Definitions

ataxia - trouble walking

blinded - an experimental technique where a rater is not aware of aspects of the experiment to prevent bias

CCOS - Chicago Chiari Outcome Scale

dysesthesia - unpleasant sensations

dysphagia - trouble swallowing

cerebellar tonsils - portion of the cerebellum located at the bottom, so named because of their shape

cerebellum - part of the brain located at the bottom of the skull, near the opening to the spinal area; important for muscle control, movement, and balance

cerebrospinal fluid (CSF) - clear liquid in the brain and spinal cord, acts as a shock absorber

Chiari malformation I - condition where the cerebellar tonsils are displaced out of the skull area into the spinal area, causing compression of brain tissue and disruption of CSF flow

decompression surgery - general term used for any of several surgical techniques employed to create more space around a Chiari malformation and to relieve compression

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

Source

A Novel Scoring System for Assessing Chiari Malformation Type I Treatment Outcomes.Aliaga L, Hekman KE, Yassari R, Straus D, Luther G, Chen J, Sampat A, Frim D. Neurosurgery. 2011 Aug 12. [Epub ahead of print]

New Scoring System Proposed For Chiari

September 30th, 2011 -- Two major issues with Chiari outcomes research today are the lack of a consistent, reasonably objective, quantitative outcome measure, and too short clinical follow-up times. The lack of a useful outcome measure makes it difficult to assess and compare the scant research that is published on patient outcomes and surgical techniques. For example, how do you weigh differing publications on the benefits of opening the dura when the research on the subject uses different measures of success. In addition, it is not very informative for patients facing the prospect of surgery to read about "significant improvement", since each person might interpret that differently.

In a recent publication in the journal, Neurosurgery, a group from the University of Chicago proposed a new scoring system for Chiari outcomes, called, not surprisingly, the Chicago Chiari Outcome Scale (CCOS). The new scale uses four categories: Pain Symptoms, Non-Pain Symptoms, Functionality, and Surgical Complications, and both pre and post-surgical information to rate patients on a 4 point scale within each category. The scores can then be added for a total score with 16 being a perfect score and 4 being a score that no one reading this article would want.

Table 1: Proposed Outcome Scale

Chicago Chiari Outcome Scale				
Pain	Non-pain	Functionality	Complications	Total Score
1 - Worse	1 - Worse	1 - Unable to attend	1 - Persistent complication, poorly controlled	4 - Incapacitated outcome
2 - Unchanged and refractory to medication	2 - Unchanged or improved but impaired	2 - Moderate impairment (<50% attendance)	2 - Persistent complication, well controlled	8 - Impaired outcome
3 Improved or controlled with medication	3 - Improved and unimpaired	3 Mild impairment (>50% attendance)	3 - Transient complication	12 - Functional outcome
4 - Resolved	4 - Resolved	4 - Fully functional	4 - Uncomplicated course	16 - Excellent outcome

The Pain category captures symptoms such as headaches, neck and shoulder pain and dysesthesia. A score of 4 is given if after surgery all pre-surgical pain symptoms are completely resolved and no new Chiari type pain symptoms have developed. A score of 3 means that pre-surgical pain symptoms have improved (decreased in intensity, frequency, or duration) or can now be controlled with medicine; or if pre-op symptoms improved, but post-op pain symptoms persisted. A score of 2 means that there are still either pre or post op pain symptoms, which are not helped by medicine, of about the same severity. Finally, a score of 1 means that any pain symptom got worse.

The Non-Pain Symptom category is scored similarly to the Pain category, but focused on major Chiari related symptoms, such as dysphagia, ataxia, sensory disturbances, vertigo, weakness, drop attacks, etc. This category also includes objective neurological signs.

The Functionality category assesses a patient's ability to attend to their daily responsibilities such as work, school, family care, etc. Full participation rates a score of 4, while impairment of less than 50% scores a 3. A score of 2 means a person is restricted from participating more than 50%, and a score of 1 means a person is not able to attend to their daily responsibilities at all.

The Complications category is, coincidentally, the most complicated to describe. It covers not only complications such as CSF leaks and infections, but also the need for more surgery and lumbar punctures to address increased pressure. Glossing over the specifics, a score of 4 in this category naturally means that there were no complications throughout the follow-up period. A score of 3 means that there were short-term complications that resolved fairly easily. A score of 2 means that were persistent complications that were still able to be controlled. Finally, a score of 1 means that there were persistent complications that were not able to be controlled through medicine or additional surgery (for example, sustained intracranial hypertension).

To get a feel for how the new scoring system would work with real patients, and to compare it to a more traditional system of Improved, Unchanged, Worse, the researchers applied the CCOS scale to 146 patients who had undergone first-time Chiari surgery, and for whom there was at least 1 year of information available sufficient to reasonably assign scores. They used 5 raters who were blinded to each other's ratings to assess each case - a statistical analysis showed the raters agreed with each other 88% of the time.

Table 2: CCOS and I/U/W Scores For 146 Surgical CM Patients

CCOS Score	Improved	Unchanged	Worse	Total
13-16	93	1	0	94
9-12	8	36	1	45
4-8	0	2	5	7
Total	101	39	6	146

Under the new system, 94 of the patients had total CCOS scores of 13-16, 45 were in the 9-12 rants, and 7 were in the 4-8 range. There was good overlap with a broader Improved,. Unchanged, Worse scale, with most of the Improved falling in the highest CCOS range, Unchanged in the middle, and Worse at the bottom. Scientifically validating a new scale when there is no existing gold standard to compare it to can be challenging, and the CCOS will have to be studied and used extensively before gaining broad acceptance. However, if it proves out, it should provide clinicians with more detailed data about individual patients and provide researchers with a new, quantitative tool to use in Chiari research. It is also exciting to speculate how a scale such as this could be combined with some of the new quantitative MRI measures that are being developed through Conquer Chiari funded research to really begin to develop an accurate picture of Chiari severity.

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