One may consider that the child with hydrocephalus complicating a primary brain tumor has two distinctly different diseases: 1) a neoplasm, and 2) hydrocephalus. The hydrocephalus may be obstructive, communicating, or (in the case of choroid plexus papilloma) hypersecretory. This paper presents the incidence, symptomatology, and management of hydrocephalus associated with infratentorial brain tumors in 156 children and with pineal tumors in 21 children, all treated by the first author during the period 1967 to 1979. Medulloblastoma was the most commonly encountered tumor. Of 117 children with cerebellar-fourth ventricle tumors, 110 had hydrocephalus, whereas 11 of 34 with brain-stem tumors and all 21 with pineal tumors presented with hydrocephalus. Of the total 143 patients with hydrocephalus, shunts were inserted before craniotomy in 123, with the interval between shunt insertion and craniotomy for tumor removal ranging from 1 to 21 days. Eighteen of these children required shunt revisions after craniotomy; 36 of 80 patients followed for 6 months or more had their shunts permanently removed, but it was necessary to reinsert the shunt from 5 days to 13 months later in 30% of these patients. Only 25 patients remained permanently shunt-free. It is considered that the placement of shunts before craniotomy is justified by the extraordinarily high incidence of papilledema and visual impairment associated with hydrocephalus. "Upward herniation" and dissemination of neoplastic cells through the shunting system are theoretical contraindications to precraniotomy shunt insertion, although the former is rare (3%), and there is no statistical evidence to support the latter.

Key Words • hydrocephalus • shunting procedure • posterior fossa tumor • upward brain herniation • childhood brain tumor • shunt metastasis • systemic metastasis
effects of hydrocephalus. Accordingly, many surgeons,\textsuperscript{7,46} including Cushing,\textsuperscript{3} have recommended routine placement of a single occipital burr hole prior to opening the posterior fossa, so that the ventricular system could be punctured to decompress the enlarged ventricles. Ventricular needles were once taped to the bedside or to patients' charts during transportation, to be used for emergency ventricular drainage and intracranial decompression. The two obvious disadvantages of this procedure are uncertain decompression and increased risk of subdural bleeding secondary to rapid decompression. With the increase in diagnostic techniques that permit the preoperative diagnosis of the associated hydrocephalus, and the significant improvement in shunting procedures, neurosurgeons began to suggest the insertion of a shunting device, external\textsuperscript{30,32,45} or internal,\textsuperscript{1,2,16-18,35} before proceeding with a craniotomy directed toward the definitive treatment of the tumor. All of these authors have reported excellent results, and many have commented on the remarkable improvement in the general condition of the child after the shunting procedure, along with a disappearance or improvement of symptoms, such as somnolence, headache, vomiting, loss of appetite, and double vision.

Several reasons have been given in support of the use of a shunting device before the definitive craniotomy: 1) false localizing signs, resulting from the associated hydrocephalus or subsequent herniation, diminish or disappear; 2) preoperative air study, in the rare instances that it may be necessary, may be carried out without risk of precipitating severe complications; 3) compensating the ICP by inserting a shunt allows adequate time for stabilization of the intracranial contents, particularly cerebral blood flow, and diminution of the cerebral edema secondary to the hydrocephalus; 4) at surgery, the operative field is slack, thus easing the approach to the tumor and eliminating the need for hypertonic solutions, steroids, ventricular cannulation, or continuous spinal drainage; 5) a smooth postoperative course is more likely.

The main theoretical disadvantage of inserting a shunt in a child with an infratentorial tumor is that it may produce or aggravate “upward herniation.” The likelihood of intracranial hypotension, which may lead to postshunt subdural hematoma, the introduction of a foreign body as a focus for a possible infection, and the possible dissemination of neoplastic cells via the shunt represent the theoretical reasons for removing the shunting device as soon as possible. Conversely, one may consider that the associated hydrocephalus may not always be secondary to simple mechanical obstruction of the CSF pathways, and that the child may indeed have permanent hydrocephalus. The reported incidence of medulloblastoma tumor dissemination along the shunt track\textsuperscript{32} has not been confirmed.\textsuperscript{34}

We shall, in this work, present the incidence, symptomatology, and management of hydrocephalus associated with brain tumors in the posterior fossa and pineal-quadrigeminal region, and attempt to answer the above questions.

Clinical Material

During the period 1967 to 1979, 332 infants and children with brain tumors were treated at the Northwestern University-Children's Memorial Hospital in Chicago. Ages ranged from newborn to 16 years at the time of diagnosis. Tumors were located infratentorially in 156 patients, in the pineal-quadrigeminal region in 21, and supratentorially in 155. A precise neuroradiological diagnosis was made in all infants with cerebral angiography, pneumoencephalography, ventriculography, radionuclide scan, and/or computerized transmission tomography (CTT). The size of the ventricular system was evaluated, and the cases were classified into hydrocephalic (Group I) and non-hydrocephalic groups (Group II).

Among 156 patients with infratentorial tumors, 122 (78.2\%) were found to have hydrocephalus. All of the 21 patients with pineal-quadrigeminal tumors were hydrocephalic. The infratentorial tumors were subdivided into lesions in the cerebellar-fourth ventricle, brain-stem, and clival-cerebellopontine angle regions. Table 1 presents data on the histological types of tumor in each location, the number of cases, and the incidence of hydrocephalus. Among 117 cerebellar-fourth ventricle tumors, medulloblastoma was the most common (53 cases), followed by benign astrocytoma (38 cases). Of these 117 patients, 110 (94.0\%) suffered hydrocephalus. The second most common site of posterior fossa tumor was the brain stem. In our series, 18 of 34 brain-stem tumors were histologically verified. Eleven patients (32.4\%) were found to be hydrocephalic by the time of diagnosis. Five clival-cerebellopontine angle tumors were operated on. Only one patient, who had a meningioma, suffered hydrocephalus. The incidence of hydrocephalus is independent of the tumor type, benign or malignant. Rather, it is related to tumor location. The hydrocephalus was invariably symmetrical.

Clinical Findings

Signs and Symptoms

Table 2 lists the symptoms encountered in these patients. The main symptoms of the hydrocephalic group (Group I) are those of increased ICP. Ninety-
A. J. Raimondi and T. Tomita

TABLE 1
Incidence of hydrocephalus according to tumor type and location

<table>
<thead>
<tr>
<th>Histology</th>
<th>Cerebellar-IV Ventricle</th>
<th>Brain Stem</th>
<th>Clival-Cerebellopontine Angle</th>
<th>Pineal</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Total Hydrocephalic</td>
<td>Total Hydrocephalic</td>
<td>Total Hydrocephalic</td>
<td>Total Hydrocephalic</td>
</tr>
<tr>
<td>medulloblastoma</td>
<td>53 51</td>
<td>0 0</td>
<td>0 0</td>
<td>3 3</td>
</tr>
<tr>
<td>benign astrocytoma</td>
<td>38 36</td>
<td>11 5</td>
<td>0 0</td>
<td>1 2</td>
</tr>
<tr>
<td>ependymoma</td>
<td>11 10</td>
<td>0 0</td>
<td>0 0</td>
<td>0 0</td>
</tr>
<tr>
<td>malignant astrocytoma</td>
<td>4 3</td>
<td>6 3</td>
<td>0 0</td>
<td>1 1</td>
</tr>
<tr>
<td>dysembryoma</td>
<td>0 0</td>
<td>0 0</td>
<td>0 0</td>
<td>4 4</td>
</tr>
<tr>
<td>ependymoblastoma</td>
<td>3 3</td>
<td>0 0</td>
<td>0 0</td>
<td>1 1</td>
</tr>
<tr>
<td>glioblastoma multiforme</td>
<td>0 0</td>
<td>1 1</td>
<td>0 0</td>
<td>1 1</td>
</tr>
<tr>
<td>meningioma</td>
<td>0 0</td>
<td>0 0</td>
<td>2 1</td>
<td>0 0</td>
</tr>
<tr>
<td>metastasis</td>
<td>2 1</td>
<td>0 0</td>
<td>0 0</td>
<td>0 0</td>
</tr>
<tr>
<td>undifferentiated</td>
<td>2 2</td>
<td>0 0</td>
<td>1 0</td>
<td>0 0</td>
</tr>
<tr>
<td>other</td>
<td>4 4</td>
<td>0 0</td>
<td>2 0</td>
<td>5 5</td>
</tr>
<tr>
<td>unverified</td>
<td>0 0</td>
<td>16 2</td>
<td>0 0</td>
<td>4 4</td>
</tr>
<tr>
<td>total</td>
<td>117 110</td>
<td>34 11</td>
<td>5 1</td>
<td>21 21</td>
</tr>
</tbody>
</table>

three patients (65%) of this group suffered headaches and 73 (51%) vomiting. Unsteady gait was also common. It is of interest that seven children in this group presented with seizures!

The symptoms of the non-hydrocephalic group (Group II) are mainly those of brain-stem glioma, representing 68% of the group. Unsteadiness was the most common symptom (38.2%), with headache and/or vomiting second (29.4%). Symptoms of intracranial hypertension were less common, and symptoms of cerebellar or brain-stem involvement more common. One patient with medulloblastoma was first admitted because of spinal cord symptoms resulting from spinal cord seeding, confirmed by a laminectomy. Posterior fossa medulloblastoma was diagnosed subsequently; hydrocephalus was not present.

TABLE 2
Symptoms associated with infratentorial tumors

<table>
<thead>
<tr>
<th>Symptoms</th>
<th>Group I (Hydrocephalic)</th>
<th>Group II (Non-Hydrocephalic)</th>
<th>Total</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>No. Percent</td>
<td>No. Percent</td>
<td>No. Percent</td>
</tr>
<tr>
<td>headache</td>
<td>93 65.0</td>
<td>10 29.4</td>
<td>103 58.2</td>
</tr>
<tr>
<td>nausea/vomiting</td>
<td>73 51.0</td>
<td>10 29.4</td>
<td>83 46.9</td>
</tr>
<tr>
<td>unsteady gait</td>
<td>52 36.3</td>
<td>13 38.2</td>
<td>65 36.7</td>
</tr>
<tr>
<td>oculs quinqua</td>
<td>23 16.1</td>
<td>6 17.6</td>
<td>29 16.4</td>
</tr>
<tr>
<td>lethargy</td>
<td>22 15.4</td>
<td>3 8.8</td>
<td>25 14.1</td>
</tr>
<tr>
<td>head tilt</td>
<td>10 7.0</td>
<td>6 17.6</td>
<td>16 9.0</td>
</tr>
<tr>
<td>muscle weakness</td>
<td>6 4.2</td>
<td>6 17.6</td>
<td>12 6.8</td>
</tr>
<tr>
<td>irritability</td>
<td>8 5.6</td>
<td>2 5.9</td>
<td>10 5.6</td>
</tr>
<tr>
<td>increased head circumference</td>
<td>10 7.0</td>
<td>0 0</td>
<td>10 5.6</td>
</tr>
<tr>
<td>seizures</td>
<td>7 4.9</td>
<td>0 0</td>
<td>7 4.0</td>
</tr>
<tr>
<td>total</td>
<td>143 96.9</td>
<td>34 2.4</td>
<td>177 98.3</td>
</tr>
</tbody>
</table>

Papilledema, the most common sign in children with posterior fossa tumors (Table 3), was present in 69.9% of Group I patients, but in only one patient (2.9%) in Group II, a child with diffuse astrocytoma of the cerebellum. Papilledema was seen in 36 (67.9%) patients with medulloblastoma, 28 (66.7%) with cerebellar astrocytoma, eight (23.5%) with brain-stem glioma, 10 (71.4%) with fourth ventricular ependymoma, and 16 (76.1%) with pineal-quadrigeminal tumors.

Ataxia was the second most common sign and dysmetria was the third, followed by such pyramidal tract signs as motor weakness and abnormal reflexes. Twelve children with cerebellar tumors, either vermian or hemispheric, had motor weakness, with five having contralateral and five ipsilateral hemiparesis; two children with vermian tumors had hemiparesis and two other children with hemispheric-vermian tumors had paraparesis or quadriparesis. Impairment of consciousness was relatively uncommon, occurring in only 15%. Fifteen of 19 children with decreased deep tendon reflexes had medulloblastomas. Parinaud's syndrome was present in 10 of 21 cases of pineal-quadrigeminal tumors.

Of 143 patients in Group I (122 posterior fossa and 21 pineal-quadrigeminal tumors), shunts were inserted before craniotomy in 123. With only two exceptions, the infratentorial tumors were operated on through a posterior fossa craniotomy, whereas of the 21 pineal-quadrigeminal tumors, seven were approached through the posterior fossa, seven via a parasagittal, two via an occipital, and five via a combined approach.

The interval between the insertion of the precraniotomy shunt and the craniotomy ranged from 1 to 21
hydrocephalus and infratentorial tumors

### TABLE 3

<table>
<thead>
<tr>
<th>Signs*</th>
<th>Hydrocephalic</th>
<th>Non-Hydrocephalic</th>
<th>Total</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Total</td>
<td>Percent</td>
<td>Total</td>
</tr>
<tr>
<td>papilledema</td>
<td>100</td>
<td>69.9</td>
<td>1</td>
</tr>
<tr>
<td>cerebellar ataxia</td>
<td>63</td>
<td>44.1</td>
<td>16</td>
</tr>
<tr>
<td>cerebellar dysmetria</td>
<td>47</td>
<td>32.9</td>
<td>10</td>
</tr>
<tr>
<td>abducens palsy</td>
<td>36</td>
<td>25.2</td>
<td>16</td>
</tr>
<tr>
<td>Babinski sign</td>
<td>37</td>
<td>25.9</td>
<td>8</td>
</tr>
<tr>
<td>other cranial nerve signs</td>
<td>27</td>
<td>18.9</td>
<td>17</td>
</tr>
<tr>
<td>facial palsy</td>
<td>27</td>
<td>18.9</td>
<td>15</td>
</tr>
<tr>
<td>motor weakness</td>
<td>22</td>
<td>15.4</td>
<td>17</td>
</tr>
<tr>
<td>nystagmus</td>
<td>27</td>
<td>18.9</td>
<td>10</td>
</tr>
<tr>
<td>increased DTR’s</td>
<td>33</td>
<td>23.1</td>
<td>2</td>
</tr>
<tr>
<td>lethargy</td>
<td>23</td>
<td>16.1</td>
<td>4</td>
</tr>
<tr>
<td>decreased DTR’s</td>
<td>19</td>
<td>33.3</td>
<td>0</td>
</tr>
<tr>
<td>Macewen’s sign</td>
<td>18</td>
<td>12.6</td>
<td>0</td>
</tr>
<tr>
<td>increased head circumference</td>
<td>10</td>
<td>7.0</td>
<td>0</td>
</tr>
<tr>
<td>irritability</td>
<td>8</td>
<td>5.6</td>
<td>2</td>
</tr>
<tr>
<td>total</td>
<td>143</td>
<td>34</td>
<td>177</td>
</tr>
</tbody>
</table>

*DTR = deep tendon reflex.

days (mean 8 days, median 10 days). The timing of the craniotomy depended upon the disappearance of papilledema, which we used as a clinical indication of disappearance of cerebral edema and cerebral complications of intracranial hypertension. Seventy-eight children had papilledema before insertion of the precraniotomy shunt. In this group, papilledema resolved completely or subsided markedly in 62 patients, and did not change in eight. Information on the status or change in papilledema was not recorded in eight patients.

Of 57 patients with headache, 45 improved and 12 showed no change after the precraniotomy shunt. Of 25 in whom ataxia was not noted before shunt placement, three developed ataxia after shunting, and this sign was noted to become more prominent in five of 41 who had exhibited it before shunting. Similar changes were noted in cerebral dysmetria. Of 20 patients with initially impaired consciousness, after shunting the level of consciousness improved in 19 and worsened in one. Of the 103 patients who were alert before the precraniotomy shunt, five showed transient lethargy and three persistent lethargy in the postshunt period.

**Illustrative Case Reports**

Three of these patients merit specific comment.

**Case 1.** This 18-month-old boy was referred to us from Venezuela. At the age of 9 months, he was diagnosed by ventriculography to have hydrocephalus and a posterior fossa tumor. A ventriculoperitoneal (VP) shunt was inserted on May 10, 1976. Because no improvement followed placement of the shunt, he was transferred to us on August 10, 1976, when he was found to be somewhat lethargic with a marked quadripareisis. Angiograms on the same day showed symmetrical hydrocephalus with a right cerebellar hemisphere tumor. The shunt was revised on August 11. Following this procedure, he developed Cheyne-Stokes respiration, anisocoria, and a decorticate posture. His respirations became progressively worse. Two days after shunt revision, he underwent a right occipital craniotomy, and a medulloblastoma of the right cerebellar hemisphere and superior vermis was removed by the transtentorial approach. Postoperatively, he did well, with only mild residual left hemiparesis persisting. He improved after discharge and is well at the time of this writing.

**Case 2.** This boy was first seen on October 10, 1975, at the age of 4 years, because of headaches, vomiting, and cerebellar ataxia. Angiography the next day revealed symmetrical hydrocephalus and a posterior fossa mass. A VP shunt was inserted on the same day. Subsequently, he became lethargic and vomited intermittently. At the 6th day after placement of the precraniotomy shunt, he developed right hemiparesis with left facial palsy and became stuporous. On October 21, he underwent posterior fossa craniotomy, and an ependymoma was found in the fourth ventricle, extending into the right cerebellar hemisphere,
cerebellopontine angle, and spinal canal. The tumor was resected. At the same time, patency of the aqueduct of Sylvius was confirmed. He remained stuporous postoperatively. The VP shunt was removed on October 27, 1975. Subsequently, his consciousness gradually improved; he started to talk, and regained full neurological function.

**Case 3.** This little girl was first seen at the age of 2 years for evaluation of progressive staggering gait, headaches, and vomiting of 2 months' duration. Skull x-ray films revealed splitting sutures, and CTT scanning and angiograms showed hydrocephalus with possible pineal lesion. A right VP shunt was inserted without difficulty after cerebral angiography. The ventricular fluid was noted to be clear. However, immediately after recovery from anesthesia, the patient had shallow respirations, constricted and fixed pupils, divergent ocular deviation, and minimal doll's-eye movements. Because of the development of intermittent decorticate and decerebrate posture, CTT scanning was repeated, showing a hyperdense lesion in the pineal region, the brain stem, and the diencephalon, although the ventricular size had returned to normal. Because of the patient's deteriorating condition during the 2 days after insertion of the precraniotomy shunt, she underwent posterior fossa craniotomy.

Surgery revealed hemorrhage in the fourth ventricle, pushing the brain stem forward, and intratumoral hemorrhage into the pineal and superior vermian portions of the tumor. Pathological examination confirmed hemorrhage into the pineal region medulloblastoma. After craniotomy, the patient gradually improved. After prolonged coma, she first became hyperkinetic, then started to talk and walk. At the present, 2 years following craniotomy, her neurological status is normal, except for mild ataxia.

**Precraniotomy Shunt Insertion: Results**

In these three instances, the deterioration in neurological status was, in our opinion, a direct result of dynamic changes that occurred secondary to the shunting procedures. Although deterioration was severe, it was completely reversible, accentuating the necessity to observe children closely during the postshunting period, and to act in a timely manner if signs of upward herniation appear.

Other minor adverse responses to the precraniotomy shunts were shunt infection in two cases; shunt malfunction in four; clinically silent supratentorial subdural hematoma in one; the onset of hemiparesis in four and its worsening in one; and a vocal cord palsy in a child with a vermician tumor. In no instance did a permanent deficit result from the shunting procedure.

Fourteen patients (Group I or Group II) required shunt insertion after craniotomy. Five Group II patients were known to have had normal-sized ventricles before the craniotomy: two of these five required shunting during the same hospitalization, the remaining three from 4 months to 2 years later. Nine Group I children were operated on without previous shunting, but in all of them it became necessary to insert a shunt from 3 months to 2 years after craniotomy. We are presently unable to evaluate the effects of these prolonged periods of hydrocephalus on ultimate survival or function.

Twenty-seven patients did not have a shunt inserted before or after craniotomy. Thirteen of them had hydrocephalus prior to craniotomy.

Among the 123 patients who had a shunt placed before craniotomy, 18 required revisions after craniotomy. Of 80 patients who were followed more than 6 months after craniotomy, 36 had their shunts removed. Eleven of this latter group (30%) required reinsertion of the shunt from 5 days to 13 months later. The remaining 25 patients remained shunt-free.

Fourteen patients with cerebellar astrocytoma had elective removal of the precraniotomy shunt, at the termination of the craniotomy in five, and within 2 weeks in the other nine. Three patients with medulloblastoma had elective shunt removal on the 5th and 6th days after craniotomy. The other seven who had their shunts removed had it done between 11 days and 2 years postoperatively, for subdural hematoma, shunt disconnection or infection, or because it was considered no longer necessary.

Out of 53 medulloblastoma patients, nine required revision of the precraniotomy shunt after craniotomy; four required reinsertion of the shunt after elective removal; six required insertion of a shunt after craniotomy. Nineteen of our 53 medulloblastoma patients (35.8%) required permanent shunts after craniotomy and radiation therapy.

Of 42 patients with cerebellar astrocytoma, two required reinsertion of a shunt, two a shunt revision, and one a postcraniotomy shunt. Six of these 42 cases (14.3%) are shunt-dependent and 25 of 42 (59.5%) became shunt-free after craniotomy.

With the intent of learning whether the posterior fossa was tense or slack at the time of craniotomy, we reviewed 90 operative reports. Sixty-seven of these patients had had a shunt placed before craniotomy and 23 did not. In the former group, 49 were operated on in the sitting position and 18 prone. The surgeon reported the posterior fossa to be “full,” “tight,” or “bulging” in only four cases (6%). On the contrary, in
Hydrocephalus and infratentorial tumors

children without a precraniotomy shunt, 21 of whom were operated on in the sitting position and two prone, the posterior fossa was reported to be “full,” “tight,” or “bulging” in seven cases (33%).

Eight (7.9%) supratentorial subdural hematomas occurred 9 days to 3 months after craniotomy in the 101 cases with precraniotomy shunts. Seven had undergone craniotomy in the sitting position, and one prone. All cases were treated by removal of the shunt and evacuation of the subdural hematoma; four required reinsertion of the shunt within 2 weeks to 1 year. There was no difference in the incidence of this complication in the non-shunted group: three of 36 cases without precraniotomy shunt (8.3%) had postoperative supratentorial subdural hematomas.

Discussion

Although the association of hydrocephalus with brain tumors is well known, very few papers have been published on its incidence, nature, natural history, and management. In 1959, Gol and McKissock reported that cerebellar astrocytoma produces “clinical” hydrocephalus in 50% of tumors along the midline and in 22% of the cerebellar hemispheres. Brain-stem glioma, the least common infratentorial tumor to produce hydrocephalus, has been reported in association with hydrocephalus in 20% to 30% of the cases. However, others have reported a higher incidence of hydrocephalus associated with brain-stem tumor. In 1968, Poppen and Marino observed 36 cases of symmetrical hydrocephalus proven by ventriculography among 45 patients with pineal tumor. All of 19 cases with pineal tumors reported by Suzuki and Iwabuchi were treated with preoperative external ventricular drainage. Cerebellar and fourth ventricular tumors have been described as invariably causing hydrocephalus.

The complicating hydrocephalus is responsible, in most instances, for the symptoms and signs that bring the child with a brain tumor to the neurosurgeon, and this increase in ICP so dominates the clinical picture that it “masks” the discrete neurological deficits resulting from tumor compression or destruction of neural centers, pathways, and nuclei. This becomes evident after a precraniotomy shunt compensates the hydrocephalus, at which time symptoms such as nystagmus, ataxia, and diplopia appear or “worsen.” Persistence of headache, especially positional, Bruns’ ataxia, vomiting, and pulse and respiratory changes are probably the result of mass effect, especially in midline posterior fossa tumors. It has been noted that the incidence of papilledema is proportional to the incidence of complicating hydrocephalus, and that it is present in 87% of infratentorial and 58% of supratentorial tumors in the pediatric age range.

It is this extraordinarily high incidence of papilledema and visual impairment, and many cases with pre- and postoperative (craniotomy) blindness or permanent visual disturbances, that puts the child at risk and justifies precraniotomy shunting to treat the hydrocephalus first. Cerebellar tumors cause papilledema in 90% of cases. Cushing stated, “It is the hydrocephalus that is responsible for the most serious symptoms of all from the standpoint of a useful life after a successful operation: namely, blindness.” In his large series of brain tumors, among 61 patients with cerebellar medulloblastomas, impaired sight was noted at the time of admission in 26, nine of whom were already blind. Forty of 76 patients with cerebellar astrocytoma had impaired vision at the time of admission, 22 of them having already become blind, or nearly so.

When a successful decompression is performed, the improvement of papilledema is usually rapid and dramatic. In the present study of charts written by neurosurgical residents, significant regression of papilledema was noted several days to 2 weeks after the insertion of the precraniotomy shunt. According to Duke-Elder, it may take as long as 2 months for papilledema to disappear (he was not reporting on postshunt children). Torkildsen reported that the papilledema subsided in 5 weeks in 28% of cases with a third ventricle to cisterna magna shunt, although in some it took 3 to 4 months.

It is of interest to note that seizures related to posterior fossa tumors only occur in patients with hydrocephalus. Backus and Millichap described a series of 165 children with infratentorial tumors, of whom 19 (12%) had seizures and hydrocephalus complicating a posterior fossa tumor. Seventy-nine percent of the 19 patients had an increase in ICP at the time of the first seizure. Wray stated, “Seizures, not usually present in pineal region tumors, occurred in 4 of the cases with severe hydrocephalus, all of whom required a shunt procedure.” Significant changes in cerebral blood flow, associated with acute obstructive hydrocephalus, as well as in the size and electrical characteristics of the extracellular space, changes secondary to the hydrocephalic process, likely play an active role in the genesis of seizures. Control of the hydrocephalus has a positive influence on control of the seizures.

The practical and theoretical disadvantages of shunting as a treatment modality to control hydrocephalus complicating a brain tumor are “upward herniation” and dissemination of neoplastic cells through the shunting system. “Upward herniation” is
a well known term but a poorly defined clinical picture, about which little has been written. Anatomically, this consists of anterosuperior displacement of the cerebellum and brain stem through the tentorial notch, compressing the dorsal surface of the mesencephalon, and deforming the posterior third ventricle. The veins of Rosenthal and Galen are compressed and distorted, raising the supratentorial pressure. 3,11 Although Plum and Posner 23 described the clinical symptoms of “upward herniation” as progressive obtundation, hyperventilation, conjugate downward gaze, or loss of upward gaze, they could not conclude with certainty that upward herniation produces a consistent clinical syndrome. Rubinstein 59 theorized that the removal of a large supratentorial mass may cause upward cerebellar herniation. Emery 12 described two autopsy cases of long-standing shunts in hydrocephalus and meningomyelocele, in which there was marked upward movement of the brain stem and cerebellum, lifting of the hypothalamus, and stretching of the pituitary stalk. It has been postulated that in children with posterior fossa tumors this phenomenon is more accelerated because the posterior fossa contents already tend to prolapse through the tentorial notch. Hoffman, et al., 17 described upward herniation in six of 96 patients (6%) who underwent precraniotomy shunting for posterior fossa tumor. They noted that symptoms usually occur 24 to 38 hours after insertion of the precraniotomy shunt and that they are remedied by a prompt posterior fossa decompression. Their reported incidence of the “upward herniation” is slightly higher than ours.

Another serious complication that we encountered after the insertion of a shunt before craniotomy was hemorrhage into the tumor, although we doubt that there is a causal relationship between the two events. Spontaneous hemorrhage into brain tumors occurs rarely; the majority of cases involve glioblastoma 26 or pituitary adenoma. Epstein and Mural 13 reported two cases of intratumoral hemorrhage (both posterior fossa astrocytomas) in 30 patients with precraniotomy shunts, and quite a few cases of intratumoral hemorrhage into medulloblastoma have been published. 24 Increase of cerebral blood flow and shift of brain structures after shunt insertion may play a role in intratumoral hemorrhage. There seems to be no real difference in supratentorial hydrodynamic changes resulting in subdural hematoma, whether the ventricles are decompressed by a shunt or by a posterior fossa craniotomy, with tumor removal. The incidence of postoperative subdural hematoma was insignificantly different between these groups in our series.

Cushing commented that, “Preliminary emptying of the cerebral ventricles is disadvantageous in one respect for it may so lower tension in the posterior fossa that surface indications of a deep median wholly subcortical tumor are effectually concealed and may be overlooked.” This comment, since the advent of preoperative diagnostic procedures, must be considered only historic in value. The application of intraventricular cannulation has several problems: risk of intracranial bleeding secondary to acute decompression of ICP, danger of irreversible brain damage due to increased ICP at the time of induction of anesthesia, and unreliable 9,25 decompression effect.

Following insertion of a shunt, the posterior fossa becomes quite slack, irrespective of whether the child is operated on in a sitting or prone position. This renders opening of the dura and separation of the tumor from the cerebellum easy and safe, and diminishes venous stasis and cerebellar prolapse.

We recommend removal of the shunt after the CSF pathway obstruction has been cleared. On occasion, we have removed the shunt directly, and at other times we have occluded it for varying periods of time before removal to be sure the child was not permanently hydrocephalic.

We are aware of the reports in the literature on dissemination of tumor cells through shunting systems 17,19,22 but have never observed it in a posterior fossa tumor (medulloblastoma or ependymoma). We have documented metastases outside the central nervous system in four of 166 hydrocephalic children with brain tumors who had been shunted, and in four of 166 similar children without shunts. Shunt-related metastases occurred in two children with primitive neuroectodermal tumors (both located in the pineal region). In both instances, the metastases were to the peritoneal cavity. One of these children had an infratentorial medulloblastoma; a ventriculoperitoneal shunt was placed, but the tumor metastasized to lung and bone. In the other child, who had a supratentorial atypical teratoma, the tumor metastasized to the intraperitoneal cavity. We concluded that three of the four children indeed had shunt-related metastases, the only exception being the child with the medulloblastoma. This number is not statistically significant, since four children without shunts suffered systemic metastases (three with medulloblastoma and one with malignant ependymoma). We conclude that, unlike medulloblastomas, it is possible that primitive neuroectodermal tumors, ectopic pineal, and pineal dysgerminomas may metastasize through a shunting system and that every effort should be made to remove the shunt as soon as possible after x-ray therapy to the primary tumor site.

Despite radical resection of cerebellar tumors, with removal of the obstructing tumor from the fourth
ventricle and/or the aqueduct, the patient may suffer permanent hydrocephalus from arachnoiditis secondary to subarachnoid bleeding or local arachnoidal adhesions that were present before the operation. 8,25,40

Nine (20%) of 45 children with medulloblastoma required postoperative insertion of shunts in a series described by Mealy and Hall. 26 Stein, et al., 41 reported that 30% of their patients with astrocytoma developed hydrocephalus in the immediate postoperative period, and 20% showed persistent postoperative hydrocephalus. None of their cerebellar astrocytoma patients had a precraniotomy shunt. In all brain-stem glioma patients, once hydrocephalus develops, it is chronic, even after radiation therapy. Pineal-quadrigeminal tumors tend to produce chronic hydrocephalus. We have observed this with permanent stenosis of the aqueduct of Sylvius, meningeal adhesions, and cystic formation in the quadrigeminal cisterns, which are the main route of CSF circulation.

In summary, we conclude that precraniotomy shunting for hydrocephalus complicating infratentorial tumors provides the child a significant margin of safety from the negative effects of cerebral edema, papilledema, and rapid decompression of both the supra- and infratentorial compartments when the tumor is removed without previous shunting. Steroids are not necessary in these children, thus eliminating the serious complications, such as hyperglycemia, increased risk of infection, and gastrointestinal bleeding, that are associated with megadose steroid management. “Upward herniation” is a real, although very rare (3%), complication. There is no statistical evidence to support the theoretical concept that “seeding” may occur along the shunting system or that this latter event increases the risk of systemic metastases from medulloblastoma or ependymoma.

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