Surgical treatment of Chiari I malformation: indications and approaches

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Chiari I malformation is a well-described entity characterized by hindbrain herniation through the foramen magnum. Although the exact origin of congenital Chiari I malformation is unknown, it appears to be caused by a mismatch between the volume of the posterior fossa neural elements and the posterior fossa cranial content. Several theories have been proposed to describe the resultant pathophysiology of this mismatch. It is clear, however, that abnormal cerebrospinal fluid flow and velocity play a role in the symptoms and signs associated with this disorder. The authors will review the pathophysiology, clinical presentation, and treatment options for patients with Chiari I malformation.

KEY WORDS • Chiari malformation • syrinx • posterior fossa • decompression surgery • dural graft

In 1891, Chiari presented his series conducted in autopsy examinations of patients with cerebellar ectopia in which he classified Type I as “elongation of the tonsils and medial parts of the inferior lobes of the cerebellum into cone-shaped projections, which accompany the medulla oblongata into the spinal canal.”10,35 Chiari I malformation is often described as caudal displacement of the cerebellar tonsils below the level of the foramen magnum with or without syringomyelia. There are several theories that have been developed to explain both the cerebellar tonsillar descent and the frequently associated syringomyelia. 1–3,5,7,8,17,26,28,37,41,42,44–47,51,53,55 Analysis of the data in the literature suggests several factors: 1) the ratio of the posterior fossa neural element volume to posterior fossa cranial volume is increased. This is related to either an underdeveloped occipital somite originating from the paraaxial mesoderm, which results in a smaller posterior fossa, or to an overgrowth of the supratentorial component and consequent shallow posterior fossa.1,3,5,7,8,17,26,28,37,41,42,44–47,51,53,55 2) The CSF flow across the foramen magnum is abnormal during systole and diastole and, thus, increased tonsillar velocity and pulsation occur.2,5,47,53 3) The normal CSF flow in the spinal compartment is prevented.

Gardner 20 proposed his hydrodynamic theory in which CSF pulsations are present in the developing brain. Failure of rhomboid roof perforation or medial and lateral foramina development results in a noncommunicating hydrocephalus. Increased pressure in the supratentorial compartment causes a shallow posterior fossa to develop due to displacement of the tentorium. Hindbrain herniation ensues because the mismatch volume and pulsations of CSF force open the central spinal canal.20

Oldfield, et al.,47 have suggested a different mechanism by which syringomyelia forms. Hindbrain herniation prevents the CSF flow to and from the spinal compartment that otherwise occurs during the cardiac cycle. Obstruction at the cervicocephalad junction results in increased CSF pressure in the spinal compartment during systole, and fluid is forced into the spinal cord through its surface.47

All of the aforementioned theories are supported, in part, through the success of surgical intervention performed to correct these pathological abnormalities. That new theories continue to be proposed nearly 30 years after Gardner's publication confirms that the origin of Chiari I malformation remains controversial and incompletely understood.

The pathophysiology of acquired Chiari I malformations, in most cases, is better characterized. These malformations are thought to be caused by a pressure gradient across the cranial and spinal CSF compartments, most commonly due to CSF diversion in the spinal component or increased cranial pressure, as seen in cases of supratentorial mass lesions or hydrocephalus11,29,34,49

PRESENTATION AND EVALUATION OF PATIENTS WITH CHIARI I MALFORMATION

There have been case reports of patients with unusual presentations, including hypoglycemia,30 developmental delay,4 paroxysmal rage,30 precocious puberty,31 glosso-
mental delay. Ten patients presented with a syrinx. We
could all be exacerbated by physical exertion or neck movement. 

The majority of patients present with symptoms and signs 
that can divided into two subsets: 1) posterior fossa compres-
sonally as dizziness, sleep apnea, respiratory dysfunction, 
aspiration, vocal cord dsmotility, extracoccer eye move-
ment abnormality, hearing loss, dysphagia, and dysarthria. 

Cerebellar compression manifest as nystagmus, ataxia, 
dizziness, and dyarthria. Spinal cord dysfunction is caused 
by canal compression or syringomyelia. Generally, in ap-
proximately 50% of patients with Chiari I malformation 
and in up to 90% of those with spinal cord symptoms an 
associated syrinx will be present. 

Initial workup includes examination of supratentorial 
structures to ensure the absence of a mass lesion or hydro-
cephalus. In a patient in whom a shunt of any kind is in 
place, it is important to determine that the device is func-
tioning adequately. To assess causation and to allow pre-
surgical planning, it is important to evaluate spinal sta-
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The use of cine cardiac–gated magnetic resonance im-
aging to assess CSF flow can be helpful, especially in a 
patient in whom a decompressive procedure has been pre-
viously performed. 

OUR EXPERIENCE 

Between 1998 and 2001, we examined 21 patients sur-
gerically treated for Chiari I malformation. The average age 
was 9 years (range 1–20 years), and the mean follow-up 
period was 16 months (range 1–48 months); 20 cases were 
congenital and one was acquired after the placement of a 
lumbar peritoneal shunt. Twelve (57%) were female and 

used the following surgical approach (Fig. 1): a limited 
suboccipital craniectomy and C-1 laminectomy with 1) 
skorning of dura but leaving it intact (three patients [14%]); 
2) durotomy with duraplasty (four patients [19%]); or 3) 
intradural exploration, resection of cerebellar tonsils, and 
duraslapping (12 patients [57%]). One patient, initially treated 
elsewhere, undergone a repeated posterior fossa decom-
pressure and cranioplasty for cerebellar subsidence into 
the previously made decompression site. In one patient 
with acquired Chiari I malformation, the lumboperitoneal 
shunt was replaced with a ventriculoperitoneal shunt sys-
tem. As of last follow-up examination symptoms had re-
solved in 14 patients (67%), improved in six (29%), and 
remained the same in one patient. Scoliosis was stable in 
two and progressive in one; no patient required spinal 
fusion. Of the 10 patients with syringomyelia, the syrinx 
resolved in eight, decreased in one, and remained the same 
size in one. There were two patients with complications: 
one with pseudomeningocele and meningitis and one with 

SURGICAL APPROACH 

Many different approaches, which have evolved over 
time, have been described in the literature. In the past, 
plugging of the obex, placement of a stent in the fourth 
ventricle, extensive posterior fossa craniectomy, and mul-
tiple cervical laminectomies have been suggested as vi-
able treatment options of Chiari I malformations. In an 
effort to define the current surgical strategies, the mem-
bers of the Pediatric Section of the American Association 
of Neurological Surgeons were surveyed as to the surgical 
approach they used in pediatric patients with Chiari I mal-
formation. Only 9% recommended performing decom-
presive surgery in asymptomatic patients. For the treat-
ment of symptomatic patients, various approaches were 
used: approximately 20% recommended only osseous de-
compression; 30% recommended osseous decompression 
with dural grafting; 25% performed osseous decompre-
sion with dural grafting and intradural dissection of ad-
hesions; and 30% recommended osseous decompression 
with dural grafting, intradural dissection, and tonsillar 
manipulation and resection. Those surveyed tended to per-
form a more aggressive decompression and intradural dis-
section when a syrinx was present. Although these data 
indicate the current practice regimen among neurosur-
geons, there have been no prospective studies in which 
one treatment form is directly compared with another. 

Simple decompression is advocated based on histologi-
ical analysis of the dural band. Compared with healthy 
controls, the authors found an increase in collagen fibers, 
hyalin sup nodule, and calcification in the dural band of 
patients with Chiari I malformation. The success of the 
compressive procedure should be confirmed using in-
traoperative ultrasonography and further decompression 
should be performed if the ultrasonographic findings 
prove it to be inadequate. Further decompression can 
be achieved by scoring the dura’s outer leaf and thus pro-
moting expansion. The extent of osseous decompression 
is important. If too much bone is removed, patients are 
placed at risk for cerebellar subsidence and recurrence of 
symptoms. Tokuno, et al., attempted to address the small 
posterior fossa by conducting an expansive suboccipital 

TABLE 1

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<thead>
<tr>
<th>Factor</th>
<th>Signs &amp; Symptoms</th>
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<tr>
<td>posterior fossa compression</td>
<td>headache, neck pain, cranial nerve dysfunction, cerebellar signs, respiratory difficulty &amp; sleep apnea, dysphagia dysorthesia, dissociated sensation, sensory level, scoliosis, spasticity, loss of fine motor function, bladder &amp; bowel dysfunction</td>
</tr>
<tr>
<td>spinal cord dysfunction</td>
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We recommend performing a limited suboccipital craniectomy to enlarge the foramen magnum and allow for both decompression and maintenance of the posterior fossa neural elements.

Krieger, et al., 36 recommend durotomy without duraplasty and have reported surgery-related outcome and complications comparable with those achieved by authors who advocate duraplasty, plugging of the obex, and tonsillar resection. Other authors have emphasized that duraplasty is essential for the prevention of scar formation and recurrent symptoms.13,14,16,18,21,41,43,48 Debate still exists as to whether, once the dura is opened, arachnoid dissection should be performed. One benefit of arachnoid dissection, in the setting of Chiari I malformation, is that it allows the surgeon to release adhesions that could potentially contribute to obstruction of CSF flow from the fourth ventricle to the spinal canal.

Resection of the cerebellar tonsils has been advocated as a way to improve the volume mismatch and to increase communication between the fourth ventricle and the spinal compartment.19,23,48 No neurological deficit has been demonstrated as a result of tonsillar resection; however, the exact function of this structure is largely unknown.

There has been a recent resurgence in the initial use of a syringosubarachnoid shunt in the treatment of patients with Chiari I malformation and syringomyelia.22,27,32,33 Although these reports suggest that this therapy leads to an increased rate of recovery and improved resolution of the syrinx, it must be noted that the underlying pathophysiology is not addressed. In the setting of symptoms not related to spinal dysfunction, this approach is not recommended.

**OUTCOME AND COMPLICATIONS**

The results of treatment are difficult to assess given the absence of uniform outcome measures as well as a randomized controlled trial in which the different treatments are compared. In most papers in which outcome is discussed the authors have reported an 80 to 90% rate of good outcome, with either resolution of the symptoms or cessation of progression. Presenting symptoms associated with a good outcome include headache, cervical pain, mild scoliosis, and sleep apnea;44 those associated with poor outcome include increased length of symptoms, muscle atrophy, ataxia, and nystagmus.16,40 The most common complications include postoperative hemorrhage, CSF leakage, aseptic or bacterial meningitis, and recurrence. Other reported complications include cervical instability and cerebellar subsidence.

**CONCLUSIONS**

In light of the many theories of pathophysiology, broad clinical presentations, and multitude of surgical interventions with variable outcomes, it is no wonder that a single
surgical approach does not exist. We recommend tailoring the surgical approach to treat the dominant clinical problem. Proper patient selection is critical to prevent unnecessary procedures and maximize outcome.

References


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