Isolated ventricles following intraventricular hemorrhage

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Panventricular enlargement often follows intraventricular hemorrhage in the premature neonate. During a recent 12-month period, the authors identified five infants who required shunting because of symptomatic post-hemorrhagic hydrocephalus, which was progressive despite serial lumbar punctures and the use of indwelling subcutaneous ventricular reservoirs. In the first 6 months following placement of the initial shunt, four of the infants required an additional shunt for isolated ventricles. Two children had isolation of the lateral ventricles and required bilateral shunts. Two other children had isolation of the fourth ventricle from the lateral ventricular system and required posterior fossa shunts. All of the children were treated successfully using multiple shunts. In all cases, progressive dilation of the isolated ventricle was unaccompanied by the usual clinical signs of shunt malfunction. It appears that isolated ventricular systems are common following post-hemorrhagic hydrocephalus and these children must be followed closely with ultrasound and computerized tomography scanning.

KEY WORDS: hydrocephalus □ intraventricular hemorrhage □ neonate □ isolated ventricle

In the past decade major advances in neonatal intensive care have allowed the survival of a large number of low birth-weight premature infants. Intraventricular hemorrhage (IVH) arising from the area of the germinal matrix is frequently associated with prematurity.1,13 Panventricular enlargement will occur in about half of those children with IVH.14 This may be a transient phenomenon, not requiring treatment, or there may be progressive signs and symptoms of hydrocephalus. The incidence of post-hemorrhagic hydrocephalus that eventually requires definitive shunting varies from study to study.13,14 The underlying pathophysiology and contributing factors in the development of IVH have been reviewed.15

Treatment is undertaken in those children who exhibit progressive ventricular enlargement accompanied by apnea, bradycardia, lethargy, vomiting, and increasing spasticity in the lower extremities. These signs are usually associated with a bulging fontanel and a rapidly increasing head circumference. The initial management of these patients includes serial lumbar punctures and the administration of drugs such as glycerol and Diamox (acetazolamide). Those infants who continue to suffer progressive ventricular enlargement are further managed with a ventricular reservoir.12,13

In a recent 12-month period, we performed ventriculoperitoneal (VP) shunting in five infants whose hydrocephalus was refractory to more conservative measures. During a relatively brief follow-up period, four of these infants went on to develop isolated ventricles which required the placement of separate additional shunts.

Previous reports of isolated ventricles have identified a heterogeneous group of patients with diverse etiologies.3,7,9,16 In most of the cases, this finding was noted at least several years after the initial shunting procedure. This present clinical report details our experience with isolated, or loculated, ventricles in the preterm infant with IVH.

Summary of Cases

Over a continuous 12-month period in 1982 and 1983, five premature infants in the neonatal intensive care unit at Evanston Hospital required VP shunting for post-hemorrhagic hydrocephalus. The four infants who subsequently required multiple shunts for treatment of isolated ventricular systems form the basis of this report.

The diagnosis of IVH was made clinically and was confirmed by ultrasound or computerized tomography (CT) scanning. The hemorrhages were graded using the criteria established by Papile, et al.15 Serial lumbar

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TABLE 1
Summary of clinical data in cases of isolated ventricles

<table>
<thead>
<tr>
<th>Clinical Data</th>
<th>Case 1</th>
<th>Case 2</th>
<th>Case 3</th>
<th>Case 4</th>
</tr>
</thead>
<tbody>
<tr>
<td>estimated gestational age (wks)</td>
<td>31</td>
<td>27</td>
<td>32</td>
<td>30</td>
</tr>
<tr>
<td>weight (gm)</td>
<td>1850</td>
<td>920</td>
<td>1920</td>
<td>1640</td>
</tr>
<tr>
<td>Apgar score (1 min/5 min)</td>
<td>8/9</td>
<td>6/8</td>
<td>4/8</td>
<td>2/7</td>
</tr>
<tr>
<td>hemorrhage grade*</td>
<td>III</td>
<td>IV</td>
<td>IV</td>
<td>III-IV</td>
</tr>
<tr>
<td>age at 1st lumbar puncture (days)</td>
<td>14</td>
<td>15</td>
<td>15</td>
<td>24</td>
</tr>
<tr>
<td>reservoir inserted (day)</td>
<td>19</td>
<td>48</td>
<td>35</td>
<td>37</td>
</tr>
<tr>
<td>ventriculoperitoneal shunt placed</td>
<td>31</td>
<td>92</td>
<td>51</td>
<td>54</td>
</tr>
<tr>
<td>diagnosis of isolated ventricle</td>
<td>270</td>
<td>141</td>
<td>135</td>
<td>121</td>
</tr>
</tbody>
</table>

* Degree of hemorrhage was graded according to the criteria of Papile, et al.15

Punctures were initiated when signs or symptoms of post-hemorrhagic hydrocephalus became apparent, and ventricular size was monitored with repeated ultrasound examinations. All of the infants were further managed with ventricular reservoirs when lumbar punctures failed to maintain adequate decompression. All of the infants underwent insertion of a low-pressure (opening pressure 2 to 5 cm H2O) distal slit-valve VP shunt before discharge from the hospital. The estimated gestational age, Apgar score, grade of hemorrhage, and the various intervals between date of birth and treatment were all recorded (Table 1).

Case Reports

Case 1

This 1850-gm infant was born after a 31-week gestation to a mother with a history of preterm labor. Apgar scores were 8 and 9 at 1 and 5 minutes, respectively. The child developed a severe respiratory distress syndrome; on Day 5 of life, a Grade III IVH was diagnosed by ultrasound scanning. On Day 14, a repeat ultrasound study showed marked enlargement of both lateral ventricles, and serial lumbar punctures were begun. Over the next 5 days the child's head circumference increased by 1.75 cm and he developed apnea, bradycardia, and vomiting, associated with a bulging fontanel. Daily lumbar punctures produced only 1 to 2 cc of cerebrospinal fluid (CSF), and the symptoms did not improve. On Day 19, a ventricular reservoir was inserted. Initially, 20 cc of CSF was removed every 8 hours to control symptoms of apnea, bradycardia, and lethargy. After several days the frequency of taps could be decreased to once daily. Removal of ventricular fluid was halted on Day 29 to assess the child's ability to absorb fluid. Rapid head growth ensued over the next 48 hours and a VP shunt was placed. His subsequent course was complicated by a shunt infection with Staphylococcus epidermidis. This was identified promptly and treated successfully with an extraventricular drain and antibiotics. The VP shunt was reinserted without difficulty 10 days later. The child was discharged from the hospital in excellent condition and with normal ventricular size documented by CT scanning and ultrasound.

His development proceeded well and his head circumference remained in the 50th percentile. When he was seen at 9 months, his head circumference continued to be at the 50th percentile, but negligible development had occurred over the preceding 3 months. He was now noted to be hypotonic and did not follow objects with his eyes. His mother confirmed that, in retrospect, he started to lose interest in his visual environment at about 6 months of age. He was noted to be vaguely irritable and had occasional vomiting, especially at night. An ultrasound study showed panventricular dilation, including the fourth ventricle (Fig. 1 left). The

FIG. 1. Case 1, at age 9 months. Left: Ultrasound scan showing panventricular enlargement and a nonfunctioning shunt in the right frontal horn. Right: Computerized tomography scan after shunt revision showing the hugely dilated fourth ventricle and left temporal horn.
VP shunt was revised, and the immediate follow-up CT scan showed collapse of the ventricles except for the left temporal horn and the fourth ventricle, both of which remained markedly dilated (Fig. 1 right). Separate low-pressure shunt systems were placed in these isolated ventricles, resulting in good decompression (Fig. 2). The child's apathy and visual inattention gradually improved. He began to smile and reach for objects over a period of several weeks. Another subsequent episode of fourth ventricular shunt malfunction was again heralded by apathy, vomiting, and visual inattention, all of which improved after revision.

Case 2

This 920-gm infant was born after a 27-week gestation period. Apgar scores were 6 and 8 at 1 and 5 minutes, respectively. He had severe respiratory distress which required mechanical ventilation. A sudden drop in hematocrit and loss of spontaneous movements heralded the onset of bilateral Grade IV hemorrhages, which were confirmed by ultrasound scanning on Day 6. Serial lumbar punctures were initiated on Day 15. Because of the limited volume of CSF that could be obtained at each puncture, a ventricular reservoir was inserted on Day 48. All attempts to wean the patient from daily ventricular taps were met with rapid head growth, a bulging fontanel, and clinical symptoms of increased pressure. A CT scan confirmed severe panventricular hydrocephalus (Fig. 3 left). A VP shunt was placed in the right frontal horn on Day 92, and ultrasound examination confirmed good decompression of the entire ventricular system. The child was seen in the clinic on Day 135, where his head circumference was noted to be at the 2nd percentile and he exhibited no signs of increased intracranial pressure. Ultrasound and CT scans revealed remarkable unilateral hydrocephalus.
with bulging across the midline and enlargement of the ipsilateral hemicranium (Fig. 3 center). A second shunt was placed in the enlarged left lateral ventricle and this resulted in prompt decompression (Fig. 3 right). The child has been followed with CT scanning at 3-month intervals and has had no further problems.

Case 3

This 1920-gm infant was the second of twins delivered by Caesarean section. He developed a severe respiratory distress syndrome and a pneumothorax while being mechanically ventilated. A Grade IV hemorrhage with extension into the right cerebrum was noted on ultrasound scanning on Day 7. Serial lumbar punctures were effective temporarily, but apneic spells, bradycardia, and vomiting ensued as the volume of CSF obtained decreased. Serial ultrasound studies demonstrated progressive enlargement of the ventricular system. A reservoir was placed on Day 35. The child continued to exhibit rapid head enlargement and a bulging fontanel each time ventricular taps were discontinued. A VP shunt was placed on Day 51. The fourth ventricle was noted to be slightly enlarged on repeat ultrasound examination but appeared stable in size since the previous examination. A routine follow-up CT scan performed on Day 135 showed collapsed lateral ventricles (Fig. 4 left). However, marked enlargement of an apparently isolated fourth ventricle had occurred in the interim (Fig. 4 center). The child had no symptoms directly attributable to this finding. A separate shunt was placed in the fourth ventricle via a midline posterior fossa approach. The fourth ventricle subsequently collapsed (Fig. 4 right).

Case 4

This 1640-gm infant was delivered by emergency Caesarean section at 30-weeks gestational age because of abruptio placentae. A bilateral Grade III IVH was diagnosed by ultrasound at 48 hours. Extension to bilateral Grade IV was noted by the next day. Progressive ventricular dilation ensued by Day 24, necessitating serial lumbar punctures. Because only 5 to 7 cc of CSF could be obtained at each puncture, a ventricular reservoir was inserted on Day 37. Serial ultrasound studies documented symmetrical enlargement of the lateral ventricles. A definitive VP shunting procedure was performed on Day 54. Prior to discharge from the hospital, a follow-up ultrasound examination revealed good reduction in the size of both ventricles. The child’s head circumference continued to parallel the 50th percentile, and he had no further symptoms of increased intracranial pressure. A routine CT scan performed 2 months after discharge revealed marked unilateral hydrocephalus (Fig. 5 left). The fourth ventricle appeared normal. Because of our previous experience with Case 2, a separate VP shunt was inserted into the enlarged left lateral ventricle, with subsequent drainage of the entire system (Fig. 5 right).

Discussion

Ventricular enlargement is common following IVH in the premature neonate and may be related to the degree of hemorrhage. The pathogenesis of post-hemorrhagic hydrocephalus is probably complex; it is not possible to explain all of the observations on the basis of a single mechanism.

Larroche found evidence of adhesive arachnoiditis in the posterior fossa and occasional obliteration of the aqueduct by necrotic debris. A more recent study with ultrasound implicates large free-floating clots, which may obstruct the foramen of Monro. Hill, et al., proposed that small mobile particulate matter suspended in the ventricular fluid may cause obstruction.
Isolated ventricles after IVH

Fig. 5. Case 4. Computerized tomography scans obtained 2 months after placement of a ventriculoperitoneal shunt in the right lateral ventricle. Left: Before placement of the second shunt, there is marked enlargement of the opposite side. Right: After placement of a second shunt on the left side, collapse of the entire ventricular system could be seen.

to the outflow of CSF at the level of the arachnoid villi. This would be consistent with the observed efficacy of serial lumbar punctures in the clinical course of these infants.

Isolation of the fourth ventricle has been reported following IVH, neonatal meningitis, multiple shunt revisions, and myelomeningocele with Arnold-Chiari malformation. In the largest reported series of isolated fourth ventricles, only four of the 16 cases followed an IVH at birth. The development of an isolated fourth ventricle may follow a sequence that starts with obliterative arachnoiditis in the posterior fossa, followed by obstruction of the aqueduct, possibly from clots and cellular debris. It is interesting that in the case reported by Hill, et al., aqueudctal occlusion occurred 20 days after the initial onset of communicating hydrocephalus.

Loculation of the lateral ventricular system with resultant unilateral hydrocephalus has, to our knowledge, not previously been reported as a complication of neonatal IVH. In a very interesting paper, Fawer and Levene documented large free-floating clots in the lateral ventricles. They reported transient unilateral ventricular dilation in their Case 2 and proposed that the observed ventricular asymmetry was due to the position of the infant’s head. They hypothesized that a freely communicating ventricular system allows hydrostatic pressure to distend the lowest ventricle. This ventricular asymmetry was observed for 1 week only and was a transient phenomenon. There were no clinical signs of hydrocephalus.

It is apparent that large pieces of clot or other debris might directly cause obstruction at the foramen of Monro. However, the unilateral hydrocephalus in our cases developed well after one would expect the dissolution of such clots. Ependymal flaps or chronic inflammatory changes, with the formation of membranes, could also account for the observed isolated ventricle. It is of some concern that all four of our patients had prior temporary indwelling ventricular reservoirs. There is no clear evidence linking the use of these reservoirs to a higher risk of isolated ventricles, although the observed location of the catheter tip at or in the foramen of Monro is potentially contributory.

The type of shunt system may play a role in the development of isolated ventricles. We used a distal slit-valve catheter with an opening pressure of 2 to 5 cm H2O in all of our cases. The low opening pressure was chosen because of the low pressures observed in some of these infants, despite progressively enlarging ventricles. Anti-siphon devices were not used. It is possible that over-drainage of CSF might result in unilateral ventricular collapse or in obstruction of the aqueduct. Replacement of a low-pressure shunt with a valve of higher opening pressure, or the inclusion of an anti-siphon device, may prevent some cases of isolated ventricles, but we have not tested this hypothesis.

Gram-negative bacterial meningitis has been reported as a frequent cause of lateral ventricle entrapment. In our Case 1, the child experienced a brief Staphylococcus epidermidis ventriculitis which was cured promptly with external ventricular drainage and penicillin. We are unaware of this organism leading to ventricular septations and loculations.

In three of our cases (Cases 1, 2, and 4), the isolated ventricle was detected as an “incidental” finding. This may be due in part to the buffering effect of the other shunted ventricles. Subtle signs of shunt malfunction may be difficult to diagnose in these infants, many of whom are already severely neurologically compromised by their hemorrhages. We believe that it is especially important to identify and treat the infants with isolated ventricles because of the relentlessly progressive course; the visual and motor disturbances arising from these large ventricles under pressure may not be reversible if corrected late in the clinical course.

The CT scans in two of our infants (Cases 2 and 4) with trapped lateral ventricles showed definite effects of pressure; the septum was bowed to the contralateral side and the skull contour was noted to bulge on the side of the enlarged ventricle. Again, internal decompression via the other shunted ventricles is probably responsible for the observed lack of rapid head growth and the flat or sunken anterior fontanel. It may be that the dilated and trapped fourth ventricle is more likely to cause symptoms because of distortion of the brain stem.

The rapidly diminished size of the anterior fontanel after initial VP shunting in all cases made follow-up with ultrasound scanning a difficult task. Although we strongly advocate early follow-up examination with ultrasound, we also believe that it is essential to obtain CT scans at 3- to 6-month intervals following placement of the first shunt, even if the patient has a normal head size, a sunken fontanel, or no clinical signs.
The generally disappointing neurological outcome in these patients confirms the experience of others in treating patients with Grade III or Grade IV hemorrhages. We are hopeful that close follow-up evaluation and aggressive treatment of isolated ventricular systems will help preserve the potential for modest development in at least some of these infants.

References
