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EDITORIAL (BY INVITATION) - NEUROSURGERY GENERAL

Clinical Chiari syndrome or anatomical Chiari malformation? A conundrum revisited

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Despite about one hundred years of academic work, the general use of the term *Chiari malformation* remains unclear and continues to cause disagreement, if not confusion, among experts [1]. The term has been employed variably and is based on an anatomical definition of usually 5 mm or more of caudal displacement of the cerebellar tonsils through the foramen magnum. It is also employed for a clinical syndrome that comprises one or more signs and symptoms such as headaches, neck pain, nausea, vertigo, diplopia or paresthesias, which may be aggravated by Valsalva-like manoeuvres. Beyond this, a multitude of pain, disability and psychological correlates have been associated with this entity [2].

One could therefore view their interrelation in a Venn diagram where there is some consensus in those patients who present with both anatomical and clinical criteria fulfilled. However, measurements of the anatomical metrics as well as predictions of treatment outcomes remain a matter of ongoing investigations [5, 7, 8].

Still, the clinical syndrome as such is not clearly established in all cases, as it appears that there are patients who meet the "clinical core criteria" but fail to meet the expected anatomical criteria. Other patients meet the criteria, but fail classic treatment approaches [6]. There hence is a true gap of knowledge of underlying causes for the symptoms of a Chiari syndrome.

In their extensive single-institution cohort study in this volume, Dan Heffetz and colleagues from the USA [3, 4] investigate the radiographic correlation of the extent of tonsillar ectopia to clinical symptomatology in patients. Interestingly, these are patients who self-referred to their institution for the evaluation of a possible Chiari malformation. The key message of their study is that the extent of tonsillar descent does not correlate with clinical symptomatology. In fact, symptoms appeared more frequently in patients with lesser caudal displacement of the measured radiographic anatomical reference point.

There are of course limitations to this study. Admittedly, certain selection criteria and even some bias in inclusion criteria for this study can be discussed, but it appears that the diagnosis was made and meticulously documented by a team of physicians dedicated, or specialized, in this condition. One has to recognize that the mechanism of self-referral and the availability of an institutional service with possible financial gain can influence the selection somewhat. Self-referred patients after all are not a random sample. This may contribute to some selection bias (e.g. patients with higher anxiety scores than seen in the general population and in Chiari patients), which might influence the results, yet professionalism of this established center and national oversight would mitigate against any deliberate unethical practice.

It is evident and well documented that a strict selection process was employed among referred patients. The secondary referral structure probably introduces some bias to a more complex patient population, since "easier" cases (conforming to the classic definitions) would be treated at local institutions. It is also relevant to note that institutions in health care systems (e.g. UK's NHS, Swedish health care, Canada) where patients represent a cost to the health care institutions rather than

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revenue (e.g. USA) will have an incentive to decrease cost and may hence not offer treatment to patients unless clinical indications are posing an undisputable need.

It can therefore be seen as a strength of this study that a sizable group of patients with less overwhelming presentations has been scrutinized. The enormous amount of work that went into this project deserves therefore acknowledgement for the diligent approach the authors have chosen to investigate this problem that bears clinical significance in clinical practice.

The dataset is strong for the inclusion in this unselected study cohort, while follow-up aspects were considerably weakened by the significant loss of patients during follow-up. This creates some caveat that the reader needs to be aware of, since in this setting, two mechanisms affect the interpretation: (i) In all surgical series, and especially in those without a strictly defined control group, a "regression to the mean" occurs. This means that patients suffering from a chronic disease with fluctuating severity are more likely to undergo interventions when symptoms are most severe. (ii) This leads to the problem that natural history alone could explain improvements seen after surgery. Beyond that, professional patient management with good patient-physician interaction can provide for a notable therapeutic benefit through "placebo". Hence, benefit is demonstrated in the patients that are being followed, but it is not strictly possible to evaluate the cause of benefit.

The submitted work however is of considerable value, since it clearly puts the very concept and terminology of a Chiari 1 malformation into question. To this end, the authors present novel possible mechanisms that may explain the observed symptoms in patients who present with a clinical "Chiari syndrome". These are important mechanisms that need further validation. One option for future studies is to choose an approach that employs patient selection based on radiographic criteria alone (e.g. one might pick patients with tonsillar ectopia found on head MRI scans performed for screening purposes only or for patients who had a single symptom (e.g. non-Valsalva headache)) that can be quantified and normalized across the study population. Then, mail-out questionnaires and specific MRI assessments during follow-up appointments in each group. This would circumvent the problem that asymptomatic patients do usually not come to our attention.

Through their admittedly controversial study, the authors have provided important data that further underline how problematic work on the "Chiari complex" is: clearly, the symptoms of a Chiari 1 type syndrome are multifactorial and coexist with an abnormal tonsillar position in many patients. One methodological aspect that needs to be addressed in the future is the fact that the midsagittal plane was chosen for radiographic measurements. As the authors acknowledge, the tonsil is a structure that resides parasagittally. However, there is no clear standardized set of measures—since most practicing neurosurgeons use maximal extent of ectopia, which may not reflect the volumetric determinants of CSF flow, and which does not allow to account for vascular-tonsillar interactions. It also does not take into account asymmetric tonsils. One way to account for such aspects is to include both the maximal decent as well as measures of the cross-sectional area at the level of the FM and compare the fraction that is being occupied by the tonsils and brain stem (+vessels) versus the remaining space that is available for CSF flow. Yet, the position of the tonsils alone cannot exhaust the possible causalities of the symptoms encountered. Subgroups of symptom constellations may exist and need to be compared with strictly defined mechanisms to improve our future understanding and definition of Chiari syndromes.

These manuscripts are a valuable contribution to the literature, since they put conventional thought patterns into question. This can be appreciated, even if we do not support all aspects of the methods and conclusion. Taken together, we believe that the papers are diligently written and have great merit for the patient population under investigation.

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