Surgical decompression for symptomatic Chiari II malformation in neonates with myelomeningocele

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Between January, 1981, and July, 1991, 17 infants under 1 month of age were admitted to The Hospital for Sick Children with the signs and symptoms of a Chiari II malformation. These patients' presentation included swallowing difficulty (71%), stridor (59%), apneic spells (29%), aspiration (12%), weakness of cry (18%), and arm weakness (53%). Decompression of the Chiari II malformation was performed in all patients, with a time interval between onset of symptoms and surgery ranging from 1 to 121 days. Fifteen patients (88%) remain alive, all of whom have shown a complete recovery. The mean follow-up period in this group of patients was 65 months. Two patients died, one due to respiratory arrest 8 months after decompression and the other because of shunt infection and peritonitis 7 years after decompression. These results support the concept that compressive forces, rather than a primary intrinsic disorder of the brain-stem nuclei, play a crucial etiological role in the development of a symptomatic Chiari II malformation. Early recognition of the symptoms of Chiari II malformation should be followed by immediate decompressive laminectomy in order to promote a prompt and full neurological recovery.

Key Words • Chiari malformation • myelomeningocele • decompression surgery • laminectomy

Virtually all children with a myelomeningocele have a Chiari II malformation. A significant percentage of these patients will develop symptoms and signs attributable to this malformation. The clinical picture in older children comes on gradually with upper-extremity weakness and long-tract signs; adequate decompression of the Chiari II malformation in these children consistently gives good results. However, during the neonatal period, the clinical picture is much more severe and is dominated by dysfunction of the lower cranial nerves, the caudal brain stem, and the cerebellum, which leads to apneic spells, vocal cord paralysis, and frequently death. Treatment of these neonates remains controversial largely because the operative results in this age group have been very poor. Often the symptoms of the Chiari II malformation are precipitated by increased intracranial pressure due to a malfunctioning shunt, in which case a simple shunt revision will usually result in prompt recovery. However, some children develop symptoms of lower cranial nerve and brain-stem dysfunction even with their hydrocephalus well controlled. The fact that these symptoms and signs are not present at birth but develop after a period of well-being strongly suggests that the patients' condition results from detrimental factors taking place after birth rather than from intrinsic disorganization or dysfunction of the brain stem itself. In the absence of shunt dysfunction, compressive forces have been found at surgery to be an important etiological factor; therefore, early surgical decompression should result in complete and rapid recovery. In this report, we review our experience with 17 neonates with closed myelomeningoceles, adequately controlled hydrocephalus, and symptomatic Chiari II malformations.

Clinical Material and Methods

Patient Population

Of 84 children admitted to The Hospital for Sick Children with myelomeningoceles and symptomatic Chiari II malformations between January, 1981, and July, 1991, 17 were aged 1 month or younger at the...
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TABLE 1
Clinical features of Chiari II malformation in 17 neonates

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Age (days)</th>
<th>Presenting Symptoms &amp; Signs</th>
<th>Time to Surgery (days)</th>
<th>Level of Pathology*</th>
<th>Tracheotomy†</th>
<th>Gastrostomy†</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>5</td>
<td>difficulty swallowing, weakness of cry, arm weakness</td>
<td>1</td>
<td>L-S</td>
<td>-</td>
<td>-</td>
</tr>
<tr>
<td>2</td>
<td>9</td>
<td>difficulty swallowing, stridor, arm weakness</td>
<td>2</td>
<td>L</td>
<td>-</td>
<td>-</td>
</tr>
<tr>
<td>3</td>
<td>14</td>
<td>difficulty swallowing, arm weakness</td>
<td>3</td>
<td>L-S</td>
<td>-</td>
<td>-</td>
</tr>
<tr>
<td>4</td>
<td>20</td>
<td>difficulty swallowing, apneic spell, arm weakness</td>
<td>4</td>
<td>L</td>
<td>+</td>
<td>+</td>
</tr>
<tr>
<td>5</td>
<td>13</td>
<td>difficulty swallowing, stridor</td>
<td>4</td>
<td>L</td>
<td>+</td>
<td>+</td>
</tr>
<tr>
<td>6</td>
<td>22</td>
<td>stridor, arm weakness</td>
<td>4</td>
<td>T-L</td>
<td>-</td>
<td>-</td>
</tr>
<tr>
<td>7</td>
<td>24</td>
<td>difficulty swallowing, stridor, apneic spell</td>
<td>5</td>
<td>L</td>
<td>-</td>
<td>-</td>
</tr>
<tr>
<td>8</td>
<td>28</td>
<td>difficulty swallowing, stridor, weakness of cry</td>
<td>5</td>
<td>T</td>
<td>+</td>
<td>-</td>
</tr>
<tr>
<td>9</td>
<td>24</td>
<td>difficulty swallowing, weakness of cry, arm weakness</td>
<td>8</td>
<td>T-L</td>
<td>-</td>
<td>-</td>
</tr>
<tr>
<td>10</td>
<td>5</td>
<td>difficulty swallowing, aspiration</td>
<td>9</td>
<td>T-L</td>
<td>-</td>
<td>-</td>
</tr>
<tr>
<td>11</td>
<td>24</td>
<td>difficulty swallowing, stridor, arm weakness</td>
<td>11</td>
<td>T</td>
<td>-</td>
<td>-</td>
</tr>
<tr>
<td>12</td>
<td>26</td>
<td>difficulty swallowing, stridor</td>
<td>13</td>
<td>L-S</td>
<td>-</td>
<td>+</td>
</tr>
<tr>
<td>13</td>
<td>4</td>
<td>stridor, aspiration</td>
<td>31</td>
<td>T-L</td>
<td>-</td>
<td>-</td>
</tr>
<tr>
<td>14</td>
<td>28</td>
<td>arm weakness</td>
<td>121</td>
<td>L</td>
<td>-</td>
<td>-</td>
</tr>
<tr>
<td>15</td>
<td>27</td>
<td>apneic spell, stridor</td>
<td>7</td>
<td>L</td>
<td>+</td>
<td>+</td>
</tr>
<tr>
<td>16</td>
<td>1</td>
<td>apnea, arm weakness</td>
<td>1</td>
<td>T-L</td>
<td>-</td>
<td>-</td>
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<tr>
<td>17</td>
<td>7</td>
<td>stridor, apneic spell, difficulty feeding, facial paralysis, tongue deviation</td>
<td>37</td>
<td>L-S</td>
<td>+</td>
<td>+</td>
</tr>
</tbody>
</table>

* Level of myelomeningocele: L = lumbar; S = sacral; T = thoracic.
† + = procedure performed; - = procedure not performed.

The time of presentation. Five of these patients were 7 days old or younger, three were 8 to 14 days old, and nine were 15 to 28 days old. The myelomeningocele was located in the thoracic region in two patients, the thoracolumbar region in five, the lumbar region in six, and the lumbosacral region in four (Table 1). The common presenting signs and symptoms in these infants were swallowing difficulty (71%), stridor (59%), apneic spells (29%), aspiration (12%), weakness of cry (18%), and arm weakness (53%). Most patients presented with a combination of these symptoms (Table 1).

All patients had adequately controlled hydrocephalus and were treated with decompressive laminectomy and duraplasty. The time interval between presentation and decompression varied from 1 to 121 days (mean 15.6 days). The follow-up period ranged from 8 months to 9 years (mean 65 months) (Table 2).

Surgical Technique

All operations were performed with the patients in a prone position with the neck flexed. The occipital bone in this group of patients was never removed because the foramen magnum was enlarged, and the compression of the brain stem was below that level. The upper cervical laminae were removed, and a tight fibrous band under the arch of C1 was found in 10 patients (59%) (Table 2). The dura was opened in the midline from below and the incision extended up to the level of the foramen magnum. The laminectomy with dural opening was then extended in a caudal direction until normal spinal cord could be identified. The lowest level of the cerebellar tongue or the medullary kink was located at C2 in one patient, at C-3 in one, at C-4 in four, at C-5 in nine, and at C-6 in two (Table 2). The dura was closed using fascia or cadaver freeze-dried dura.

Results

Fifteen patients (88%) survived, all showing a complete recovery. Two patients died, one due to respiratory arrest 8 months after decompression and the other because of shunt infection and peritonitis after almost 7 years.

None of the four infants who underwent decompression within 3 days after presentation required a trache-
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ostomy or gastrostomy for persistent lower cranial-nerve dysfunction, and all made a complete recovery within 5 days (Table 2). Of these four patients, arm weakness was noted in four, swallowing difficulty in three, stridor in one, prolonged apnea in one, and weakness of cry in one. At presentation of symptoms, these four patients were aged 1, 5, 9, and 14 days. The lowest level of the cerebellar tongue or medullary kink was at C-5 in two patients and C-6 in the other two. No tight fibrous band under the arch of C-1 was noted at operation in three of the four patients.

Among the 13 patients who were treated beyond 3 days after presentation, the time of recovery varied from 1 week to 2 years for the survivors; two patients died. One patient required a tracheostomy, another needed a gastrostomy, and four required both a tracheostomy and a gastrostomy (Table 1). Three of the five tracheostomies were later removed; one was removed 9 years after insertion by which time the vocal cords were moving normally. A fourth patient will have her tracheostomy removed as soon as both vocal cords are functioning normally again. The fifth patient with a tracheostomy died at the age of 8 months. All the gastrostomies were closed.

Discussion

Pathogenesis of Symptoms

Although the Chiari II malformation is present in almost all patients with a myelomeningocele, it is thought that only a small proportion of these patients becomes symptomatic. However, in a review of 262 patients with a myelomeningocele, Park, et al., found that 18% of the patients developed signs of the Chiari II malformation despite good control of hydrocephalus. During the 10-year period (1981-1991) of our study, 84 (21%) of 405 patients with a myelomeningocele presented with a symptomatic Chiari II malformation. These significant percentages may be caused not only by an increased awareness on the part of physicians who manage these patients, but also perhaps because of the increasing availability of magnetic resonance imaging, leading to an earlier and more accurate diagnosis. In older children and young adults, direct compression of the brain stem and cervical medulla often leads to syringomyelia, producing a different clinical picture of upper-extremity weakness and long-tract signs, which respond favorably to posterior fossa decompression and cervical laminectomy. In comparison to the older children, the combination of direct compression and vascular compromise leads to a much more devastating clinical picture in young infants.

The treatment of the infant with clinical symptoms of lower cranial-nerve dysfunction, brain-stem dysfunction, or both still remains controversial as the pathogenesis of the clinical symptomatology has yet to be fully resolved. Various theories have been postulated, including: traction or pressure on the lower cranial nerves caused by caudal displacement of the brainstem; vascular compromise to the brain stem caused by direct compression, leading to infarction, hemorrhage, and necrosis; or primary intrinsic disorganization or dysfunction of brain-stem nuclei. Furthermore, in infants with a Chiari II malformation, the cerebellar tongue appears quite healthy at operation, and there is little in the way of gliosis. However, beyond infancy the cerebellar tongue becomes markedly gliotic, and there is frequently thickening of the arachnoid, all bearing witness to the effects of long-standing compression.

Histological examination of postmortem material has shown that the protruded cerebellar tissue can be dysplastic and can show neuronal loss and gliosis. The choroid plexus may be atrophic and fibrotic, and the ependyma lining the distorted ventricle may be atrophic and discontinuous. Such gliotic tissue and fibrosis is caused by vascular compromise due to direct chronic or intermittent compression. Histologically, the cervical medullary oblongata usually does not show the gliosis and neuronal loss that is found in the protruding cerebellar tissue, although a detailed study with glial fibrillary acidic protein quantitative morphometry has not yet been reported. Distortion of the intramedullary nuclei and fiber tracts has been a common finding, but it is difficult to document the previously described atrophy of brain-stem nuclei because of the distorted anatomical relations in histological sections.

The fact that most patients develop symptoms that are not present at birth supports the concept that intrinsic abnormalities do not play a major role in the pathogenesis of symptomatic Chiari II malformation. Our results support the notion that even in the very young it is the compressive forces leading to secondary neuronal destruction that play a crucial role in the development of a symptomatic Chiari II malformation.

Treatment Recommendations

Prompt decompression led to a complete and early recovery. However, many of our patients were not brought in for treatment until many days, weeks, or even months had elapsed since the onset of their symptoms. Thus, they presented with paralyzed vocal cords which necessitated tracheostomy, and impaired swallowing mechanisms which required gastrostomy. Despite the poor condition of these infants following decompression, they made a full recovery although, for some, recovery took months and even years. Furthermore, our results show that it is not necessary to perform an occipital craniectomy or to extend the dural incision above the level of the foramen magnum. The unusually tight and dense fibrotic band, when present, occurs at the C-1 level. It is essential that the decompression be carried out as far caudally as is necessary until normal spinal cord is identified. In conclusion, we believe that early recognition of the symptomatic Chiari II malformation should be followed by immediate decompressive laminectomy in order to promote a prompt and full neurological recovery.
References


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