Tonsillectomy without craniectomy for the management of infantile Chiari I malformation

JORGE A. LAZAREFF, M.D., MARCELO GALARZA, M.D., TOORAJ GRAVORI, M.D., AND THEODORE J. SPINKS, M.D.

Division of Neurosurgery, University of California at Los Angeles School of Medicine, Los Angeles, California

Object. The authors report their experience with 15 pediatric patients who underwent resection or shrinkage of the cerebellar tonsils without craniectomy or laminectomy, for the management of Chiari I malformation.

Methods. The procedure was performed in six boys and nine girls with a mean age of 10 years. Thirteen patients presented with the congenital form of this disorder and two patients with Chiari I malformation caused by lumboperitoneal shunting. Clinical complaints included headaches (seven patients), scoliosis (four patients), numbness of the extremities (four patients), and upper-limb weakness (two patients). Two patients presented with failure to thrive and one with vocal cord palsy. Eight patients (six girls and two boys) had syringomyelia. The patients’ symptoms had developed within a mean time period of 21 months (range 1–70 months). In all patients the cerebellar tonsils were exposed through a dura mater–arachnoid incision at the occipitoatlantal space. In seven patients the tonsils were resected and in the remaining eight patients the tonsils were shrunk by coagulating their surfaces.

All patients improved postoperatively. Gliosis with cortical atrophy was observed in the resected neural tissue. Syringomyelia was reduced in seven of eight patients. The mean length of the follow-up period was 7 months.

Conclusions. Removal of herniated cerebellar tonsils can be sufficient for alleviating symptoms in patients with Chiari I malformations.

KEY WORDS • Chiari malformation • syringomyelia • cerebellar tonsil

CHIARI I malformation is a congenital disorder resulting from developmental abnormalities of the paraaxial mesoderm.31,33 It is characterized by a reduced volume of the posterior fossa and often by caudal displacement of the cerebellar tonsils through the foramen magnum in the cervical canal.5,9–11

Surgical treatment of Chiari I malformation aims at expanding the volume of the posterior fossa through a suboccipital craniectomy. Following this common approach, surgeons opt for different procedures: cervical laminectomy, duraplasty, and/or tonsillectomy. All these treatment alternatives have proved to be successful in relieving the symptoms associated with this malformation.5,8,11,12,20,21,25,27,30,33

Nonetheless, considering that most, if not all, clinical manifestations of Chiari I malformation, as well as of the syringomyelia that is present in 40% of patients, can be attributed to the caudal displacement of the cerebellar tonsils,5,9,24,28,30,34 we hypothesized that a procedure that would selectively remove or shrink the cerebellar tonsils would be sufficient for achieving proper brainstem decompression and restitution of normal CSF flow. Such a limited approach could reduce postoperative discomfort and the eventual risk of descent or sagging of the cerebellar hemispheres into the cervical canal.

In the present study we report on 15 patients in whom this selective tonsillar technique was performed.

Abbreviations used in this paper: CSF = cerebrospinal fluid; MR = magnetic resonance.
Tonsillectomy without craniectomy for Chiari I malformation

**TABLE 1**

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Age (yrs)</th>
<th>Main Clinical Complain</th>
<th>Other Neurological Symptom(s)</th>
<th>Tonsillar Displacement</th>
<th>Preop Syringomyelia</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>9, F</td>
<td>5 scoliosis</td>
<td>none</td>
<td>C-2</td>
<td>cervical</td>
</tr>
<tr>
<td>2</td>
<td>10, F</td>
<td>5 scoliosis</td>
<td>none</td>
<td>C-2</td>
<td>cervical</td>
</tr>
<tr>
<td>3</td>
<td>12, F</td>
<td>5 scoliosis</td>
<td>none</td>
<td>C-1</td>
<td>thoracic</td>
</tr>
<tr>
<td>4</td>
<td>10, F</td>
<td>4 headache</td>
<td>neck &amp; back pain</td>
<td>C-2</td>
<td>cervical</td>
</tr>
<tr>
<td>5</td>
<td>18, M</td>
<td>50 scoliosis</td>
<td>none</td>
<td>C-1</td>
<td>cervical</td>
</tr>
<tr>
<td>6</td>
<td>14, F</td>
<td>70 headache</td>
<td>numbness in upper extremities</td>
<td>C-1</td>
<td>cervical</td>
</tr>
<tr>
<td>7</td>
<td>14, F</td>
<td>3 weakness in upper extremities</td>
<td>numbness in upper extremities</td>
<td>C-1</td>
<td>cervical</td>
</tr>
<tr>
<td>8</td>
<td>2, M</td>
<td>12 failure to thrive</td>
<td>numbness in upper extremities</td>
<td>C-1</td>
<td>cervical</td>
</tr>
<tr>
<td>9</td>
<td>15, M</td>
<td>1 occipital headache</td>
<td>back pain</td>
<td>C-1</td>
<td>none</td>
</tr>
<tr>
<td>10</td>
<td>2, M</td>
<td>6 failure to thrive</td>
<td>retroocular pain</td>
<td>C-2</td>
<td>none</td>
</tr>
<tr>
<td>11</td>
<td>4, M</td>
<td>1 occipital headache</td>
<td>weakness in lower extremities</td>
<td>C-1</td>
<td>cervical</td>
</tr>
<tr>
<td>12</td>
<td>4, M</td>
<td>2 weakness in upper extremities</td>
<td>retroocular pain</td>
<td>C-2</td>
<td>none</td>
</tr>
<tr>
<td>13</td>
<td>12, F</td>
<td>50 headache</td>
<td>tinnitus</td>
<td>C-2</td>
<td>none</td>
</tr>
<tr>
<td>14</td>
<td>17, F</td>
<td>6 occipital headache</td>
<td>retroocular pain</td>
<td>C-2</td>
<td>none</td>
</tr>
<tr>
<td>15</td>
<td>15, F</td>
<td>6 occipital headache</td>
<td>numbness in upper extremities</td>
<td>C-2</td>
<td>none</td>
</tr>
</tbody>
</table>

* Length of preoperative symptoms.

Surgical Technique

The patient was placed prone, the head immobilized with a three-pin headholder, and the posterior occipital area was prepared for surgery following a standard surgical technique. A midline incision extending from the inferior portion of the occiput down to the level of C-2 was performed. The skin was retracted laterally. Dissection progressed along the midline, and the occiput and the arch of C-1 were exposed. The rectus capitis posterior minor muscle was removed bilaterally from the tubercle of the posterior lamina of C-1, and the posterior atlantooccipital ligament was sectioned. The C-1 lamina was retracted caudally with fishhook clamps. With the aid of microscopic magnification, we performed a cruciform incision with the midline component extending from the edges of the foramen magnum to the superior edge of the C-1 lamina, and a bilateral incision of 1 cm at each side of the midline. Immediately, we were able to visualize the underlying cerebellar tonsils.

**TABLE 2**

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Postop Clinical Status</th>
<th>Postop Syringomyelia</th>
<th>Follow Up (mos)</th>
<th>Pathological Findings in Tonsils†</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>improved</td>
<td>decreased</td>
<td>5</td>
<td>NA</td>
</tr>
<tr>
<td>2</td>
<td>improved</td>
<td>disappeared</td>
<td>6</td>
<td>atrophic</td>
</tr>
<tr>
<td>3</td>
<td>improved</td>
<td>decreased</td>
<td>7</td>
<td>NA</td>
</tr>
<tr>
<td>4</td>
<td>improved</td>
<td>decreased</td>
<td>8</td>
<td>NA</td>
</tr>
<tr>
<td>5</td>
<td>improved</td>
<td>decreased</td>
<td>8</td>
<td>gliotic</td>
</tr>
<tr>
<td>6</td>
<td>improved</td>
<td>decreased</td>
<td>9</td>
<td>NA</td>
</tr>
<tr>
<td>7</td>
<td>improved</td>
<td>decreased</td>
<td>20</td>
<td>NA</td>
</tr>
<tr>
<td>8</td>
<td>improved</td>
<td>decreased</td>
<td>3</td>
<td>sclerotic</td>
</tr>
<tr>
<td>9</td>
<td>persisting headaches</td>
<td>NA</td>
<td>3</td>
<td>sclerotic</td>
</tr>
<tr>
<td>10</td>
<td>improved</td>
<td>NA</td>
<td>5</td>
<td>sclerotic</td>
</tr>
<tr>
<td>11</td>
<td>improved</td>
<td>NA</td>
<td>7</td>
<td>sclerotic</td>
</tr>
<tr>
<td>12</td>
<td>improved</td>
<td>NA</td>
<td>7</td>
<td>NA</td>
</tr>
<tr>
<td>13</td>
<td>improved</td>
<td>NA</td>
<td>9</td>
<td>NA</td>
</tr>
<tr>
<td>14</td>
<td>improved</td>
<td>NA</td>
<td>4</td>
<td>NA</td>
</tr>
<tr>
<td>15</td>
<td>improved</td>
<td>NA</td>
<td>3</td>
<td>sclerotic</td>
</tr>
</tbody>
</table>

* NA = not applicable.
† Findings only appropriate in cases in which tonsils were resected.

At first we carefully used a bipolar coagulator to cauterize the pial surface of the cerebellar tonsils, aiming at complete cephalic retraction with exposure of the posterior and lateral aspects of the cervical cord and obex. When this objective was not achieved, the redundant cerebellar tonsils were resected.

After having decompressed the cervical canal and the obex, we proceeded to close the wound by using a synthetic dura mater substitute, sized according to the dural opening. This graft was sutured to the surrounding dura by using No. 5-0 nylon thread in continuous stitches. We further sealed the incision between the dura mater and the patch by using fibrin glue. After that, we proceeded to close the muscle, nuchal ligament, and subcutaneous tissue with absorbable sutures and the skin with nylon thread. All patients were extubated while in the operating room and then transferred to the recovery room. The estimated blood loss was negligible in all patients. The resected cerebellar tonsils were sent to the neuropathology laboratory. The length of the surgical procedure ranged from 2 to 3 hours, being shorter in younger patients.

Results

Outcomes of the patients are summarized in Table 2. None of the patients experienced intraoperative complications. The mean postoperative hospital stay was 3.6 days, with 1 day spent in the neurosurgical intensive care unit and 2 days in the ward.

In no cases did we observe thick arachnoid adhesions or other impediments to manipulation of the tonsils. In seven cases the cerebellar tonsils were resected, and in the remaining eight patients coagulation of the pia mater was sufficient to retract the herniated cerebellar tissue from the cervical canal. Among the seven patients whose tonsils were resected, in three the tonsils were found to have descended down to the level of C-2. In contrast, in four of the eight patients in whom pial coagulation was sufficient, the cerebellar tonsils reached the posterior arch of C-2 (Fig. 1).

Headaches improved within the 1st week after surgery.
in all patients except one child whose Chiari I malformation was secondary to prolonged lumboperitoneal shunting (Case 9). In this patient the headaches abated 2 months after surgery. The weight of the two children who were experiencing failure to thrive due to defective swallowing improved significantly. The preoperative stridor exhibited by one child disappeared after surgery. In this child, flexible fiberoptic nasopharyngolaryngoscopic examination revealed minimal paresis of the bilateral vocal cords during abduction, with no evidence of vocal cord paralysis after the procedure.

Motor and sensory symptoms in the upper extremities disappeared in the months following surgery. A similar evolution was observed in two children who experienced low-back pain. Syringomyelia was reduced in seven of eight patients. In six children this was evident on the first postoperative spine MR image obtained between 2 and 6 months after surgery. In one child the cervical syrinx was noticeably reduced 20 months after surgery (Fig. 2). In one child the syrinx had not decreased in size at the 4-month follow-up MR imaging examination. Nonetheless, in this patient the upper-extremity weakness had resolved, and this was evidenced by the rapid improvement in his penmanship. One patient presented 1 week after surgery with a CSF leak through the operative wound, which required a reoperation to repair an imperfectly closed dural patch.

Neuropathological examination of the resected tonsils was performed in seven patients. Histopathological studies disclosed fragments of cerebellar tissue with partial loss of Purkinje cells and overlying dense fibrous connective tissue in all cases. In three cases histological sections were found to contain portions of mildly fibrotic leptomeninges. The granular cell layer was preserved overall, but focally thinned in three cases. The molecular cell layers were gliotic with occasional Rosenthal fibers in four cases. The Purkinje cells were sparse and atrophic in all cases.

Postoperative cine phase–differential mode MR imaging was performed in five patients, two of whom had undergone tonsillar resection and three of whom had undergone tonsillar shrinkage. In all studies a bidirectional pulsatile flow of CSF was observed around the basal cisterns, anterior to the spinal cord and posteriorly in the region of the foramen of Magendie. Only one patient, whose MR image is shown in Fig. 1A, underwent a preoperative CSF flow study. The flow was reported to be normal anterior to the brainstem, but no flow was observed in the posterior cerebellum or in the spinal canal.

Discussion

The surgical technique that we have described selectively removes the herniated cerebellar tonsils without the need to perform an occipital craniectomy or a C-1 laminectomy. Initially discredited by Gardner and Angel, resection of the cerebellar tonsils was shown to be safe by Bertrand and Williams, who emphasized its usefulness for restoring the CSF pathway. Since then, removal of tonsils after occipital craniotomy has been considered to be a reasonable approach to treat Chiari I malformation. In a recent survey among pediatric neurosurgeons, 31% favored performing tonsillar manipulation.
Tonsillectomy without craniectomy for Chiari I malformation

Because the herniated cerebellar tonsils seem to be responsible for the great majority of symptoms of Chiari I malformation, their removal from the cervical canal was the single goal of the procedure that we performed. In our series we resected or shrank the tonsils in an almost equal number of patients. Some authors choose to opt for the former when they presume that the tonsils are gliotic; we were not able to differentiate between gliotic and normal cerebellar tissue in this series.

The tonsils are neocerebellar structures located in the inferior pole of the posterior lobe of the cerebellum and are connected to the cerebellum by the tonsillar peduncle, but so far no specific function has been attributed to them. Their downward displacement is considered to be a hallmark of Chiari malformation. It is presumed that this downward displacement is caused by a normally developed cerebellum that exceeds the capacity of the underdeveloped posterior fossa. In addition, different pressure gradients between the supratentorial and infratentorial compartments are causative factors in the caudal displacement of the cerebellar tonsils. The histopathological examination of resected tonsils provided further support to their role as a compressing factor. We noted that most of them appeared to have a sclerotic or an atrophic pattern. To our knowledge, only a few reports exist in which the histopathology of tonsils in Chiari I malformation are described. The displaced cerebellar tissue may exhibit some disorganization of normal lamination with disoriented or heterotopic Purkinje cells or commonly, loss of Purkinje, granule, and basket cells and extensive gliosis of the cerebellar folia. Such gliotic tissue may be barely recognizable cerebellar tonsils and by itself does not provide evidence of dysplasia. It can be hypothesized that this is correlated to the compression exerted between the brainstem and the bone rim of the foramen magnum and dural transverse bands, with a compromised vascular supply that potentially causes cellular loss, shrinkage, and gliosis.15

Among our patients were children with congenital hydrocephalus and children in whom the cerebellum sagged following prolonged lumboperitoneal shunting; thus it can be argued that our group did not represent the classic cohort of patients with Chiari I malformation. Nonetheless, there was radiological and clinical evidence compatible with this malformation in these patients and, although their symptoms did not improve after shunt placement or removal of the lumboperitoneal systems, their symptoms were relieved after the cerebellar tonsils had been removed from the upper cervical canal.

Some of our patients presented with occipital headaches that were most likely the consequence of direct compression of the first cervical nerve by caudally displaced cerebellar tonsils. Three of our patients presented with severe symptoms such as sleep apnea, hoarseness, and swallowing difficulties—clinical conditions that can be attributed to lower brainstem compression. Conversely, some patients complained of retroocular pain—a symptom that could be attributed to stretching of the inferior portion of the tentorium by the crammed cerebellum. After surgery all patients experienced relief from their symptoms. This was achieved either by directly decreasing pressure to nervous structures or by improving compliance of the CSF pathways in the posterior fossa. There was one child whose preoperative stridor disappeared after selective tonsillectomy, but who still displayed a partial paresis of the vocal cords. This may represent either incomplete decompression or structural damage to the brainstem nuclei similar to the one reported in patients with Chiari II malformations. The postoperative complication that we observed can be attributed to a deficient surgical technique, although the narrowness of the space between the occiput and C-1 has to be taken into consideration.

The association between descended tonsils and the development of syringomyelia has been extensively discussed in the medical literature. Cardiac-gated cine-mode MR images confirm that CSF flow is abnormal at the level of the foramen magnum in patients with Chiari I malformations. The descended cerebellar tonsils may induce abnormal CSF flow into the spinal cord along the Virchow–Robin space. Oldfield, et al., have proposed that systolic pressure waves in the spinal cord, in the presence of tonsillar impaction, may force CSF into the cord along perivascular and interstitial spaces. That the syrinx volume decreased in all but one of our patients adds to the significance of the effect that descended tonsils have on obstructing CSF flow and, by extension, on the cause of the syringomyelia. In one patient, although syrinx volume was not reduced, his upper-extremity deficits were noticeably decreased.

We acknowledge that a shortcoming of our paper is, with the exception of one patient, the lack of preoperative CSF flow studies. We can only rely on the decrease in the volume of the syrinx to presume that, after removal of the tonsils, CSF flow in the posterior fossa and in the cervicomedullary junction has improved.

Conclusions

Either tonsillectomy or shrinkage through the atlanto-
to occipital interspace allows the removal of a significant pathological factor associated with Chiari I malformation without altering the bone anatomy of the region, thus preventing putative complications such as cerebellar ptosis and spinal deformity. This new selective surgical technique has yielded good preliminary results in this limited series.

References


Manuscript received June 15, 2001. Accepted in final form June 18, 2002.
Address reprint requests to: Jorge A. Lazareff, M.D., Division of Neurosurgery, University of California at Los Angeles Medical School, Box 957039, Los Angeles, California 90095-7039. email: jlazareff@mednet.ucla.edu.