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REVIEW PAPER

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Treatment and management of the Chiari II malformation: an evidence-based review of the literature

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Abstract *Objective:* Multiple surgical strategies exist for the management of the symptomatic Chiari II malformation. To date, no comprehensive analysis of this medical literature in an attempt to seek out standards or guidelines has been performed, thus serving as the impetus for this present review. *Methods:* A computerized search of the database of the National Library of Medicine was performed on the English-language medical literature between 1966 and 2003. Terms searched for included hindbrain hernia, Chiari II, Arnold-Chiari, surgery, decompression, syringomyelia, and CSF shunts. All literature found was reviewed with associated references from these sources. Each piece of literature reviewed was scrutinized and guidelines developed utilizing the methodology used by the Guidelines for the Management of Acute Cervical Spine and Spinal Cord Injuries. *Results:* All data reviewed were deemed to be Class III in nature. No standards or guidelines could be established from the available literature. There is significant debate and variable results in the current neurosurgical literature regarding the evaluation of shunt function vs. Chiari II decompression in patients with symptomatic hindbrain herniation. *Conclusions:* Multi-institutional prospective randomized trials are needed before any conclusions can be drawn regarding symptomatic Chiari II and its treatment paradigm. Until this time, clinical practice will be based on individual surgeons' experience, training, and personal beliefs.

Keywords Hindbrain herniation · Spina bifida · Treatment · Management

Introduction

The Chiari II malformation (CIIM) can present in a protean fashion. Today the problem lies not in evaluating the anatomical extent of the caudal herniation of the cerebellum, but in determining which patients should be considered for operative intervention, and the extent and choice of initial and subsequent surgeries. Should all symptomatic children be subjected to a decompressive operation, and what criteria should the surgeon use to verify shunt function? Chiari II patients are now frequently operated on at the first detectable symptom or evidence of a syrinx, and yet medullary dysfunction from the CIIM remains the leading cause of death in treated myelomeningoceles today [26, 27, 33]. Our knowledge of the natural history of the untreated conditions and the increased safety of surgery has made surgical intervention a much more viable option for this group of patients. However, to date, all evidence regarding the management and treatment of the symptomatic CIIM is Class III data.

We review the current medical literature regarding the treatment and management of the CIIM. Of note, much of the earlier literature regarding hindbrain hernia and its treatment collectively groups Chiari I and Chiari II malformations together as Arnold-Chiari malformations, which often clouds the results of such studies.

Materials and methods

A computerized search of the database of the National Library of Medicine was performed on the English medical literature between 1966 and 2003. Terms searched for included hindbrain hernia, Chiari II, Arnold-Chiari, surgery, decompression, syringomyelia, hydrosyringomyelia, myelomeningocele, and CSF shunts. All appropriate literature found was thoroughly reviewed including associated references from these sources. Each piece of literature reviewed was scrutinized and guidelines developed utilizing the methodology used by the Guidelines for the Management of Acute Cervical Spine and Spinal Cord Injuries (Neurosurgery 50:S1–S199, 2002) developed by the American Association of Neurological Surgeons/Congress of Neurological Surgeons Joint Section on Disorders of the Spine and Peripheral Nerves with the collaboration of the Joint Section on Trauma.

Scientific foundation

Among the theories of pathogenesis of the CIIM, the development of a differential craniospinal pressure gradient across the foramen magnum most adequately accounts for the clinical and anatomical findings observed [2, 40, 41]. Altered cerebral spinal fluid (CSF) circulation at the foramen magnum prevents instantaneous pressure equilibration between the intracranial and spinal subarachnoid space. There then develops an intermittent vector of force with each Valsalva, which can lead to the progressive downward movement of developing tissue through the foramen magnum. In utero, with the leakage of CSF through the open placode the intraspinal pressure may be maintained below intracranial pressure. This differential pressure occurs prior to the development of the cerebellar tonsils but after vermian development. This could help explain why the vermis and not the tonsils are caudally displaced in the CIIM. The data above serves as the basis for the therapeutic options for treating the CIIM. Additionally, some [22, 27] have found that the posterior cranial fossa is smaller in this cohort of patients, as is often the case in patients with Chiari I malformation [10]. This confinement of the structures of the posterior fossa may cause or exaggerate herniation of the cerebellum and brainstem. The small size of the posterior cranial fossa is one reason why some neurosurgeons advocate suboccipital craniectomy in addition to cervical laminectomy over the herniated brain tissue. This bony enlargement may further aid in the reestablishment of CSF flow at the craniocervical junction.

Symptomatic CIIMs most commonly affect infants. A third of myelomeningocele patients may develop brainstem dysfunction by the age of 5 years, and of those, as many as a third may die before infancy [11, 21]. Both brain stem dysgenesis and compression with resultant chronic ischemia are plausible causes. Holschneider et al. [16] have found a 20% mortality in children with cranial nerve deficits, including swallowing difficulties, while Hesz and Wolraich [11] reported a 60% mortality in those with central apnea [7, 8, 14].

Apneic spells have been noted to spontaneously resolve without treatment [42]. Cochrane et al. [8], in a small number of patients, have delineated between obstructive and central apnea and have found that obstructive apnea is usually reversed with an optimally functioning ventriculoperitoneal shunt, whereas central apnea does not respond to cervical decompression. These data are supported by the findings of Mori et al. [24], who found prompt improvement of brainstem conduction following ventricular shunting, although responses were still far from normal. Holliday et al. [15] have shown decreased brainstem conduction in a patient with a functioning shunt who, following cervical laminectomy, had a dramatic improvement in brainstem auditory evoked potentials. Abnormal brainstem auditory evoked potentials (BSAEVP) are associated with brainstem dysfunction but do not appear to be predictive of clinical outcome in this cohort [3]. Feeding and swallowing difficulties are reported in 59–71% of symptomatic Chiari II patients [9, 13, 30, 37, 38]. Progression of symptoms is usually over months rather than days compared with that in neonates [30, 31].

The formation and propagation of hydrosyringomyelia that is often seen in the patient with the CIIM may be explained by the same mechanism that interferes with the equilibration of CSF pressure between the intracranial and intraspinal compartment [27, 40, 41]. These cavitations are seen in 48–88% of patients with the CII [6, 9, 26].

Treatment and outcome

The natural history of the CIIM seems to be age dependent. In the retrospective series of Pollack et al. [32] of 25 symptomatic Chiari II patients, 13 neonatal patients presented with symptoms of brainstem compression before 3 months of age and 12 patients presented after this period. In the older group, none of the patients died or had a poor outcome. In the neonates, 23% died and 16% had a poor outcome; the remainder had a good outcome. The reason for the unfortunate outcome in neonates was thought to be because of intrinsic brainstem abnormality in this subset of patients with the CIIM. These data are somewhat confounded in that patients thought to also have significant hydrosyringomyelia had a conduit placed between the fourth ventricle or syrinx and the subarach-

noid space. Adequate shunt function was mentioned but not defined in all patients. In the prospective series of Pollack et al. [32], approximately 77% of symptomatic patients recovered normal or nearly normal brainstem function following decompression (occipital craniectomy and laminectomy). Presentation of bilateral vocal cord paralysis was found to have the worst prognosis. Again, adequate shunt function was not defined by these authors. The results of Pollack et al. [31, 32] are contrasted with the experience of Bell et al. [4], who described 17 infants with a symptomatic CIIM (primarily lower cranial nerve dysfunction) who underwent occipital craniectomy and cervical laminectomy. The success rate in these patients was only 30%. The conclusion from this study is that decompressive procedures for the symptomatic CIIM have little effect on the natural history of this clinical entity. However, these data are of some patients who underwent duraplasty and some who did not. Teo et al. [34] have reported that in primarily older children (mean = 11 years) complete resolution of symptoms were noted in 73% of patients following Chiari II decompression. This number was found 6 weeks after surgery whereas 80% of patients were asymptomatic after 1 year. Interestingly, 1 out of 3 of patients underwent reoperation. Vandertop et al. [36] have discussed their results in 17 neonates with lower cranial nerve dysfunction. All patients received only cervical laminectomies, and complete resolution was noted in all but two. These data are nonspecific in that the timing of surgery ranged from days to months following the beginning of symptoms. Hoffman et al. [12] have discussed similar findings but with no mention of the assessment of shunt function. Park et al. [28], in a large series, have concurred that early cervical laminectomy in symptomatic Chiari II patients is essential in deterring further neurologic compromise, although at last follow-up 38% of their patients had died. It is possible that in the third of myelodysplastics that become symptomatic before the age of 3, there is a greater degree of brainstem dysmorphism; consequently, the brainstem is more susceptible to injury from compression and/or ischemia.

Early surgical intervention may prove life sustaining in symptomatic Chiari II patients where symptoms are referable to the medullary dysfunction [21, 22]. Timing of decompression prior to bilateral vocal cord paralysis may predict a better outcome [10]. Pollack et al. [30] have described favorable outcomes for patients undergoing early surgical intervention, i.e., occipital craniectomy and cervical laminectomy for neurogenic dysphagia. Each of these Chiari II patients had adequate shunt function per head CT only.

Indications for surgical intervention are based on clinical symptoms. Narayan et al. [25] have found that neither the level of vermian herniation nor the level of cervicomedullary deformity predict which Chiari II patients will respond to decompression. In their series of 14, eight (57%) patients responded to posterior fossa decompression with improvement of preoperative symptoms. The presence of a significant hydrosyringomyelia or one that has progressively enlarged should be considered for therapy. Given the safety of current procedures in experienced hands, the criteria for operation can be expanded to include those patients whose symptoms are not immediately life threatening. These symptoms include: inspiratory stridor at rest or progressive by history, aspiration pneumonia due to palatal dysfunction or gastroesophageal reflux, central apnea with or without cyanosis, especially during sleep, opisthotonus, functionally significant or progressive spasticity of the upper extremities, and functionally significant or progressive truncal or limb ataxia [26, 27]. Yamada et al. [42] have discussed that posterior fossa decompression for stridor has not had a favorable outcome in their experience. Questionably, some patients received shunt revision first and others were decompressed first with no delineation between these groups.

The natural history and indications for surgery for Chiari II patients with hydrosyringomyelia are better understood today. If the patient is symptomatic, the chances for improvement are favorable following surgical intervention [12, 13, 28, 29]. Asymptomatic patients with extensive cavitation comprising 50% or more of the cross-sectional diameter of the spinal cord are likely to progress and should be considered as surgical candidates [26, 27].

Ventricular shunts

Before considering surgical decompression, symptomatic Chiari II patients must have physiologic intracranial pressure. This assurance requires a functioning shunt. It has been our recent experience and that of others that a properly functioning ventricular shunt can often obviate the need for decompression of hindbrain herniation. Caldarelli et al. [5] reported that of 11 symptomatic CIIM patients, 5 had resolution of their symptoms following shunt revision alone. Shunt patency studies, intracranial pressure monitoring, or ultimately shunt revision is appropriate if questions are raised regarding shunt function. In the presence of a shunt, progressive hydrosyringomyelia is shunt malfunction until proven otherwise. Milhorat et al. [23] have found in a retrospective study of a small number of patients (5 out of 5 patients with Chiari II and syringomyelia) that improvement in the size of their syrinx was observed following only ventriculoperitoneal shunting or revision. LaMarca et al. [19] have found that ventriculoperitoneal shunt revision alone resolves symptoms in the majority of Chiari II patients with hydrosyringomyelia. Of note, some patients in this study had mixed symptomatology (i.e., symptoms referable to brainstem compression or tethered cord syndrome) and were seem-

ingly arbitrarily offered posterior fossa decompression or tethered spinal cord release. Interestingly, Hoffman et al. [13] have discussed 16 out of 22 patients who had improvement in their hydrosyringomyelia following laminectomy over their vermian herniation without occipital bone removal. Each of these patients had plugging of their obex (i.e., iatrogenic obstruction at the level of the obex) a technique that has now been abandoned. Each of these patients were said to have functioning ventriculoperitoneal shunts. Some have found that lower cranial nerve findings are not improved following the confirmation of a functioning shunt, but only after posterior fossa decompression [37, 38]. Contrastingly, Charney et al. [7] found that lower cranial nerve dysfunction was shown to resolve following ventricular shunt revision in 50% of their patients. Shunt function was determined by head CT alone. Tomita and McLone [35], in a small number of patients, have concluded that shunt revision can reverse acute respiratory arrest. One important crux that is not addressed in the current literature is what defines an adequately working ventricular shunt? Is it radiological stable ventricles, normal shunt taps, operative visualization of flow from a ventricular catheter, or something else? Indeed, if merely imaging is relied upon, Iskandar et al. [17] found that in 20% of a study group of myelodysplastic children with shunted hydrocephalus that CT studies were prospectively interpreted as unchanged, stable, or no shunt malfunction, whereas with continued clinical decline, e.g., brainstem dysfunction, surgical observation was that of a malfunctioning shunt. Following shunt revision, all patients in the myelodysplasia group had resolution of preoperative symptoms.

Chiari II decompression

The surgical technique for Chiari malformations usually consists of bony decompression followed by dural expansion. The degree of intradural exploration will vary with the surgeon and the anatomy of the patient. In a CIIM with a large foramen magnum and a low lying torcula and transverse sinus, occipital craniectomy is not required, and may prove fatal if the sinus is entered inadvertently. However, many still perform occipital craniectomy in this group. Intraoperative ultrasonography may be useful to avoid mistaking the medullary kink for the cerebellar vermis. Variations from this technique are bony decompression without opening the arachnoid and the dura [18, 20]. Delayed cervical instability may occur and patients need postoperative cervical spine evaluation [1]. Symptomatic patients with hydrosyringomyelia need to establish normal intracranial pressure followed by a posterior fossa decompression the same as in those patients without hydrosyringomyelia. Posterior fossa decompression relieves symptoms of syringomyelia in more than 75% of the patients, and is preferred over syrinx shunting [23]. If a syrinx persists by MRI and the patient remains symptomatic, a syringosubarachnoid, syringopleural, or a syringoperitoneal shunt could be considered, as well as reasons why the posterior fossa decompression failed. Syringopleural shunting for patients with the CIIM was found to be preferable in the series of Vernet et al. [39], although the comparison syringosubarachnoid shunt was performed in only two patients. Table 1 summarizes the description and conclusions of larger series of treated and managed Chiari II patients. Each series consists of class III data.

Summary

Literature reviews are limited by the constraints of the literature on which they are based. In the end, a clinician must use his own experience coupled with what is widely accepted practice in peer-reviewed medical literature. Medical literature is at once imperfect with potential bias related to personal experience, training differences, and subjectivity.

Treatment of the symptomatic infant with clinical evidence of brainstem dysfunction is quite controversial and problematic. There is currently no consensus of what is the most appropriate initial therapy in symptomatic Chiari II patients and the data available are quite confounding. Shunt malfunction is known to mimic symptoms that are more characteristic of other pathologies seen in the myelodysplastic population, such as hindbrain herniation and tethered spinal cord, and may be the less invasive entity to address in the intervention for a symptomatic patient. Multi-institutional prospective randomized trials are needed before any conclusions can be drawn regarding symptomatic Chiari II and its treatment paradigm.

Key issues for future investigation

Future questions that should be addressed in a multi-institutional randomized manner for the patient with a symptomatic CIIM are:

1. What is the definition of a functioning ventricular shunt? e.g., CT parameters, shunt tap characteristics, and operative inspection
2. Is there utility in surgical decompression of the occiput, cervical spine, or both?
3. What treatment paradigm should be instituted in these children? e.g., verification of shunt function followed by decompression followed by syrinx shunting, etc.
4. In cases of hydrosyringomyelia, what is the ideal area to shunt to: pleura, subarachnoid, peritoneum?
5. How often should attempts be made at each step in a potential paradigm before moving to the next clinical step? i.e., how many attempts should be made at providing an ideally functioning shunt?

Table 1 Summary of major reports of the treatment and management of the Chiari II malformation (CIIM). *BAEPs* brainstem auditory evoked potentials, *PF* posterior fossa, *VP* ventriculoperitoneal

Reference	Description of study	Conclusions
[4]	Retrospective review of 22 symptomatic CIIM patients	Decompression may be effective in children but not always in infants
[5]	Retrospective review of 11 symptomatic CIIM patients	Improvement in 2 after shunt placement, 4 after decompression, 3 died after shunt revision, 2 died prior to any surgical intervention
[7]	Review of the management of 19 symptomatic CIIM patients	Half of symptomatic patients improved after shunt revision, 1out of 5 with decompressions improved
[8]	Review of 9 CIIM patients with principal symptom of apnea	None of the patients responded to VP shunt revision and/or cervical laminectomy
[12]	Review of symptomatic CIIM patients	Authors advocate immediate PF decompression, 100% resolution of symptoms in children treated immediately
[13]	Review of 30 CIIM patients with syrinx	State that PF decompression with obex plugging results in 70% improvement
[19]	Retrospective review of 231 MRIs of neurologically compromised CIIM patients	MRI indicated only in compromised patients
[23]	Retrospective review of 5 patients with CIIM and syrinx	Improvement in all patients following VP shunt revision only
[24]	BAEPs used to evaluate stem function in 16 CIIM patients pre- and postshunt placement	All patients had improvement in stem function following shunt procedures, although responses still far from normal
[25]	To correlate stem deformity to postoperative outcome	No correlation between stem deformity and either surgical outcome or presenting symptom
[28]	Review of 85 patients undergoing PF decompression for CIIM	Prompt decompression essential for successful outcome. Although roughly half had died by most recent follow-up
[31]	Retrospective univariate and multivariate review of 25 CIIM patients to assess the relationship between preoperative clinical factors and postoperative outcome	Early PF decompression with 17 patients improving postoperatively. Vocal paresis is a bad prognostic indicator for postoperative improvement
[32]	Prospective nonrandomized study in which 13 symptomatic CIIM patients underwent PF decompression at first signs of brainstem compression	Ten out of 13 patients recovered normal or nearly normal brainstem function following operation. Vocal cord paresis has a poor prognosis for recovery especially if bilateral
[30]	Review of 9 CIIM patients with neurogenic dysphagia	Rapid deterioration in all patients. Best results when PF decompression was performed promptly
[33]	Review of 17 symptomatic CIIM patients	Surgical intervention (shunt or PF decompression) improves symptoms in the majority of older children. Newborns and infants have a mortality rate of approximately 46%
[34]	Review of 30 CIIM patients who underwent PF decompression for symptoms referable to brainstem compression	Presenting symptoms resolved in 74% after 6 weeks and 80% after 1 year
[35]	Review of 3 CIIM patients who presented with neck pain, opisthotonos, and apnea	Prompt surgical revision of the VP shunt results in alleviation of symptoms in all patients
[36]	Review of 17 symptomatic CIIM patients	Prompt decompressive laminectomy promotes full neurologic recovery. Shunt malfunction must be ascertained prior to decompression as this results in recovery in the majority of patients
[39]	Review of syringopleural vs. syringosubarachnoid shunting for syrinx in CIIM patients	Syringopleural shunting is a valuable option for controlling syringomyelia
[38]	Review of experience with 14 CIIM patients	Recommend PF decompression and internal shunting for symptomatic patients
[42]	Review of 12 CIIM patients presenting with stridor and respiratory distress	VP shunt revision is a better treatment than PF decompression, which did not show a favorable effect

6. How many patients with CII simply have dysplastic brainstem nuclei where symptoms will never be addressed with clinical intervention?

These questions again can only be answered with appropriate data gleaned from prospective randomized multiinstitutional trials. Once data are available, recommendations can be made based on substance and not on single clinical experiences.

Recommendations

Standards: there is insufficient evidence to support treatment standards.

Guidelines: there is insufficient evidence to support treatment guidelines.

Options:

- Symptomatic CIIM patients who are shunted should have shunt function verified.
- Decompressive procedures at the craniocervical junction may lessen the symptoms of patients.
- Syrinx-induced symptoms may be stabilized or lessened by the shunting of the syrinx to an accepting cavity.

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