly dilated and contain a large massa intermedia. The fourth ventricle is typically small or nonvisualized, flattened and elongated, and extends into the cervical canal. The lateral ventricles may be asymmetrically dilated, with prominence of the atria and occipital horns (colpocephaly), and the septum pellucidum is frequently absent. This sharpness of the frontal horn (lemon sign) and the caudal displacement of the fourth ventricle (banana sign) are relatively easily seen on in utero ultrasound examinations.[43,44]

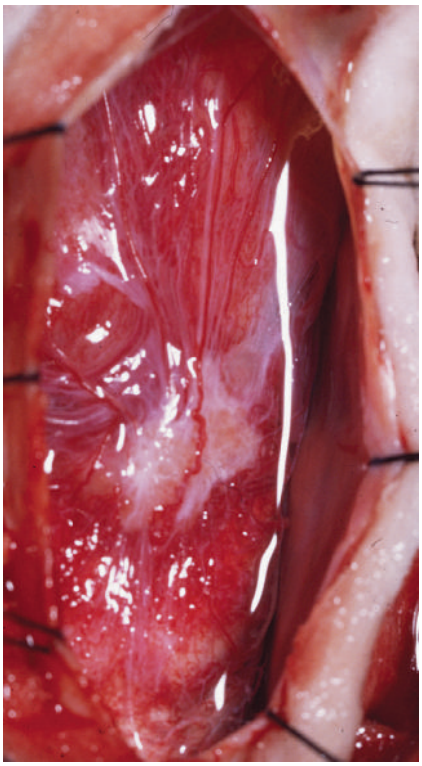

Figure 190-5. Intraoperative Chiari II malformation showing the exposed herniated cerebellar vermis. Note the extraventricular location of the choroid plexus in the center of the field.

The upper cervical canal also displays several bony and spinal cord anomalies in association with the CIIM. The posterior arch of the atlas is often missing. Klippel-Feil fusion anomalies of the cervical spine are present in a small group of patients. Basilar impression and C1 assimilation are uncommon in CIIM. Significant shortening and scalloping of the clivus can be seen.[45] Other radiographic signs of CIIM include Lückenschädel, scalloping of the posterior surface of the petrous pyramid, falx hypoplasia, falx cerebri fenestration, tentorial hypoplasia with a wide incisura and tiny posterior fossa, and enlargement of the foramen.[40,41]

Cerebrospinal Fluid Flow Studies and Cine-Mode Magnetic Resonance Imaging

One technique to assist in the surgical decision making of patients with Chiari malformations is motion-sensitive MRI, or cine-mode MRI. As opposed to static MRI, cine MRI[46-50] may demonstrate lack of CSF flow patterns that can occur in the setting of CIM. The presence of a CIM and a syrinx resulting from disturbed CSF flow, which can be demonstrated by this technique, adds helpful information to the clinical assessment.[51] Repeating the study postoperatively may help in evaluating the adequacy of the decompression.[52] Having said this, we find cine MRI of minimal utility in clinical practice and especially in patient selection for operation. Therefore, this imaging modality is no longer used at our institution.

Electrophysiologic Studies

The role of brainstem auditory evoked potentials (BAEPs) in Chiari patients has been investigated, especially in the Chiari II population. Worley and coworkers[28] studied 37 newborn infants with myelomeningoceles at a median age of 8 days and followed them for 30 months. None of the infants clinically had brainstem dysfunction at initial testing. Of 12 infants who subsequently developed brainstem dysfunction at a median age of 3 months, 11 had abnormal BAEPs on the first evaluation. Conversely, of the 25 patients who did not develop clinical signs of brainstem dysfunction, 10 had abnormal BAEPs early on in life. Multiple case reports have noted clinical and electrophysiologic improvement after posterior fossa decompression in symptomatic Chiari II patients, demonstrating objective confirmation of the observed clinical improvement.[51,53,54] The potential uses of BAEPs to evaluate neonatal myelomeningocele patients is still evolving, so this test alone is not a reliable indication for surgical intervention but instead should only be used as added criteria to a detailed history and physical examination.[55]

PATHOPHYSIOLOGY

The theories of pathogenesis of the Chiari malformations may be grouped into the hindbrain dysgenesis and developmental arrest theory, the caudal traction theory, the hydrocephalus and hydrodynamic theory of Gardner, the small posterior fossa/hindbrain overgrowth theory, and the lack of embryologic ventricular distention theory.[56]

It is unlikely that Chiari types I and II malformations share a common pathoembryologic origin. Nonetheless, the result of cerebellar descent, crowded foramen magnum, and impaired CSF flow across the craniocervical junction may share a common etiology in the development of syringomyelia.

Chiari I Malformation

Chiari's belief that hydrocephalus causes CIM has been abandoned because hydrocephalus is present in only a minority of cases. Idiopathic CIM may be the result of mesodermal defects that create a congenitally small posterior fossa, leading to compression of the neural elements and herniation through the foramen magnum. Neural dysfunction and deleterious alterations of CSF flow may ensue, resulting in the symptoms associated with CIM.

Nishikawa and coworkers[57] suggested that underdevelopment of occipital somites within the paraxial mesoderm creates a small posterior fossa and CIM. This contention is supported by the association of CIM with other spine, skull, somatic, and craniofacial abnormalities, which are the result of mesodermal maldevelopment. The association of craniosynostosis and CIM is known and appears strongest in cases of syndromic, multisuture, and lambdoid synostosis. Lambdoid suture closure, typical in Crouzon's syndrome, can directly reduce posterior fossa volume. Cephalocranial disproportion in multisuture synostosis can elevate intracranial pressure and promote herniation of posterior fossa elements.

Other medical conditions may promote formation of an abnormally small posterior fossa. Familial vitamin D–resistant rickets causes bony overgrowth of the posterior fossa, thus reducing its volume. Up to 20% of patients with growth hormone deficiency have CIM. Development of a craniospinal pressure gradient across the foramen magnum may cause or hasten the development of CIM. The gradient results from impaired CSF flow across the foramen magnum. Negative CSF pressure in the spinal compartment relative to the intracranial compartment creates a "sump effect" that forces the tonsils down through the foramen magnum. Once CSF flow is blocked at the foramen magnum, low intraspinal pressures can be accentuated and perpetuated by continuous absorption of CSF through spinal pathways, further worsening the clinical situation. Lumboperitoneal shunting, repetitive lumbar punctures, lumbar drainage, and chronic spinal CSF leaks of an iatrogenic nature are all familiar causes of an acquired CIM.[58,59] Small posterior fossae have not been identified in all patients with CIM.[60-62]

Although CIM is thought to occur sporadically, inheritable genetic factors may be responsible for a minority of cases. This conclusion is based on studies in which disease incidence is abnormally higher in some families than in the general population.

Chiari II Malformation

Because the cerebellar vermis develops before the tonsils, an abnormal pressure differential that develops in utero would cause abnormal displacement of the vermis and brainstem structures, without any tonsillar involvement. Such a pressure differential could occur with fluid leakage from the myelomeningocele.[6,63]

McLone and Knepper[56] have advanced a "unified theory," which would explain the cause of the CIIM along with most, if not all, of the associated anomalies. This theory is based on the previously mentioned assumption that the neural tube defect occurs first and that all the other manifestations, including the Chiari malformation and hydrocephalus, follow secondarily. Leakage of CSF through the spinal defect causes a lack of distention of the primitive cranial ventricular system. In experimental animals, venting fluid from the embryonic ventricular system can cause, for example, disorganization of the developing cerebral cortex and abnormal development of the pontine flexure.

Syringohydromyelia

In the 1960s, Gardner[63] presented the hydrodynamic theory. Gardner's theory stated that in normal embryology, CSF pulsations from the choroid plexus play a significant role in the expansion of the neural tube.[64] According to Gardner, these pulsations help with the development of the arachnoid pathways as well as with modeling of the expanding brain. He believed that the balance between the pulsatile flow in the supratentorial and fourth ventricular choroid plexus directed brain growth differentially; therefore, if the fourth ventricular pulsations were overactive, the tentorium would be pushed upward, and a Dandy-Walker malformation could develop. Conversely, if the supratentorial pulsations were overactive, tentorial migration would become such that the posterior fossa is small, allowing the development of a Chiari malformation.[64,65]

Based on experimental evidence using manometric measurements in normal and Chiari patients, Williams[66] expanded on Gardner's theory by suggesting that Valsalva maneuvers resulted in epidural venous congestion and intracranial as well as intraspinal pressure elevation, causing fluid to flow both cranially and caudally. Although flow into the cranial compartment meets no resistance, caudal flow is delayed by hindbrain adhesions and outlet obstruction, thus creating a pressure differential between the cranial and spinal compartments. This pressure differential may last a few seconds and cause worsening hindbrain impaction and syringomyelia. Repeat measurements were made after surgical decompression, showing equilibration of the pressures in the two compartments, which, in turn, correlated with clinical improvement. However, spinal cord cavitation is often acquired (as in posttraumatic syringomyelia), and a connection between the cyst and the fourth ventricle is not always present, which raises doubts about the adequacy of this theory.

Oldfield and associates[67] investigated the anatomy and dynamics of movement of the cerebellar tonsils and CSF during the respiratory and cardiac cycles to explore the mechanism of syringomyelia progression in patients with CIM. During systole, there is normally movement of CSF in a caudal direction across the foramen magnum to counter the increased intracranial volume of blood and maintain physiologic intracranial pressure. This flow reverses in diastole. Dynamic movement of subarachnoid fluid is mirrored by caudal and cranial pulsations of fluid within the central canal during systole and diastole, respectively. In patients with CIM, the cerebellar tonsils are forced down and obstruct CSF flow across the foramen magnum during systole. This piston-like movement of the cerebellar tonsils imparts a systolic pressure wave in the spinal CSF that acts on the surface of the spinal cord, forcing fluid into the cord through perivascular and interstitial spaces. Oldfield and collaborators[67] demonstrated the dynamic CSF flow into the syrinx in patients with CIM preoperatively by dynamic MRI and intraoperatively by ultrasound and further demonstrated the resolution of pathologic flow following bony and dural decompression. Postoperatively, adequate decompression of the foramen magnum allows resolution of the syringomyelia.

TREATMENT

Chiari I Malformation

There is no effective nonsurgical alternative to operative decompression for patients with symptomatic CIM. However, a detailed understanding of the natural history is not objectively known because the true incidence and prevalence are not well recognized. The challenge is identifying which patients will benefit most from posterior fossa decompression. Appropriate selection of patients likely to respond to surgical therapy can be challenging. Surgical indications vary among surgeons, especially for subjective symptoms such as headache.[68] A 2004 opinion survey of neurosurgeons concluded there was widespread agreement to treat patients with a syrinx and progressive scoliosis or symptoms.[69] Opinions were mixed on how to manage individuals with asymptomatic syrinxes as well as on the most appropriate surgical technique. In our experience, the farther away (e.g., lower thoracic, lumbar) from the craniocervical junction a syrinx is, the less likely it is to respond to posterior fossa decompression.

Acquiring adequate imaging and clinical data is the first step in managing this diverse group of patients, with MRI clearly the study of choice. Patients suspected of having CIM should undergo MRI of the posterior fossa and entire cervical spine to assess tonsillar ectopia and screen for a syrinx. Furthermore, patients with long-tract signs and no evidence of a cervical syrinx should also have their thoracic and lumbar spine screened. Also necessary is imaging to rule out hydrocephalus. Flexion-extension radiographs of the cervical spine are suggested for individuals in whom there is a concern for instability. This is especially relevant in this group because cervical spine anomalies are very common.

Patients determined to have idiopathic CIM are divided into two broad categories based on the presence or absence of a syrinx. All patients with a syrinx (*not* a patent central canal), regardless of the size, location, or other associated symptoms, are offered surgical intervention. This strategy is based on the belief that a syrinx is indicative of pathologic forces acting on the spinal cord that should be corrected to prevent permanent cord damage. However, a recently published study by Nishikawa and coworkers[57] challenges this notion. They followed nine asymptomatic patients who had incidentally discovered syrinxes for 11 years with serial MRIs and clinical examinations. In this period, only one patient showed any deterioration that required surgery. The syrinxes in the others did not change. The authors concluded that small, asymptomatic syrinxes can be safely followed with serial examinations and imaging.

The decision about whether to operate is a bit more complicated in patients without a syrinx. Individuals with minimal tonsillar ectopia, no objective neurological findings, and mild headaches that are not lifestyle limiting are observed. Surgery is deferred until symptoms worsen and become unacceptable to the patient and headaches are refractory to multidisciplinary management. Patients with lifestyle-limiting headaches or objective neurological abnormalities, especially respiratory or cranial nerve dysfunction, receive earlier surgical intervention.[70]

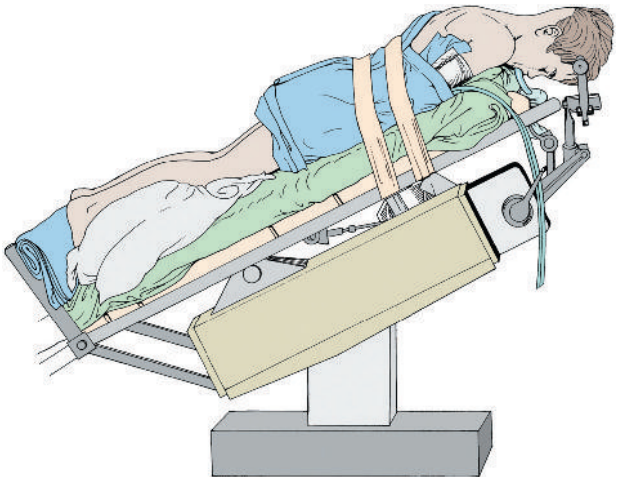

Figure 190-6. Surgical positioning for the decompression of a Chiari malformation.

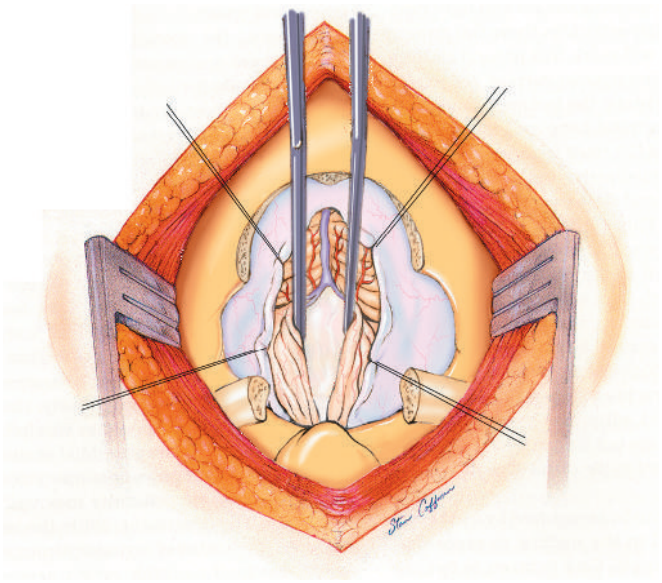

Figure 190-7. Operative exposure of the Chiari I malformation. The cerebellar tonsils are shown being spread apart so that the floor of the fourth ventricle is visualized.

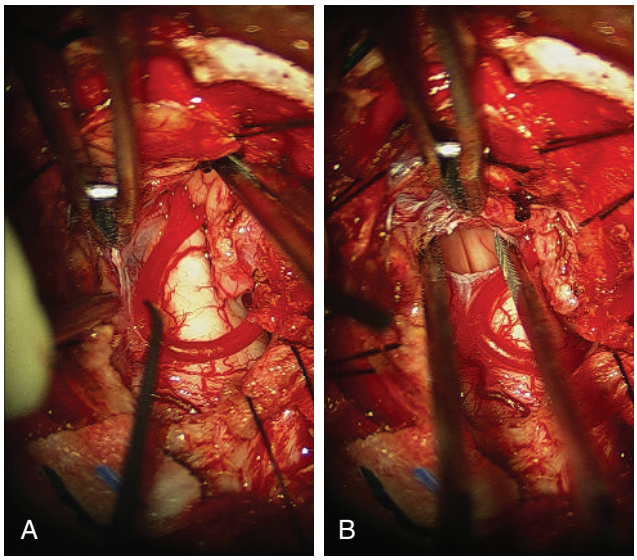

Figure 190-8. Intradural exploration of the fourth ventricle in a child with Chiari I malformation and a huge holocord syrinx. Note the arachnoid veil (at the tip of the forceps) occluding cerebrospinal fluid egress from the fourth ventricle **(A)** and penetration of this veil and visualization of the fourth ventricular floor **(B).** At 6 months' follow-up, the patient's holocord syrinx had resolved.

Some basic principles are generally agreed on. If hydrocephalus coexists or any question of raised intracranial pressure is present, this situation should be resolved before consideration is given to a decompression. Only a minority of patients (<10%) fall into this category. After shunting or third ventriculostomy, if symptoms persist or an associated syrinx is large and unchanged for months, then consideration should be given to a Chiari decompression.

After a decision has been made to do a Chiari decompression, the patient is positioned prone with the head held flexed in a pin fixation device (Fig. 190-6). The skin is clipped about 5 cm wide in the midline from the midocciput, just below the inion, down to the second cervical vertebra. A skin incision is then made along this same region, and the soft tissues and occipital musculature are separated with monopolar cautery in the midline through a relatively avascular plane (Video 190-1). The foramen magnum and posterior arch of C1 are exposed the entire width of the dura. There is no reason for exposure more lateral than this because exposure of the vertebral artery and surrounding vein on each side of C1 carries risk without benefit. Therefore, we generally remove about 2.2 cm of the posterior arch and a 2.2-cm wide × 2.5-cm long piece of occipital bone. The bone of the occiput is removed first, followed by the posterior arch of the atlas, which is removed with a bone rongeur after dissection of the soft tissues off the bone. Because we use a craniotome to remove the occipital bone, leaving the posterior arch of the atlas protects the underlying cord in cases in which the craniotome "skips." By leaving the muscle attachments and laminae of C2 intact, postoperative pain and potential spinal instability are minimized. Rarely, it may be necessary to remove the superior aspect of the C2 laminae to fully visualize the caudal tip of the cerebellar tonsil. By minimizing the opening and maximizing the decompression, we believe we have enhanced our surgical results and have not seen any cervical kyphosis or postoperative instability.

The purpose of the operation is to enlarge the bony area of the craniocervical junction and expand the dura surrounding the brainstem. By doing this, the surgeon wishes to relieve any direct compression and facilitate adequate outflow of CSF from the fourth ventricle. Some have championed the idea of bony decompression alone, or alternatively, some surgeons split the dura, opening only the outer layer. Because we have so often identified intradural pathology (e.g., arachnoid webs at the fourth ventricular outlet) as a cause of an associated syrinx, we always open the dura. After the dura is opened in the midline, the tonsils are gently separated to visualize the fourth ventricular floor (Fig. 190-7). This reestablishes free flow of CSF from the foramen of Magendie.[71] We consider visualization of the relatively avascular floor of the fourth ventricle and free flow of CSF into the subarachnoid space as evidence of adequate decompression. Occasionally, in cases with severe tonsillar ectopia, the foramen of Magendie remains occluded (e.g., from arachnoid veils) (Fig. 190-8A and B), and CSF egress is limited. In our review of 500 surgically treated CIM cases, 12% were found to have an intradural veil. Extrapial coagulation of one or both tonsillar tips shrinks the tonsils sufficiently to restore CSF flow. It is imperative to maintain complete hemostasis to minimize arachnoiditis and the risk for chemical meningitis. Rarely, and usually in cases of

surgically recalcitrant syringomyelia in which a primary operation failed to diminish the size of the syrinx, a small piece of Silastic tubing is placed from the fourth ventricle to the upper cervical subarachnoid space and secured with a suture to the pia of the cerebellum. Care must be taken to make sure that the distal tubing is placed anterior to the denticulate ligaments to avoid irritation of the dorsal roots from the tubing and resultant postoperative pain.

Finally, the need for a dural graft has been debated.[72] We believe that such a graft allows for protection of the neural tissues from chemical contamination while providing a capacious dural sac, which probably optimizes the decompression and minimizes postoperative adhesions. We prefer pericranium as the graft and harvest this tissue in a separate vertical incision just cephalad to the decompression wound. After decompression, the fasciae and skin are closed in a routine manner. Postoperative pain control is addressed with a running schedule of alternating acetaminophen and ibuprofen. In general, patients are observed in the intensive care over night and then moved to the ward for 1 to 2 days before discharge.

The likelihood of a craniocervical decompression resolving appropriate symptoms is quite high with minimal operative risk. In a similar vein, significantly sized syrinxes should clearly show evidence of decreasing in size or totally resolving within weeks to months of surgery. It is so likely that Chiari decompression will resolve the situation that an inadequate clinical outcome almost always is due to an inadequate decompression. This has been true in the cases referred to us operated on elsewhere as well as our own initial failures. In patients who initially improved clinically and radiologically with decompression, and then worsened, the most likely explanation is a reclosure of the outlet foramen, which will respond to repeat decompression and possibly resection of a portion of the tonsil rather than any other surgical approach. Raised intracranial pressure must be excluded, or if the patient has shunted hydrocephalus, adequate shunt function must be verified. In the rare patient who fails adequate decompression, reexploration of the operative site should be performed to verify an unobstructed fourth ventricular outlet. If reexamination of the operative site does not result in improved symptoms and syrinx size, consideration should be given to insertion of a syringopleural or syringoperitoneal shunt. Care should be given not to resort to this option except to resolve a syrinx that is large or is growing despite adequate decompression.

A challenging group of patients includes those in whom there is a significant component of ventral compression. We have defined significant ventral compression as greater than 9 mm of reclination of the odontoid process from a line connecting the basion to the posterior aspect of the body of the axis (Fig. 190-9).[73,74] In these patients, spinal stability should be ensured by performing dynamic cervical radiographs in flexion and extension before decompression. These patients often present with a greater degree of brainstem symptoms and may not benefit from posterior decompression alone. In our hands, dorsal decompression is addressed first with close observation in an intensive care setting postoperatively. Symptoms and signs of respiratory compromise, swallowing difficulty, and hemodynamic instability herald ongoing brainstem compression and warrant occipitocervical stabilization and possibly ventral decompression.

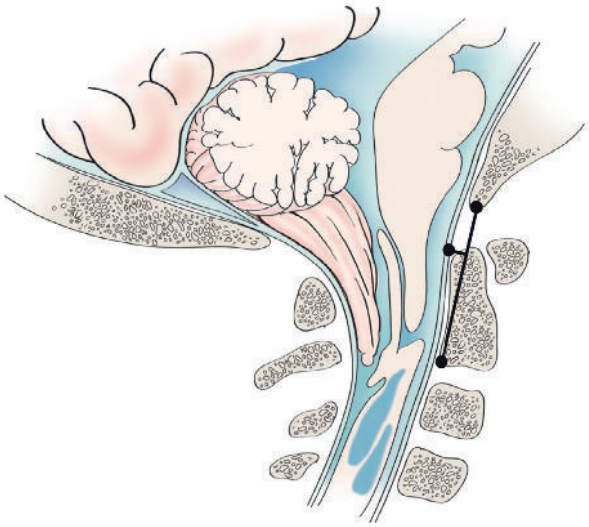

Figure 190-9. Schematic of sagittal magnetic resonance image through the craniocervical junction. Measurements used to examine for ventral compression. A line is drawn from the basion to the posterior lower border of C2, and then a perpendicular line from there to the anterior dural edge. A distance of more than 9 mm would be concerning.

Chiari II Malformation

Early surgical intervention may prove life sustaining in Chiari II patients whose symptoms are referable to the medullary dysfunction.[21,22] Timing of decompression before bilateral vocal cord paralysis may predict a better outcome.[73] Pollack and colleagues[75] have described favorable outcomes for patients undergoing early surgical intervention, that is, occipital craniectomy and cervical laminectomy for neurogenic dysphagia.

Indications for surgical intervention are based on clinical symptoms. The presence of a significant syrinx, or one that has progressively enlarged, should be considered for therapy. Symptoms include inspiratory stridor at rest or progressive by history, aspiration pneumonia due to palatal dysfunction or gastroesophageal reflux, central apnea with or without cyanosis, especially during sleep, opisthotonos, functionally significant or progressive spasticity of the upper extremities, and functionally significant or progressive truncal or limb ataxia.[27,28]

Before surgical decompression can be considered, patients with symptomatic Chiari II malformation must have physiologic intracranial pressure. This assurance requires a functioning shunt. It has been our experience, and that of others, that a properly functioning ventricular shunt can often obviate the need for decompression of hindbrain herniation. Many times, this can only be determined by surgical inspection of the shunt. Caldarelli and colleagues[24] reported that of 11 symptomatic CIIM patients, 5 had resolution of their symptoms after shunt revision alone. Milhorat and associates[76] found in a retrospective study of a small number of patients that improvement in the size of their syrinx was observed after only ventriculoperitoneal shunting or revision. Tomita and McLone[77] concluded that shunt revision can reverse acute respiratory arrest. In contrast, lower cranial nerve findings may not improve after the shunt revision but rather only after posterior fossa decompression.

One important caveat is that stable ventricular size is not a reliable indicator of a functioning ventricular shunt. Iskandar and colleagues[78] found that in 20% of myelodysplastic children with shunted hydrocephalus, CT studies were unchanged in the presence of shunt malfunction, as demonstrated by surgical exploration following continued clinical decline. After shunt revision, all patients in this myelodysplasia group had resolution of preoperative symptoms.

The Chiari II anomaly is a challenging surgical entity with unusual and highly variable anatomy. Unlike the Chiari I operation, which can be performed by any experienced neurosurgeon with a good knowledge of the anatomy of the region, a Chiari II decompression requires significant experience with this particular entity for the procedure to be done safely and efficaciously. The

main challenge in this procedure is the variable anatomy. For instance, the cerebellar tissue usually extends into the lower cervical spine; it may be very adherent to the medulla, and occasionally the two tissues may even seem indistinguishable or fused. The confluence of sinuses can be as low as the rim of the foramen magnum, and the dura may contain large venous sinuses.

After adequate shunt function is ensured, and if the symptoms are progressive or serious and due to the CIIM, consideration should be given to surgical decompression. Helpful preoperative diagnostic studies include BAEPs, swallow study, direct vocal cord visualization by an otolaryngologist, and assessment of pulmonary function including obstructive and central apnea (sleep study) as well as hypercapnic ventilatory drive by a pulmonary specialist. Thorough study of the preoperative MRI is important in planning the procedural details. The position of the confluence of sinuses, cerebellar vermis, cervicomedullary kink, and choroid plexus should be specifically identified. The extent of the bony opening should encompass the cerebellar hindbrain hernia, but need not include the medullary kink or the occipital bone—especially if the confluence of sinuses is low lying. The foramen magnum is generally enlarged in this condition, and the patient is unlikely to benefit if it is made even larger. Minimizing the bony decompression is important to reduce the concern for delayed cervical instability and postlaminectomy kyphosis.[79]

The salient aspects of the procedure are that the patient is first placed prone in a pin fixation device with the neck slightly flexed. The skin, fascia, and muscle opening are similar to the Chiari I procedure. The dura is opened in the midline, and the neural structures are exposed. Dense arachnoidal adhesions are common, as is striking superficial hypervascularity. The choroid plexus is identified by its yellow-orange color and granular appearance. It maintains its early embryologic extraventricular location and marks the entrance into the fourth ventricle. The interface between vermis and medulla is usually densely adherent and difficult to separate. The procedure is not thought to be finished until the avascular floor of the fourth ventricle is well visualized. Use of intraoperative ultrasound may be helpful in some cases.[80] The tip of the vermis is coagulated to maintain an opening out of the fourth ventricle to the subarachnoid space. The dura is grafted with periosteum and the wound closed in routine fashion.

COMPLICATIONS

Posterior fossa decompression is relatively safe but not without complications.[81] Direct vascular or neural injury, pseudomeningocele, CSF leaks, and meningitis are well recognized. Bleeding from dural venous lakes can be profuse at times. Less common complications include occipital-cervical instability, acute postoperative hydrocephalus secondary to infratentorial hygromas, and anterior brainstem compression from a retroflexed odontoid.[73,82] A complication unique to posterior fossa craniectomies is cerebellar slump or ptosis, which results from extending a craniectomy so far laterally that the cerebellum herniates through the craniectomy defect. This can cause headaches (different from typical Chiari I headaches), obstruction of CSF flow with syrinx formation, and a variety of motor, sensory, and cranial nerve deficits. Cranioplasty to buttress the cerebellum into place is the most definitive treatment. We have not seen this complication in more than 700 patients by limiting the bony removal to the width of the spinal dura. Complications in our group of 500 surgical CIM patients included persistent symptoms or syrinx found in 3%. In an attempt to more specifically categorize these complications in Chiari I patients and syringomyelia, Menezes[83] reviewed a series of 35 children and identified a set of complications associated with surgery. These included excessive bleeding from venous lakes, failure to get into the fourth ventricle secondary to adhesions, persistent variation of blood pressure and heart rate, failure to awaken, respiratory compromise, and weakness. Although any of these can occur, many can be avoided by meticulous preparation and surgical execution and a thorough understanding of the pathology.

RESULTS AND PROGNOSIS

Chiari I Malformation

With an incomplete knowledge of the natural history, surgical data accumulated from a series of retrospective studies claiming to improve on the natural history should be looked at cautiously.[84] Saez and associates[85] attempted to classify patients into preoperative prognostic categories. The poorest prognosis was seen in patients with central cord signs; the best prognosis was found in patients with paroxysmal intracranial hypertension. In our series of 500 surgical cases,[86] there were no acute returns to the operating room or blood transfusions.

Chiari II Malformation

The natural history of these lesions can be disastrous. Therefore, the idea of surgical intervention early in a child with symptoms is widely accepted, although only a few studies exploring the role of decompression in Chiari II patients are available in the literature. Pollack and colleagues[75] have prospectively treated neonates and young infants displaying symptomatic CIIM with urgent laminectomies and limited suboccipital craniectomies. Ten of the 13 children recovered normal or almost normal neurological function postoperatively, whereas the other 3 exhibited bilateral vocal cord paralysis and severe central hypoventilation. The authors concluded that early recognition and treatment of brainstem compromise in Chiari II patients produces prompt and long-term clinical recovery.

FOLLOW-UP

Chiari I Malformation

The follow-up regimen is determined by preoperative pathology and the postoperative clinical course. Our clinical paradigm includes seeing patients without a syrinx and symptomatic improvement at 1, 6, and 12 months, then every 12 to 24 months thereafter without repeat imaging. Patients with a preoperative syrinx receive a follow-up MRI in 6 to 12 months. No further imaging is obtained if symptoms improve or the syrinx decreases in size significantly. If the syrinx improves minimally, additional imaging is obtained at the surgeon's discretion. As long as the syrinx progressively shrinks and no additional symptoms or signs occur, no matter how slowly, we continue to follow the patient conservatively with imaging. If the syrinx fails to improve or symptoms referable to a persistent syrinx are present, a second surgery is performed.

From our series, up to 3% of patients required a second posterior fossa exploration for syrinx persistence.[8] Again, no attempt was made to manipulate the syrinx. The second surgery is more aggressive than the first. Unilateral tonsillar coagulation was performed for each patient. Rarely, stents are placed when free egress of CSF from the fourth ventricle is not achieved after lysis of subarachnoid adhesions and tonsil coagulation, or when no obvious pathology explaining syrinx persistence was identified. We stress that reexploration of the posterior fossa is the best strategy for dealing with a recalcitrant syrinx.

Chiari II Malformation

Although all patients with myelodysplasia are followed in our multidisciplinary spina bifida clinic, children with symptomatic CIIM require closer follow-up. Preoperative and postoperative

sleep studies are valuable in assessing the severity of central sleep apnea and response to surgical decompression. Frequently, these children require gastrostomy tubes for management of significant dysphagia. A surveillance fast-sequence MRI is performed at 3 months to evaluate ventricular size, whereas any clinical relapse is managed as a presumed shunt malfunction before any direct surgical approach to persisting syrinx. Direct shunting of persisting symptomatic syrinxes is frequently not necessary.

CONCLUSION

Chiari presented his series of hindbrain herniations more than 100 years ago and attempted to elucidate a unifying theory as to their formation. Today, we understand the Chiari malformations to involve an abnormality at the craniocervical junction resulting in impaired neural function and CSF hydrodynamics. However, the pathophysiology of each malformation is likely very different, and the management is tailored to each individual. There are some consistent simple tenets in the management of these patients: (1) patient selection is important in determining who will benefit from surgical intervention; (2) assurance of normal intracranial physiology before suboccipital decompression is paramount; and (3) restoration of normal CSF dynamic flow from the fourth ventricle to the subarachnoid space and relief of direct brainstem compression are the goals of surgery. Adequate decompression can be accomplished in a variety of ways but must allow free flow of CSF between the intracranial compartment and spinal subarachnoid space to successfully treat this patient population.

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