

## Key Points

- Before the Chiari Severity Index (CSI) was designed, most classification systems relied solely on clinical or neuroimaging evidence-- the CSI allows for both systems and their data to be used in unison.
- The CSI can be utilized to moderately predict the long-term outcomes of patients after decompression surgery.
- The children engaged in this study were asked two questions about their overall post-op outcomes: how much of an improvement in your general health did you experience after your surgery? -AND- how satisfied are you with your overall outcome from surgery?
- 111 of the 158 pediatric patients (70%) stated that they recovered well after decompression.
- At this time, the CSI should be solely used to assist physician and patient discussions surrounding Chiari and its associated symptoms.

## Definitions

**basilar invagination** - condition, sometimes associated with Chiari, where the C2 vertebra is displaced upward, potentially compressing the brainstem

**central sleep apnea** - a disruption of breathing during sleep which lasts longer than 10 seconds, due to a delay in the nerve signal from the brain to breathe

**dysphagia** - trouble swallowing

**nystagmus** - involuntary, rapid eye movements

**syringomyelia** - neurological condition where a fluid filled cyst forms in the spinal cord

## Uniting Patient-Based Outcomes: The Chiari Severity Index

**February 2015** - Since the 1970's, numerous professionals within the medical community have attempted to predict which patients would be most likely to benefit from Chiari decompression surgery. However, it wasn't until 1995 that a limited classification system was created to catalogue patients with *syringomyelia* as well as symptoms stemming from brainstem compression ([Bindal et al. – Bindal AK, Dunsker SB, Tew JM Jr. Chiari I malformation: classification and management. Neurosurgery. 1995;37\(6\):1069-1074.](#)) Additionally, previous endeavors have mainly concentrated on clinical or neuroimaging (ex. CT, CAT-scan, MRI) findings, but efforts to combine both of these methods were not investigated until the summer of last year.

Ten dedicated specialists from the Washington University School of Medicine devoted their time to try to define patient-based quality of life (QOL) guidelines for those with Chiari 1. For many years, there has been an urgency to create a system which evaluates a patient's post-op status after medical intervention and thanks to Jacob Greenberg et al., the Chiari community now has the Chiari Severity Index (CSI) to moderately predict the long-term outcomes after decompression surgery.

In the initial stages of this study, 314 pediatric patients were contacted, but 158 decided to participate—providing the researchers with 93 female patients and 65 males. Among the 158 adolescents, syringomyelia was recorded in 119 before surgery was performed. Considering the presence of a syrinx, most of the children underwent posterior fossa decompression (74%) while others opted for extradural decompression (26%), where the dura was not opened. The median age at the time of surgery was 12 years and the average post-surgical follow-up period was 4.8 years.

After this primary information was collected, the analysts reviewed the patients' pre-surgical medical records. Clinical data (related to characteristics and locations of symptoms), such as persistent headache, numbness, weakness, unsteady gait, *central sleep apnea*, *nystagmus*, *dysphagia*, and spinal cord injury, were recorded along with neuroimaging specifications (e.g. tonsillar herniation).

From the compiled data, Greenberg et al. then created a clinical grading system consisting of common Chiari 1 symptoms such as headache location and signs of spinal cord or brainstem compression—and a neuroimaging grading system which measured anatomical abnormalities such as cerebral herniation and syrinx length to gauge improvement after decompression.

Additionally, participants were asked two questions pertaining to their overall outcome after surgery: "How much of an improvement in your general health did you experience after your surgery?" and "How satisfied are you with your overall outcome from surgery?" (Greenberg et al.). The analysts reported that 111 of the 158 pediatric patients (70%) recovered well after decompression.

Once the information was analyzed, the specialists found that there was less of an improvement in adolescents with syrinxes larger than 6 mm and basilar invagination (55%), or the upward movement of the upper cervical spine (C1, C2) into the base of the skull, compared to children with syrinxes less than 6mm (74%). However, in the case of *basilar invagination*, Greenberg et al. stated that they could not accurately assess its full significance in the Chiari population considering their study's small patient size.

The last step of this study consisted of merging the clinical and neuroimaging systems together to create the Chiari Severity Index. Currently, the CSI consists of three graded categories and provides doctors and patients with an estimated improvement percentage after surgery. By taking the collected clinical and neuroimaging information, the team was able to calculate ranges that simultaneously relied on Chiari symptoms and MRI evidence, which had never been purposed in the past. When compared to the independent clinical and neuro-visual models, the researchers found that the CSI fared noticeably well as a new system that gauges improvement in quality of life for Chiari patients after surgery.

Greenberg et al. found that their new diagram detailed considerably accurate predictors of patient-reported outcomes. Although the team successfully combined clinical and patient data, they stated that the CSI should, at this time, be solely used to assist physician and patient discussions surrounding Chiari and its associated symptoms.

The development of the CSI has produced some staple guidelines in relation to Chiari, but some limitations did exist. For example, the analysts could not find a substantial improvement rate as follow-up time progressed because patient health perceptions appear to change over time when recalling information is requested. Another restraint lies in the inability to have a larger population size to document and assess rare findings, such as basilar invagination.

The Chiari Severity Index will be used to further understand both the clinical and neuroimaging aspects of Chiari patients. To close, Greenberg et al. state that in the future they look forward to seeing what new

## Source

[The Chiari Severity Index: A Preoperative Grading System for Chiari Malformation Type](#)

1. Greenberg JK, Yarbrough CK, Radmanesh A, Godzik J, Yu M, Jeffe DB, Smyth MD, Park TS, Piccirillo JF, Limbrick DD. *Neurosurgery*. 2015 Jan 12. [Epub ahead of print]

systems are developed to test and further improve the performance of the current CSI.

**Author's Note:** In the interest of finding private improvement rates, all participants in this study received surgical treatment at the St. Louis Children's Hospital, which is affiliated with Washington University School of Medicine.



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Ms. Eubanks is a professional writing and researching scholar from Purdue University Northwest. After being diagnosed with a Chiari I Malformation in 2011, she quickly decided that being conquered was not an option—she was committed to fight and pursue a budding love of healthcare/medical writing. Spreading awareness and hope to others battling Chiari is her largest motivator alongside educating others who have not heard about the condition. Reporting for Ideas in Motion Media and tutoring at the Writing Center (Purdue University North Central) has been immensely beneficial to her success as well as all the remarkable individuals who helped her become the composer and analyst she is today.

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