Preoperative evaluation and surgical management of the Arnold-Chiari II malformation

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The authors report their recent experience with 14 meningomyelocele patients with the Arnold-Chiari II malformation. Three major types of fourth ventricle anomalies seen in the Arnold-Chiari II malformation are defined, based on preoperative magnetic resonance imaging and intraoperative ultrasound studies. The Type A deformity is defined as no cystic dilatation of the fourth ventricle. In the Type B anomaly, there is intracranial dilatation of the fourth ventricle. The Type C deformity involves intraspinal dilatation of the fourth ventricle, either dorsal to the cord or within the substance of the cord. The Type A deformity was most common in infants, and in two cases progression from a Type A to Type B deformity was documented. Recognition of the type of Arnold-Chiari II malformation aids in designing an operative approach more specific to that structural abnormality. Intraoperative ultrasound is a valuable adjunct in localization of the underlying anomalies and permits safe decompression of the fourth ventricle.

The authors' indications for surgery now include failure to thrive due to either early respiratory and swallowing dysfunction, progressive spasticity, or upper-extremity weakness. Nine patients significantly improved following surgery and three patients with a progressively deteriorating course were stabilized by surgery. Decompression of the fourth ventricle by fenestration and internal shunting appears to be well tolerated, even in young infants, and is recommended in the treatment of the Arnold-Chiari II deformity.

KEY WORDS • Arnold-Chiari malformation • meningomyelocele • surgical decompression • magnetic resonance imaging • ultrasonography

Surgery for relief of symptoms associated with cervicomedullary compression in patients with the Arnold-Chiari II malformation has been performed infrequently in the child with myelodysplasia and has generally been limited to decompression of bone with or without dural grafting. This approach differs significantly from management of the less severe Arnold-Chiari I deformity seen most often in the adult. Although the Arnold-Chiari I anomaly is less severe, the surgical treatment is more radical and includes not only bone decompression but drainage of the fourth ventricle and of the associated hydromyelia. We have recently applied the surgical principles outlined by Rhoton for treatment of Arnold-Chiari I to children with Arnold-Chiari II malformation. Preoperative magnetic resonance imaging (MRI) using a 0.25-Tesla unit and spine coil, and intraoperative ultrasonography allowed us to tailor the operative approach to the specific anatomical abnormality and improve the safety of the procedure.

The variable nature of the cervicomedullary abnormalities demonstrated in this report has been documented in the pathological literature, but has not been stressed in discussion of the surgical management. We present our results in 14 children with the Arnold-Chiari II deformity and suggest a categorization we have found helpful in planning the surgical approach.

Summary of Cases

Clinical Material

Fourteen children with repaired meningomyelocele and shunted hydrocephalus were operated on for Arnold-Chiari II malformation between 1983 and 1985 (Table 1). Thirteen patients were 15 years old or younger. A 20-year-old patient (Case 14) with a high lumbar myelomeningocele had several years of visual blurring treated locally by a change in lenses. Neuroophthalmological examination demonstrated periodic alternating nystagmus consistent with a cervicomedullary junction lesion. Serial testing with a hand-held ergometer showed a progressive decline in upper-extremity strength.

Three major types of fourth ventricular malforma-
### TABLE 1

**Summary of 14 patients with Arnold-Chiari II malformation**

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Age at Surgery</th>
<th>Indications for Surgery</th>
<th>Postop Status</th>
<th>Type of Deformity*</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>1 mo</td>
<td>vocal cord paralysis</td>
<td>improved</td>
<td>C2</td>
</tr>
<tr>
<td>2</td>
<td>2 mos</td>
<td>vocal cord paralysis</td>
<td>improved</td>
<td>A</td>
</tr>
<tr>
<td>3</td>
<td>3 mos</td>
<td>vocal cord paralysis</td>
<td>improved</td>
<td>A</td>
</tr>
<tr>
<td>4</td>
<td>6 mos</td>
<td>retrocollis, poor suck, failure to thrive</td>
<td>improved</td>
<td>A</td>
</tr>
<tr>
<td>5</td>
<td>7 mos</td>
<td>retrocollis, poor suck</td>
<td>improved</td>
<td>B</td>
</tr>
<tr>
<td>6</td>
<td>1 yr</td>
<td>retrocollis, failure to thrive, psychomotor retardation</td>
<td>stable</td>
<td>A</td>
</tr>
<tr>
<td>7</td>
<td>2½ yrs</td>
<td>vocal cord paralysis</td>
<td>improved</td>
<td>C1</td>
</tr>
<tr>
<td>8</td>
<td>3 yrs</td>
<td>ataxia, obtundation</td>
<td>improved</td>
<td>B</td>
</tr>
<tr>
<td>9</td>
<td>3 yrs</td>
<td>vocal cord paralysis, poor swallowing, chewing ataxia</td>
<td>stable</td>
<td>C2</td>
</tr>
<tr>
<td>10</td>
<td>5 yrs</td>
<td>rt upper-extremity pain, progressive spasticity</td>
<td>died</td>
<td>C1</td>
</tr>
<tr>
<td>11</td>
<td>13 yrs</td>
<td>pain &amp; progressive right upper-extremity monoparesis</td>
<td>improved</td>
<td>B</td>
</tr>
<tr>
<td>12</td>
<td>15 yrs</td>
<td>posterior head &amp; neck pain, spasticity</td>
<td>improved</td>
<td>B</td>
</tr>
<tr>
<td>13</td>
<td>15 yrs</td>
<td>progressive spasticity</td>
<td>stable</td>
<td>A</td>
</tr>
<tr>
<td>14</td>
<td>20 yrs</td>
<td>visual blurring, upper-extremity weakness</td>
<td>improved</td>
<td>B</td>
</tr>
</tbody>
</table>

* For definition of deformity types see text.

Surgical Indications

Respiratory or swallowing dysfunction associated with poor feeding and failure to thrive was considered an indication for surgery in the infant. This represents a modification of our prior indications in which dysfunction had to be severe enough to cause respiratory distress or aspiration. In the older child, progressive spasticity and upper-extremity weakness were the most common indications for surgery. All children over 3 years of age are tested at each clinic session using a hand-held ergometer.* Surgery was designed to decompress the fourth ventricle and spinal cord, promoting relief of pressure symptoms resulting from either internal expansion or dorsal compression of the medulla and cervical spinal cord.

Surgical Principles

Preoperative x-ray films of the lateral cervical spine with the neck in flexion should be obtained to demonstrate the position of the odontoid. Even when minimal flexion is used, significant anterior decompression can occur (Fig. 5).

A standard laminectomy is performed taking great care to preserve the articular facets. We believe that the problems to be attacked lie at the midline and do not require exposure of the lateral gutters. We remove the midline spinous process and enough bone so as to just visualize the dorsal root entry zone. The laminectomy is carried down to the most inferior portion of the lesion demonstrated radiographically. Not uncommonly, the decompression must be extended an additional vertebral level to visualize the normal cord. In our series, the vermian peg has extended as low as T-1. The rim of the foramen magnum is removed to the level of the transverse sinus. The transverse sinus may

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* Vigorimeter manufactured by Martin, Inc., West Germany.
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be attached just above or at the level of the foramen magnum. The ultrasound apparatus is brought into the wound and the position of the fourth ventricle and central canal is identified. The value of ultrasound studies in determining the operative approach cannot be overstated.

A graft of cadaver dura lined with dural substitute is used routinely. The tense dura and absent pulsations noted during laminectomy are indicative of the high degree of spinal canal compromise in these patients. The absence of cortical bone density noted on MRI may give a false impression of the width of the spinal canal.

Type A: No Cystic Dilatation. The fibrovascular adhesions occluding the foramen of Magendie are lysed using microdissection. A radical tissue removal is not attempted. There is a risk for later encystment of the fourth ventricle, so in recent cases we have used ultrasound monitoring to guide entry into the fourth ventricle through a wide midline fenestration of the vermian peg. A catheter is directed cephalad through the fenestration and secured with a single 6-0 Prolene stitch to the pia-arachnoid. We have had no complications with this procedure.

Type B: Intracranial Fourth Ventricle Dilatation. Ultrasonography is used to identify the most su-

FIG. 2. Type B deformity visualized on sagittal magnetic resonance imaging. Left: Intracranial dilatation of the fourth ventricle and aqueduct is revealed, with no apparent communication between the third (small arrow) and fourth (large arrow) ventricles. Right: The dilated fourth ventricle (arrow) may extend into the spinal canal.

FIG. 3. Type C1 deformity. Left: Intraoperative ultrasonograph showing cystic expansion of the fourth ventricle (small arrow) arising from the caudal portion of the vermian peg (large arrow) lying dorsal (extra-axial) to the cord. The central canal is indicated by the arrowhead. Right: Intraoperative photograph showing a cyst (arrow) lying caudal to the vermian peg (p).
perforated extension of the cystic cavity in the midline. A wide fenestration is accomplished. We have thus far been unsuccessful in attempts to perforate the membrane separating the cystic fourth from the third ventricle. Postoperative computerized tomography (CT) has demonstrated complete obliteration of the cystic space. These children have not been followed long enough to know whether or not decompression of the cyst will remain without placement of a shunt into the peritoneal cavity. Remarkable symptomatic improvement is seen in these children, and serial clinical evaluation should provide early indication of reexpansion of the fourth ventricle.

Type C1: Fourth Ventricle Expansion. The cystic expansion of the fourth ventricle dorsal to the cervical spinal cord begins just distal to the most caudal extent of the vermis peg. If it can be readily mobilized, the cyst is removed. Otherwise it is widely opened to the floor of the medulla. The choroid plexus, which is usually entrapped within the cyst, is excised.

Type C2: Fourth Ventricle Dilatation Within Cord Substance and/or Hydromyelia. To treat dilatation of the fourth ventricle within the cord substance with or without hydromyelia, myelotomy guided by ultrasound is performed through the thinnest portion of the cord and is followed by placement of a stent from the cavity into the subarachnoid space. The stent is secured to the pia with a single 6-0 nylon suture.

Operative Results

Ten patients improved, three remained the same, and none were made worse by surgery. One patient died on the 5th postoperative day. Three patients required preoperative tracheostomy. Two infants (Cases 1 and 3) were seen early in the series, and decompression surgery was not performed until apnea intervened at the age of 1 month and 3 months. Both children are now decannulated and have normal vocal cord function. Two years postoperatively there is no indication of cervical spine instability. The third child (Case 7) had tracheostomy performed in infancy. She underwent bone decompression and resection of a Type C1 ventricular cyst at 2 1/2 years of age. She did well postoperatively and was successfully decannulated. Direct laryngoscopy 3 months postoperatively demonstrated full recovery of vocal cord function, but 1 1/2 years after surgery she had a series of “breath-holding spells” associated with temper tantrums. Otolaryngological evaluation was unremarkable, and a CT scan showed no ventriculomegaly. She died suddenly following such a breath-holding episode. This child was treated early in the series with only a bone decompression and dural grafting. With the exception of Case 13, all other children have had surgery according to the principles outlined above.

Three infants in whom surgery was indicated for failure to thrive, poor suck, and retrocollis improved rapidly and dramatically following decompression. One child (Case 5) with associated hydromyelia had return of lower-extremity function which had been lost in the first weeks of life. This child had been followed elsewhere until his admission for diagnostic evaluation, and it was not realized until the mother remembered after his surgery that he had been able to move his legs as a newborn.

In Case 8, cystic dilatation of the fourth ventricle was first noted on CT scanning for minor head trauma and ataxia. He precipitously deteriorated within 36 hours of injury. Postoperatively, his bilateral facial palsies and profound ataxia slowly resolved. His sixth nerve palsies required muscle resection procedures. He is
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the only child in whom a shunt from the cyst to the peritoneum was placed at the time of posterior fossa decompression, and his decompression persists satisfactorily 2 years after surgery.

Four older children (Cases 10 to 13) have been operated on for either progressive upper-extremity weakness and/or extremity spasticity, and only one of these has been followed for longer than 6 months. In that patient (Case 13), an adolescent, progression of spasticity appears to have been halted but no significant improvement has been noted. He had a decompression with plugging of the obex. One child (Case 11) had rapid recovery of upper-extremity function and relief of recent severe neck and shoulder pain. One child (Case 12) had unexpected improvement in a facial nerve palsy which had been present for 4 years. Neither of these cases has been followed for a sufficient length of time to predict maintenance of improvement. The only operative death in the series occurred in this group (Case 10) and was due to massive cerebral infarction on the 5th postoperative day associated with left middle cerebral and renal artery emboli of undetermined origin. Patients with marginal basilar impression must be positioned for surgery with care to avoid carotid artery injury, although in this particular case the occurrence of a concomitant renal embolus makes primary carotid injury an unlikely inciting event.

One child (Case 9) deserves special mention because of the severe degree of his Type C2 malformation. This youngster had suffered stridor and respiratory distress as an infant, which gradually improved. By 3 years of age he was still eating semi-solid foods because he was unable to chew, and his mother reported impaired swallowing. He had profound truncal ataxia and bilateral upper-extremity weakness. At surgery, the fourth ventricle was found at the C-6 vertebral level. Preoperatively this was mistakenly identified as hydromyelia on the MRI (Fig. 4). Intraoperative ultrasonography provided the correct interpretation by demonstrating a Type C2 abnormality. This child has made little improvement postoperatively, and this may reflect either a severe intrinsic abnormality, as indicated by the degree of cerebellar dysgenesis, or failure to operate early in the course. A youngster (Case 1) with a very similar anatomical abnormality operated on in the first months of life has done extremely well, with good upper-extremity function and no evidence of ataxia.

The Type A deformity is commonly seen in infants, whereas Type B and C malformations appeared more common in our series after 3 years of age (Table 1). In one child (Case 5), a second preoperative CT and MRI showed a cystic enlargement of the aqueduct and fourth ventricle not present on the first MRI scan, thus documenting a progression of the deformity from Type A to Type B. Deterioration in patients with the Type B deformity can be rapid, and we suggest that identification of a trapped fourth ventricle on CT is an indication for MRI if it is available.

Discussion

A number of necropsy studies delineate the anatomy of the posterior fossa in children with Arnold-Chiari II malformation.\textsuperscript{1,2,6,13,15,18} Several features are of particular interest to the surgeon\textsuperscript{2} and are particularly well demonstrated by MRI.

The posterior fossa is much smaller than it is in normal skulls of similar ages. The tentorium cerebelli is attached very low on the occipital bone so that it often appears to spring almost from the margin of the foramen magnum. The transverse sinus therefore is often encountered at or just above the rim of the foramen magnum (Fig. 4). Decompression of the posterior fossa is limited by the tentorium and, as noted by Park and coworkers,\textsuperscript{11} the cervical laminectomy is the major component of the bone decompression.

There is marked elongation and downward displacement of the brain stem. The medulla and fourth ventricle may be twice as long as in a normal baby and always extend into the spinal canal, irrespective of whether the cerebellar vermian “tail” is long or short. At the site normally occupied by the gracile and cuneate tubercles there are two partly fused protuberances which may extend for several centimeters dorsal to the spinal cord. This region is the site of “kinking” of the spinal cord. The long narrow fourth ventricle hollows out the upper end of these protuberances and the beginning of the central canal of the spinal cord can usually be found in the floor of this hollowed out pouch (Fig. 6). The tail of the cerebellum (nodulus pyramid) is bound to the medulla by fibrovascular tissue which also covers its dorsal aspect. The choroid plexus is usually found within this tissue (Fig. 7).

Peach\textsuperscript{12} described a cystic prolongation of the fourth ventricle in 25% of Arnold-Chiari II cases coming to

Fig. 6. Sagittal magnetic resonance imaging showing medullary kinking (arrow). The fourth ventricle is indicated by the arrowhead.
necropsy, and suggested that this cystic anomaly occurred in association with the kinking deformity of the medulla. The dorsal margin of the cyst wall originates from the caudal portion of the cerebellar component of the malformation, and the ventral wall arises from the caudal extremity of the medullary kink. The apex of the cyst is directed caudally so that the long axis of the cyst is in line with the spinal cord in a dorsal position. Peach reported that the cyst cavity communicates with the fourth ventricle. This cystic prolongation of the fourth ventricle corresponds to our Type C1 (Figs. 3 and 6). As stated by Peach, all of our Type C1 patients had medullary kinking.

The Type C2 deformity with prolongation of the fourth ventricle into the spinal cord dorsal to the central canal may in fact represent the syringomyelia-like anomaly associated with the Arnold-Chiari deformity and described by Lichtenstein, a local expansion within the cervical cord should prompt intraoperative ultrasonographic evaluation.

Naidich, et al., described the CT signs of the Chiari II malformation. Their description of the ventricles and cisterns added another dimension to the morphological data obtained at necropsy. They described a patient with cystic dilatation of the suprapineal recess with extension into the quadrigeminal plate. Their reference photograph is quite similar to the CT findings in our Case 8 (Fig. 8). In this instance, MRI demonstrated that the presumed dilatation of the suprapineal recess was in fact a dilatation of the aqueduct extending into the fourth ventricle (Fig. 2 left). The beaking deformity of the tectum (mesencephalic spur) is well illustrated in Fig. 9.

Perhaps the first report of posterior fossa exploration in infants with Arnold-Chiari II malformation was by D’Errico in 1939. He performed surgical exploration in eight infants following closure of meningomyelocele. The operation was limited to a suboccipital craniectomy and cervical laminectomy of C1–2 with intradural and (in a few cases) intraventricular exploration. There were three deaths, and two children in whom the hydrocephalus progressed. However, in three children relief of the obstruction and long-term good results were achieved. Little was done to attack the lesion directly until 1968 when Wickramasinghe, et al., published their results in 14 children treated because of severe respiratory dysfunction. Five children were “cured.”

Although control of the hydrocephalus usually relieved the lower cranial nerve palsies responsible for the bilateral abductor vocal cord paralysis, several other authors have reported that posterior fossa decompression was helpful when ventricular decompression by a functioning shunt was not successful in relieving the symptoms. Crosby, et al., first advocated direct surgical attack with dissection of displaced cerebellar tissue from the medullary surface working either from a caudal direction or, less frequently, opening the fourth ventricle and proceeding caudally with dissection. In their view excision of the abnormal cerebellar tissue was necessary to relieve obstruction and allow normal CSF circulation. Carmel cautioned against dissection in the region of the medullary floor, and most authors recommend simply a decompression of the cervical canal with placement of a dural graft and lysis.
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Fig. 9. Sagittal magnetic resonance imaging showing a Type B deformity. A mesencephalic spur (arrow) is well demonstrated.

of the thickened dural band present at the cranio-cervical junction.

We were impressed with the dichotomy between the aim of surgery in patients with Arnold-Chiari I deformity (that is, "decompress the cerebellar tonsils and permit egress of CSF from the fourth ventricle") and in patients with Arnold-Chiari II (that is, decompression by bone removal and dural grafting). It appeared to us that providing egress of CSF from a dilated fourth ventricle and spinal cord would be equally important in the child with the more severe type of abnormality in whom the malformed brain stem might have a low threshold for injury from pressure. We broadened our indications for surgery to include those commonly used in evaluating patients with Arnold-Chiari I malformation: namely, progressive spasticity and upper-extremity weakness as well as the more widely accepted respiratory and swallowing dysfunction. A recent paper by Park, et al.,10 documents the beneficial results in eight of 12 patients with progressive spasticity and upper-arm weakness in whom "posterior fossa decompression" was accompanied by plugging of the obex.

Magnetic resonance imaging has made us more cognizant of the variable nature of the fourth ventricle abnormality. The ventricle may be slit-like throughout its length, or it may present with caudal cystic dilatation or even with diffuse dilatation involving the more proximal aqueduct. It is readily apparent that this type of information is invaluable in planning a surgical approach to these lesions.

Type B malformation is the commonest anomaly seen in the infant with symptomatic Arnold-Chiari malformation. Progression to Type C1 or C2 has been documented in our series and can be inferred from reports in the literature of late occurrence of isolated or trapped fourth ventricles in these children. Deterioration in patients with a Type C abnormality can be rapid, and we suggest that identification of a trapped fourth ventricle on CT scanning is an indication for MRI if available. If MRI is not feasible, then metrizamide CT with coronal and sagittal reconstruction offers an alternative method. The noninvasive nature of MRI, its ability to clearly demonstrate structures in the sagittal plane, and its avoidance of the need for general anesthesia clearly make this the diagnostic procedure of choice in these lesions. It would seem that there is little decompression to be gained by opening the narrowly elongated fourth ventricle, whereas relief of the pressure symptoms associated with cystic expansion of the fourth ventricle is to be expected when the cyst is drained. However, opening into the narrow fourth ventricle with placement of a shunt into the subarachnoid space may prevent later cystic expansion.

Although most authors have advised a conservative approach to surgical intervention in patients with Arnold-Chiari II, Park, et al.,10 suggests that a more aggressive approach is indicated. We would agree and urge the use of preoperative MRI and intraoperative ultrasonography to allow the operative approach to be tailored to the specific pathological anatomy. While dissection of the vermian peg from the floor of the medulla remains prohibitively risky, decompression of the involved structures can be accomplished safely and may provide long-lasting relief.

Addendum

Since this article was submitted for publication, a patient has been referred to us who was operated on for decompression of a Arnold-Chiari malformation. The dramatic cystic dilatation of the ventricle occurring postoperatively was accompanied by increased swallowing difficulty and the onset of apneic episodes. Failure to improve or gradual deterioration following surgery for Arnold-Chiari malformation should prompt early reexamination to rule out encystment of the fourth ventricle. Placement of a shunt tube from the fourth ventricle to the subarachnoid space accompanied by wide fenestration of the vermian peg may reduce the likelihood of this complication.

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References


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