Letter to the editor: Management of Chiari I Deformity in Children and Adolescents: A Report from the Consensus Taskforce of the Brazilian Society of Pediatric Neurosurgery

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Members from the Brazilian Neurosurgery Society Spine Department (BNSSD) read with enthrallment your manuscript and wish to complement the authors for the initiative in discussing a complex and controversial issue such as the Chiari I disease.

The panel's consensus stated that a yearly MRI follow-up interval is indicated for children that harbor a 5 mm tonsils foramen magnum caudal displacement. However, careful literature analyses disclose a normal tonsillar position range that varies with age as posted by Mikullis et al.[1] These authors reported normal tonsillar position change with age and suggested the following criteria as lower limits for ectopia: in the first decade of life a 6 mm descent is considered normal, in the second and third, 5 mm, from the fourth to the eighth, 4 mm and beyond this age, 3 mm. All these distances were more than 2 standard deviations (SDs) over the normal range. Despite that, we also believe that the position of the tonsils is only an epiphenomenon, generally found concomitant with the disease, characterized by altered craniocervical transition cerebrospinal fluid (CSF) flow. Other authors have deeply questioned[2,3] a 5 mm cutoff to consider the diagnosis of Chiari disease since tonsillar descent measurements vary accordingly to other neglected parameters, such as asymmetric tonsillar positions, anatomic vessels relationships and CSF flow. Thus, what actually defines Chiari disease are the morphometric parameters of the posterior cranial fossa and concomitant typical clinical findings (8). Barkovich et al.[4] made this concept even clearer comparing the mean tonsillar position between normal and Chiari patients. They concluded that the most important factor for diagnosis would not be tonsillar descent, but compressions of the structures of the posterior fossa. Beijani[5], reviewing aspects of historical definitions before and after the advent of MRI, made clear that "there is no single test that allows a clear-cut distinction between clinically significant tonsillar ectopia and incidental tonsillar descent". Therefore, we consider two main issues in the diagnosis of Chiari 1 disease: an absent cisterna magna, filled with tonsillar

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tissue and clinically relevant symptoms associated with Chiari disease.

It is even more paradoxical to designate the condition as a deformity. Considering a small posterior cranial fossa volume as the main cause of the disease,[6] we can postulate that the Chiari type 1 condition could be a developmental disease and not congenital, since the patient is not born with the disease, and certainly not a deformity, as many asymptomatic persons might present a small posterior fossa and tonsillar descent and should not be diagnosed with Chiari disease (or Chiari deformity!).

Furthermore, we do not agree that the Chiari type 0 and Chiari type 1,5 terms brought confusion to the literature since these terms shed light on important issues regarding underlying mechanisms involved in this disease.[7,8]

Regarding the mentioned clinical indications for surgery, authors divide symptoms into major and minor and state that major symptoms are a strong surgical indication. Even though typical occipital headaches might improve with surgical treatment, there are huge variations in headache intensities and different impacts in quality of life. Thus, when headache symptoms are satisfactorily controlled with medication, surgery can be postponed or avoided. Besides, headache evaluation without other symptoms in children is even harder. Therefore, suggesting surgery for all these patients, as a strong recommendation, seems controversial.

The presence of hydrocephalus as a strong surgical recommendation is also questionable, since the tonsillar ectopia might be secondary to the hydrocephalus and this should be treated before even considering the diagnosis of Chiari type 1.

Regarding the use of intraoperative ultrasound to decide whether to perform duroplasty versus only bone decompression, there was a not a clear statement. Bond et al[9] had demonstrated that the position of the head alters the craniocervical junction CSF flow; therefore, the use of ultrasound is not conclusive when selecting between opening versus not opening the dura during the surgery. There are still many controversies in the literature and, therefore, we cannot routinely recommend this method to decide between opening or not the dura.

Finally, the theory that cerebellar tonsil caudal migration occurs due to a supposed "caudal traction" of cranial nerve structures in a so-called occult tethered cord syndrome is also controversial. A recent systematic review published by our group showed that filum terminale sectioning intended to treat Chiari 1 disease is still an experimental treatment. Furthermore, after 20 years of its proposal, it has still not been proven to be effective. Thus, so far, this procedure should not be recommended for Chiari type 1 treatment.[10]

We congratulate the authors for raising important information regarding the diagnosis and the surgical decision-making process in Chiari disease type 1, and hope that our considerations might contribute in the continuous understanding and management of this challenging disease.

Conflict of Interests

The authors have no conflict of interests to declare.

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