Outcomes for the Surgical Management of Chiari I and Chiari II Malformations

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Abstract

An up-to-date, evidence-based review of the neurosurgical literature reveals that clinical outcomes following the operative management of Chiari I and II malformations have improved dramatically since these congenital disorders were first recognized as surgical diseases. A detailed assessment of major measurable postoperative parameters, including improvement in clinical signs and symptoms, resolution of syringomyelia, and progression of scoliosis, proves these procedures to be safe and effective when performed in a timely manner by an experienced neurosurgeon. Patients with CM-I routinely report a significant reduction in headache, neck pain, apnea, and syrinx-related symptoms and encounter low rates of complication or reoperation after posterior fossa decompression using a bone-only or intradural approach. Neonates and infants with CM-II have higher rates of symptomatic improvement and reversal of impairment when an operative intervention is made at the first sign of brainstem dysfunction. The current trend of less invasive bone-only surgical approaches, if shown in larger prospective trials to be superior to traditional decompressions with dural opening, will only add to the modern-day neurosurgeon's ability to achieve excellent clinical outcomes with minimal risk in the treatment of patients with Chiari I and II malformations.

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The details regarding classification, embryology, epidemiology, pathology, presentation, evaluation, and management of these entities will be discussed elsewhere in this text. The objective of this chapter is to provide a literature-based description of the clinical outcomes observed and reported following the surgical treatment of Chiari I and II malformations (CM-I and CM-II). The major measurable parameters include the improvement of clinical signs and symptoms, resolution of syringomyelia, and progression of scoliosis. Endpoints such as duration of surgery, findings on

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intraoperative ultrasonography, and length of hospital stay, each of which has been evaluated in more recent investigations, will be discussed as well. Complications and rates of reoperation will be noted but have been addressed in another chapter. The differences in results documented in patients with CM-I or CM-II that have undergone either bony decompression alone or in conjunction with duraplasty will also be outlined. In light of the fact that Chiari I and II malformations are congenital disorders truly distinct from one another, they will be addressed separately here.

Chiari I Malformation (CM-I)

First described in 1891 by Hans Chiari, Chiari malformation type I (CM-I) refers to a caudal descent of the cerebellar tonsils through the foramen magnum – sometimes as low as the mid-cervical spine – that results in a variety of clinical signs and symptoms [1]. Multiple theories exist regarding the proposed pathogenesis of CM-I, the most routinely cited being that of an anomalous differential craniospinal pressure gradient across the foramen magnum; the lack of pressure equilibration between the intracranial and spinal subarachnoid spaces in this location permits the development of a caudal vector of force that results in worsening downward displacement of posterior fossa tissues [2–10].

The diagnosis of CM-I, typically made by magnetic resonance imaging (MRI) criteria, is being delivered with increasing frequency in both children and adults as the threshold for obtaining radiologic studies in the setting of minor clinical complaints has continued to decline. Unlike CM-I patients of a half-century ago that presented with severe ataxia, quadriplegia, and signs of elevated intracranial pressure, patients today are frequently diagnosed earlier with minor deficits, permitting more elective surgical management [11–13]. An excellent retrospective review of the 20-year institutional experience with pediatric CM-I at a major children's hospital conducted by Tubbs et al. revealed the two most common presentations to be headache/neck pain (40 % of patients) and scoliosis (18 %); they also found that only 20 % of patients referred with radiological CM-I actually had symptoms likely to be improved by surgical intervention [14]. Numerous authors have similarly determined headache (exertional, Valsalva-induced) and pain to be the dominant presenting complaints in adults [15–19]. In the large pediatric study population evaluated by Tubbs and colleagues, less than 10 % of patients presented with cranial neuropathies and fewer than 5 % had central sleep apnea. Other common findings included irritability, opisthotonus, upper extremity pain, paresthesias and weakness, ataxia, and lower extremity hyperreflexia. Among associated diagnoses, shunted hydrocephalus, retroversion of the dens, and scoliosis were observed most often [14].

Syringomyelia (SM), a condition caused by the abnormal accumulation of fluid within the spinal cord, is seen in up to 20 % of asymptomatic patients with CM-I and 75 % of those with symptoms [5, 20–26]. Although difficult to separate from the clinical findings in CM-I itself, these patients typically complain of suboccipital headaches and neck pain that may occur in conjunction with uni- or bilateral numbness, weakness or atrophy, and spasticity depending on the size and location of the syrinx. It is well-recognized from the Boman and Iivanainen study from the 1960s describing the natural history of untreated cervical SM that the condition will gradually progress and ultimately lead to both early disability and death if a timely intervention is not made [27].

Although no causal relationship has been definitively proven, the association between CM-I, SM, and scoliosis is well established and has been extensively studied [28–30]. It is believed by many that impairment of the lower motor neurons in the setting of syringomyelia results in aberrant innervation of the trunk musculature and creates an imbalance that directly contributes to the development of scoliotic deformity [31–33]. Several reviews have determined that not only is scoliosis often the earliest presenting sign of SM in children and teens but that it may be present to varying degrees in up to 85 % of young patients with SM [34, 35]. The likelihood of an individual case of idiopathic scoliosis (coronal spinal curve with Cobb angle > 11°) being associated with CM-I

and SM is increased in the setting of left-sided thoracic or otherwise atypical curves, hyperkyphosis, loss of thoracic apical segment lordosis, rapidly progressive curves, male gender, pain, and neurological deficits; the evaluation of scoliotic patients with such findings must therefore include spinal MRI [28, 30, 32, 36–46].

Operative Management of CM-I

With the exception of medical pain management, surgery is the only proven treatment available for CM-I. Although traditional approaches have included operations to address the syringomyelia itself via syrinx fenestration and shunting, the mainstay of therapy involves procedures directed at the presumed mechanism of syrinx development. Since some of the first descriptions in the literature of successful surgical management of CM by McConnell and D'Errico in 1938, studies by Fischer, Galarza et al., Krieger et al., and Navarro et al. have demonstrated the safety and efficacy of multiple techniques for decompression of the posterior fossa [47–52]. More recently, as the debate regarding the advantages and disadvantages of performing a "bone-only" extradural decompression via removal of the suboccipital bone with or without cervical laminectomy and lysis of fibrous epidural bands (PFD) versus the more invasive intradural maneuvers (PFDD) has grown stronger, Durham et al. and Hankinson et al. published meta-analyses of the current literature comparing the results of both approaches [53, 54]. Mutchnick and colleagues added to this growing body of class IIb and III data in 2010 with a single-institution retrospective review comparing 121 CM-I patients that underwent either PFD or PFDD [55]. As has been mentioned previously, however, there is no level I or IIa evidence comparing posterior fossa decompression without dural opening (PFD) to posterior fossa decompression with duraplasty (PFDD).

Clinical Outcome

In their meta-analysis composed of five retrospective cohort studies and two prospective cohort studies in which both surgical techniques were directly compared, Durham and Fjeld-Olenec found that 65 % of patients undergoing PFD experienced clinical improvement as compared to 79 % of the PFDD patients [53]. Hankinson and colleagues also reviewed the relatively limited database of studies retrospectively assessing the efficacy of PFD and PFDD separately. Some of these included the use of intraoperative ultrasonography to determine whether or not to perform intradural maneuvers, while others used electrophysiological evidence or preoperative factors to support the selection of PFD versus PFDD [52, 56–61]. Two retrospective studies from Italy reviewed by Hankinson et al. in which patients underwent PFD-only demonstrated complete symptom resolution in 81.3 % of patients [24] and a significant improvement in clinical condition at nearly 5 years of follow-up in 93.3 % of patients [62].

A wealth of class III evidence exists in the form of retrospective, single-institution studies analyzing the outcomes in patients with CM-I managed primarily with PFDD. Reports from the 1980s and 1990s by Paul, Nagib, and Nohria, respectively, showed that the majority of patients treated with PFDD experienced either improvement or stabilization of symptoms related to CM-I following surgery [25, 63, 64]. Outcomes were better when the onset of symptoms occurred less than 2 years prior to operative intervention [17]. Numerous small studies have shown a range of clinical improvement from 92 to 100 % with fairly low complication rates [51, 56, 57, 65–70]. In their extensive analysis of 500 pediatric patients treated for CM-I, Tubbs et al. demonstrated relief of preoperative symptoms or signs in 83 % of patients; headache (particularly Valsalva-induced and occipital in location), sleep apnea, and syringomyelia were affected more reliably than were preoperative motor or sensory abnormalities [14].

Syrinx Resolution

In the Durham and Fjeld-Olenec meta-analysis reviewing studies in which PFD and PFDD patients were directly compared, radiological

syrinx improvement rates were 56 % in the PFD patients and 87 % in those undergoing PFDD, although this finding did not reach statistical significance [53]. In the study by Genitori et al., eight of ten patients that presented with syringomyelia achieved complete syrinx resolution following PFD alone; Caldarelli and colleagues showed that 50 % had a decrease in syrinx size following bony decompression and 16.7 % experienced postoperative syrinx growth and persistent or worsening symptoms [24, 62].

Among studies looking at PFDD alone, investigators have reported rates of syringomyelia reduction ranging between 55 and 100 %, though no universal criteria defining improvement in syrinx exist [51, 56–58, 65, 67, 69–71]. Tubbs et al. found that of 285 patients with syringomyelia who underwent decompression with duraplasty, only 4 patients were found to have syrinx progression at follow-up 6 months to 1 year postoperatively; 80 % of patients had resolution of syringomyelia symptoms following the first operation and 95 % of patients achieved relief following a second operation [14]. Zhang and colleagues reviewed 200 cases and demonstrated collapse or diminished size of syrinx in 60 % of CM-I patients following PFDD [72]. Although case reports exist in the literature, the likelihood of delayed syrinx resolution is low, and reoperation is recommended for persistent symptomatic syringomyelia at the 3–6-month postoperative time point [73].

Scoliosis Improvement

There is a paucity of literature regarding the management of scoliotic CM-I patients with PFD alone. Genitori et al. documented radiologic improvement in two of three patients, and Caldarelli's paper reported mild improvement in two of two patients [24, 62]. Attenello et al. detailed a single patient who had progression of scoliosis requiring reoperation with duraplasty following an initial PFD [74]. The likelihood of improvement in CM-I patients with syringomyelia and scoliosis is better defined for the PFDD approach. A detailed search of the literature reveals at least 15 published clinical studies retrospectively evaluating scoliosis outcomes in patients treated primarily with PFDD. Though confounded by a lack of uniformity in surgical criteria and approaches across these series, rates of scoliosis improvement and progression range between 0–73 and 18–72 %, respectively [75–86]. An association has been made between better outcomes and both a younger age at intervention and a smaller presenting Cobb angle [75, 76, 82–84]. Isu and colleagues demonstrated that two-thirds of patients with CM-I-related syringomyelia and scoliosis might have both a postoperative reduction in the Cobb angle as well as lower rate of scoliosis progression when preoperative Cobb angles were less than 40° [32]. Nagib found that 6 of 10 patients with Cobb angles less than 30° improved and 4 patients with preoperative angles greater than 30° stabilized after PFDD [63]. Tubbs et al. observed that 18 % of patients in their large series had scoliosis, 82 % of whom had syringomyelia; 40 patients (8 % of all subjects) ultimately required spinal fusion for deformity correction. The authors observed that a preoperative Cobb angle of more than 40° was associated with higher rates of scoliosis progression even in the setting of decreased syrinx size following surgery [14]. Attenello et al. found that in addition to a larger preoperative Cobb angle, scoliosis located at the thoracolumbar junction and a lack of radiographic improvement in syrinx size following surgery were predictive of scoliosis progression [74]. Most recently, Krieger and colleagues published a 10-year retrospective review of 79 pediatric patients found to have CM-I and syringomyelia greater than 6 mm in diameter during an evaluation for scoliosis [86]. Each patient underwent PFDD, and none of the 49 patients with curves less than 20° had progression of their curves postoperatively; 70 % of the patients with curves between 25 and 80° required either bracing or spinal instrumentation and fusion for scoliosis after the Chiari decompression. In total, 87 % of the 79 patients had a significant size reduction of the syrinx following PFDD, but this, along with the magnitude of the preoperative curvature (in patients with Cobb angle > 20°), did not predict the need for subsequent deformity correction. Krieger and colleagues concluded appropriately

that timely intervention was the key to improving neurological signs and symptoms and to preventing the need for later spinal fusion surgery [86].

Reoperation Requirement and Complications

In the Durham and Fjeld-Olenec meta-analysis, patients who underwent duraplasty were less likely to require reoperation for persistent or recurrent symptoms (2.1 % vs. 12.6 %) but were more likely to sustain cerebrospinal fluid (CSF)-related complications (18.5 % vs. 1.8 %) [53]. McGirt et al. published a 3 % incidence of CSF leak in a 2009 retrospective review of 393 adult patients undergoing PFDD [87]. Mutchnick et al. found that 12.5 % of PFD patients needed a subsequent PFDD for symptomatic recurrence, though none suffered a complication; only 2 (3.1 %) patients receiving an upfront PFDD in their series underwent a repeated PFDD for lack of symptom improvement, and 3 patients suffered minor complications [55]. Tubbs and colleagues reported a complication rate of 2.4 % in 500 patients; these included posterior fossa extra-axial fluid collections causing acute hydrocephalus (managed with external ventricular drainage), severe brain stem compression within 48 h of surgery requiring transoral odontoidectomy and occipitocervical fusion, two aborted operations due to excessive occipital sinus bleeding, one case each of chemical and bacterial meningitis, and one patient with CSF leak secondary to untreated hydrocephalus that resolved with shunt placement [14]. Fifteen of 500 patients required reoperation (3.2 %). It is estimated that the annual expected mortality rate following CM-I decompression is between 2.5 and 4.5 % [88].

Operative Time and Length of Stay

In the retrospective review of their own institutional experience published in 2011, Tubbs et al. reported the mean operative duration to be 95 min for PFDD [14]. The average hospital stay for their patients (all but 1 of whom underwent PFDD) was 2–7 days with a mean of 3 days; the length of time away before returning to school ranged between 7 and 16 days, with a mean of 12 days. Mutchnick and colleagues found that those patients in their series undergoing PFDD spent a longer time in the operating room (201 ± 34 min vs. 127 ± 25 min) and in the hospital (4.0 vs. 2.7 days) than the patients who underwent PFD [55]. The 2005–2008 national normative data showed mean lengths of stay between 4.5 and 6 days [88].

Chiari II Malformation (CM-II)

Chiari malformation type II (CM-II) is a disorder of hindbrain development observed in the setting of myelomeningocele that was initially described by Hans Chiari in 1891 and is now known to include a variety of supra- and infratentorial anomalies [89]. In addition to caudal displacement of the cerebellar vermis, brain stem, and fourth ventricle, CM-II may include cerebellar inversion, a small posterior fossa, low-lying torcular Herophili, enlargement of the massa intermedia, shallow to absent cerebellar folia, a medullary "kink," and heterotopias [90–94]. The hypothesis currently favored by most neurosurgeons that best explains the myriad of findings in CM-II is the unified theory championed by McLone and Knepper; the combination of cranial constriction and settling, spinal cord tethering or traction, intracranial hypertension and intraspinal hypotension present in this malformation leads to the aforementioned spectrum of anatomical abnormalities [95].

Clinical signs of CM-II include apnea and respiratory stridor, neurogenic dysphagia, aspiration, hypotonia or spasticity, and para- or quadriparesis. Symptoms of the disease, which occur in one-third of patients with CM-II, range from very subtle to life threatening; symptomatic CM-II is the leading cause of death in children less than 2 years old with myelomeningocele, and surgical decompression is required in up to one-third of symptomatic patients with CM-II [96–100]. Although the malformation is present to variable degrees in every child born with a myelomeningocele and the diagnosis is straightforward, some patients become symptomatic only

later in adolescence with deficits or pain related to the more chronic effects of syringomyelia or scoliosis; these older children will manifest classic effects of cervical myelopathy with upper extremity weakness, spasticity, loss of dexterity, ataxia, and occipital headaches and are treated operatively in a more elective fashion [83, 101].

Operative Management of CM-II

The evaluation of symptomatic CM-II in a young child begins with a determination of the presence or absence of hydrocephalus, as many of these children require shunting at birth or shortly thereafter; in those patients with a shunt, the possibility of a malfunction must be addressed first. Because of the potentially fatal nature of the symptoms with which these young children present, whether secondary to cranial nerve traction, lower brain stem compression, or congenitally malformed cranial nerve nuclei, the workup and, if necessary, surgical decompression must be completed in an urgent manner [102–108].

Once hydrocephalus and/or shunt malfunction has been eliminated as the etiology of the CM-II patient's symptoms, the options for surgical intervention include suboccipital craniectomy, cervical laminectomy, and durotomy with or without dural augmentation [109–111]. As in CM-I operative management, controversy exists regarding the decision to perform a bony decompression only versus the more invasive durotomy and even fourth ventricular fenestration; each technique has been shown in separate investigations to be safe and effective for the treatment of CM-II, but the data remains class IIb or III [112–114]. The advantages of staying outside the intradural space include reduced risk of bleeding and decreased exposure to general anesthesia, while avoidance of a suboccipital craniectomy eliminates the chance of violating the low-lying torcular in these patients [115, 116].

Clinical Outcome

Overall, the prognosis for patients with symptomatic CM-II remains guarded, as up to 15 % of these patients die by 3 years old and an additional one-third suffer a permanent neurological disability [98]. Prior to the recognition of hindbrain compression as the cause of apnea, bradycardia, and cranial neuropathies and the establishment of an effective and aggressive surgical treatment, mortality rates for patients presenting with brain stem dysfunction that underwent "less urgent" surgical decompression ranged between 50 and 70 %; more recent studies in which surgery was undertaken early in an attempt to reverse the signs of brain stem compression reported postoperative mortality rates between 15 and 23 % [105, 108, 117, 118]. Conversely, outcomes in children and adolescents presenting with symptoms related to myelopathy or syringomyelia may mirror those of CM-I patients, with mortality rates near 0 % and clinical improvement in 79–100 % after surgery [105, 119].

As stated earlier, controversy exists regarding the optimal approach for craniovertebral decompression in these patients, in particular whether to include a suboccipital craniectomy and the utility of durotomy with dural augmentation. Tubbs and Oakes found in a 2004 evidence-based review of the literature regarding CM-II evaluation and management that all data were class III in nature and no reliable conclusions or recommendations could be made at that time [111].

With regard to the more invasive techniques, Pollack et al. published in 1992 on the use of a suboccipital craniectomy, cervical laminectomy, dural decompression, and, in patients with syringomyelia, a fourth ventricular shunt, in 25 CM-II patients with symptoms of increasing brain stem compression and deterioration [105]. The authors found that this approach resulted in near-complete or total reversal of clinical symptoms in 17 patients, while 3 others had mild-moderate residual deficits and 5 experienced no change. They established an association between worse preoperative neurological status, particularly bilateral vocal cord paralysis, and poorer outcomes, with an emphasis on the importance of expeditious treatment. Pollack and colleagues subsequently published a prospective report in 1996 in which children underwent the aforementioned decompression in a protocolized manner at the earliest

signs of CM-II-related brain stem dysfunction [104]. Ten of 13 patients returned to normal or near-normal brain stem function shortly after surgery, and only one required a temporary gastrostomy with no tracheostomies in the group. The remaining three patients presented with bilateral vocal cord paralysis and severe central apnea prior to operative intervention and achieved no meaningful recovery of function following surgery.

In 1992, Vandertop et al. retrospectively reviewed the management of 17 CM-II neonates over a decade with cervical laminectomy and duraplasty alone, finding that 88 % of patients achieved complete recovery with a mean follow-up of 65 months; one patient expired from respiratory arrest 8 months after surgery and the other died from a remote shunt infection 7 years later [108]. The authors argued that the relatively spacious size of the foramen magnum in CM-II patients eliminated the need for routine suboccipital craniectomy as part of the decompression.

With regard to the least-invasive end of the surgical spectrum, a 1996 investigation by Yundt and colleagues found that two children presenting with CM-II and stridor experienced clinical improvement following osseous decompression alone [116]. A later retrospective review by James et al. of 22 patients with CM including 18 children with CM-II that underwent a bony decompression only reported no surgical morbidities or mortality and partial or total symptomatic improvement in 86 % [115].

Most recently, Akbari, Limbrick, and colleagues conducted a retrospective analysis of 33 patients that underwent bony decompression with or without dural augmentation for the treatment of symptomatic CM-II and compared outcomes in patients managed with each approach [120]. Twenty-six patients had an osseous decompression alone, including 21 with cervical laminectomy and 5 others with both laminectomy and suboccipital craniectomy; seven patients underwent cervical laminectomy with or without suboccipital craniectomy and upfront duraplasty. At a median follow-up of 5 years, nearly 70 % of patients had symptomatic improvement, 62 % of those undergoing bone-only decompression compared to 57 % of the patients with dural augmentation (though this did not reach statistical significance). Signs including apnea, opisthotonus, stridor, and dysphagia were most responsive to surgical intervention, and the intraoperative blood loss, time under general anesthesia, and length of hospital stay were less in the bony decompression group, though statistical significance was not achieved. Rates of repeat surgery for lack of improvement or symptomatic recurrence were higher but not statistically significant in the bone-only cohort (19.2 % vs. 14.3 %); outcomes were not different between the patients that underwent cervical laminectomy alone compared to those that also had a suboccipital craniectomy. Overall, 6 of 33 patients required tracheostomies after surgery, and one patient died secondary to fungal sepsis unrelated to the Chiari decompression. The authors concluded that the less invasive approach of cervical laminectomy and sectioning of the dural band alone avoided the inherent risks of performing a suboccipital craniectomy and durotomy, including injury to the torcular herophili, CSF leak, pseudomeningocele, and meningitis, and should be considered a first-line option in the operative management of children with CM-II. Emphasis must also be placed on the critical need to evaluate each CM-II patient for active hydrocephalus or shunt malfunction, whether through radiographic imaging, shunt tap, or exploration, prior to undertaking a decompressive surgery. Undoubtedly, the need exists for a larger retrospective series or randomized controlled trial comparing the aforementioned approaches in order to make an outcomes-based decision regarding the optimal technique for CM-II treatment.

Fetal Myelomeningocele Repair and Improvement in Hindbrain Herniation

Finally, no discussion of CM-II outcomes would be complete without mention of the recently published prospective, randomized controlled trial of prenatal versus postnatal repair of myelomeningocele [121]. Though the primary findings of this

study included reduced need for shunting and improved motor outcomes at 30 months, the multi-institutional investigation also revealed that the proportion of infants without evidence of hindbrain herniation was higher (36 %) in the prenatal surgery cohort than in the postnatal surgery group (4 %) at 12 months of age. Similarly, the rate of moderate or severe herniation was lower (25 %), as were brain stem kinking, abnormal fourth ventricle location, and syringomyelia, in the prenatal surgery group than in the postnatal surgery patients (67 %). These data suggest that interruption of CSF flow through the myelomeningocele neural placode in utero, if performed early enough, may halt or even reverse abnormal hindbrain development. Although more work remains to be done, the impact of these findings on the future neurosurgical management of CM-II may be enormous.

Summary

An up-to-date, evidence-based review of the neurosurgical literature reveals that clinical outcomes following the operative management of Chiari I and II malformations have improved dramatically since these congenital disorders were first recognized as surgical diseases a century ago. A detailed assessment of major measurable postoperative parameters including the improvement of clinical signs and symptoms, resolution of syringomyelia, and progression of scoliosis proves these decompressive procedures to be safe and effective when performed in a timely manner by an experienced neurosurgeon. Patients with CM-I now routinely report a significant reduction in headache, neck pain, apnea, and syrinx-related symptoms and encounter low rates of complication or reoperation whether a bone-only or intradural posterior fossa decompression is performed. Neonates and infants with CM-II, though facing more significant deficits and frequently presenting in an emergent fashion, have higher rates of symptomatic improvement and reversal of impairment when an operative intervention is made at the first sign of brain stem dysfunction. The current trend of less invasive and faster bone-only surgical approaches, if shown in larger prospective trials to be truly superior to traditional intradural decompressions, will only add to the modern-day neurosurgeon's ability to achieve excellent clinical outcomes with minimal risk in the treatment of patients with Chiari I and II malformations.

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