Editorial

Basioccipital hypoplasia in Chiari malformation
Type I

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The understanding of chronic tonsillar herniation (CTH), which has been called Chiari malformation Type I (CMI), is hampered to some extent by the lack of an accepted nomenclature and classification. Noudel and colleagues1 assessed the relationship between the descent of the cerebellar tonsils and dimensions of the basiocciput, specifically the length of the clivus. They studied 17 patients who had symptoms and signs related to the descent of the cerebellar tonsils more than 5 mm below the foramen magnum. These patients were selected because they had no obvious causative factors that would otherwise explain their tonsillar herniation such as basilar invagination or multisutural craniosynostosis. The authors referred to these patients as having “classic” CMI.

While training in neurosurgery at Case Western Reserve University in Cleveland, Ohio, my thinking regarding the Chiari I or adult Chiari malformation was greatly influenced by the work of our 2 neuropathologists, Drs. Uros Roessmann and Reinhart Friede.2 They studied all cases of CTH that had been published to that point. They postulated that all of the cases of herniated cerebellar tonsils were related to distortion of the brainstem and cerebellum at the foramen magnum from multiple causes: brain distortion from above, as in the case of multiple suture closure in syndromic craniosynostosis;3 suctioning of the brain from below, as in response to lumboperitoneal shunting;4 or direct distortion by bony abnormalities of the craniovertebral junction, as in basilar invagination.5

Based on their influence, I have always thought that tonsillar herniation has a specific cause and that with further study we would be able to define the cause in any situation. To this point the cause of more than one-half of the cases of CMI remains unknown. The work presented by Noudel and colleagues is likely to lead to an understanding of the cause of any tonsillar herniation. The patients were all symptomatic, and all had a herniation of at least 5 mm. There is no description of the shape of the tonsils, which I have found to be related to symptoms. If substantiated by others, this finding could improve our understanding of the mechanism underlying herniation in such patients and lead to an objective measurement to guide treatment and ensure that each investigator is actually studying the same phenomena.

Novegno and colleagues6 have studied the natural history of hindbrain herniation and have proposed that the name be changed to “Chiari Type I anomaly.” In general, I favor this term for defined causes that are not clearly malformative such as lumboperitoneal shunting. I would like to see objective measurements of clival length in asymptomatic or minimally symptomatic patients to determine whether this parameter offers a prognostic benefit.

References

Response

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We thank Dr. Rekate for the very thoughtful comments and perspective on the results of our study.

Concerning the semantic discussion about CMI, we agree with Dr. Rekate about the need to define more accurately and ubiquitously the clinical entity we are talking about. To this point, he refers to a discussion in a recently published editorial: should CTH be named an “anomaly” or a “malformation”? The unaccepted terminology

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reflects the complex and polymorphic etiopathogenicity of CTH, which can lead to a certain degree of confusion in some reports in which different conditions are grouped under the term “CMI.” The results of such studies can therefore be misinterpreted and become hardly reproducible, which could explain the lack of understanding about some aspects of the CMI.

We are grateful to Dr. Rekate because he believes that our work may improve the understanding of the pathophysiology of CMI and contribute to defining acceptable terminology for CTH. We used the term “classic CMI,” which was introduced by Dr. Milhorat to describe cases of CMI related to an isolated constriction of the posterior cranial fossa (PCF). This term depicts a condition in which there is no other inductive cause of tonsillar descent such as multiple faciocraniosynostosis, hydrocephalus, lumboperitoneal shunting, or intracranial hypertension. The results of our study support the notion that a cranioencephalic disproportion between the normal cerebellum and a too-small PCF, even when restricted to the basioccipital portion, is a sufficient condition to explain the development of a symptomatic CMI. New insights gleaned from our data confirm the main responsibility of basioccipital hypoplasia that had yet to be suggested by experimental works and previous morphometric studies.

There exists accumulating evidence that occipital hypoplasia results from a disorder of the paraxial mesoderm, justifying the terminology “malformation.” However, CMI is a dynamic disease in which the number of symptomatic patients increases with age. Moreover, the complex and prolonged development of the infratentorial part of the occipital bone, resulting from the tardive fusion of 4 cartilaginous precursors—the basioccipital portion anteriorly, the 2 exoccipitals laterally, and the supraocciput posteriorly—may explain its vulnerability to growth impairment. Because of the tardive closure of the sphenooccipital suture, the clivus represents a significantly weak growing zone, not only in the embryological period but also later in infancy.

We therefore hypothesized that the predominance of basioccipital hypoplasia could proceed either from an early paraxial mesodermal insufficiency due to a failure of notochordal induction in utero or from premature stenosis of the sphenooccipital synchondrosis occurring later in infancy. This theory is likely to explain both congenital or very early symptomatic CMI, probably related to a true abnormal formation of the occipital bone precursor during the intrauterine life, and later CMIs caused by a sphenoioccipital synchondrosial growth anomaly.

Given the difficulty of distinguishing these two origins, both “anomaly” and “malformation” may be representative of the potential pathophysiological mechanisms of CTH and can be accepted.

Another potential application of our results is that basioccipital measurement could help in establishing the diagnosis of classical CMI. Indeed, the expanding use of MR imaging has increased the number of cases of incidentally diagnosed CTH. The demonstration of basioccipital hypoplasia could be a means of differentiating true cases of CMI from incidentally diagnosed cases, because a short clivus or basiocciput and a shallow posterior fossa, which can be considered part of the same group of bone deformities of the skull base given the vertical orientation of these structures, are ubiquitous findings in CMI. In addition, because of its early development and central position, basioccipital hypoplasia could affect the subsequent development of nearly every cranial bone of the cephalic region. Posteriorly, it articulates with the exoccipital bones and could determine the location and orientation of the posterior cerebral fossa, as well as the position of the temporal bones laterally and the orofacial and pharyngolaryngeal cavities anteriorly. Cases of CMI related to basioccipital hypoplasia may represent a specific subtype in which orofacial skeleton malformations resulting from the adaptation of the facial skeleton to a primarily short and lordotic axial basicranium are more frequently observed.

We are also grateful to Dr. Rekate because he emphasizes the perspective that basioccipital hypoplasia may be utilized as a prognostic factor in patients with CMI. Morphometric and volumetric studies have been considered useful tools in increasing the understanding of the pathophysiological conditions at play in the development of CTH. It is attractive to speculate that morphometric parameters of the posterior cranial fossa, such as basioccipital dimension, could influence the natural history of patients with CMI as well as their prognosis after surgical treatment and could reduce the risk of postoperative complications such as cerebellar ptosis. Interestingly, Tubbs et al. have shown that children with Chiari 0 malformations improved after posterior fossa decompression because they had a distorted or smaller posterior fossa. Badie et al. demonstrated that patients with smaller posterior fossa volumes presented at a younger age and had a better response to surgery. Cardiac-gated phase-contrast cine MR can be a valuable tool in identifying patients who are less likely to respond to suboccipital decompression for CMI.

Based on the presence or absence of a basioccipital anomaly, further insights raised by our study may concern the ability to distinguish patients requiring a wait-and-see strategy from those requiring early surgical intervention for posterior fossa reconstruction. Further studies will also attempt to identify preoperatively the steps of the surgical treatment and the bone resection needed for decompression of the posterior fossa in the individual. Batzdorf also suggests that approximate measurements for optimal bone removal in an individual patient can be made on the basis of preoperative MR imaging.

Finally, our results emphasize the need for further studies of the developmental aspects of the basiocciput and neuroradiological evaluation of the normal values of basioccipital dimensions. Indeed, modern and adequate management of patients with CMI requires an accurate understanding of the embryological aspects and growth of the skull base components. Therefore, the sphenoioccipital synchondrosis should be assessed in cases of CMI, especially in children, by performing not only MR imaging but also bone CT scans with sagittal views of the clivus in order to identify a premature and progressive synostosis. A more extensive study, using repeatable measures with a larger number of patients, is necessary to