Analysis of the Posterior Fossa in Children with the
Chiari 0 Malformation

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OBJECTIVE: We previously reported the resolution of syringohydromyelia without cerebellar tonsillar ectopia in five
patients after posterior fossa decompression of the so-called Chiari 0 malformation. A sixth patient is described.
In this study, the anatomy of the posterior fossa is analyzed using radiological imaging, enabling features of the
posterior fossa in this uncommon subgroup of children to be characterized.

METHODS: Multiple measurements were made on magnetic resonance imaging studies in six children with Chiari
0 malformation to determine the position of the brainstem relative to the foramen magnum. Fifty children with
normal magnetic resonance imaging studies of the brain were used as controls.

RESULTS: All children with a Chiari 0 malformation were found to have the following positive results: obices that
were located more than 2 standard deviations below normal, an increase in the anteroposterior midsagittal
distance of the spinomedullary junction at the level of the foramen magnum, an increase in the angle between
the floor of the fourth ventricle and clivus, and an increase in the anteroposterior midsagittal distance of the
foramen magnum.

CONCLUSION: The findings of this study suggest that the contents of the posterior fossa are indeed compromised
and/or distorted in patients with syringohydromyelia but no tonsillar ectopia. In this group, the brainstem was
caudally displaced more than 3 standard deviations below normal. (Neurosurgery 48:1050–1055, 2001)

Key words: Chiari malformation, Posterior fossa, Syrinx, Tonsillar ectopia

The resolution of syringohydromyelia in children with-
out cerebellar tonsillar ectopia after posterior fossa de-
compression—the so-called Chiari 0 malformation—
was described previously by our group (4, 13, 20). In the
1890s, Hans Chiari described four cerebellar abnormalities
that were later termed Chiari malformations I, II, III, and IV
(2, 3). Many authors have attributed the Chiari I malformation
to overcrowding of the contents of the posterior fossa because
of a small posterior fossa (1, 11, 14, 17). In some patients with
Chiari I malformation, however, the posterior fossa is of nor-
mal dimensions (16, 19). This study analyzes various radi-
ological measurements of the posterior fossa and cranioce-
rvical junction in 50 children whose brains were deemed normal on
the basis of magnetic resonance imaging (MRI) studies. These
data are compared with measurements found in a population
of six children with Chiari 0 malformation. The goal of this
study was to determine whether the posterior fossa is com-
promised and/or distorted, and because of this goal, appro-
priate drainage of the fourth ventricle was restricted in a
group of children with no tonsillar ectopia but with resolution
of the syrinx after posterior fossa decompression. Also, we
attempted to develop additional radiological criteria to assist
the neurosurgeon in evaluating patients with syringohydro-
myelia without herniated tonsils to strengthen the preopera-
tive assumption that posterior fossa decompression will ade-
quately resolve the patient’s symptoms.

PATIENTS AND METHODS

This study retrospectively analyzed 50 children (30 boys
and 20 girls) ranging in age from 2 months to 17 years (mean,
6.9 yr; median, 7 yr) with normal brain imaging on 1.5-T MRI
scanning (General Electric Medical Systems, Milwaukee, WI).
The measurements in this group were compared with corre-
sp ending measurements obtained by using the same MRI
scanner preoperatively in pediatric patients with no tonsillar
Chiari 0 Analysis

RESULTS

The results of the measurements for the six patients with Chiari 0 malformation are listed in Table 1. In this group, the distance from the basion to the opisthion ranged from 30 to 40 mm (mean, 34 mm). The tip of the obex was found to be at the level of the foramen magnum in two patients, 2.0 mm superior to the foramen magnum in two patients, 1.0 mm superior to the foramen magnum in one patient, and 9.0 mm inferior to the foramen magnum in one patient. The midsagittal horizontal distance of the spinomedullary junction at a midpoint on a line connecting the basion to the opisthion ranged from 23 to 35 degrees (mean, 28 degrees). At approximately 115 to 118 degrees, the clival angle was believed to be within normal limits in all six patients (7). The distance from the fastigium to the floor of the fourth ventricle ranged from 9.0 to 11.0 mm (mean, 10 mm). The horizontal distance from the sphenop-occipital synchondrosis to the basis pontis was measured to further evaluate for anterior and/or inferior displacement of the brainstem (Fig. 1).

FIGURE 1. Drawing of the midsagittal region of the cranio cervical junction. a, the distance between the basion and the opisthion; b, the anteroposterior distance of the spinomedullary junction in the horizontal plane on a midpoint of a; c, the relationship of the obex to the plane of the foramen magnum (from a midpoint of a); e, the orthogonal distance between the fastigium and the floor of the fourth ventricle; f, the orthogonal distance between the sphenop-occipital synchondrosis and the basis pontis.

FIGURE 2. Drawing of the midsagittal region of the cranio cervical junction showing the angle between intersecting lines drawn between the clivus and the floor of the fourth ventricle (d).
be within normal limits. None of our patients had basilar invagination. One patient had midline bone defects of the posterior arches of C1 and C2, and one had assimilation of the posterior arch of C1 on the right. No other obvious bony anomalies of the posterior fossa or upper cervical spine were noted. No statistically significant differences in sex or age were noted in the patients in this group (P < 0.05).

The results for the 50 pediatric patients with normal MRI scanning of the brain are listed in Table 2. In this group, the distance from the basion to the opisthion in the sagittal plane ranged from 21 to 40 mm (mean, 28.6 mm). The midsagittal horizontal distance of the spinomedullary junction at a midpoint on a line connecting the basion to the opisthion ranged from 4.0 to 14 mm (mean, 9.5 mm). The angle created by intersecting lines of the clivus and floor of the fourth ventricle ranged from 5.0 to 32 degrees (mean, 13.8 degrees). The clival angle was believed to be within normal limits in all 50 patients. No bony anomalies of the posterior fossa or upper cervical spine were noted. No statistically significant differences were noted between the sexes among the patients in this group (P < 0.05), nor were any significant differences found when groups of similar age were examined.

**DISCUSSION**

We previously reported the resolution of syringohydromyelia in five patients with posterior fossa decompression but absence of cerebellar tonsillar ectopia (4, 6). In those patients, clinical and radiological improvement with presumably idiopathic syringohydromyelia was documented. The patients continue to have good clinical results after an average follow-up of approximately 4 years. We have added a patient with this clinical presentation who has been followed for 1 year. In two of the original five patients, a surgical observation was made that the posterior fossa appeared to be "crowded" at the level of the foramen magnum. Of interest, all three patients described in the original article who underwent both preoperative and postoperative cine MRI demonstrated decreased flow preoperatively, and two of the three demonstrated improved flow postoperatively (6). In a study of syringohydromyelia conducted before the availability of computed tomography or MRI, Newton (12) described drainage anomalies of the fourth ventricle in which some patients with syringohydromyelia were without hindbrain herniation. These patients were treated with posterior fossa decompression and drainage of the syrinx, with good results (12).

The measurements obtained in our study were chosen to directly and indirectly assess the bony compactness of the posterior fossa in and around the level of the foramen magnum and to determine whether the brainstem may have been displaced on midsagittal MRI scans. Although linear and nonvolumetric, these measurements on sagittal MRI scans may assist the neurosurgeon in evaluating patients with syringohydromyelia and the absence of tonsillar ectopia. First, by measuring the midline anteroposterior distance of the foramen magnum in the sagittal plane, a determination can be made regarding whether the outlet of the posterior fossa was compromised. Next, by assessing the anteroposterior distance of the spinomedullary junction at the foramen magnum, the relationship of the amount of soft tissue to bony encasement at the level of the foramen magnum can be determined. The normal range of the diameter of the spinal cord at C1 is 7.0 to 11.0 mm (7). Measurement of the angle between the clivus and floor of the fourth ventricle, the distance

**TABLE 1. Measurements in the Chiari 0 Group**

<table>
<thead>
<tr>
<th>Patient No.</th>
<th>Age (yr)</th>
<th>a (mm)</th>
<th>b (mm)</th>
<th>c (mm)</th>
<th>d (degrees)</th>
<th>e (mm)</th>
<th>f (mm)</th>
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</thead>
<tbody>
<tr>
<td>1</td>
<td>9/F</td>
<td>40.0</td>
<td>15.0</td>
<td>0</td>
<td>24.0</td>
<td>9.0</td>
<td>5.0</td>
</tr>
<tr>
<td>2</td>
<td>13/F</td>
<td>31.0</td>
<td>12.0</td>
<td>2.0 sup</td>
<td>24.0</td>
<td>10.0</td>
<td>4.0</td>
</tr>
<tr>
<td>3</td>
<td>16/M</td>
<td>30.0</td>
<td>14.0</td>
<td>0</td>
<td>35.0</td>
<td>9.0</td>
<td>4.0</td>
</tr>
<tr>
<td>4</td>
<td>10/M</td>
<td>35.0</td>
<td>14.0</td>
<td>2.0 sup</td>
<td>30.0</td>
<td>10.0</td>
<td>6.0</td>
</tr>
<tr>
<td>5</td>
<td>3/F</td>
<td>30.0</td>
<td>12.0</td>
<td>9.0 inf</td>
<td>23.0</td>
<td>11.0</td>
<td>5.0</td>
</tr>
<tr>
<td>6</td>
<td>13/F</td>
<td>31.0</td>
<td>13.0</td>
<td>1.0 sup</td>
<td>35.0</td>
<td>10.0</td>
<td>5.0</td>
</tr>
</tbody>
</table>

**TABLE 2. Measurements of Control Group**

<table>
<thead>
<tr>
<th>Group</th>
<th>Age (yr)</th>
<th>a (mm)</th>
<th>b (mm)</th>
<th>c (mm)</th>
<th>d (degrees)</th>
<th>e (mm)</th>
<th>f (mm)</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>2–4</td>
<td>22.0–40.0</td>
<td>4.0–10.0</td>
<td>8.5–17.0</td>
<td>5.0–25.0</td>
<td>8.0–15.0</td>
<td>2.0–8.0</td>
</tr>
<tr>
<td>2</td>
<td>8–10</td>
<td>21.0–33.0</td>
<td>6.0–14.0</td>
<td>8.5–16.5</td>
<td>5.0–20.0</td>
<td>6.0–12.0</td>
<td>5.0–10.0</td>
</tr>
<tr>
<td>3</td>
<td>12–14</td>
<td>30.0–35.0</td>
<td>9.0–12.0</td>
<td>7.5–16.0</td>
<td>6.0–32.0</td>
<td>6.0–12.0</td>
<td>4.0–12.0</td>
</tr>
<tr>
<td>4</td>
<td>15–17</td>
<td>28.0–33.0</td>
<td>9.0–12.0</td>
<td>8.0–17.0</td>
<td>12.0–20.0</td>
<td>11.0–19.0</td>
<td>4.0–10.0</td>
</tr>
</tbody>
</table>

* Values are ranges (means) for associated ages at presentation (±1 yr) of the Chiari 0 group; a, distance from basion to opisthion; b, midsagittal horizontal distance of the spinomedullary junction at the midpoint of “a”; c, the height of the obex superior to the plane “e” of the foramen magnum; d, the angle (in degrees) created between intersecting lines between the clivus and floor of the fourth ventricle; e, the orthogonal distance between the fastigium and the floor of the fourth ventricle; f, the horizontal distance between the sphenoid-occipital synchondrosis and the basis pontis.
from the fastigium of the fourth ventricle to the floor of the fourth ventricle, and the distance from the sphen-o-occipital synchondrosis to the basis pontis allowed us to ascertain whether the fourth ventricle and/or the brainstem were displaced anteroinferiorly, considering that all clival angles were found to be within normal limits.

The results of our study suggest that the bony posterior fossa in patients with a Chiari 0 malformation may be distorted at the level of the foramen magnum as compared with a control group. This finding held true when comparing data from people of all ages with our group of six patients and also when comparing the patients of different ages in our group with children of the same age in the control group. Our measurements demonstrated that in all six patients with this entity, the sagittal anteroposterior distance of the spinomedullary junction is increased, implying caudal displacement of the more rotund medulla oblongata. This was statistically different when compared with our control individuals \( (P = 0.00121) \). The concept of caudal descent of the brainstem is further supported by the finding that, in all patients in this group, the tip of the obex was more than 3 standard deviations below the normal position; this migration implies a compromised posterior fossa in these patients \( (10, 15) \). The Chiari I malformation occasionally has been associated with caudal migration of the brainstem and is sometimes termed the Chiari 1.5 malformation \( (5, 13, 14, 20) \). The anteroposterior distance of the foramen magnum in our group of patients was significantly larger \( (P = 0.00458) \) \( (7, 15) \). Of interest, patients with Chiari I malformation have been described as often having a smaller than normal foramen magnum \( (13, 15) \). The angle between the clivus and the floor of the fourth ventricle had statistically significant differences \( (P = 0.00001) \), with means of 14 degrees for the control group and 28 degrees for the Chiari 0 group. This implies some degree of posterior tilt to the pons and medulla, because the clival angles of the Chiari 0 group were believed to be within normal limits. In addition, we found no instance in which the spinomedullary junction appeared kinked, nor did we find any retroversion of the odontoid process, both of which would most likely decrease this angle. The distance between the sphen-o-occipital synchondrosis and basis pontis and the orthogonal distance between the fastigium and floor of the fourth ventricle were not significantly different between the Chiari 0 group and the control group \( (P = 0.46126 \) and \( P = 0.18149, \) respectively). This suggests that, in patients with Chiari 0 malformation, the contents of the posterior fossa compressed caudally, and a laterally compromised posterior fossa would help explain the increase in the basion-to-opisthion distance as well as the increased angles noted between the clivus and the floor of the fourth ventricle.

Many authors have discussed the findings that the posterior fossa volume tends to be decreased and more shallow in the Chiari I population \( (1, 8, 9, 11, 17, 18) \). In this infrequently encountered group of patients with syringohydromyelia and without tonsillar ectopia, the measurements demonstrated that the posterior fossa revealed signs of being compromised. Our assumption is that, in this rare group of patients, compactness at the foramen magnum disturbs the normal circulation of cerebrospinal fluid (CSF), thereby causing syringohydromyelia. This mechanism of restriction of CSF flow is the same cause of the syringohydromyelia observed in many patients with a Chiari I malformation. Of note, the cerebellar tonsils are paramedian structures and may have been minimally caudally displaced. This displacement was not appreciated on our midsagittal evaluations. Also of interest is that the angle between the clivus and the floor of the fourth ventricle was significantly increased in our Chiari 0 group. This is a curious observation because all clival angles were within normal limits, as was the prepontine space in each patient \( (6) \). Although this study did not evaluate posterior fossa volumes, the radiological measurements imply that, in this select group, the posterior fossa is smaller than normal. We now have a protocol for MRI analysis of prospective patients with the Chiari 0 malformation by which to compare the posterior fossa volumes and ratios with those of patients with Chiari I malformation.

**CONCLUSION**

We have demonstrated that the contents of the posterior fossa demonstrate signs of being compromised in patients described as having a Chiari 0 malformation. These findings indicate that syringohydromyelia may be a result of a smaller than normal posterior fossa’s compromising normal CSF egress out of the cranium and that actual cerebellar tonsillar herniation is not essential to disturb the delicate balance that is involved with CSF circulation. Although limited by not having volumetric measurements, abnormal measurements of the variety we obtained on midsagittal MRI scanning may assist the neurosurgeon to choose patients more confidently for posterior fossa decompression when a syrinx is present without cerebellar ectopia. Findings of particular note include the following: obices that were abnormally descended when measured from a midpoint on a midsagittal line connecting the basion to the opisthion, an increase in the midsagittal horizontal anteroposterior distance of the spinomedullary junction on the line described above, an increase in the angle between intersecting lines of the clivus and the floor of the fourth ventricle, and an increase in anteroposterior dimensions of the foramen magnum. It is hoped that these findings will add to current knowledge regarding hindbrain herniation and syringohydromyelia and perhaps lead to new nomenclature aimed not at degrees of tonsillar herniation in the currently termed Chiari I malformation but rather at capaciousness at the foramen magnum.

**ACKNOWLEDGMENT**

We thank Bermans J. Iskandar, M.D., for his pioneering findings and writings on the topic of the Chiari 0 malformation.

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**REFERENCES**

15. Quisling RG, Quisling SG, Mickle JP: Obex/nucleus gracilis po-

COMMENTS

Dr. Oakes and his colleagues previously reported the successful treatment of syringohydromyelia without Chiari malformation by posterior fossa decompression (1). Five of the six children in this study were included in that publication. At the time of operation, the surgeon had the impression of “crowding” of posterior fossa structures at the foramen magnum, although a true Chiari malformation could not be identified. This article is an attempt to document quantitatively whether evidence of a small posterior fossa and/or caudal displacement of the brainstem is present in these patients. Their findings support that hypothesis. Volumetric analysis of the posterior fossa and its structures would be a better way to determine whether the posterior fossa is too small. It is hoped that this type of analysis can be done in the future.

The authors suggest that such measurements may be helpful in deciding whether to perform posterior fossa decompression on a particular patient. This assumes that another group of children with syringohydromyelia exists without the changes described. Such patients would not benefit from posterior fossa decompression. Until this can be determined, I suspect that the presence of syringohydromyelia per se will dictate the decision for surgery. In my experience, only a fraction of the patients examined for evaluation of a Chiari I malformation have symptoms relevant to that finding. I am a bit unsettled by the prospect of contending with an even larger group of patients with nondescriptive symptoms such as headache or dizziness whose anthropomorphic measurements suggest a need for a posterior fossa decompression. Finally, I agree with the authors’ concluding comment that a better term is needed to describe this phenomenon. Because of the euphony of the term and the ease with which it can be related to a recognized entity, however, I am afraid that the term will stick.

Paul H. Chapman
Boston, Massachusetts


It is sometimes difficult not to be a skeptic with regard to the many draftsperson’s lines sketched through the cranium, particularly as they apply to structures within the posterior cranial fossa. How does one reconcile fixed cranial measurements with those of the contained “pliable” brain tissue, and at which point, if any, might the respective measurements negate each other?

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intent is to “assist the neurosurgeon” when evaluating patients who have syringohydromyelia in the absence of cerebellar tonsillar ectopia.

Following up on this, the posterior fossa is defined as being compromised or distorted, which brings about tissue shifts and the caudal migration of the brainstem. This is the essential message. In the end, it may be that pure geometry is correct. Certainly, one cannot argue with success (i.e., the resolution of clinical symptoms after posterior fossa decompression in patients with the Chiari 0 malformation).

Robin P. Humphreys
Toronto, Ontario, Canada

The authors of this article previously reported five patients with hydrosyringomyelia but no cerebellar tonsil displacement beyond the level of the foramen magnum—the so-called Chiari 0 malformation—that resolved after a craniocervical decompression. The authors have added a sixth patient to this group, all of whom had multiple measurements obtained from imaging studies to determine the existence of objective findings that could be used to guide the decision-making process with regard to whether surgical decompression would likely benefit similar patients. From their measurements, the authors determined that the brainstems of the patients who underwent decompression were caudally displaced more than 3 standard deviations below the normal level. Another finding was that the angle between the clivus and the floor of the fourth ventricle was significantly increased in the Chiari 0 group as compared with control individuals.

Because this study was retrospective, the authors did not have adequate information to determine the volume of the posterior fossa in their patients, although they had evidence that the volume was reduced. It is hoped that the authors and other investigators will obtain subsequent volume measurements to confirm this presumptive finding. It would also be of value for the patients to undergo cine magnetic resonance imaging studies to determine whether abnormal cerebrospinal fluid (CSF) flow patterns are usually or always present preoperatively, as well as their response to a craniocervical decompression.

Of interest, the authors were successful in resolving the presence of hydrosyringomyelia in this group of six patients after a craniocervical decompression procedure. With this rate of success, why worry about measurements? One wonders whether this success rate will continue. Given the alternatives for treating hydrosyringomyelia (i.e., myelotomy, various forms of CSF diversion), cervicomedullary decompression in general seems to be the best initial approach because it addresses the underlying pathophysiology of an alteration in normal CSF flow patterns.

J. Gordon McComb
Los Angeles, California

This important study is described by a distinguished group of investigators who previously reported the resolution of syringohydromyelia after posterior fossa decompression in five children lacking magnetic resonance imaging (MRI) evidence of cerebellar tonsillar ectopia. The MRI findings in these patients seemed to exclude cranial base abnormalities consistent with Chiari I malformation, although two of five patients were believed to have a “crowded” foramen magnum at the time of surgery. To emphasize the possibility that patients with idiopathic syringohydromyelia might benefit from posterior fossa decompression, the authors introduced the interesting and provocative concept of the Chiari 0 malformation.

In this study, the authors reviewed the MRI findings in the original study population and added a sixth patient. Unfortunately, none of the patients in the original report had undergone complete brain MRI before surgery, which made it impossible for the authors to discuss anatomical features of Chiari I malformation, such as a reduced volume of the posterior fossa and an increased slope of the tentorium. To address this issue, the authors developed a number of radiographic measurements to assist in establishing the position of the brainstem relative to the foramen magnum. These measurements are easy to obtain and seem to provide a reliable index of brainstem displacement. On the basis of a comparison of findings in six patients with Chiari 0 malformation and 50 children with normal MRI scans, the authors conclude that children with Chiari 0 malformation have a “compromised and/or distorted” posterior fossa and caudal displacement of the brainstem.

Although the term Chiari 0 malformation is certainly attractive, it is called into question by the demonstration of subtle but distinct hindbrain and cranial base abnormalities. Perhaps a more apt description would be “borderline Chiari I malformation characterized by low-lying cerebellar tonsils, caudal displacement of the brainstem, and underdevelopment of the posterior fossa.” The absence of tonsillar ectopia on midsagittal MRI scans may underestimate the incidence of minimal tonsillar herniation. The cerebellar tonsils are paramedian structures, and the apices of the tonsillar tips are found lateral to the midline. More work is required to fully understand the pathophysiology and clinical manifestations of this interesting condition.

Evidence has accumulated in recent years that noncommunicating syringomyelia is caused by an obstruction of the CSF circulation at or below the foramen magnum, which increases the pulsatile systolic pulse wave in the spinal subarachnoid space and drives CSF into the central canal of the spinal cord. In patients with idiopathic syringomyelia, it is advisable to obtain an exhaustive neuroradiological workup that is focused on the detection of occult CSF blocks. Cine MRI and computed tomographic myelography are often helpful in this regard. It is important that pediatric patients undergo MRI scanning of the thoracolumbar spine to rule out spinal cord tethering, which is a recognized cause of chronic tonsillar ectopia and is commonly associated with idiopathic syringomyelia, presumably on the basis of occult obstruction of CSF flow at the level of the foramen magnum. The authors have revised their original conclusions and expanded our understanding of the Chiari 0 phenomenon.

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