Chiari Malformation and Syringomyelia

6

Ambre' L. Pownall

6.1 Introduction

Chiari malformations are a group of structural abnormalities of the hindbrain which were originally described by John Cleland in 1883 and then classified in 1891 by Hans Chiari, a German professor. His work, based on autopsy results, created the classic definitions of hindbrain herniation now described as Chiari I (CM-I), Chiari II (CM-II), and Chiari III (CM-III). Although named similarly to the other Chiari malformations, Chiari IV malformation (CM-IV) is now recognized as cerebellar hypoplasia and unrelated to the others; thus, it will not be discussed (Greenberg 2010; Khoury 2015; Oakes et al. 2011; Weprin and Oakes 2001).

Syringomyelia refers to the development of a cyst or a cavity filled with cerebrospinal fluid (CSF) within the spinal cord. The cyst is also known as a syrinx. Despite advances in neuroimaging and embryological work, the natural history of Chiari malformation and syringomyelia remains incompletely understood.

CM-I consists of displacement of the cerebellar tonsils below the foramen magnum and is often associated with syringomyelia. CM-II, also known as the Chiari malformation, is associated with myelomeningocele (MM) and includes caudal displacement of the inferior cerebellar vermis, the fourth ventricle, and the medulla into the cervical canal. CM-III, the rarest and most severe form, includes a low occipital or high cervical encephalocele in combination with downward displacement of most of the cerebellum, the fourth ventricle, and possibly portions of the brainstem. Two other subtypes have been described. The Chiari 0 malformation exhibits normally located cerebellar tonsils in the presence of syringomyelia, abnormal posterior fossa anatomy, and altered CSF dynamics, analogous to Chiari I malformation. The Chiari 1.5 malformation mimics Chiari II in the absence of spina bifida (Khoury 2015). The CM-0,1.5 and III are not often seen, with CM-I and CM-II being the most common.

6.2 Chiari I Malformation

Historically, Chiari malformations were described as developmental anomalies. However, there is currently evidence to indicate that some CM-Is are acquired (Oakes et al. 2011). In addi-

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tion, debate exists about whether the term malformation, implying faulty formation and supporting the etiology as a developmental process, accurately describes the range of the Chiari phenomena (Novegno et al. 2008; Rekate 2008). Although the true incidence of CM-I is unknown, studies have reported approximately 0.1–0.05% in diagnosis since the availability of MRI (Milhorat et al. 2007).

CM-I has historically been considered to occur sporadically. However, familial clustering suggests inheritable genetic factors may be present in a small number of cases. Other genetic syndromes have been associated with CM-I, such as achondroplasia and Williams syndrome. To identify potential inheritable cases, it is important to obtain a thorough family history and consult with a genetics specialist, when needed (Fig. 6.1).

6.2.1 Developmental Anomaly

CM-I is anatomically the simplest of the Chiari malformations. Magnetic resonance imaging demonstrates descent of the cerebellar tonsils 5 mm or more below the foramen magnum and occasionally below the second cervical (C2) level (Figs. 6.2 and 6.3). Cerebellar tonsils that enter the cervical canal but descend less than 5 mm are considered cerebellar ectopia, not meeting the criteria for the diagnosis of Chiari malformation.

Skull-based deformities, such as a small posterior fossa and steep incline of the tentorium, may be present. Basilar impression or invagination, concavity of the clivus, and atlantoaxial assimilation have been associated with CM-I (Weprin and Oakes 2001). Although this historically was considered a condition of adulthood,

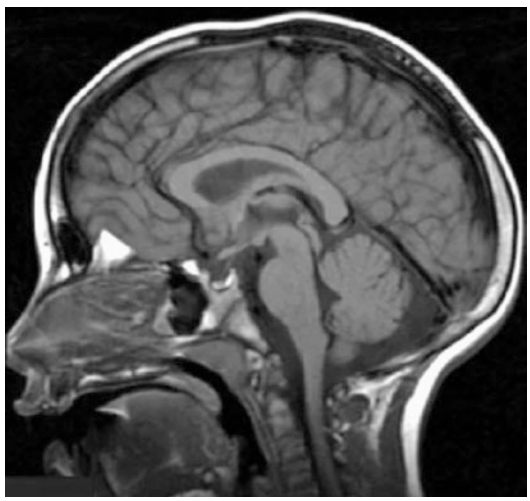


Fig. 6.1 Normal T1 sagittal MRI of a 5-year-old

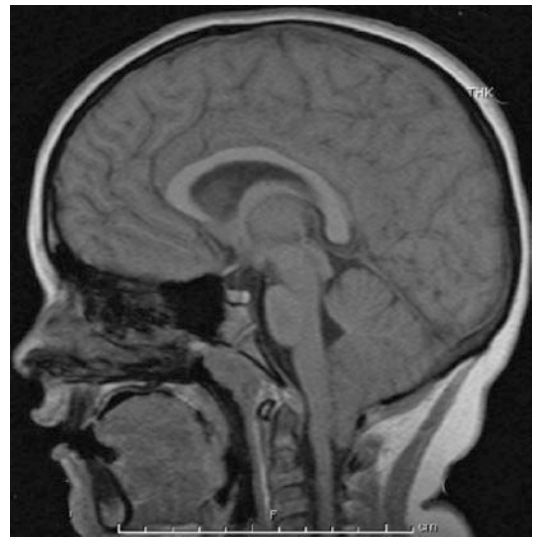


Fig. 6.2 Chiari I

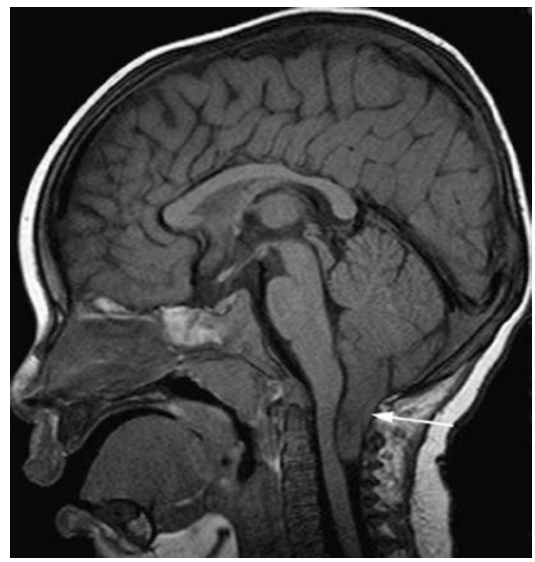


Fig. 6.3 Chiari I

CM-Is have been identified in all age groups, including the neonatal population (Lazareff et al. 2002; Menezes 1995; Nohria and Oakes 1991; Yundt et al. 1996).

The prevalence of hydrocephalus associated with CM-I is approximately 10% and may be caused by fibrous adhesions or scarring that develop between the dura, the arachnoid, and the cerebellar tonsils (Nohria and Oakes 1991). This in turn may cause obstruction of the flow of CSF from the fourth ventricle.

6.2.2 Acquired Anomaly

CM-I may develop in patients treated for hydrocephalus or pseudotumor with a ventriculoperitoneal shunt or lumboperitoneal shunt (Payner et al. 1994; Weprin and Oakes 2001). Chronic shunting of CSF from the lumbar subarachnoid space to the peritoneal cavity may cause the cerebellar tonsils to move caudally below the foramen magnum. This descent of the cerebellar tonsils may be reversed by removing the shunt. In patients with ventriculoperitoneal shunts, it has been reported that the overdrainage of the ventricles caused increased CSF in the subarachnoid space, theoretically changing the pressure gradient and contributing to the downward movement of the cerebellar tonsils. Other authors report that, with specific patients and techniques to prevent overshunting, this phenomenon can be avoided (Rekate and Wallace 2003).

6.3 Chiari II Malformation

The Chiari II malformation is present in nearly all children with myelomeningocele (MM) (Dias 1999). CM-II is probably a primary dysgenesis of the brainstem associated with the neural tube defect and multiple other developmental anomalies present in these patients (Greenberg 2010). However, there is evidence that patients undergoing intrauterine repair of the MM may not have the typical low-lying tonsils of the CM-II (Adzick et al. 2011; Sutton et al. 1999; Tulipan et al. 1998, 1999), thus placing into question the theory that this is a primary dysgenesis and giving support to the hydrodynamic theories of Chiari malformations. Indeed, the Management of Myelomeningocele Study (MOMS) (Adzick et al. 2011) demonstrated that 36% of the prenatal surgery group had no evidence of hindbrain herniation at the age of 12 months compared to 4% of the postnatal surgery group. Up to 90% of MM patients also develop symptomatic hydrocephalus, with 50% of infants showing evidence of hydrocephalus at birth (Detwiler et al. 1999). In the MOMS study, fewer CSF shunts were placed in infants in the prenatal surgery group by 12 months (40%) compared to the postnatal group (82%) ($p < 0.001$) (Adzick et al. 2011).

For these patients, the Chiari malformation appears to be more than hindbrain herniation but also includes anatomic changes in the supratentorial structures and the skull as well. The posterior fossa abnormalities include caudal descent of the pons, medulla, cerebellar vermis and fourth ventricle, "kinking" of the brainstem, "beaking" of the tectum, and aqueductal stenosis (Fig. 6.4). Some associated anomalies of the cerebral hemispheres include polymicrogyria, cortical heterotopias, dysgenesis of the corpus callosum, and a large massa intermedia. Skull deformities include "luckenschadel" or craniolacunia, shortening of

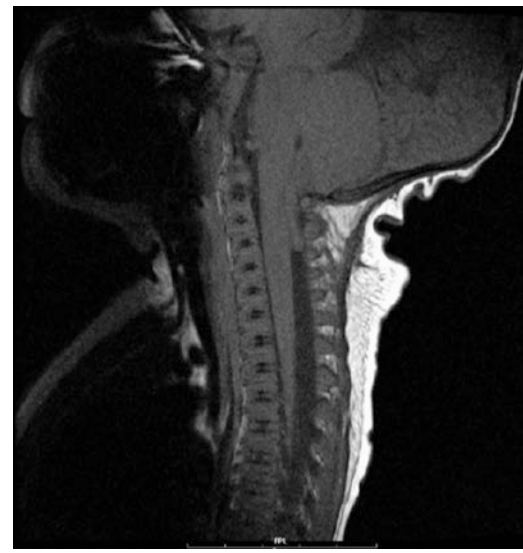


Fig. 6.4 Chiari II

bony clivus, and enlargement of the foreman magnum (Greenberg 2010). Hindbrain and lower cranial nerve dysfunction is the leading cause of death in children with myelodysplasia (Oakes et al. 2011).

6.4 Chiari III Malformation

The Chiari III malformation involves descent of most of the cerebellum and brainstem below the foramen magnum and may be associated with a cervical or occipital encephalocele. The encephalocele may contain cerebellum, occipital lobes, and brainstem. Herniation of the fourth and lateral ventricles may occur. Hydrocephalus is often present (Oakes et al. 2011; Weprin and Oakes 2001).

6.4.1 Etiology

Despite being identified in the 1800s, a debate still continues about the cause of Chiari malformations. Although these malformations have abnormalities of the cerebellum and the craniocervical junction in common, they are thought to be distinct conditions with differing etiologic factors (Greenberg 2010; Strayer 2001). Many theories about the etiology have been proposed. Ongoing research brings hope for information that will help in determining best treatment options for this challenging spectrum of disorders (Table 6.1).

Table 6.1 Etiology of Chiari malformations Nohria and Oakes (1991), Oakes et al. (2011), Fic and Eide (2015)

Theory	Mechanism
Hydrodynamic	Hydrocephalus the primary cause
Mechanical	(a) Spinal cord tethering causing abnormal development
	(b) Abnormal bony structures not providing enough space in the posterior fossa (cephalocranial disproportion)
Variation in pressure gradient	Pressure gradient between the intracranial and spinal compartments forcing the cerebellar tonsils to migrate caudally (craniospinal pressure gradient)
Traumatic birth	Birth trauma causing tonsillar edema and arachnoid scarring

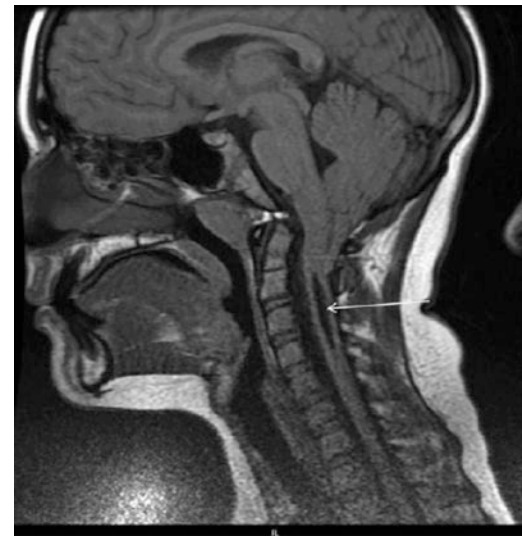


Fig. 6.5 T1 sagittal MRI showing cervical syrinx

6.4.2 Syringomyelia

Syringomyelia (or syrinx) refers to a cavitation or cyst within the substance of the spinal cord extending over many spinal levels (Figs. 6.5 and 6.6). Hydromyelia is a term that describes a distended central canal lined by ependymal tissue. The technical difference between these two terms has little clinical significance because the hydrodynamics of both types of cavitations are identical as evaluated by MRI. Therefore, medical literature currently uses the term syringomyelia to describe all intramedullary cysts with cerebrospinal fluidlike content (Oakes et al. 2011).

Although syringomyelia most often occurs in association with a posterior fossa abnormality, a syrinx can also be associated with tumors, injury, and inflammatory processes or may simply be idiopathic.

Syringomyelia is present in 30–85% of patients with Chiari I malformation (Schijman 2004) and is found most often in the older female child who has a larger degree of tonsillar descent with CSF flow impairment (Strahle et al. 2011). Oftentimes the syrinx is found in the cervical spine (Strahle et al. 2011).

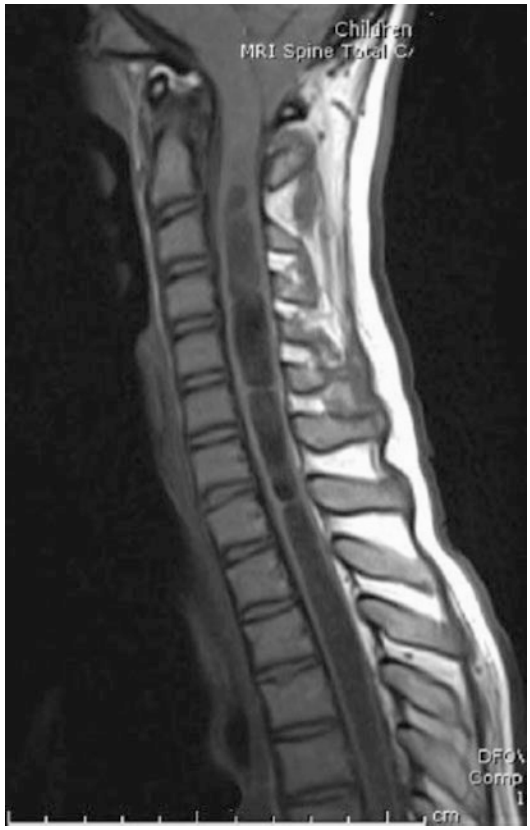


Fig. 6.6 T1 sagittal MRI showing cervical and thoracic syrinx

The medical literature has posed a variety of mechanisms for the development of the syrinx in patients with Chiari malformations. In general, there is agreement that the abnormal CSF dynamic associated with Chiari malformations produces a net bulk flow of CSF into the central canal (rather than a balanced bidirectional flow through the parenchyma) that creates the syrinx. The presence of syringomyelia will have an impact on symptom presentation, treatment options, and long-term outcomes (Dias 1999; Oakes et al. 2011; Weprin and Oakes 2001). Also, it is important to realize that a new syrinx in patients with CM-II with a ventriculoperitoneal shunt may represent a shunt malfunction causing altered spinal cord CSF dynamics.

Presyrinx, first described in 1999, is a reversible state of spinal cord edema caused by alterations in CSF flow, typically in the cervical region. Ongoing clinical examination and serial MRI imaging are used to monitor progression. The presyrinx may advance to a syrinx if untreated (Goh et al. 2008; Khoury 2015).

6.5 Presentation

6.5.1 Chiari I Malformation

Before the use of MRI, Chiari I malformations were thought to be a condition that is presented in late childhood or adolescence. Occipital and upper cervical headache is the most common presenting symptom in this age group, occurring in 63–69% of patients (Dias 1999; Hida et al. 1995). The headache may be triggered or exacerbated by Valsalva maneuver, extreme neck movement, or during exertional activities such as sports. Headaches may progress over time and, primarily in younger children, may cause nighttime awakening. Weakness or numbness of one or both arms may be present. Some patients report gait unsteadiness, sensory changes, and dysphagia.

On physical exam, nystagmus, facial hyperesthesia, dysarthria, palatal weakness, or tongue atrophy may be present. Vocal cord paralysis may be present in rare cases. Other possible findings include hyperactive upper extremity reflexes, positive Babinski, weakness of the upper and lower extremities, scoliosis, spasticity, and ataxia (Oakes et al. 2011; Weprin and Oakes 2001). The literature indicates, however, that 10% of all patients with CM-I present with headache only and have a normal neurological examination. This percentage may be higher in the pediatric population. A recent study reported findings of about 130 children with CM-I, of whom 21% presented with headache only and a normal neurological examination (Yeh et al. 2006). Diagnoses of CM-I in children and adolescents are often based on history, symptoms, and radiographic studies, in the absence of focal neurological findings.

In rare cases, ventral brainstem compression (VBSC) can occur when there is compression on the brainstem and upper spinal cord (Fig. 6.7). These patients will present with signs and symptoms similar to a CM-I such as neck pain and occipitalcervical headache. Other signs and

symptoms include myelopathy or quadraparesis, brainstem dysfunction, lower cranial nerve abnormalities, basilar migranes, ataxia, facial pain, and nystagmus (Menezes 2008; Rider et al. 2015). Treatment for VBSC includes close observation as well as surgery. Two different surgical approaches, open transoral and endoscopic transnasal, may be performed depending on the size of the patient and their imaging studies.

The availability of MRI has assisted in the identification of Chiari malformations in the younger child. Infants and the nonverbal child may present with persistent crying and irritability as well as arching of the neck. Respiratory irregularities and recurrent aspirations may, in addition to the brainstem signs noted above, mark the presentation of the youngest patients (Benglis et al. 2011; Oakes et al. 2011). One recent study identified significant differences in the presentation of children 2 years and younger compared to those 3–5 years of age. The younger age group was more likely to present with oropharyngeal symptoms (77.8–38.1%, $p = 0.01$), while the 3- to 5-year-old subjects were more likely to present with scoliosis (38.1–16.7%, $p = 0.03$) or with syrinx (85.7–27.8%, $p = 0.002$). Although more of their older subjects (3- to 5-year-olds) presented with headache, this difference was not statistically significant (Albert et al. 2010) (Table 6.2).

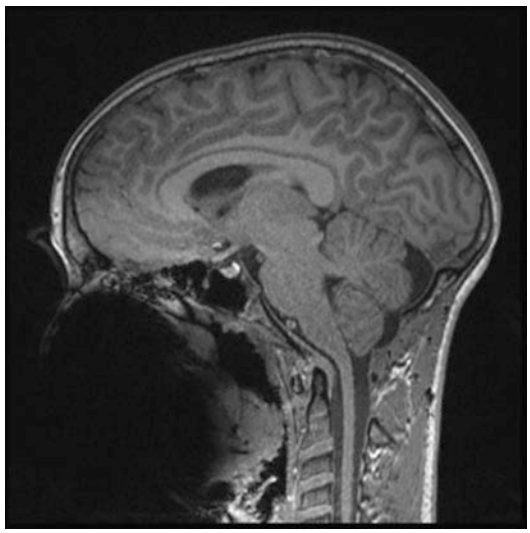


Fig. 6.7 Sagittal MRI of 12-year-old boy showing ventral compression and Chiari I. He presented with new onset of progressive left-sided esotropia and blurry vision

6.5.2 Chiari II Malformation

Chiari II malformations are present at birth in patients with an open neural tube defect. The literature reports that 18–33% of these patients will demonstrate Chiari II symptoms (Dias 1999; Weprin and Oakes 2001). The MOMS study provides evidence that prenatal repair of open neural tube defects may lessen the incidence of CM-IIs (Adzick et al. 2011). Infants with a symptomatic CM-II have a more rapid and severe onset of symptoms than those who present later in childhood. Symptom presentation early in life is related to higher morbidity and mortality.

Most patients are asymptomatic at birth, but a small group of neonates have respiratory distress. These patients demonstrate a poor respiratory drive, likely related to brainstem dysfunction. In infancy, respiratory distress including cyanotic spells, central or obstructive apnea, inspiratory stridor, and hoarse or high-pitched cries are the most common presenting signs. New or worsening stridor, accompanied by oxygen desaturation, in an infant with CM-II is considered a medical emergency.

Swallowing dysfunction is the second most common sign of a symptomatic CM-II (Dias 1999). Infants demonstrate poor suck and swallow coordination, nasal regurgitation, projectile emesis, choking, drooling, or pooling of food in the posterior pharynx. As a result, these children may suffer from failure to thrive, repeated episodes of aspiration pneumonia, and chronic gastroesophageal reflux. Nystagmus and vocal cord paralysis may also be present. This combination of symptoms reflects brainstem and lower cranial nerve dysfunction (Dias 1999; Weprin and Oakes 2001). Decreased upper extremity tone is another common sign in young children with a symptomatic CM-II.

The clinical presentation of a symptomatic CM-II in the older child is usually more gradual, with milder symptoms that are often responsive to surgical intervention. Symptoms in this age group include upper extremity weakness, spasticity, decreased function of the lower extremities, headache, neck pain, nystagmus, ataxia, and

Table 6.2 Categories of patients with Chiari I malformation based on predominant clinical symptoms

Headache	98%	Arm pain	27%
Dizziness	84%	Adbdominal pain	23%
Difficulty sleeping	72%	Photophobia	21%
Wekanes of an upper extremity	69%	Decrease or loss of hearing	16%
Neck pain	67%	Tachycardia	16%
Numbness/tingling of an upper extremity	62%	Fever	15%
Fatigue	59%	Word-finding problmes	14%
Nausea	58%	Vision loss	7.2%
Shortness of breath	57%	Blackout spells	6.8%
Blurred vision	57%	Apnea	5.7%
Tinnitius	56%	Vertigo	5.6%
Difficulty swallowing	54%	Peripheral vision loss	5%
Weakness of a lower extremity	52%	Nystabmus	5%
Depression	47%	Earache	4.5%
Vomiting	15%	Epistaxis	3.8%
Diplopia	15%	Increased snoring	3.7%
Generalized body weakness	46%	Thoracic pain	2.6%
Disequilibrium	46%	Hypotension	1.9%
Memory problems	45%	Wakes up choking	1.9%
Numbness/tingling of a lower extremity	43%	Leg pain	1.7%
Hoarseness in voice	41%	Palpitations	1.5%
Chest pain	39%	Hypertension	1.5%
Numbness in the face	32%	Absent gag	1.1%
Anxiety	30%	Face pain/tingling	0.3%
Slurred speech	28%		

Mueller and Oro (2004)

scoliosis. This group of symptoms is related to dysfunction of the cerebellum and spinal cord. Because these symptoms may progress very slowly, a complete history to identify subtle and gradual changes is vital. Presentation in adulthood is rare but would mimic the progression of symptoms of the older child (Table 6.3).

6.5.3 Chiari III Malformation

Chiari III malformations are present at birth and are identified by an occipital or high cervical encephalocele. Multiple anomalies of the cerebellum and brainstem accompany the encephalocele, which contains varying amounts of brain tissue. This anomaly is associated with poor prognosis due to the severity of the cranial nerve deficits and developmental and neurological impact. Even with supportive treatment, patients have a short life expectancy (Oakes et al. 2011; Weprin and Oakes 2001).

6.5.3.1 Syringomyelia

The neurological examination should include a thorough sensory evaluation and testing of the reflexes, in addition to strength testing. Syringomyelia should be suspected in patients that present with scoliosis, leg or foot asymmetries, or abnormal sensory examination. Dysesthetic pain of the trunk or extremities may be present. New or progressive spasticity is another symptom of concern for syrinx. Clumsiness, weakness, and atrophy of the upper extremities also may occur. In myelomeningocele patients, a worsening of urodynamics or changes in baseline motor function should be noted. In patients with CM-1, urinary incontinence may be a late sign of syringomyelia (Nohria and Oakes 1991; Oakes et al. 2011; Weprin and Oakes 2001).

6.5.3.2 Diagnostic Tests

The creation of the MRI provided a breakthrough in the diagnosis of Chiari malformations, which often present with vague and nonspecific signs and

Table 6.3 Comparison of Chiari I and II malformations

	Chiari I malformation	Chiari II malformation
Brain	Caudal descent of cerebellar tonsils > than 5–7 mm below foramen magnum	Caudal descent of cerebellar vermis, brainstem, and fourth ventricle below the foramen magnum
	Peg like or pointed	
	Often asymmetric	
Common associated radiographic findings		
Skull	Underdeveloped occiput	Craniolacunia luckenschadel
	Small posterior fossa	Lemon sign on fetal ultrasound
	+/− Enlarged foramen magnum	Small posterior fossa
	Basilar impression	Enlarged foramen magnum
		+/− Basilar impression
Spine	Assimilation of the atlas	+/− Assimilation of the atlas
	Progressive scoliosis (10% in those who also have syringomyelia)	Enlarged cervical canal
	Klippel-Feil deformity	Klippel-Feil deformity
		Scoliosis
Ventricles and cisterns	Hydrocephalus (3–10%)	Hydrocephalus (90%)
		Intrinsic malformation of ventricles including asymmetry, pointed frontal horns, and colpocephaly (enlarged occipital horns)
Spinal cord	Syrinx (40–75%)	Syrinx (20–95%)

Khoury (2015), Menezes (1999), Nohria and Oakes (1991)

symptoms. Identifying the compression of the hindbrain and cervical spine as the possible cause of discomfort in these pediatric patients, especially those that are nonverbal, aided clinicians in providing useful treatment options. Recognition of Chiari I malformation in the very young children provides them with an opportunity to benefit from advances made in the surgical approach to this condition. Cine MRI may be used to assess CSF flow around the cerebellar tonsils. The location and extent of syringomyelia is best defined by a non-contrasted spinal MRI (Sherman et al. 1999).

CT is of limited value in diagnosing Chiari malformations but provides information about the presence of hydrocephalus. In addition, cerebellar tonsillar ectopia may be noted as an incidental finding on a CT scan obtained for new symptoms such as headache or head injury. Sleep and swallow studies may be indicated prior to surgery to further evaluate the signs of brainstem or cranial nerve compression. Vocal cord motility may be evaluated if indicated.

Cervical radiographs can identify potential bony instability of the neck. Ultrasonography may provide identification of Chiari malformations and syringomyelia in the neonate and infant, but decisions about surgical intervention are based on MRI findings. Intraoperative ultrasound is used to identify whether bony decompression establishes adequate CSF flow. If CSF flow remains impaired with bony decompression, the surgery may proceed to include duraplasty and fourth ventricular stent (Sherman et al. 1999).

6.6 Treatment Options for Chiari I Malformation

6.6.1 Medical

A child diagnosed with CM-I presents a variety of challenges related to developmental considerations and the nonspecific symptoms often associated with this condition. Because the CM-I

may present with only headache, care must be taken to confirm that the malformation itself is causing the headaches. Children, as well as adults, are subject to a variety of types of headaches. Taking a thorough history of the type, pattern, and location of the headache and evaluating the effect of conservative treatment are key components of the medical management of these patients. If the headaches can be managed medically, the child may avoid a major surgical procedure. One recent review concluded that children with Chiari I malformation who are not clearly symptomatic and do not have scoliosis or syrinx can be followed conservatively. The development of symptoms and new neurological deficits were extremely uncommon in a group of 124 children followed retrospectively for 1.0–8.6 years (mean 2.83 years) without surgery (Benglis et al. 2011).

Children with known CM-I should be followed annually for evaluation of symptom development or progression. MRI imaging with cine of the craniocervical junction to assess CSF flow may be indicated. The parents and child should be advised that the child should avoid lumbar punctures that could worsen the herniation of the cerebellum tonsils.

6.6.2 Surgical

Early surgery is recommended for symptomatic patients (Hida et al. 1995). Patients who have CM-I identified on MRI, and have occipital headaches unrelieved by medical management and/or other signs/symptoms associated with Chiari I malformation, are candidates for surgery. MRI evidence of a syrinx is an additional reason for surgical intervention. Common goals are improvement of presenting symptoms, radiographic reduction of syringomyelia, and arrest or remission of associated scoliosis (Greenberg 2010). If the patient also has hydrocephalus, treatment with a CSF diversionary shunt should precede surgery to treat the CM-I.

The surgical procedure is planned to decompress the posterior fossa sufficiently to allow room for CSF to circulate around the cerebellum and the cervical spinal cord (Boxes 6.1 and 6.2). Electrophysiologic neuromonitoring is generally employed. A vertical occipital incision is made to

Box 6.1 Chiari I Malformation: Case Study

LL is a 4-year-old boy with a 2-month history of suboccipital headaches with vomiting.

He was evaluated by his primary care provider who ordered an MRI of the brain and spine. The MRI showed herniated tonsils projecting 6 mm below the foramen magnum, upper cervical cord compression, and medullary kink. Cine images demonstrated restricted CSF flow at the cervicomedullary junction. MRI of the spine showed no syrinx. LL was referred to the pediatric neurosurgery clinic for further evaluation.

Upon evaluation in the pediatric neurosurgery clinic, the patient was found to have a normal neurological exam except for an absent gag reflex. He was doing well with potty training, and his suboccipital headaches had increased in frequency, and he was now vomiting daily.

The patient was scheduled for surgery, and a suboccipital craniectomy with C1–C2 laminectory was performed. The patient was found to have large cerebral tonsils with severe compression at the craniocervical junction. Appropriate bony decompression and dural opening were achieved; the patient then had CSF flowing freely from the fourth ventricle through the craniocervical junction. Spinal monitoring was done during the entire procedure. After the surgery, the patient was transferred to the pediatric intensive care unit in stable condition.

Box 6.2 One Family's Chiari Malformation Story

It was October 31, 2001 when we first got the diagnosis of Chiari malformation for our 1-year-old son. When I heard the words "brain surgery," I felt like the air was sucked right out of my lungs. I can honestly say I remember nothing else that was told to us that day at the doctor's office. Looking back though, I guess I knew all along something was wrong but was not sure what to do because at that time Jacob was our only child and I had never been a mother before. As a baby, Jacob never really slept well, and there would be periods of crying with his eyes closed or banging his head on things that would last for hours over night. After numerous visits to our pediatrician and being told that our son was just a bad sleeper, I began to assume I was maybe not the best mother. One afternoon after a morning of crying, I decided to put Jacob down for nap. I went downstairs and heard a very loud noise. Upon entering his room, I realized Jacob had fallen out of his bed and knocked himself unconscious. In the emergency room, we were told that his CT scan looked fine from the fall but that there was a malformation at the base of his brain. Further testing was necessary, and we were told to follow up with our pediatrician to get those things scheduled. As a parent, you believe that you can protect your child from anything, but in this circumstance, that is not true. I found myself totally helpless and lost. I would be holding my son as he was put to sleep for an MRI and having no knowledge as to what they were looking at. All I needed was someone to show me a little compassion and knowledge about what was coming next, to take the time to answer my questions and put some of my fears to rest. Surgery was scheduled, and I was introduced to an angel that will always be a part of my life. A nurse at the neurosurgeon's office who was our surgeon's right-hand lady began to take the time and explain in so much detail about the steps we were beginning to take. She spent hours (it felt like) listening and answering questions. Whenever we called scared, nervous, or lost, she made us feel like no one was more important at that moment than our family. If we had not been prepared for surgery and the days after, I do not honestly think we could have survived it. For Jacob, it took two surgeries to create excellent CSF flow, and today he is just like any other 11-year-old boy. Both surgeries were different though; in the second one, we had an idea of what to expect but still spent every night watching the monitors on Jacob to make sure that he was breathing. The fear after the second surgery was not about hoping we got him home. It was about wondering if we were going to have to do this again in the future. Two and a half years after Jacob had his last surgery, our second son, Dylan, was beginning his Chiari journey. Dylan's symptoms were totally different than Jacob's, but this time I knew in my gut without a doubt what was going on. Dylan never really spoke or made sounds as an infant, had extreme difficulty drinking his bottles, and, when began moving, always dragged his left leg. As soon as I saw the leg dragging, I called this nurse who I trusted as much as our neurosurgeon. Instead of telling me I was just seeing things or that I was jumping to a conclusion too quickly, she listened to me and offered me resources to find out what was going on. Once we had an MRI showing his Chiari, we scheduled surgery. Dylan's case has always been much worse than Jacob's. To date, he has had four decompressions, and it looks like things have finally resolved. Every night though in the hospital, I would sit by his bed and cry because I felt so guilty that I could not make things better with a kiss (like moms are supposed to), and the following day, that same nurse would check in and remind me that things will get better. Chiari malformation is a frightening diagnosis to any parent and is not an easy recovery the first couple of days after surgery. At that moment in a family's life, the only thing they focus on is their child and getting him or her healed. Compassion, knowledge, and recognition are things that can assist every family during their journey.

allow for bony decompression of the foramen magnum. Initial suboccipital craniectomy may be followed by cervical laminectomy (Greenberg 2010). If the removal of bone allows for adequate CSF flow, as determined by intraoperative ultrasound, the procedure may be completed at this stage (Sherman et al. 1999). If there is continued evidence of impingement on the brainstem and cerebellum, the surgeon may perform a variety of procedures to further decompress the space (Fig. 6.8). This may include intradural exploration, partial dural removal or scoring, duraplasty with graft material or pericranium, plugging of the obex, shunting of the fourth ventricle, and coagulation of the cerebellar tonsils (Dias 1999; Narvaro et al. 2004; Sherman et al. 1999).

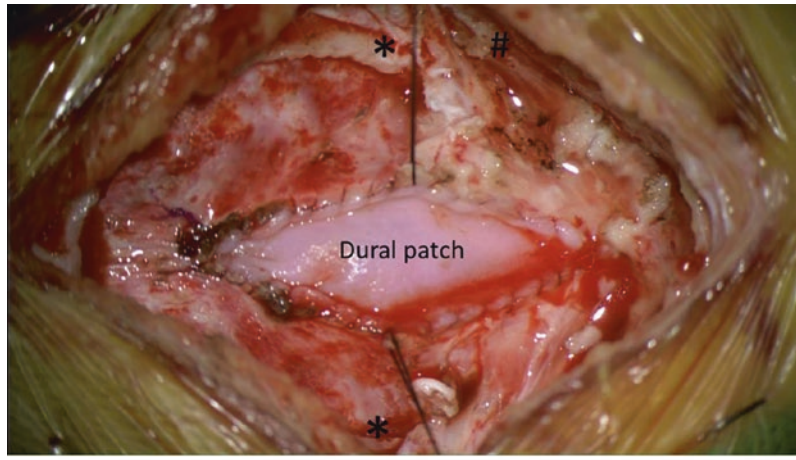
6.6.3 Nursing Care

The main concerns for nurses taking care of these patients postoperatively are pain management and respiratory compromise, which may be aggravated by narcotic pain medications. In addition, if the dura was opened, the patient is at risk for CSF leak and infection. Surgical treatment without disrupting the dura is limited to the pediatric population and has decreased the incidence of postoperative complications (Sherman et al. 1999). The patient is monitored in the intensive care unit (ICU) for prevention and early detection of potential complications and initiation of pain management.

Pain and stiffness of the neck are due to the incision through the semispinalis capitis and splenius capitis muscles, as well as from opening the dura. Pain management in the early postoperative period includes use of narcotics, either from "as-needed" medications or by patient (or parent)-controlled analgesia (PCA). When the patient is able to tolerate oral medications, adding nonsteroidal anti-inflammatory medication scheduled around the clock can improve pain scores and decrease the need for narcotics for breakthrough pain. Antispasmodics for neck spasm may also be indicated. Encouraging the patient to turn his or her head frequently so it doesn't stay in a "fixed" position is recommended.

When surgery includes duraplasty and a fourth ventricular stent, intraoperative stimulation of the area postrema located near the fourth ventricle often causes nausea and vomiting. Scheduled antiemetic medications given around the clock are indicated.

Monitoring patients for respiratory compromise is vital. The combination of potential irritation to the brainstem and the need for narcotics can make these patients susceptible to having a



* Bone edges of suboccipital bony decompression at foramen magnum; # one end of C1 laminectomy;

Fig. 6.8 Dural patch graft (Courtesy of Dr. Rongsheng Cai)

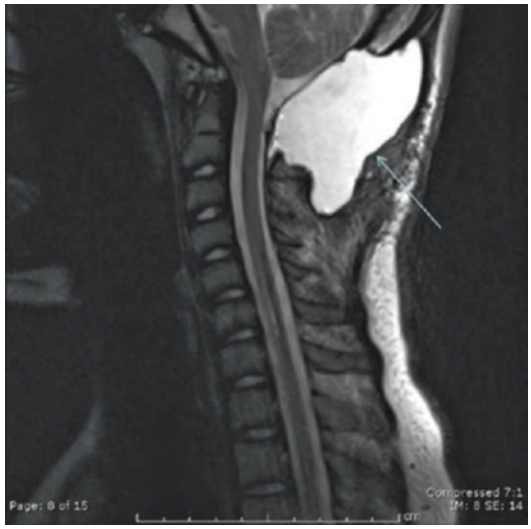


Fig. 6.9 T2 sagittal MRI of pseudomeningocele

decreased respiratory drive. ICU monitoring until most of anesthesia effects are eliminated allows for close assessment and early intervention to decrease the risk of complications.

Pseudomeningocele is the most common surgical complication when the dura has been disrupted (Fig. 6.9). This occurs when CSF leaks into the subcutaneous space, causing a fullness of the surgical site (Sherman et al. 1999). To minimize the risk of CSF leak in patients with dural compromise, the operative site should be closely monitored. A short course of dexamethasone may minimize symptoms from postoperative edema. Another possible complication is chemical meningitis (or aseptic meningitis). The symptoms include nuchal rigidity, low-grade fever, and headache. If bacterial meningitis has been ruled out, a short course of dexamethasone is the treatment of choice. Chemical meningitis after surgery for Chiari I malformation may be related to the use of dural graft material and/or tissue sealants (Parker et al. 2011).

The usual hospital length of stay after a Chiari decompression is 3–5 days. Discharge criteria include normothermia, adequate oral fluid intake, and pain control with oral medications. In addition, it is particularly important for patients who have undergone duraplasty to have a bowel regime that keeps their bowel movements soft and regular to prevent disruption of the surgical site by straining.

Resolution of symptoms such as headache may be immediate, but other symptoms may take up to 3 months to begin to resolve. Symptoms resulting from long-standing brainstem compression do not always completely resolve (Oakes et al. 2011).

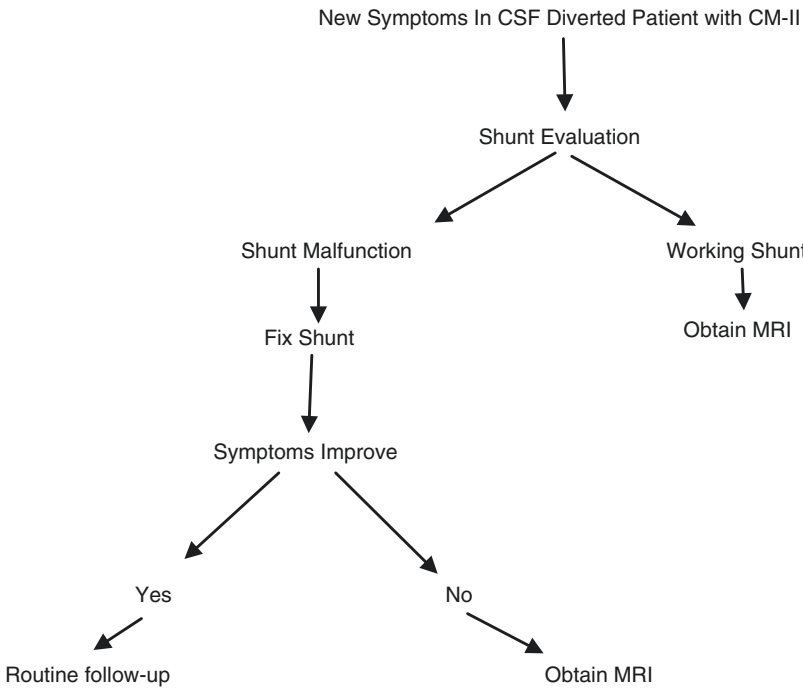
6.7 Treatment Options for Chiari II Malformation

6.7.1 Medical

Imaging for the Chiari II malformation is indicated only when new symptoms occur or when baseline status deteriorates. Symptoms of concern may include swallowing difficulties, weakness or increased weakness of the upper extremities, new spasticity, or occipital headaches. If the child has shunted hydrocephalus, the shunt should be evaluated first and revised if it is malfunctioning. In the presence of a functioning CSF shunt, evaluation of the CM-II by an MRI of the brain and craniocervical junction is the next step. If the symptoms persist and the MRI shows brainstem compression or obstruction of CSF flow, surgical treatment is indicated. In CM-II, early surgical intervention in the child with symptoms may prevent significant morbidity and mortality (Oakes et al. 2011).

6.7.2 Surgical

The surgical intervention for the CM-II parallels that for the Chiari I malformation. Based on the need for extensive dissection of the brainstem and cranial nerve structures, electrophysiologic neuromonitoring is generally employed. A suboccipital incision is made to allow for removal of the posterior arch of C1 and excision of any extradural constrictive band. Laminectomy is performed to the level of descent of the cerebellar tonsils, which may require a 1-, 2-, or 3-level laminectomy. Myelomeningocele patients, unlike the Chiari I malformation patients, have an elongated



foramen magnum and, thus, do not require further expansion. The dura should be opened to create CSF flow around the CM-II. Commonly, extremely dense arachnoidal adhesions require lysis. The herniated tonsils may require fulguration (cauterization). A stent spanning the obex, lying within the fourth ventricle proximally and the cervical subarachnoid space distally, is frequently placed. Finally, a dural augmentation graft (allograft or autograft pericrania) is typically sutured into the opened dural margins.

6.7.3 Nursing Care

As with the CM-I, these patients are monitored after surgery in the ICU or the neonatal intensive care unit. Extensive microsurgical manipulation, involving multiple lower cranial nerves and brainstem structures, places the patient at risk for postoperative neurological deterioration, especially regarding swallowing and ventilation. These patients must be monitored for late extubation, apnea, swallowing dysfunction, and feeding problems after surgery. The risk for CSF leak and infection exists when the dura has been opened. Neck movement limitation and steroids may be indicated to minimize symptoms related to dural opening and postoperative edema. Neck pain and stiffness occur in these patients and must be managed carefully in light of the presence of respiratory compromise preoperatively, especially in the very young patients.

Like CM-I patients, discharge criteria include normothermia, adequate oral fluid intake, and pain controlled with oral medications. A bowel regime is needed to keep stool soft and regular to prevent disruption of the surgical site by straining.

6.8 Treatment Options for Syringomyelia

Left untreated, a syrinx can enlarge or elongate over time causing damage to the spinal cord. When syringomyelia is associated with a Chiari malformation, treatment by posterior fossa decompression of the hindbrain malformation may result in resolution of the syrinx. Primarily in CM-II, symptomatic syringomyelia may persist despite decompressive surgery. In the absence of Chiari malformation, asymptomatic syringomyelia may

be observed clinically with yearly clinical examinations and intermittent MRI.

Direct shunting of the syrinx may improve symptoms in those patients who have persistent symptoms after successful posterior fossa decompression, or in those patients who have a symptomatic syrinx without a Chiari malformation. Options include syringoperitoneal, syringopleural, and syringosubarachnoid shunts. The shunt acts to decompress the fluid buildup within the spinal cord, diverting the fluid to another space for reabsorption (Menezes 1999). Similarly, stenting across the obstructed fourth ventricular obex can prevent "water hammering" of CSF into the proximal cervical central canal.

6.8.1 Nursing Care

Postoperative care includes incision care, pain management, and evaluation of shunt function. With syringoperitoneal shunting, there will be an incision over the spine at the level of the syrinx and an incision over the abdomen for insertion of the distal catheter. Abdominal pain and bowel function are key areas for nursing assessment.

The syringopleural shunt will have a similar back incision with the distal catheter incision in the lateral chest. Observation of respiratory status is important with this treatment option. Decreased breath sounds and oxygen desaturation may indicate a symptomatic pleural effusion. Indeed, small pleural effusions are typical and generally well tolerated. The patient may have mild tachypnea and low oxygen saturations and may require nasal cannula oxygen supplementation for up to 1 week. If tachypnea or desaturations worsen, the patient may need more intervention including thoracentesis or removal of shunt from the pleural space. Serial chest radiographs may be used to evaluate the patient's ability to accommodate the pleural fluid being diverted by the shunt.

The syringosubarachnoid shunt requires only one incision to accommodate both the proximal and distal catheters and may be effective in symptom relief. The use of a shunt to treat syringomyelia requires ongoing follow-up to observe for signs of shunt failure (Boxes 6.3, 6.4, and 6.5).

Box 6.3 Case Study Progress Note

S: Pt's mom and dad at bedside, state he did well after surgery but has really struggled with pain since around 0200. N/V has been better controlled since MN. Pt is resting comfortably at the moment, in NAD.

O: HR: 1150, BP: 102/72, RR: 19.

Pt wakes easily, states pain is a 5/10, mainly at incision area.

Dressing has a scant amount of old blood on it, no fluid collection noted.

Medications: acetaminophen 420 mg IV, every 6 h.

Valium 3 mg IV, every 6 h.

Oxycodone 3 mg PO, every 4 h, PRN mild pain (received three doses in past 24 h).

Morphine 3 mg IV, every 2 h, PRN severe pain (received five doses in past 24 h).

Zofran 4 mg IV, every 6 h PRN nausea/vomiting (received two doses in past 24 h).

A: 4-year-old CM with CM-I, POD 1 Chiari decompression.

P: Keep in the PICU this morning.

Will add ketorolac (Toradol) 15 mg IV every 6 h, PRN moderate pain, if pain is better controlled, O2 sats stable and requiring less IV narcotics and then pt. can tx to floor.

Encourage clear liquid diet as tolerated, keep MIVF. Will add stool softener.

Box 6.4 Cont. Case Study Progress Note

S: Pt's mom at bedside, pt. is on his way back from the playroom. Mom states he had a great night and is much improved. Minimal n/v (only when taking pain medications on empty stomach).

O: HR: 80, BP: 100/69, RR: 18.

Pt AAO x3. States pain is a 2/10, mainly at incision area.

Dressing was removed, no fluid collection. Incisions is clean, dry, and intact. No

redness, drainage, or swelling noted. Sensate to light touch. Stiff neck with ROM.

Medications: acetaminophen 350 mg PO, every 4 h, PRN (received three doses in past 24 h).

Valium 3 mg PO, every 6 h, PRN (received two doses in past 24 h).

Oxycodone 3 mg PO, every 4 h, PRN mild pain (received one doses in past 24 h).

Morphine 3 mg IV, every 2 h, PRN severe pain (received 0 doses in past 24 h).

Zofran 4 mg IV, every 6 h, PRN nausea/vomiting (received two doses in past 24 h) docusate 50 mg, once daily.

A: 4-year-old CM with CM-I, POD 3 Chiari decompression, doing well with limited n/v, good pain control.

Limited ROM of neck.

P: Ok to D/C home with mom and dad.

Will send home with valium and oxycodone in limited quantities since he is not taking many at this time. Encouraged patient to take tylenol and colace as needed for pain and constipation.

Encouraged oxycodone to be taken with food to avoid upset stomach and for severe pain.

Incision care instructions were given.

Return to school note, and sports and activities restrictions were given. Follow-up appointment was made. PT consult prior to D/C for home exercise.

Box 6.5 Cont. Case Study

At LL's first postoperative appointment 2 weeks after surgery, his parents reported improved sleep and headaches as well as a resolution of the vomiting. His surgical site was healing well without redness, drainage, or swelling, and the absorbable sutures were beginning to fall out spontaneously. He had full ROM of his neck and was taking acetaminophen one to two times a day.

Six months after surgery, a follow-up brain MRI revealed "near-complete resolution of the brainstem and upper cervical cord compression with resolved medullary kink, adequate CSF at the craniocervical junction, and resolution of low-lying tonsil".

LL will have follow-up with the neurosurgeon again in 18 months.

Patient and Family Education

1. Informed consent: major risks of surgery include bleeding, CSF leak, infection, persistence of symptoms, neurological deficit, and anesthesia complications.
2. Preoperative history and physical examination.
3. Preoperative diagnostic tests that may include swallow evaluation, sleep study, MRI, and developmental assessment.
4. Educational handouts about Chiari malformations and website information recommendations.
5. Incision care after dressing removed.
6. Sutures either dissolvable or removed in about 2 weeks.
7. Activity restrictions: no driving while on narcotics or while neck is stiff; return to school or work once cleared by neurosurgeon, usually between 2 and 4 weeks.
8. Follow-up imaging: MRI in 4–6 weeks and then annually for 5 years (more frequently if syrinx present).
9. Signs and symptoms of shunt failure, for patients requiring shunting of the syrinx.
10. Discharge instructions: incision care with observation for infection or pseudomeningocele; call surgeon's office for headache not responsive to medication and fever greater than 101 °F.

6.9 Outcomes: Short and Long Term

CM-I: Successful decompression can provide relief of headache. Symptoms due to cranial nerve or brainstem dysfunction can show improvement over several weeks to months. Follow-up swallow studies are useful to evaluate the effects of treatment when done 6 or more weeks postoperatively. Ataxia or weakness may also gradually improve. Patients with symptoms other than isolated headaches on presentation benefit from appropriate therapies postoperatively, such as occupational, physical, and/or speech therapy.

MRI imaging should demonstrate improvement in CSF flow around the craniocervical junction by approximately 6 weeks after surgery. A syrinx should radiographically resolve or decrease in size within 3–6 months of posterior fossa decompression. Symptoms may persist in spite of the radiographic improvement (Dias 1999).

CM-II: Better outcomes occur with older children who present with cerebellar dysfunction, spasticity, and weakness. Results in the neonatal and infant population have been varied, but in general, their outcomes are poorer. CM-II may cause death by respiratory failure (Dias 1999). The rapidity of neurological decline and immediate preoperative neurological status are the most important factors affecting prognosis.

Conclusion

The spectrum of Chiari malformations and syringomyelia present a continuum of challenges to the pediatric patient. The range of effect on quality of life varies from mild, with effective treatments available, to very severe, with minimal or no benefit from medical intervention. Advances in radiographic imaging and surgical techniques have provided opportunities to improve the health status of many of these patients. Advances in nursing research provide the opportunity for nurses and the allied health professionals to further enhance functional level and optimal development of children with this varied spectrum of disorders. Incorporating best practice for the pediatric neurosurgical patient in the areas of wound healing, pain management, prevention of postoperative complications, and effects of hospitalization on development and psychosocial wellness will further enhance the quality of life of this young population.

Pediatric Practice Pearls

1. For pain management after posterior fossa decompression, start nonsteroidal anti-inflammatory medications when the patient is taking fluids orally. Starting scheduled IV acetaminophen immediately postoperatively can significantly reduce the amount of opioids taken and controls pain well.
2. May need antispasmodics ordered due to muscle spasm after Chiari decompression.
3. Straining with constipation can disrupt the surgical site and is particularly a risk when the dura has been disrupted. Start a bowel regime when the patient is taking fluids orally to avoid constipation.
4. Relaxation techniques and gentle massage can be helpful during recovery from posterior fossa decompression. Muscle spasms often complicate the pain cycle.
5. For patients recovering from Chiari decompressions, place devices such as the TV remote, tablets, and phones on the side of the bed to encourage the patients to turn their heads once outside of the acute postoperative stage to avoid their necks becoming stiff.
6. Involve physical therapy prior to discharge for range of motion exercises to be done at home to avoid a stiff neck.
7. Avoid wearing hats, especially in summer months, that could result in skin breakdown and introduce bacteria on sweaty skin until incisions are well healed.

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