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In Reply: Chiari Severity Index: A Novel Grading System Intended for Pre-operative Counseling

Jacob K. Greenberg, MD, MSCI¹, Chester K. Yarbrough, MD, MPHS¹, Alireza Radmanesh, MD⁴, Jakub Godzik, MD, MSCI⁵, Megan Yu, BA¹, Donna B. Jeffe, PhD³, Matthew D. Smyth, MD¹, Tae Sung Park, MD¹, Jay F. Piccirillo, MD², and David D. Limbrick Jr, MD, PhD¹

¹Departments of Neurological Surgery, Washington University School of Medicine in St. Louis, St. Louis, Missouri

²Departments of Otolaryngology, Washington University School of Medicine in St. Louis, St. Louis, Missouri

³Departments of Medicine, Washington University School of Medicine in St. Louis, St. Louis, Missouri

⁴Department of Radiology and Biomedical Imaging, University of California, San Francisco, San Francisco, California

⁵Barrow Neurological Institute, Phoenix, Arizona

We appreciate Dr Meybodi's interest in our study, "The Chiari Severity Index: A Preoperative Grading System for Chiari Malformation Type 1."¹ In his Letter to the Editor, Dr Meybodi raised several questions and concerns that we would like to address.

First, we agree with Dr Meybodi that the CSI is optimally designed for use in children who are able to articulate their headache symptoms. In our sample of 119 patients used to develop the integrated CSI, only 3 patients (2.5%) were under 2 years of age and presumably nonverbal or minimally verbal. Twenty others (16.8%) were younger than 5 years, and most were in fact able to articulate headache symptoms, either verbally or with specific body gestures (eg, pointing to the back of the head). Specifically, 11 patients were thought to have Classic Chiari Headaches (see original publication for definition¹) by the treating neurosurgeon, while six had Poorly Localized Headaches, one had a Frontotemporal Headache, and seven had no headache. Some patients had more than one headache type. Among the three very young children, only one was thought to have a headache, characterized as occipital based on observed body gestures and behavior. Particularly in this very young group, we recognize that patients' headache experiences may not have been fully characterized, emphasizing the need for a modified index designed specifically for very young, preverbal children. Nonetheless, given that most young children in our study were able to convey headache symptoms and very young children (< 2 years) accounted for a very

Correspondence: Jacob K. Greenberg, St. Louis Children's Hospital, One Children's Way, 4S20, St. Louis, Missouri 63110, greenbergj@wusm.wustl.edu.

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small proportion of the study cohort, the CSI should accurately predict outcomes for verbal children.

Second, Dr Meybodi correctly points out that patients may have both headaches and signs of myelopathy. According to the sequential sequestration technique we used to develop the clinical grading system,² the lowest (worst) category is defined by the variable associated with the worst treatment outcome, regardless of the values of other variables. In this case, patients with both myelopathic symptoms and headaches would be considered equivalent to those with myelopathic symptoms and no headaches, and thus assigned to Clinical Grade category 3.

Third, although timing of surgery might influence treatment outcomes in CM-1 patients, this factor was unlikely a significant influence of outcome in our study. Dr Meybodi cites a key treatment latency of 5 years for primarily adult CM-1 patients with ventricular dilation.³ In our cohort of pediatric patients, most of whom had no history of ventriculomegaly, 99% (118/119) of patients had surgery within 5 years of initial neurosurgical consultation, and the median time from initial neurosurgical consultation until surgery was 29 days (interquartile range 13–85 days). While we are unable to report the precise timing of symptom onset in each patient, these data suggest that the timing of surgery was likely not a significant confounding factor in the creation of the CSI.

In summary, the CSI is the first iteration of a rigorously developed preoperative stratification tool for CM-1, but it is also a framework for further development. Undoubtedly, future research efforts will yield new discoveries in neuroimaging and clinical evaluation that may allow additional refinement of this important tool, and prospective external validation studies will further support its utility. Nonetheless, we believe the CSI remains a potentially valuable adjunct for patient counseling and an effective means to stratify patients for comparative effectiveness research.

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