CHIARI malformations (CMs) are well-known structural defects that most often consist of downward displacement of the cerebellar tonsils through the foramen magnum, causing either obstruction of CSF or compression of the brainstem. Because of the significant variability in the structure of the posterior fossa (PF) and related congenital anomalies, there have been a number of different classification subtypes that have been described. Initially, there were 4 subtypes delineated that were related to different anomalies of the brain and hindbrain as well as other associated features. CM Type 1 (CM-1) is the most common and has been defined as cerebellar tonsillar descent of greater than 5 mm. Two further subtypes have been more recently described, including Type 0 (CM-0, lack of cerebellar herniation with or without syringomyelia) and Type 1.5 (CM-1.5, brainstem and tonsillar herniation). These different structural anomalies may result in symptoms related to the obstruction of CSF and/or compression, such as headaches aggravated by Valsalva maneuvers, vertigo, dizziness, etc., or to injurious changes in the spinal cord (specifically signal changes on imaging, or worse, syringomyelia) that may be asymptomatic or symptomatic, i.e., pain, weakness, or numbness in the back, arms, or legs. CM is believed to be fairly prevalent (1–3/1000 births), and women are 3 times more likely to have CMs compared with men. However, with our society’s frequent use of imaging for minimal or unrelated symptomatology, the incidental diagnosis of CMs has become more common.

CM Types 3 and 4 are rare, and it is abundantly clear as to their specific and related deficits. Type 2 is by definition accompanied by a myelomeningocele, with a number of other congenital anomalies frequently associated with this syndrome. These different subtypes have different presentations and have specific criteria for intervention. For the rest of the different subtypes, the decision-making involved for proceeding with surgery may vary from surgeon to surgeon, but for the most part there have been two criteria that have served as justification for surgical intervention: 1) a symptomatic syndrome of headache and associated other symptoms interfering with quality of life; or 2) significant (frequently defined as > 3 mm in diameter) and/or progressive syringomyelia with or without symptoms. For CM-0, CM-1, and CM-1.5, however, the challenge is defining objective criteria for surgical intervention for the vast majority of those that are found incidentally.

In this issue, Moncho and colleagues utilize a prospectively collected, retrospectively reviewed cohort of 200 adolescent and adult patients (from an overall cohort of almost 550 patients) with CM-0, CM-1, and CM-1.5 over a 9-year period who had preoperative electrophysiological studies as part of their “routine workup” for symptomatic and asymptomatic CMs. Their objective was to begin to define the relationship between preoperative electrophysiological studies—including brainstem auditory evoked potentials (BAEPs) and somatosensory evoked potentials (SSEPs)—and the clinical findings, associated abnormalities in patients with syringomyelia, and the clinical and neuroradiological risk factors in CM. They also obtained normative data from 50 healthy volunteers that were age-, sex-, and height-matched controls and that had been previously published by the authors. These authors found that 60% of their patients showed alterations in electrophysiological recordings with variable differences between the different subtypes, findings of syringomyelia, and whether the CM was discovered incidentally.

While there is no question that CM-0, CM-1, and CM-1.5 likely represent a continuum of structural findings and variability of compression and/or obstruction of CSF out-

**EDITORIAL**

**Evoked potentials and Chiari malformation Type 1**

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flow, previous studies and commentaries have led to the present classification scheme and definition of the different subtypes of CM. There is also some confusion that despite the fact that more than 40% of the cohort was patients with CM-0 and CM-1.5, there is a focus on CM-1, based on the title of the paper. Obviously, there is more recent literature suggesting the specific reasons for these new subtypes of CM and unless there is new evidence indicating such, they should not be represented as subtypes of CM-1. Using the current literature, it is also difficult to interpret the validity of the extent of tonsillar herniation, whether 3 mm or 5 mm, to define the specific CM subtype. The authors have chosen 3 mm as their cutoff to define their subtypes, which may confuse what ultimately is the issue, that of obstruction and/or compression. By making this choice, they lessen the size of the cohort of patients with CM-0, a subtype that remains controversial as to its existence and/or its significance, creating a very small subgroup for analysis. Similarly, because not all patients with CM-1.5 underwent electrophysiological studies that indicated compression, this may represent a different pathological finding as well as a different indication for surgical intervention. Additionally, it is unclear how an asymptomatic patient with CM-0 and no syringomyelia would even need to undergo electrophysiological studies and then potentially have a surgical indication for intervention if the electrophysiological studies were “abnormal.”

While this study has attempted to answer the question that electrophysiological studies can provide additional information, especially to patients with incidental findings of CM, and despite the normative electrophysiological data provided, it is well recognized that there can still be significant variability and subjectivity from electrophysiological study to electrophysiological study technically in how the study is completed and interpreted, resulting in differences within the same patient, variability in response from one patient to the next, and variability in the interpretation of clinical “significance” by the neurophysiologist. Because of this significant variability, it would be very difficult to conclude that a single electrophysiological abnormality, either BAEP or SSEP (especially if not impacting both), indicates a high probability of progression of disease and/or being able to define the damage to the spinal cord either with or without syringomyelia. The literature is replete with studies of the clinical impact of abnormal electrophysiology that have been quite variable and inconsistent, especially related to surgical decision-making. While a study such as the one by Moncho et al. in this issue helps to define abnormality at any given point in time, it still does not indicate “damage” or progression of disease. We must be careful in this type of interpretation because these patients do not have a baseline to make comparisons longitudinally in the preoperative setting to be able to state definitively that there is progression or further damage, as no repeated studies were conducted. Particularly if we were going to define surgical indication based on “progression,” longitudinal and progressive abnormality of the electrophysiological studies would need to be defined.

Lastly, as the authors concur, this study suffers from being a retrospective review, even though they utilized a prospectively collected cohort. Ultimately, this was just a clinical registry of patients with CMs since it lacked a prospective set of criteria for inclusion, exclusion, diagnosis, management and/or assessment of outcome. Future studies should consider other methods for optimizing the prospective collection of data so as to optimize the eventual conclusions. While a prospective clinical registry can be quite useful to begin to raise new clinical questions, if specific hypotheses for diagnosis, management, and outcomes are not delineated up front, then the data do not provide sufficient power to answer specific clinical questions (such as posed in this study) and provide understanding as to the nuances of a particular diagnosis and its management. This study highlights this weakness because the data were collected over a 9-year period, the indications for obtaining electrophysiological studies are not clearly indicated or delineated, and as a result it is unclear why almost two-thirds of the patients did not obtain these studies as part of the routine workup, as the number of physician decision-makers and the evolution in their thinking would have to be considered as to the number and types of patients receiving the diagnostic studies. Similarly, it is presumptive to be able to conclude that there was no further assistance of these studies in the surgical indication and ultimately surgical decision-making without understanding the bias in the patient selection criteria for obtaining these studies as part of this routine diagnostic evaluation, and then ultimately what were the indications for surgical intervention.

Overall this is a well-done, large, retrospective study that further contributes to our understanding of the pathophysiology of adult CM and syringomyelia. It also begins to suggest a potential clinical pathway for the evaluation and then decision-making for surgical intervention in patients with these incidentally found lesions utilizing electrophysiological studies, which should be further explored. Obviously, defining a clinical pathway for this diagnostic evaluation with specific indications, and then performing a prospective, hypothesis-driven clinical study to establish objective evidence for the diagnosis of subclinical dysfunctions and actual neurological progression, as well as the surgical indications for these patients, would be useful.

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References

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