A critical analysis of the Chiari 1.5 malformation

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Object. Although the traditional nomenclature used to describe hindbrain hernias is useful, there are certainly patients in whom these morphological entities do not strictly apply. One such group is composed of patients with the more recently described Chiari 1.5 malformation in which a Chiari I malformation is seen in combination with brainstem herniation through the foramen magnum. In an attempt to elucidate further the best surgical strategy and to refine the descriptive terminology, the authors retrospectively analyzed all cases at their institutions in which this form of hindbrain herniation was diagnosed.

Methods. The authors reviewed the database for all patients in whom Chiari 1.5 malformation had been diagnosed. Each patient had undergone a posterior fossa decompressive surgery. Magnetic resonance images were evaluated for the extent of caudal descent of the brainstem, amount of tonsillar ectopia, inclination of the odontoid process, and any brain or brainstem abnormalities. Clinical presentations and postoperative results were correlated to the aforementioned radiological findings.

Twenty-two patients were identified. The obex was a mean 14.4 mm inferior to the foramen magnum, and the medulla exhibited a flattened appearance in the mid-sagittal plane in all patients. Syringomyelia was documented in 50% of the cases. The cerebellar tonsils were found to lie at C-1 and C-2 in nine and 13 patients, respectively. The mean angulation of the odontoid process was 84.4°. No abnormalities or caudal descent of the midbrain or pons was identified. Eighteen patients experienced resolution of preoperative symptoms. Persistence of syringomyelia prompted a second posterior fossa operation secondary to progressive scoliosis in 13.6% of the patients.

Conclusions. No single sign or symptom was found to be peculiar to the Chiari 1.5 malformation, although all patients in whom this diagnosis was established had undergone a posterior fossa decompressive surgery. A significant number (13.6%) of patients required repeated operation for persistent syringomyelia. Neurosurgeons may wish to consider that many patients may not respond as well to posterior fossa decompressive surgery especially if syringomyelia is present.

KEY WORDS • Chiari malformation • hindbrain hernia • tonsillar ectopia • posterior fossa • pediatric neurosurgery

CHIARI’S original description of the four pathological entities that now bear his name involved structures of the posterior cranial fossa. Three of these (Types I–III) involved herniation of rhombencephalic derivatives out of the posterior fossa. The fourth variety (Type IV) that did not involve rhombencephalic ectopia through the foramen magnum was described as cerebellar hypoplasia or aplasia. Although the traditional nomenclature (Chiari I–III malformation) is useful in defining various forms of hindbrain herniation, there are certainly patients in whom these morphological entities do not strictly apply. One such group is composed of patients with the more recently described Chiari 1.5 malformation. The essential difference between this cohort and patients with the traditional Chiari I malformation is that in addition to tonsillar ectopia patients with Chiari 1.5 malformation also exhibit caudal descent of the brainstem. In an attempt to further elucidate the best surgical strategy and refine the descriptive terminology, we retrospectively analyzed data obtained in all patients at our institutions with a diagnosis of this form of hindbrain herniation.

Clinical Material and Methods

The database comprising detail of all patients with Chiari 1.5 malformation treated at Children’s Hospital (Birmingham, AL [20 cases]) and Pediatric Neurosurgery (University of Wisconsin, Madison, WI [2 cases]) was reviewed. Magnetic resonance imaging data were collected in all cases in which tonsillar ectopia was greater than 5 mm. Each patient underwent evaluation to determine the degree of caudal descent of the brainstem, as exhibited by an inferiorly...
displaced obex beneath the foramen magnum (basion-opisthion line), the extent of tonsillar ectopia, inclination of the odontoid process, and any brainstem abnormalities. Moreover, clinical presentation and postoperative data were correlated to the aforementioned neuroimaging findings. No patient had a history of lumbar puncture or other intervention that violated the distal subarachnoid space. History of intracranial lesions, craniosynostosis, meningitis or encephalitis, violation of the subarachnoid space, or head trauma was absent in all cases. One observer (R.S.T.) made all calculations to avoid interobserver error, which can increase the coefficient of error to greater than 5%. Statistical analyses were performed using commercially available software (version 8.0; SPSS, Inc., Chicago, IL) for Windows.

Results

Twenty-two patients with the diagnosis of Chiari 1.5 malformation were identified. There were seven male and 15 female patients who ranged in age from 3 to 20 years (mean 12.5 years). Additional diagnoses included neurofibromatosis Type 1, asthma, pericallosal lipoma, basilar invagination, pseudotumor cerebri, cystic hygroma, and a history of atlantooccipital fusion. The angulation of the odontoid process ranged from 65 to 100° (mean 84.4° ± 9.3 [standard deviation]) (Fig. 1). The distance of the obex beneath the foramen magnum ranged from 9 to 20 mm (mean 14.4 ± 3.96 mm) (Fig. 1 center). The anteroposterior width of the medulla oblongata (< 15 mm in all patients) resulted in a flattened appearance in the middorsal region. In 11 patients (50%) syringomyelia was demonstrated: six with a holocord syrinx, two with a cervical syrinx, one with a thoracic syrinx, and two with a cervicothoracic syrinx. Scoliosis was documented in six patients. In one patient we observed both basilar invagination and assimilation of the atlas to the occipital bone. The tonsils were found to lie under the posterior aspect of C-1 and C-2 in nine and 13 patients, respectively. Caudal descent of the cerebellar tonsils ranged from 7 to 23 mm (mean 12.7 ± 4.62 mm) (Fig. 1 lower). All tonsils bilaterally exhibited a pointed configuration. No abnormalities or caudal descent of the midbrain or pons were identified (Fig. 2). Kinking of the CMJ was demonstrated in one patient (Fig. 3). Hydrocephalus was found in one patient who underwent ventriculoperitoneal shunt placement prior to posterior fossa decompressive surgery. Mild ventriculomegaly without symptoms of raised ICP was documented in one case in which preoperative ICP monitoring revealed normal results. Symptoms for the overall included headache (seven cases), valsalva maneuver–induced headache (eight cases), dyspnea (one case), and hand weakness (one case). Other signs and symptoms include scoliosis, jaw pain, dysarthria, extremity paresthesias, opisthotonos, perioral numbness, absent gag reflex, lethargy, and drop attacks. All patients underwent posterior fossa decompressive surgery and duraplasty. Intraoperatively, two patients were found to harbor arachnoid veils that occluded the foramen of Magendie. Postoperatively 18 patients experienced resolution of preoperative symptoms. Symptoms that did not resolve following the initial operation included migrainelike headache (two cases), syringomyelia (three cases), scoliosis (two cases), and frontal headache (one case). Persistence of the aforementioned syringomyelia prompted a second posterior fossa operation because of progressive scoliosis in three patients (13.6%). Each of these patients required a second posterior fossa operation, and unilateral tonsillar coagulation was performed. In two of these patients the syringes resolved after the second operation, whereas in the third patient a significant holocord syrinx persists 1 year following her second operation. This patient’s family is contemplating surgery to insert a syringopleural shunt or a possible third posterior fossa procedure. The follow-up period for the entire group ranged from 6 months to 9 years (mean 3 years). Intraoperative anomalies, other than hindbrain hernia, observed in this group included the aforementioned arachnoid veils at the foramen of Magendie and caudal looping of the posterior inferior cerebellar artery below the foramen magnum. No surgery-related complications occurred in this group. In one girl there was a documented acquired Chiari 1.5 malformation.
Chiari 1.5 malformation

Fig. 2. Midsagittal T1-weighted MR image of the craniocervical junction and cervical spine obtained in a 12-year-old girl presenting with scoliosis and holocord syringomyelia. Note the obex (arrow) well below the foramen magnum posterior to the odontoid process and the thinned medulla oblongata.

Fig. 3. Midsagittal MR image demonstrating the CMJ kink found in one patient.

formation. She developed postviral meningitis at the age of 3 years. At this time MR imaging did not reveal any form of hindbrain herniation or hydrocephalus (Fig. 4 left). At the age of 12 years she developed valsalva maneuver–induced headache and jaw pain. Neuromaging revealed a Chiari 1.5 malformation with no hydrocephalus (Fig. 4 right). She underwent ICP monitoring that demonstrated normal parameters. Following a posterior fossa decompressive surgery, her symptoms completely resolved.

The correlation coefficient between odontoid angulation and extent of brainstem ectopia was $-0.2471 (p = 0.3477)$; that between this same odontoid angulation and the degree of tonsillar ectopia was $-0.22351 (p = 0.4322)$; and finally, that between the amount of brainstem and tonsillar ectopia was $0.63072 (p = 0.0117)$. Using principal components, a linear combination of the correlation among odontoid angulation, amount of brainstem ectopia, and amount of tonsillar ectopia accounted for approximately 59% of all variation. Thus, the only significant pairwise correlation (Pearson correlation) found was between the extent of brainstem and tonsillar ectopia. In patients with syringomyelia there was no significant ($p > 0.05$) correlation between syrinx location (cervical, thoracic, holocord) and obex or tonsillar position nor was a single symptom found to be associated with any particular extent of herniation of either of these anatomical structures. For patients with syringomyelia, retroflexion of the odontoid$^{20}$ was graded as 0, I, II, and III in one, one, six, and three patients, respectively. Of note, when data in children with Chiari 1.5 malformation and syringomyelia in the present series were compared with those in the senior author’s (W.J.O.) own large series of patients with Chiari I malformation and syringomyelia,$^{14}$ the former were approximately twofold as likely not to respond favorably to a posterior fossa decompression (that is, no resolution of their syringomyelia) ($p = 0.0001$).

Discussion

Brainstem herniation through the foramen magnum is seen in various clinicopathological conditions and is often not compatible with life. Of these conditions, some may be acquired such as with concomitant encephalitis,$^{16}$ brain tumor,$^{14}$ and cerebral edema as seen with head trauma,$^{13}$ some marathon runners,$^{1}$ and hepatic failure.$^{6}$ Samii, et al.$^{15}$ have described the pseudo–Chiari I malformation in which vertical displacement of the brain (cerebellar tonsils and pons) was attributed to cerebrospinal fluid leakage either spontaneously, iatrogenically, or due to spinal trauma. Congenital diseases that may and often do produce brainstem herniation through the foramen magnum include craniosynostosis as seen in Apert and Pfeiffer syndromes$^{4}$ and myelodysplasia (that is, the Chiari II malformation). We have previously reported on a small cohort of patients in whom syringomyelia resolved following posterior fossa decompressive surgery; no tonsillar herniation (Chiari 0 malformation) was present in any of these patients.$^{17}$ These children were all found to have caudal brainstem descent as exhibited by an inferiorly displaced obex greater than three standard deviations below the normal position.

Traditionally, the decussation of the pyramids or plane of the foramen magnum has been used as the border between spinal cord and medulla oblongata. Because the pyramidal decussations are difficult to image, however, some investigators have used the position of the obex as the landmark for the CMJ.$^{12,17}$ Quisling, et al.$^{12}$ have found that the mean obex position is 10 to 12 mm superior to the plane of the foramen magnum; however, they mistakenly defined the obex as the nucleus gracilis, which is more laterally located and not seen in true midsagittal sections.$^{10}$ To be precise, the obex is a thin band of neural tissue that extends transversely between the nuclei of the area postrema. Midsagittally, this structure forms the dorsal roof of the central canal’s opening, and its apex defines the inferior margin of the foramen of Magendie.$^{13}$ Because the obex and nucleus gracilis are located in proximity to each other, however, the findings of Quisling, et al., are not grossly inaccurate. Previously, in healthy control individuals, we found that the mean position for obex was 10.5 to 11.5 mm superior to the foramen magnum in patients 2 to 17 years of age.$^{18}$ In our present cohort, the obex was found to lie 9 to 20 mm (mean 14 ± 3.96 mm) beneath the foramen magnum. The medulla was thinned midsagittally in all patients.$^{7}$
Repeated imaging demonstrating the caudal descent of the brainstem in the Chiari II malformation. Our review of the imaging studies acquired in these patients indicated an obex that seemed to approach the foramen magnum; thus, these were not Chiari 1.5 malformations as we have defined the entity (Chiari I malformation and additional brainstem herniation through the foramen magnum).

A developmental error that results in caudal descent of the brainstem would be the most attractive mechanism by which to explain our findings in the present cohort. One patient in this cohort, however, seemed to harbor a documented “acquired” brainstem ectopia without life-threatening symptoms. Furthermore, one would expect some degree of pons deformity if there was developmental derangement of the rhombencephalon. Additionally, the cerebellar tonsils form later in development than does the vermis, which is herniated in the Chiari II malformation, implying that a potential embryological anomaly would occur after or during tonsillar formation. Embryologically, does brainstem displacement in the Chiari I malformation represent a continuum of the Chiari II malformation in the absence of myelodysplasia? Again, the answer remains unclear and debated; however, features of the Chiari II malformation have been found in a small number of cases in which myelodysplasia was absent. Interestingly, one of our study patients exhibited the CMJ kinking (German knickung) seen so often in the Chiari II malformation (Fig. 3). Why brainstem kinking was not evident in more patients is curious because all patients would be expected to harbor cervical cord denticulate ligaments, which presumably inhibit caudal migration of the brainstem in the Chiari II malformation with resultant CMJ kinking.

Clinically, and of more interest to the neurosurgeon, is whether the additional brainstem descent found in patients with Chiari 1.5 malformation changes the presentation or response to posterior fossa decompression. As already noted, all patients with this diagnosis underwent posterior fossa decompressive surgery. Intuitively, more the soft tissue is affected at the foramen magnum (that is, the more corpulent medulla compared with spinal cord in concert with the cerebellar tonsils) the greater the potential for symptoms and inappropriate CSF flow is increased in patients with simultaneous tonsillar herniation. Compared with larger groups of patients with Chiari I malformation, however, symptoms associated with Chiari 1.5 malformation are similar. We did not find that brainstem symptoms were different or more prevalent in this group compared with that composed of patients with Chiari I malformation. In our experience, the incidence of unresolved syringomyelia (13.6%) in the present study was nearly twofold that (6.9%) in the previous report after the first posterior fossa decompression. Indeed, in one patient with a Chiari 1.5 malformation a large holocord syrinx persisted even after two posterior fossa surgeries that were thought to be adequate technically at the time of each operation.

Bone anomalies in the present group included basilar invagination and assimilation of the atlas to the occipital bone. Various degrees of retroverted odontoid processes were also seen in many patients. We have reported previously that increased grades of odontoid retroflexion corresponded to obices that were more inferiorly displaced in pediatric patients with a Chiari I malformation compared with controls. Increased grades of odontoid retroversion were more likely in patients with syringomyelia. This was also the case with the present cohort in which the majority (82%) of patients with syringomyelia exhibited Grade II or III odontoid retroflexion. Retroversion of the odontoid process did not correlate with the degree of tonsillar or brainstem herniation; however, the amount of tonsillar and brainstem herniation inferior to the foramen magnum did positively correlate with each other (that is, as brainstem descent increased tonsillar ectopia increased). Furthermore, in almost every instance the obex position below the foramen magnum was distal to the ectopic cerebellar tips.

**Conclusions**

We have found no single sign or symptom that is pecu-
Chiari 1.5 malformation

liar to the Chiari 1.5 malformation. Each patient with this disorder, however, required operation. Of more interest, in considering data in the present and previously treated patients with Chiari I malformation, we found that a significant number (13.6%) required repeated operation for persistent syringomyelia. The neurosurgeon who treats these patients may wish to consider that in many cases involving symptomatic tonsillar ectopia and additional medullary descent the disorder does not respond as well to posterior fossa decompression, especially those with concomitant syringomyelia.

References