Outcome following hindbrain decompression of symptomatic Chiari malformations in children previously treated with myelomeningocele closure and shunts

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Between 1975 and 1989, 25 children treated with myelomeningocele closure and shunting for hydrocephalus at the Children's Hospital of Pittsburgh developed progressive lower brain-stem dysfunction from their Chiari malformation. Retrospective univariate and multivariate analyses of these cases were undertaken to assess the relationship between preoperative clinical factors and postoperative outcome. Since earlier reports have suggested that neonates with symptomatic Chiari malformations show a less favorable response than older children to craniocervical decompression, particular attention was directed at examining the effect of age on preoperative symptoms and postoperative outcome. Patients were subdivided by age into two groups, namely: 13 patients who became symptomatic before 2 months of age (neonatal group) and 12 older infants and children who developed initial symptoms between 6 months and 10 years of age. Once symptoms developed, patients in both groups deteriorated progressively until brain-stem decompression was performed. The mode of presentation and the rate and extent of neurological deterioration differed substantially in the two groups. Whereas the neonates typically showed rapid neurological deterioration and often manifested profound brain-stem dysfunction within a period of several days, the older patients experienced a more insidious symptom progression and rarely demonstrated the severe degree of impairment seen in the neonates.

All patients underwent suboccipital craniectomy, cervical laminectomy, and dural decompression. A shunt from the fourth ventricle and/or syrinx to the subarachnoid space was placed in those with significant syringomyelia. Following surgery, 17 patients had complete or nearly complete resolution of all signs of brain-stem compression, three had mild to moderate residual deficits, and five showed no improvement. Outcome correlated closely with the preoperative neurological status. In particular, the presence of bilateral vocal cord paralysis was associated with a poor response to surgery (p < 0.001 on both univariate and multivariate analyses). Of the six patients (all neonates) who progressed to complete bilateral vocal cord paralysis before surgery, only one improved. In contrast, all patients with less profound but nonetheless severe deficits recovered function postoperatively. Although the neonates as a group had a poorer outcome than did the older patients (p = 0.02 on univariate analysis), this in large part reflected their more severe preoperative impairments; neonates who still had some preservation of vocal cord function before surgery subsequently did as well as the older patients. Accordingly, age did not prove to be an independent prognostic factor on multivariate analysis. Taken together, these results indicate that, in most patients with symptomatic Chiari II malformations (including neonates), neurological deficits are potentially reversible if hindbrain decompression is performed expeditiously.

KEY WORDS • Chiari malformation • brain-stem compression • outcome • cervical decompression surgery • cervicomedullary junction • myelomeningocele

The development of brain-stem and lower cranial nerve dysfunction is a major source of morbidity and mortality in children with myelodysplasia and Chiari malformations. This syndrome often occurs in association with untreated hydrocephalus or shunt malfunction, and generally resolves after placement of a functioning cerebrospinal fluid (CSF) shunt. However, a significant percentage of children continue to deteriorate neurologically despite having adequate CSF diversion. While the efficacy of hindbrain decompression for older children and young adults with symptomatic Chiari malformations has been well estab-
lished, the merit of brain-stem decompression in infants is still a matter of some debate.\textsuperscript{2,5,14,15,20,22,25} In some reports,\textsuperscript{2,5,6} at least 50\% of symptomatic infants have died in early childhood despite having undergone decompression.

The basis for the disappointing postoperative results in these younger patients remains speculative. To address this issue, we reviewed our experience with the treatment of symptomatic Chiari malformations in both neonates and older children with myelodysplasia. Our specific goals were to examine the relationship between age and the pattern and severity of preoperative symptoms and to determine the contribution of these factors to postoperative outcome.

Clinical Material and Methods

Patient Population

Between 1975 and 1989, 287 children underwent myelomeningocele closure at the Children's Hospital of Pittsburgh. Twenty-five of these children (14 boys and 11 girls) developed symptoms and signs of brain-stem and lower cranial nerve dysfunction due to associated Chiari malformations despite having adequate CSF diversion. Each of these patients ultimately underwent craniocervical decompression because of persistent or progressive neurological deficits. Our findings in these 25 children are the focus of this study. Patients with symptoms and signs of brain-stem compression that resolved following placement of a functioning CSF shunt or ventriculostomy were not considered in this series.

None of the 25 children showed obvious signs of brain-stem compromise at birth, although three required a brief period of ventilatory assistance after delivery. The other 22 patients had Apgar scores of 7 or higher at 5 minutes after birth and required no perinatal ventilatory support. All 25 patients underwent uneventful closure of their myelomeningoceles during the first several days of life and were extubated postoperatively. Since a major goal of this study was to examine the effect of age on preoperative symptoms and postoperative outcome, the patients were divided into two groups according to age at onset of brain-stem symptoms: the first group consisted of 13 patients who became symptomatic during the neonatal period (birth to 3 months) and the second included 12 patients who did not develop their initial symptoms until after the neonatal period. A detailed review of the presenting symptoms and signs and mode of neurological deterioration was made for each of the patients in order to facilitate comparisons between the two groups.

Presenting Symptoms and Signs

The pattern of neurological impairment at presentation differed substantially in the two groups of patients (Table 1). For example, whereas 12 of the 13 neonates manifested stridor, only two of the 12 older infants and children had this symptom at the time of surgery. Apneic spells were also more common in neonates: in two such patients apneic spells were brief and responded to stimulation; however, in eight others they were protracted and associated with profound bradycardia, necessitating ventilatory assistance. In contrast, only two of the older patients developed apneic spells, and in both cases they were brief.

Neurogenic dysphagia was common in both groups of patients, but was almost invariably more severe in the neonates. While affected patients in both groups manifested similar symptoms as poor feeding or prolonged feeding time, pooling of oral secretions, and coughing and choking during feedings, the neonates generally had more serious problems from dysphagia. Ten of the 12 patients with severe symptoms, such as frank cyanosis with feedings, nasal regurgitation, weight loss or poor weight gain, and recurrent aspiration pneumonitis, were neonates. On the other hand, older patients were more likely than neonates to present with quadripareis, sensory loss, ataxia, or pain (Table 1). This may reflect the fact that these symptoms and signs were more readily appreciated in the older patients. Although profound quadripareis was detected in three of the neonates, more subtle motor or sensory findings might have escaped detection.

Mode of Neurological Deterioration

Regardless of the patient's age, once signs of brain-stem impairment developed, neurological deterioration was progressive until surgical decompression was performed. However, the rate and extent of neurological deterioration differed substantially, depending on the age of the patient. As a group, the neonates deteriorated more rapidly and demonstrated more severe neurological impairment by the time of surgery than the older patients. Moreover, the progression of neurological impairment frequently followed an almost stereotyped pattern in the neonates. The development of neurogenic dysphagia often heralded the onset of other signs of severe brain-stem compromise. Not uncommonly, the significance of the dysphagic symptoms was only appreciated after other brain-stem signs had developed, thus delaying neurosurgical referral. By the time of surgery, eight neonates who initially manifested only swallowing impairment had begun to show evidence of more diffuse medullary injury such as stridor, apneic spells, and quadripareis. Stridor usually appeared insidiously during inspiration only, but later became more severe and generalized to both phases of respiration, often to the point of causing profound airway obstruction. Several such patients went into acute respiratory failure and required emergency intubation and assisted ventilation.

Many of the neonates progressed within several days from having only mild feeding difficulties to showing profound dysphagia, severe stridor, apneic spells, and quadripareis. In contrast, neurological deterioration in the older patients generally progressed insidiously during a period of several months. In addition, the mode of neurological progression in the older patients rarely
Outcome after Chiari malformation decompression

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Age at Onset of Symptoms</th>
<th>Vocal Cord Paralysis</th>
<th>Vocal Cord Paralysis</th>
<th>Apnea</th>
<th>Neurogenic Dysphagia</th>
<th>Quadriparesis</th>
<th>Pain or Sensory Loss</th>
<th>Cranial Nerve Impairment</th>
<th>Oosphagocystotomy</th>
<th>Preop Duration of Symptoms</th>
<th>Inferior Extent of Cerebellar Tissue</th>
<th>Outcome</th>
<th>Follow-Up Period (mos)</th>
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<td>3 mos</td>
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<td>C-3 fair</td>
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<td>6 mos</td>
<td>C-4 good</td>
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<td>+</td>
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<td>7</td>
<td>-</td>
<td>1 mo</td>
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<td>72**</td>
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<td>+ (brief)</td>
<td>mild</td>
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<td>6, 7, 9, 10</td>
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<td>6 wks</td>
<td>C-1 good</td>
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</table>

* Symbols: + = feature present; - = feature absent.
† Orotracheal intubation or tracheostomy required preoperatively.
‡ Nasogastric or gastrostomy feedings required.
§ Fundoplication performed before brain-stem decompression in Cases 1, 5, and 13 and after decompression in Case 8.
†† Patients died of progressive respiratory failure (Cases 5 and 13) and cardiorespiratory arrest of unclear etiology (Case 4).
** Lost to follow-up study subsequently.

followed the consistent pattern demonstrated in the neonates. Several of these patients manifested only slowly progressive dysphagia symptoms, quadriparesis, and/or sensory loss for prolonged periods without showing evidence of vocal cord paresis or central hypoventilation. As a result, despite the frequently long interval between the onset of symptoms and the time of surgery, these older patients only rarely sustained the severe degree of impairment seen in the neonates.

**Diagnostic Evaluation**

The adequacy of shunt function was confirmed in each child before more aggressive intervention was considered. In preparation for decompression, angiography was performed in four patients, computerized tomography in seven, metrizamide cisternography in two, and, more recently, magnetic resonance imaging in 13 to determine the inferior extent of the cerebellar tissue.

Six patients with severe neurogenic dysphagia underwent a preoperative evaluation of swallowing function that consisted of the following studies: 1) barium cinesophagography to demonstrate gross impairment of swallowing such as cricopharyngeal achalasia, nasal regurgitation, and tracheal aspiration; 2) manometry studies to identify more subtle abnormalities in pharyngoesophageal motility and upper and lower esophageal sphincter function; 3) continuous esophageal pH monitoring to determine the frequency and severity of gastroesophageal reflux; and 4) dynamic and delayed scintigraphic scanning of the lungs fields following oral or nasogastric administration of ⁹⁹Tc sulfur colloid mixed with milk to quantitate the severity of gastroesophageal reflux and aspiration of gastric contents.

In four other patients who required urgent brain-stem decompression because of rapid neurological deterioration, evaluation of swallowing function was performed during the postoperative period.

Vocal cord function was evaluated by direct laryngoscopy in each of the patients who presented with stridor. Serial examinations were performed in several of the patients. In children with protracted apneic spells and/or stridor, serial measurements of arterial blood CO₂ and O₂, or, in recent years, continuous pulse oximetry and monitoring of end-tidal CO₂ were performed to evaluate the presence and severity of central hypventilation.

J. Neurosurg. / Volume 77 / December, 1992
Operative Intervention

The timing of operative intervention varied substantially among patients in this series. This in part reflects differences in the rapidity with which neurosurgical consultation was obtained by parents and physicians involved in the care of the patient, as well as differing viewpoints over the years among neurosurgeons at our institution regarding the appropriate criteria for proceeding with craniocervical decompression.

Surgery was performed with the patient prone and the neck flexed. Somatosensory evoked potential monitoring was used intraoperatively in all procedures conducted during the last 10 years. The decompression consisted of a suboccipital craniectomy and cervical laminectomy, which was carried down past the inferior extent of the ectopic cerebellar tissue. The dural opening was begun in the cervical canal and extended inferiorty past the caudally displaced vermilian peg. The opening was then extended rostrally toward the foramen magnum. A thick, constrictive transverse dural band was commonly encountered between the C-1 arch and the foramen magnum. Upon division of this band, cerebellar tissue often immediately bulged out of the dural sheath, attesting to the severe degree of compression in these patients. Extreme caution was used in dealing with the occipital venous sinus while extending the dural opening rostrally in a Y-shaped fashion. Here, the dura was carefully divided between sequentially applied hemoclips until the decompression was deemed adequate. Fifteen patients had thick arachnoidal adhesions over the cerebellar tonsils and vermis, but dissection of the arachnoid was limited to opening the foramen of Magendie, when possible, to re-establish CSF egress. In five patients with significant syringomyelia, a Silastic catheter was placed from the fourth ventricle to the upper cervical subarachnoid space anterior to the dentate ligament. Three of these patients also received a separate syringosubarachnoid shunt. No attempt was made to plug the obex. In five patients treated early in the series, the dura was left open. In all other patients, the dura was closed with a large graft of lyophilized human dura.

Outcome Analysis

Follow-up neurological examinations were performed 6 weeks to 3 months postoperatively and at 3- to 6-month intervals thereafter for at least 1 year. Further examinations were generally conducted annually, or more frequently if warranted by the patient's clinical status. In patients with preoperative neurogenic dysphagia, vocal cord dysfunction, and/or central hypoventilation, serial evaluations of swallowing function, vocal cord mobility, and ventilatory function, respectively, were performed at varying intervals after surgery, depending on the patient's clinical status.

Outcome was considered good if the patient had resolution of all or nearly all signs of brain-stem impairment, fair if there were mild to moderate residual symptoms and signs, and poor if the child showed no improvement postoperatively. The following factors were examined for their impact on postoperative outcome (good or fair vs. poor): 1) age at onset of symptoms (neonates vs. infants and older children; 2) sex; 3) preoperative symptoms and signs such as stridor, bilateral vocal cord paralysis, protracted apneic spells, severe dysphagia, quadriparesis, pain or sensory loss, and opisthotonus; 4) duration of symptoms before surgery (≤ 4 weeks vs. > 4 weeks); and 5) inferior extent of the cerebellar tissue (at or below C-3 vs. above C-3). Univariate comparisons between subgroups of patients were made using Fisher's exact test. Multivariate stepwise discriminative analysis incorporating each of the above variables was performed using the BMDP statistical software package.*

Results

Diagnostic Evaluation

On the basis of imaging studies and confirmatory operative observations, the lower limit of the cerebellar tissue was found to extend to C-1 in one child, C-2 in five, C-3 in 10, and C-4 in nine. Each of the 10 patients with severe dysphagic symptoms who underwent an evaluation of swallowing function preoperatively or in the immediate postoperative period showed evidence on barium cine-esophagography of tracheal aspiration, nasopharyngeal reflux, and pharyngoesophageal dysmotility including dysfunctional peristalsis and isolated spasms of uncoordinated esophageal contraction. Six patients had a discrete bar at the level of the cricopharyngeus muscle that caused a functional obstruction to the passage of contrast material into the esophagus. Manometry studies confirmed the presence of severe pharyngoesophageal incoordination in each of the patients. In addition, manometry also demonstrated cricopharyngeal achalasia in eight patients, with increased resting tone of the cricopharyngeus muscle and incomplete or poorly timed relaxation of this sphincter against peak contractions of the pharyngeal constrictor muscles during the early phases of swallowing. Two patients with manometric evidence of cricopharyngeal achalasia did not have a cricopharyngeal bar on cine-esophagography, making this a less sensitive test than manometry for identifying pharyngoesophageal incoordination. Continuous pH monitoring demonstrated severe gastroesophageal reflux in six of the patients. In four neonates with particularly severe reflux, the scintigraphic milk scan also confirmed substantial aspiration of gastric contents into the lungs.

Laryngoscopy demonstrated varying degrees of impaired vocal cord mobility in each of the 14 patients who presented with stridor. In seven patients who underwent serial laryngoscopic evaluations during the period when their stridor was worsening, vocal cord movement was found to deteriorate progressively, often from

* Software supplied by BMDP Statistical Software, Inc., Los Angeles, California.
Outcome after Chiari malformation decompression

mild paresis to unilateral or bilateral paralysis. By the
time of surgery, four of the patients manifested bilateral
cord paresis, four had unilateral paresis and contralateral paralysis, and six (all neonates) showed bilateral paralysis.

All eight patients who presented with protracted apneic spells and one other child who presented with severe stridor but without documented apnea manifested spontaneous oxygen desaturation and/or hypercarbia despite intubation.

Adjunctive Treatment

Patients with severe preoperative dysphagia, stridor, and/or central hypoventilation frequently required additional therapies directed at providing nutritional support and pulmonary care. Ten children required nasogastric or gastrostomy feedings. Gastroesophageal reflux and pulmonary aspiration of feedings were generally well controlled with metoclopramide administration and postural therapy. However, four patients with intractable gastroesophageal reflux and recurrent episodes of aspiration did not respond to these measures, but improved with gastric fundoplication. Eleven patients underwent tracheostomies during the perioperative period because of paralysis or severe paresis of the vocal cords, and one other patient required an extended period of orotracheal intubation. Nine of these patients also required assisted mechanical ventilation because of persistent central hypoventilation.

Outcome

The follow-up period ranged from 6 to 128 months (median 38 months). Seventeen of the 25 patients had complete or nearly complete resolution of all signs of cervicomedullary junction compression (good outcome), three had mild residual deficits (fair outcome), and five showed no improvement in neurological function (poor outcome) (Table 1).

Table 2 summarizes the impact of the preoperative factors examined in this study on postoperative outcome. Among the variables examined, the most important predictor of postoperative outcome was the presence of bilateral vocal cord paralysis (p = 0.00011 on univariate analysis, p < 0.001 on multivariate analysis). Of six patients (all neonates) with bilateral vocal cord paralysis, only one showed significant improvement in neurological function after surgery; this patient underwent urgent brain-stem decompression within hours after this finding was detected. The other five patients all suffered permanent respiratory insufficiency from central hypoventilation and vocal cord paralysis, and all required tracheostomies for long-term mechanical ventilation. These patients also had permanent gastrostomies for persistent dysphagia, and all but one required gastric fundoplication for intractable gastroesophageal reflux. Thus, with only one exception, the group with this severe degree of preoperative brain-stem compromise showed no recovery of brain-stem function postoperatively. Further, the only deaths in this series occurred in this group; three of these five neonates subsequently died from end-stage respiratory failure and pulmonary complications 10, 12, and 35 months after operation. Stridor, which was present not only in the six patients with bilateral vocal cord paralysis but also in eight others with less severe impairment of vocal cord mobility, was a less reliable predictor of outcome (p = 0.038 on univariate analysis, p > 0.1 on multivariate analysis). Other preoperative symptoms and signs of severe brain-stem compromise, such as protracted apneic spells, profound dysphagia, and opisthotonic posturing, were also individually less predictive of outcome (p = 0.022, 0.18, and 0.064, respectively, on univariate analysis; p ≥ 0.1 on multivariate analysis). As such, all patients with these deficits who still had some preservation of vocal cord function improved postoperatively. In some, recovery of function was noted immediately after surgery; in others, improvement occurred gradually over several months. Five of these patients who ultimately made good recoveries required temporary tracheostomies and/orenteral feedings for 1 to 8 months postoperatively. Follow-up cine-esophagography and manometry studies, performed in four of these patients before reinstitution of oral alimentation, showed resolution of cricopharyngeal achalasia and tracheal aspiration and reversion to normal or nearly normal esophageal motility. Follow-up laryngoscopy was performed in seven of the eight patients with preoperative vocal cord paresis or unilateral paralysis and likewise demonstrated recovery of vocal cord mobility in all seven.

An additional factor that on univariate analysis correlated with postoperative outcome was the age at the onset of symptoms. As a group, neonates had a poorer postoperative outcome than did older infants and chil-

<table>
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<th>TABLE 2</th>
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<tr>
<td><strong>Correlation of preoperative factors with postoperative outcome</strong></td>
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<tr>
<td>Variable*</td>
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<tr>
<td>age at onset of symptoms (≤ 3 mos (n = 13) vs. ≥ 6 mos (n = 12))</td>
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<tr>
<td>sex (14 M, 11 F)</td>
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<tr>
<td>stridor (n = 14)</td>
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<td>bilat vocal cord paralysis (n = 6)</td>
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<td>protracted apnea (n = 8)</td>
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<td>severe dysphagia (n = 12)</td>
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<td>quadriparesis (n = 11)</td>
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<td>pain or sensory loss (n = 8)</td>
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<td>opisthotonus (n = 10)</td>
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<tr>
<td>duration of symptoms before surgery (≤ 4 wks (n = 10) vs. &gt; 4 wks (n = 15))</td>
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<tr>
<td>inferior extent of cerebellar tissue (at or below C-3 (n = 19) vs. above C-3 (n = 6))</td>
</tr>
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*Abbreviation: n = number of cases.
† In this univariate analysis, only variables with a p value of ≤ 0.1 were retained. In all other analyses, the p value is > 0.1.
Discussion

Although the efficacy of hindbrain decompression in older children with symptomatic Chiari malformations has long been recognized, the value of decompression in neonates remains controversial because of the disappointing postoperative results reported by some authors. For example, Wickramasinghe, et al., reported only five survivors among 14 symptomatic infants undergoing decompression, although these five recovered completely after surgery. Similarly, Bell, et al., reported that seven of 14 infants who underwent decompression ultimately died of apnea or cardiorespiratory arrest, and only four of the seven survivors made complete or nearly complete recoveries. In a larger series, Park, et al., noted that only 28 of 45 young infants with symptomatic Chiari malformations who underwent decompression were alive at the conclusion of the study, and only 24 made a complete recovery. In agreement with the results of these studies, we found that preoperative neurological impairment improved or resolved in only eight of 13 neonates after craniocecal decompression, whereas all 12 older infants or children demonstrated significant recovery of function after surgery.

Compression vs. Dysplasia

The basis for the poor postoperative results in affected neonates remains speculative. It has been suggested by several authors that congenital brain-stem dysplasia may be responsible for the refractory medullary symptoms of the Chiari malformation in neonates. In support of this hypothesis, a postmortem study by Gilbert, et al., reported brain-stem maldevelopment including defective myelination and absence or hypoplasia of cranial nerve nuclei, basal pontine nuclei, olivary nuclei, or brain-stem tegmentum in 19 of 25 children with myelomeningoceles and Chiari malformations. Furthermore, Sieben, et al., and Holinger, et al., noted a paucity of normal neurons in the ambiguous, hypoglossal, and vagal nuclei and retroolivary region. However, a contrary argument holds that at least some of these histological lesions might have resulted from chronic brain-stem compression and ischemia, or traction on cranial nerve roots, and that these acquired lesions might be the basis for the symptoms and signs in affected patients. In this regard, Park, et al., noted that symptoms virtually always began postnatally and were generally progressive, thus suggesting an ongoing acquired process rather than a congenital one. They further noted that outcome was correlated with the severity of the preoperative neurological impairment and suggested that early decompression was essential for improving postoperative results.

Several observations from our study also favor the view that postnatal insults due to compression and ischemia of the brain stem rather than brain-stem dysplasia may be responsible for these patients’ neurological deficits. First, only three patients were symptomatic from their Chiari malformations during the first 2 weeks of life, and most did not develop symptoms until some months later. Second, the symptoms in our patients were invariably progressive and not fixed, as might be expected from a congenital process. Third, even among patients with severe preoperative deficits, significant recovery of function was achieved if brain-stem decompression was performed before the onset of bilateral vocal cord paralysis, whereas poor results were almost invariably obtained in patients who had developed bilateral vocal cord paralysis by the time of surgery. This pattern strongly suggests a progressive process with an outcome depending largely on the timing of operative intervention. Presumably, the abysmal results with delayed treatment of severely affected neonates reflects the endpoint of this progressive process, which may well be the expression of irreversible tissue damage acquired in the brain stem before treatment was given.

In this context, Papazomenos and Roessmann found medullary hemorrhage, hemorrhagic necrosis, and bland infarcts in 12 of 14 children who died with severe symptoms from their Chiari malformations, and they suggested that secondary vascular injury may have been the cause of the irremediable deficits. Cameron, Charney, et al., and Morley also noted medullary hemorrhages in patients with Chiari malformations who died after developing stridor and apnea.

The fact that many neonates showed rapid neurological deterioration despite having functioning CSF shunts suggests that these patients may have more “severe” malformations (that is, a relatively greater degree of cervicomедullary compression), a tenuous brain-stem vascular supply, and/or an intrinsic immaturity in their brain stems which makes them particularly vulnerable to compression. This vulnerability may in part account for the tendency of these patients to develop profound brain-stem compromise and, ultimately, irreversible brain-stem injury. Although a percentage of such patients may improve spontaneously without undergoing decompression, the majority of conservatively managed neonates show inexorable neurological deterioration with a high rate of mortality and morbidity. Late decompression of those patients who have already developed profound fixed brain-stem def-
Outcome after Chiari malformation decompression

ictis almost never results in meaningful recovery of neurological function.

Timing of Surgery

Although it is not possible to establish definitive guidelines regarding the timing of operative intervention based on this study, given the limited size of the patient population and the retrospective nature of the review, our results indicate that the development of neurogenic dysphagia and, subsequently, the onset of stridor and apneic spells are critical warning signs of impending but still potentially reversible brain-stem compromise. If surgery is performed before the onset of bilateral vocal cord paralysis, patients with these symptoms often improve substantially. In contrast, the development of bilateral vocal cord paralysis appears to correlate with the onset of an irreversible brain-stem injury.

The management goal at our institution in recent years has therefore been to identify and treat affected patients promptly, before the development of irreversible brain-stem injury. Early recognition of the symptoms produced by the Chiari malformation by individuals caring for the patient with myelodysplasia therefore assumes paramount importance. A number of patients in this series suffered progressive neurological deterioration for several days or, in some cases, for several weeks before neurosurgical consultation was obtained. Thus, it is essential for pediatricians, family practitioners, and family members to be well aware of the distinctive constellation of symptoms and signs manifested by patients with symptomatic Chiari malformations. As noted in this series, the presenting symptoms and signs in neonates differ substantially from those of older infants and children. Neonates frequently develop severe neurogenic dysphagia as an early symptom of their Chiari malformation. Stridor, which is rarely a presenting symptom in older patients, is also common in symptomatic neonates. Stridor, which is rarely a presenting symptom in older patients, is also common in symptomatic neonates. This should not be mistakenly attributed to group or to other upper respiratory illnesses until a thorough examination of vocal cord function can be obtained. With progressive brain-stem compromise, neonates frequently show an impaired ventilatory drive, manifested by hypoventilation, inadequate responses to hypoxia and hypercarbia, and, ultimately, apnea. These patients may manifest features of both obstructive and central apnea. Neonates also differ from older patients in their rate of symptomatic progression. Once initial signs of brain-stem compromise appear, further neurological deterioration generally occurs rapidly. In contrast to the mode of symptom progression typical for older patients, where neurological deterioration occurs insidiously over a period of months, neonates may develop severe dysphagia, stridor, and apnea within days or weeks after the onset of symptoms. We therefore encourage families and primary physicians caring for patients with myelodysplasia to contact us at the first indication of brain-stem compromise.

Adjunctive Measures

A second but no less critical aspect in the management of these patients involves identifying the need for adjunctive respiratory and nutritional care in those patients with hypventilation, impaired vocal cord mobility, recurrent aspiration, and severe neurogenic dysphagia. Patients with stridor and severe abductor laryngeal palsy may require prolonged orotracheal intubation or even tracheostomy to maintain an adequate airway. Because of the associated risk of central hypventilation in these patients, blood oxygen saturation and CO2 measurements should also be monitored closely during the perioperative period. The development of hypoxia or CO2 retention should signal the need for mechanical ventilation. Patients with severe dysphagia should undergo barium cine-esophagography and pharyngoesophageal manometry to determine whether nasogastric or gastrostomy feeding should be implemented to maintain adequate nutrition and to reduce the risk of aspiration pneumonitis. Esophagel pH monitoring and scintigraphic scanning of the lung fields following administration of radiolabeled tracer are helpful in identifying patients with gastroesophageal reflux and silent aspiration, who in turn may benefit from gastric fundoplication. During subsequent follow-up periods, serial evaluations of vocal cord mobility and ventilatory and swallowing functions are useful in deciding when and if adjunctive support can be safely withdrawn.

References


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