MANAGEMENT of Chiari I malformations with or without associated syringomyelia continues to pose great challenges to physicians. Although the complete pathogenic mechanisms are yet to be elucidated, it is widely accepted that the descended cerebellar tonsils create craniospinal pressure dissociation and impaired CSF flow, which is a prerequisite for syrinx formation. Consequently, the first line of surgical therapy at many institutions is CVD. The specific surgical steps in this operation continue to undergo modification as surgeons attempt to identify the optimum procedure. The standard procedure at many institutions consists of a suboccipital craniectomy, cervical laminectomy at C-1, intradural exploration with or without reduction of the tonsils, and duraplasty.

This operation has proved to be quite effective, and both its successes and failures have been well documented in the medical literature. The most frequently mentioned complications include pseudomeningocele, meningitis, and CSF leak. However, a complication with potentially severe consequences—cerebellar ptosis, or slump—has received comparatively little attention.

Cerebellar ptosis is generally the result of a suboccipital craniectomy that is too large for the particular patient, leading to the descent of the cerebellum through the craniectomy defect. Patients with cerebellar ptosis characteristically present with intractable headaches that differ from those usually associated with Chiari I malformation. Furthermore, by reestablishing contact between the tonsils and brainstem, as well as creating dorsal adherence, the ptosed cerebellum can cause obstruction of CSF flow at the cervicomedullary junction and, thereby, participate in the maintenance of a syringomyelic cavity. In essence, this complication can potentially negate the desired effects of the operation.

The occurrence of cerebellar ptosis has previously been discussed by several authors; however, an effective form of treatment has yet to be defined. The goal of this paper is to describe our experience in the management of a series of symptomatic patients suffering from cerebellar ptosis following CVD for Chiari I malformation, and the development of an effective treatment using cranioplasty.

**Clinical Material and Methods**

**Patient Population**

Seven patients with symptomatic cerebellar ptosis following CVDs performed at other institutions were evalu-
ated at the University of California Los Angeles Medical Center between 1984 and 1999 (Table 1). All of the patients were women and they ranged in age from 30 to 44 years (mean age 37 years). The amount of time between the initial decompressive surgery and evaluation at the University of California Los Angeles Medical Center ranged from 9 months to 17 years (average 6.8 years). Three of the seven patients are known to have had a dural graft placed during the original operation (lyophilized dura, cervical fascia, and polyglactin mesh). The dura mater was not opened or was closed primarily in two patients, and the dural procedure was unknown in two patients.

**TABLE 1**
Clinical characteristics of seven women with cerebellar ptosis following CVD for Chiari I malformation*

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Age (yrs)</th>
<th>Previous Op(s)†</th>
<th>Syrinx</th>
<th>Primary Symptom</th>
<th>Treatment for Cerebellar Ptosis</th>
<th>Postop Status of Syrinx</th>
<th>Persistent Symptoms</th>
<th>Follow Up (mos)</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>42</td>
<td>1) SC, FV–SA shunt; 2) SP shunt</td>
<td>yes</td>
<td>headache</td>
<td>VP shunt refilled</td>
<td>continued headache, ND</td>
<td>24</td>
<td></td>
</tr>
<tr>
<td>2</td>
<td>41</td>
<td>1) SC; 2) SP shunt</td>
<td>yes</td>
<td>headache</td>
<td>SP shunt collapsed</td>
<td>continued headache, ND</td>
<td>96</td>
<td></td>
</tr>
<tr>
<td>3</td>
<td>38</td>
<td>1) SC, FV–SA shunt</td>
<td>yes</td>
<td>sensory deficit</td>
<td>SP shunt refilled</td>
<td>ND</td>
<td>150</td>
<td></td>
</tr>
<tr>
<td>4</td>
<td>44</td>
<td>1) SC, no intradural exploration</td>
<td>yes</td>
<td>headache</td>
<td>partial cranioplasty &amp; intradural exploration</td>
<td>collapsed resolved</td>
<td>29</td>
<td></td>
</tr>
<tr>
<td>5</td>
<td>39</td>
<td>1) SC, DP; 2) SP shunt</td>
<td>yes</td>
<td>sensory deficit</td>
<td>partial cranioplasty &amp; intradural exploration</td>
<td>collapsed resolved</td>
<td>34</td>
<td></td>
</tr>
<tr>
<td>6</td>
<td>39</td>
<td>1) SC, DP</td>
<td>collapsed</td>
<td>headache</td>
<td>partial cranioplasty</td>
<td>collapsed resolved</td>
<td>16</td>
<td></td>
</tr>
<tr>
<td>7</td>
<td>30</td>
<td>1) SC, ST, DP</td>
<td>no</td>
<td>headache</td>
<td>partial cranioplasty</td>
<td>none</td>
<td>resolved</td>
<td>8</td>
</tr>
</tbody>
</table>

*DP = duraplasty; FV–SA = fourth ventricle–subarachnoid; ND = neurological deterioration; SC = suboccipital craniectomy; ST = shrinkage of tonsils.
† Numerals indicate numbered operation.

**Signs and Symptoms of Cerebellar Ptosis**

Headache was the most frequently noted symptom and was seen in five patients. These patients stated that the pain was clearly different from the pain they had experienced before undergoing CVD. The newer pain was located in the suboccipital region but radiated into the frontal region or jaw; it was not associated with coughing, straining, or valsalva movements. Other chief physical complaints were noted primarily by the two remaining patients and included hypesthesia and weakness in the extremities.

**Surgical Treatment**

All seven patients underwent surgical management to treat their cerebellar ptosis and/or residual syringomyelia. The procedures performed included VP shunt placement in one patient, SP shunt placement in two, and partial suboccipital cranioplasty (with or without intradural exploration) in four.

**Placement of a VP Shunt**

One patient (Case 1) had previously undergone a suboccipital craniectomy with fourth ventricle– and syrinx–subarachnoid shunts. One year later she began to experience headaches, and MR images revealed ptosis of the cerebellum and recurrent syrinx formation (Fig. 1). A VP shunt was placed in the right occipital lobe to treat this condition. This was the first patient with cerebellar ptosis encountered by the senior author (U.B.).

**Placement of an SP Shunt**

Two patients (Cases 2 and 3) had previously undergone suboccipital craniectomies. In one patient (Case 3) a fourth ventricle–subarachnoid shunt was placed at the time of the original craniectomy, whereas in the other patient (Case 2) an SP shunt was implanted after CVD. These patients remained clinically stable for 11 and 3 years, respectively, but then suffered progressive neurological deterioration. The patient in Case 2 noticed severe headache and weakening of upper-extremity strength, and the patient in Case 3 began to experience lower-extremity sensory deficits. Magnetic resonance images revealed the presence of severe cerebellar ptosis and recurrent syrinx cavities in both patients. Concerned about potential risks,
we elected not to treat the ptosis per se, and SP shunts were placed for definitive management.

**Partial Suboccipital Cranioplasty With or Without Intradural Exploration**

The patient in Case 4 had previously undergone suboccipital craniectomy without intradural exploration for treatment of a Chiari I malformation with associated syringomyelia. Several years following the surgery she experienced severe headaches. The patient in Case 5 noted decreasing sensation in her upper and lower extremities 5 years after CVD and placement of an SP shunt. Magnetic resonance images obtained in both patients revealed ptosis of the cerebellum and a recurrent distended syrinx cavity. Reexploration of the craniectomy was performed along with reduction of the tonsils, pericranial duraplasty, and partial cranioplasty (accomplished using hydroxyapatite and methylmethacrylate, respectively).

Two patients (Cases 6 and 7; Fig. 2) had previously undergone CVD for Chiari I malformation, with and without syringomyelia, respectively. Severe headaches developed in both patients less than 1 year postoperatively. Magnetic resonance images displayed severe cerebellar ptosis in both patients. In one patient (Case 6), however, the syrinx remained collapsed. Both patients underwent extradural exploration of the suboccipital craniectomy and partial suboccipital cranioplasty (accomplished using methylmethacrylate).

**Results**

The mean postoperative follow-up period in this series was 51 months (range 8–150 months; Table 1). Patients were reexamined during scheduled visits at 1 month, 3 months, and 1 year postoperatively and intermittently as needed. In three patients (Cases 1–3) the results were unsatisfactory. One patient (Case 1) continued to suffer from headaches following VP shunt placement. Although an initial postoperative MR image had demonstrated a decrease in the size of her syrinx following the procedure, she suffered increasing weakness and sensory deficits 6 months later as a result of syrinx refilling, which led to cyst shunt placement performed at another institution. The second patient (Case 2) experienced persistent headaches and progressive weakening of her upper-extremity strength following placement of an SP shunt. In this case, however, the syrinx remained collapsed. The third patient (Case 3) underwent multiple SP shunt revisions because of treatment failure and infection. Her sensory deficit worsened, and she began to experience severe burning pain.

Partial cranioplasty, with or without intradural exploration, yielded successful results in all four patients who underwent the procedure. In the patient in Case 4 resolution of her preoperative headache was obtained after she underwent intradural exploration and partial cranioplasty; however, she did note having some residual neuropathic perirncisional pain at a follow-up examination conducted more than 2 years later. The patient in Case 5 noticed improvement in lower-extremity and genital sensation after she underwent the same procedure. This correlated well with her postoperative MR image, which revealed reduction of her syrinx (Fig. 3). In two patients (Cases 6 [Fig. 4] and 7) total relief of their headaches was achieved following partial cranioplasty without intradural exploration. Note that the cerebellum in the patient in Case 7 (Fig. 2) had bulged through the craniectomy defect due to stretching of a cervical fascia dural graft and, therefore, a pericranial graft sling was placed under the cerebellar hemispheres and tacked up to the superior bone edge for

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**Fig. 2. Case 7.** Left: A T1-weighted MR image obtained before the patient underwent suboccipital craniectomy for Chiari I malformation. Note the position of the cerebellum in relation to the suboccipital bone edge (arrow). Right: Image obtained after the suboccipital craniectomy demonstrating that the cerebellum now sags below the craniectomy bone edge (arrow); this patient is suffering from severe headache.

**Fig. 3. Case 5.** Sagittal (A) and axial (B) MR images obtained in a patient with cerebellar ptosis and an associated syrinx cavity following CVD for Chiari I malformation. Sagittal (C) and axial (D) images obtained after intradural exploration and partial cranioplasty, demonstrating a reduction in the size of the syrinx cavity.
additional support before we began the cranioplasty. This sling was considered essential to the procedure in this case because the inferior bulging of the existing dura extended beyond the region to be covered by the cranioplasty (Fig. 5). The average size of the previous suboccipital craniectomy in these four patients was more than $4 \times 4$ cm from the estimated edge of the foramen magnum. The partial cranioplasty reduced the craniectomy to approximately $2 \times 2$ cm in each case. In each patient, a crescent-shaped cranioplasty was prepared (Fig. 5). The contiguous bone edge was first notched with a high-speed drill so that injections of methylmethacrylate into these notches would help to secure the prosthesis. None of these patients was neurologically worse, and there were no operative complications.

Discussion

Cerebellar Ptosis

Symptomatic cerebellar ptosis following CVD was first described by Williams in 1978. He presented the case of a young woman who suffered from a new onset of severe headache after having undergone a wide suboccipital craniectomy. The headache was not associated with valsala movements and was located in the frontal and vertex regions. Air encephalography performed 1 year after the decompression revealed severe cerebellar ptosis, and it was believed that this was the cause of her pain. The majority of our patients presented in a similar fashion. We propose that the mechanism of this headache most likely involves stretching of an inadequately supported dura mater of the posterior fossa, which is caused by the sagging cerebellum. The transverse sinus had not been unroofed in any of these patients and is believed not to participate in the progressive downward migration of the cerebellum. The dura mater is amply innervated by nociceptive fibers and is thought to be a prominent source of headache pain. The contribution from the trigeminal nerve to the dural innervation may explain the pain referral to the orbital and/or jaw region observed in our patients. In patients with cerebellar ptosis, neck movement has no influence on their pain, and the pain may take from months to several years to develop. It differs from pain that is attributed to attachment of the nuchal muscle to the dura mater at the posterior fossa, which is associated with neck movement and typically begins within 1 month after surgery.

Patients with cerebellar ptosis can also present with a neurological deficit due to persistent syringomyelia. Severe ptosis results in an obstruction of CSF flow at the cervicomedullary junction in cases in which the descended cerebellar tonsils lie against the neuraxis, and the dorsal subarachnoid space is again obliterated. This presumed craniospinal pressure dissociation or flow impairment may prevent collapse of the syrinx cavity, leading to further neurological deterioration.

Causes of Cerebellar Ptosis

The primary cause of the development of cerebellar ptosis following CVD for Chiari I malformation is a suboccipital craniectomy that is too large for the specific patient. In some patients the type of material used for dural grafting may also play a role. There is usually no need to decompress the entire posterior fossa to treat a pathological process that occurs primarily at the level of the foramen magnum. Although it is recognized that some surgeons favor a wide posterior fossa craniectomy, others have opted for a small craniectomy in the treatment of Chiari I malformation to avoid the complication of ptosis. In the patients in this series who underwent repeated surgical exploration, the original average craniectomy size was greater than $4 \times 4$ cm. In our opinion this area was nearly twice as large as necessary for these patients, and we reduced the size of the defect to approximately $2 \times 2$ cm in each case.

The importance of performing a small craniectomy is magnified in patients with Chiari I malformation because it is known that a significant proportion of these patients...
have a small posterior fossa volume. The patients in our series tended to have a vertically oriented tentorium cerebelli that was inserted low on the occipital bone. We emphasize the need for a preoperative MR imaging study to obtain an approximate measurement of the amount of bone that can be removed without jeopardizing support for the cerebellum. In one particular case, a large craniectomy had been performed, even though the windows of the MR image did not extend far enough to show the point of insertion of the tentorium into the occipital bone. We believe that enough bone should be left to provide a supportive shelf extending up to the greatest diameter of the cerebellar hemisphere observed on a sagittal MR image. The cerebellum may appear to be adequately supported at the time of the initial surgery, but ptosis may occur later, over a period of months or even years, due to molding of the cerebellar tissue.

There are several other factors that may augment the risk of cerebellar ptosis following CVD in patients with Chiari I malformation. Together with bone, the dura mater provides resistance to the cerebellum, and any breach in the dura diminishes the support of the contents of the posterior fossa. Higher rates of ptosis have been described in patients in whom a duraplasty was not performed and the dura was left open. Duddy and Williams described a series of patients who underwent craniectomies that ranged from approximately 4 to 5 cm in height and 3 to 4.5 cm in width without closure of the dura. Descent of the cerebellum occurred in 50% of the patients, although in no case was the descent significant enough to produce symptoms during the follow-up period. The material used for duraplasty may also play a role in the development of cerebellar ptosis. One of the most severe examples of ptosis we encountered was seen in a patient in whom duraplasty had been performed using a cervical paraspinous fascia graft (Case 7). On surgical reexploration, the graft was found to be intact but severely stretched. To date, we have found the pericranium to be quite strong and able to provide adequate support.

Treatment of Cerebellar Ptosis

In his discussion of the first patient he treated for postoperative cerebellar ptosis, Williams mentioned that shunt placement in the ventricle might be helpful in some cases, but that this patient’s ptosis was so extreme that no operation could be offered, and her condition was left uncorrected. He subsequently reiterated this point by stating the following: “. . . if there is no hydrocephalus, there seems little to do.”

Based on this information and on consultation with Williams by the senior author (B Williams, personal correspondence, 1989), the management strategy selected for our first patients with cerebellar ptosis was to implant a VP shunt. The postoperative MR image in one patient demonstrated some decrease in the size of the ventricles and the syrinx cavity, but the patient continued to experience headaches. Furthermore, she suffered neurological deterioration due to cyst reexpansion, which led to placement of additional shunts in the syrinx at another institution 6 months later. We obtained similarly poor clinical outcomes by implanting SP shunts in two patients. Both of these patients continued to be symptomatic with persistent headaches and neurological deterioration. In retrospect, the CSF obstruction caused by the positioned cerebellum was clearly a major contributor to the filling mechanism of the syrinx, and a cyst shunt insertion procedure in this situation had a high likelihood of failure. Additionally, shunt placement did not address the issue of headaches, which are frequently found in this population.

In the patients subsequently treated, we focused our attention entirely on the positioned cerebellum, and developed a method in which structural support could be directly provided. We elected to use methylmethacrylate or hydroxyapatite (one patient) for the cranioplasty because by using these materials we can lock the prosthesis in situ without addition of hardware. Split calvarial grafts have been used in other situations for reconstruction of the posterior fossa. The four patients selected for cranioplasty were separated into two general groups. Patients who required intradural exploration in addition to the partial cranioplasty were those harboring a syrinx cavity; patients who needed only partial cranioplasty were those without a syrinx. Recurrence of the syrinx cavity suggests a continued obstruction of CSF flow at the cervicomedullary junction, which is most often due to large tonsils and obliteration of the dorsal subarachnoid space. When the tonsils have descended very significantly and have not been dealt with previously, we favor intradural exploration with reduction of the cerebellar tonsils. Before performing an intradural procedure, we obtained a confirmatory examination such as an MR cardiac-gated CSF flow study, which can aid in determining the presence and severity of the obstruction at the cranio cervical junction.

The patient in Case 4 began to suffer severe headache after she had undergone suboccipital craniectomy without opening of the dura. A postoperative MR image obtained in this patient revealed cerebellar ptosis, pointed tonsils, and a distended syrinx cavity. A CSF flow study displayed nearly complete obstruction of CSF flow dorsal to the cerebellum. The patient in Case 5 presented with cerebellar ptosis, residual syringomyelia, pointed tonsils, and a CSF flow obstruction, although her chief complaint was declining sensation in her extremities. Both of these patients underwent intradural exploration and reduction of the cerebellar tonsils, as well as partial cranioplasty. Postoperatively, their clinical symptoms resolved, and in both cases MR images revealed collapse of the syringes.

Two patients (Cases 6 and 7) had cerebellar ptosis without distended syrinx cavities, and their tonsils were rounded. In both cases CSF flow studies revealed normal flow at the cranio cervical junction. We concluded that there was no need to perform an intradural exploration in these patients, and both experienced headache resolution after undergoing partial cranioplasty.

Conclusions

Cerebellar ptosis is an infrequent and underrecognized potential delayed complication following CVD for Chiari I malformation. The condition is principally caused by a suboccipital craniectomy that is too large for the individual patient. Patients with cerebellar ptosis generally present with severe headache; however, neurological deterioration from persistent or recurrent syringomyelia can...
occur when CSF flow is obstructed. Partial suboccipital
cranioplasty is successful in treating the headache by sup-
porting the cerebellum and alleviating stretching the dura
mater. In patients with cerebellar ptosis in whom there is
CSF flow obstruction, partial cranioplasty with intradural
exploration is effective in reversing the flow obstruction
and presumed craniospinal pressure dissociation, result-
ing in collapse of the syrinx and neurological recovery.

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