Perimesencephalic cistern obliteration: a CT sign of life-threatening shunt failure

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Death from cerebrospinal fluid shunt malfunction is a rare but tragic event. The authors describe seven children who when admitted were lethargic but arousable because of shunt failure. Sudden deterioration prompted unscheduled emergency shunt revision in each case. Two children subsequently died. Although neither the history nor the physical findings predicted this life-threatening shunt malfunction, obliteration of the perimesencephalic cistern was apparent on all preoperative computerized tomography (CT) scans. For reference, the scans of 43 other hydrocephalic children were drawn randomly from the hospital files and analyzed. It is concluded that CT evidence of obliteration of the perimesencephalic cistern is a useful warning of life-threatening shunt failure.

KEY WORDS • perimesencephalic cistern • ventriculoperitoneal shunt • shunt failure

Children seldom die of shunt malfunction. This tragedy occurred in two children at our institution, prompting us to review our experience with patients who deteriorated precipitously while awaiting scheduled shunt revision or while being observed for diagnostic confirmation of shunt malfunction. Although there are many diagnostic tests that help confirm the clinical impression of shunt malfunction, there is no single test that predicts the need for urgent treatment. In this paper we present seven cases of life-threatening shunt malfunction which were associated with obliteration of the perimesencephalic cistern seen on preoperative computerized tomography (CT) scans. The incidence of acute shunt malfunction and the relative importance of cistern obliteration in patients with hydrocephalus is discussed.

Summary of Cases

A summary of the seven cases in this series is presented in Table 1. On admission, all patients were lethargic but arousable, and in each case a precipitous decline in neurological status prompted unscheduled emergency shunt revision. The spectrum of diagnoses is representative of most pediatric neurosurgical practices, and the patients' ages ranged from 1 to 17 years. Headache was the most common presenting symptom and was lacking only in the 1-year-old child and one of the 3-year-old children. The duration of symptoms averaged 4.7 days, with a range of 24 hours to 8 days. No patient manifested papilledema, and only two patients had new strabismus. Although none of the seven patients displayed decerebrate posturing on admission, five were posturing just prior to surgery. In all but one case palpation indicated that the shunt was functioning well. In that case, a proximal malfunction was predicted and a distal malfunction was found at surgery. Obliteration of the perimesencephalic cistern was demonstrated on CT in all seven cases (Fig. 1).

We reviewed 137 scans obtained in 43 hydrocephalic patients drawn randomly from our neuroradiology files. No cisterns were visible on the scans of four patients, and in each case cistern obliteration was associated with ventriculomegaly. All four patients subsequently underwent shunt revision, and the cisterns reappeared. In none of the cases was shunt revision performed as an emergency procedure. In all the other scans, the cisterns were readily visible, even though there were many cases where the ventricles were small.

To determine the frequency of fatal as contrasted to life-threatening shunt failure, one of us (D.C.M.) surveyed his experience with 407 patients followed over a 15-year period. In this group there were four deaths associated with shunt malfunction (Table 2).
Perimesencephalic cistern obliteration in shunt failure

### TABLE 1
Clinical data in seven cases of life-threatening shunt malfunction

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Diagnosis</th>
<th>Age (yrs)</th>
<th>Time (hrs): Admission to Op</th>
<th>First Symptom/Sign</th>
<th>Prior Revisions</th>
<th>Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>postop meningitis</td>
<td>3</td>
<td>8-12</td>
<td>lethargy</td>
<td>2</td>
<td>died</td>
</tr>
<tr>
<td>2*</td>
<td>aqueductal stenosis</td>
<td>8</td>
<td>12-24</td>
<td>headache</td>
<td>0</td>
<td>died</td>
</tr>
<tr>
<td>3</td>
<td>spina bifida</td>
<td>1</td>
<td>12-24</td>
<td>vomiting</td>
<td>1</td>
<td>survived</td>
</tr>
<tr>
<td>4</td>
<td>spina bifida</td>
<td>3</td>
<td>8-12</td>
<td>headache</td>
<td>1</td>
<td>survived</td>
</tr>
<tr>
<td>5</td>
<td>spina bifida</td>
<td>17</td>
<td>8-12</td>
<td>headache</td>
<td>1</td>
<td>survived</td>
</tr>
<tr>
<td>6</td>
<td>aqueductal stenosis</td>
<td>8</td>
<td>12-24</td>
<td>headache</td>
<td>12</td>
<td>survived</td>
</tr>
<tr>
<td>7</td>
<td>intraventricular hemorrhage</td>
<td>4</td>
<td>4-6</td>
<td>headache</td>
<td>0</td>
<td>survived</td>
</tr>
</tbody>
</table>

* This case is the same as Case 1 in Table 2.

**Fig. 1.** Computerized tomography scans in Case 7. **Upper:** The perimesencephalic cistern is not visible at the time of acute shunt failure. **Lower:** Three days later, after shunt revision, the cistern is clearly visible.

**Discussion**

The incidence of sudden death due to shunt malfunction in a small series of 407 patients is less than 1%. Most patients with shunt malfunction require urgent but not emergency treatment. In actual practice a shunt revision is often worked into the operative schedule the day following admission. However, every experienced neurosurgeon is aware of patients who suddenly deteriorate on the ward while awaiting surgery or diagnosis confirmation of shunt malfunction. Our seven patients are a select group in that the shunt malfunction was truly life-threatening. Three of the patients suffered cardiac arrest and two died.

The patients' history and physical examination offered few clues to separate this select group from other patients with nonurgent shunt malfunction. All patients with noncompliant skulls who could verbalize complaints had headache, and vomiting was associated with headache in four of the seven patients. No patient had papilledema or anisocoria. New sixth nerve palsies were apparent in only two of the patients on admission. Six of the shunt systems were tested by compression of the valve and thought to be functioning, but the plethora of "objective" methods of assessing shunt function is evidence of the poor correlation between shunt function and the response of the valve or flushing device to digital compression. Osaka, et al., found good correlation between the response of the flushing device and the site of the obstruction in a study of 50 revisions for obstruction. However, in 40% of patients who were clinically symptomatic and who subsequently underwent revision, the devices flushed normally. On the other hand, 50% of asymptomatic patients had devices that either compressed with difficulty or refilled slowly. Other groups have questioned the reliability of "pumping" in assessing shunt function. The present study agrees with our own experience that neither shunt malfunction nor the site of obstruction can be predicted by palpation of the valve.

Since obliteration of the perimesencephalic cisterns on CT was the only finding common to all these cases, several questions deserve discussion. Is obliteration of the cisterns the radiological correlate of cerebral herniation, or does it warn of impending cerebral herniation and thereby assume much greater clinical importance? In all but one of the seven cases (Case 1), CT scanning was performed on admission when the patient was lethargic but arousable and manifested no clinical signs of uncal or central transtentorial herniation. Of the 43 randomly selected hydrocephalic patients, the four cases with obliteration of the basal cisterns requiring nonurgent shunt revision also had signs of shunt malfunction, but no patient had signs associated with cerebral herniation.
Cisternal obliteration that occurs in children with lumboperitoneal shunts is a nonspecific finding, but in the context of severe head injuries or cerebral anoxia it has more specific implications. The association between intracranial hypertension and absence of the basal cisterns is well documented.\(^5,13,14,21\) \(\text{Van Dongen, et al.}^{20}\) were able to show a relationship between cistern obliteration and outcome after head injury. \(\text{Toutant, et al.}^{19}\) confirmed their observation in a consecutive series of 218 patients in the National Traumatic Coma Data Bank. Absent cisterns on CT scans were consistently associated with a poorer prognosis. Of the patients with a Glasgow Coma Scale (GCS) score of 6 to 8 and absent cisterns, 75% had a poor outcome (defined as dead, vegetative state, or severe disability). If cisterns were present, the risk of a poor outcome was only 20%. Of those with a GCS score of 3 to 5 and no cisterns, 92% had a poor outcome, whereas if cisterns were present, 75% had a poor outcome.

To address the question of extreme shunt dependency, we reviewed all previous revisions in our group of seven patients. There were 17 revisions (12 in one patient); none was performed precipitously and none was associated with obliteration of the basal cisterns. We cannot address the appearance of the cisterns in children who are symptomatic from small ventricles because we see so few patients with this syndrome.

In the group of 407 patients followed more than 15 years, three of the four deaths occurred after discharge. The fourth child died in the hospital while awaiting confirmation of shunt malfunction and is included in our series of seven hospitalized patients who experienced life-threatening shunt malfunction (Case 2 in Table 1). In this group of children, severe shunt malfunction could not be predicted from the history and the physical findings on admission, even in retrospect. Obliteration of the basal cisterns on CT scans proved to be the single most important indicator. We recommend that the patient who presents with shunt failure and obliteration of the basal cisterns on CT undergo immediate shunt revision.

### Table 2

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Diagnosis</th>
<th>Age  (yrs)</th>
<th>Duration of Symptoms</th>
<th>First Sign/ Symptom</th>
<th>Prior Revisions</th>
<th>Cause of Malfunction</th>
<th>Circumstances</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>1</strong></td>
<td>aqueductal stenosis</td>
<td>8</td>
<td>5 days</td>
<td>headache</td>
<td>0</td>
<td>ventricular obstruction</td>
<td>died while awaiting diagnostic confirmation and is included in our series of seven hospitalized patients who experienced life-threatening shunt malfunction (Case 2 in Table 1). In this group of children, severe shunt malfunction could not be predicted from the history and the physical findings on admission, even in retrospect. Obliteration of the basal cisterns on CT scans proved to be the single most important indicator. We recommend that the patient who presents with shunt failure and obliteration of the basal cisterns on CT undergo immediate shunt revision.</td>
</tr>
<tr>
<td><strong>2</strong></td>
<td>Dandy-Walker variant</td>
<td>2</td>
<td>&lt; 24 hrs</td>
<td>apnea</td>
<td>0</td>
<td>disconnection</td>
<td>died at home after minor head trauma and is included in our series of seven hospitalized patients who experienced life-threatening shunt malfunction (Case 2 in Table 1). In this group of children, severe shunt malfunction could not be predicted from the history and the physical findings on admission, even in retrospect. Obliteration of the basal cisterns on CT scans proved to be the single most important indicator. We recommend that the patient who presents with shunt failure and obliteration of the basal cisterns on CT undergo immediate shunt revision.</td>
</tr>
<tr>
<td><strong>3</strong></td>
<td>intraventricular hemorrhage</td>
<td>10</td>
<td>&lt; 24 hrs</td>
<td>malaise</td>
<td>8†</td>
<td>disconnection</td>
<td>died at camp and is included in our series of seven hospitalized patients who experienced life-threatening shunt malfunction (Case 2 in Table 1). In this group of children, severe shunt malfunction could not be predicted from the history and the physical findings on admission, even in retrospect. Obliteration of the basal cisterns on CT scans proved to be the single most important indicator. We recommend that the patient who presents with shunt failure and obliteration of the basal cisterns on CT undergo immediate shunt revision.</td>
</tr>
<tr>
<td><strong>4</strong></td>
<td>spina bifida</td>
<td>2</td>
<td>&lt; 24 hrs</td>
<td>apnea</td>
<td>0</td>
<td>ventricular obstruction</td>
<td>died of cardiac arrest at home and is included in our series of seven hospitalized patients who experienced life-threatening shunt malfunction (Case 2 in Table 1). In this group of children, severe shunt malfunction could not be predicted from the history and the physical findings on admission, even in retrospect. Obliteration of the basal cisterns on CT scans proved to be the single most important indicator. We recommend that the patient who presents with shunt failure and obliteration of the basal cisterns on CT undergo immediate shunt revision.</td>
</tr>
</tbody>
</table>

* This case is the same as Case 2 in Table 1. † Last revision at 4 years of age.

### References

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