

Key Points

1. CCOS is an outcome scale designed specifically to measure success of Chiari surgery
2. Study used CCOS to identify specific predictors of surgical outcome
3. Found that peripheral nerve issues was a strong predictor of poor outcomes
4. Found that children respond better to surgery than adults
5. Application of CCOS scoring to a larger patient population undergoing a variety of operative CM1 treatments should allow for better-informed decisions regarding patient selection and treatment options for CM1

Definitions

- ataxia** - trouble walking
- dysphagia** - trouble swallowing
- peripheral neuropathy** - nerve related pain or sensations in the peripheral nerves, such as tingling in the hands or feet
- Romberg sign** - swaying or falling when standing with feet together and eyes closed
- syncope** - loss of consciousness
- tinnitus** - ringing in the ears
- Valsalva** - straining, a Valsalva headache is one brought on by straining
- vertigo** - a sense of spinning, dizziness or disorientation
- 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

Surgical Predictors Based on the Chicago Chiari Outcome Scale

November 1st, 2012 -- One of the major limitations in Chiari research has been the lack of a standard outcome measure which could be used as the basis for a variety of research. The lack of this measure has limited the ability to gauge the results of different studies against each other. Recently, however, a group from the University of Chicago (Frim) has proposed that the Chicago Chiari Outcome Scale (CCOS) can become the standard measure that has been missing for so long. Even more recently, the group has published the first work using the CCOS to identify the ever elusive predictors of surgical success.

The CCOS (below) is comprised of 4 categories: Pain, Non-Pain Symptoms, Functionality, and Complications. A patient is given a score of 1-4 in each category, for a total score of 4-16. A final score of 4 means the person is incapacitated; a score of 8 means they have an impaired outcome; a score of 12 equates to a functional outcome; and a score of 16 is of course an excellent outcome.

| Chicago Chiari Outcome Scale | | | | |
|---|---|---|--|--------------------------|
| Pain | Non-pain | Functionality | Complications | Total Score |
| 1: pre-operative symptoms worse | 1: pre-operative symptoms worse | 1: unable to attend | 1: persistent complication-poorly controlled | 4: incapacitated outcome |
| 2: unchanged/refractory to meds/onset of new poorly managed symptoms | 2: unchanged/improved but impaired/onset of new poorly managed symptoms | 2: moderate impairment (< 50% attendance) | 2: persistent complication-well controlled | 8: impaired outcome |
| 3: improved/managed with meds/onset of new symptoms managed with meds | 3: improved-unimpaired/onset of new symptoms managed with meds | 3: mild impairment (>50% attendance) | 3: transient complication | 12: functional outcome |
| 4: resolved/no onset of new symptoms | 4: resolved/no onset of new symptoms | 4: fully functional | 4: uncomplicated course | 16: excellent outcome |

To identify predictors of surgical outcome, the authors retrospectively reviewed the records of 167 Chiari patients who had undergone first time decompression surgery at their institution, and for whom enough information was available to assign a CCOS score. The group was out of a total of 245 patients who had undergone surgery. The 167 was comprised of both pediatric and adult cases with slightly more than half being under 18 at the time of surgery. Each patient underwent a similar surgery with almost all of them being performed by Dr. Frim.

The subjects were randomly divided among five raters who used their latest follow-up visit (usually 1 year or more after surgery) to assign a CCOS score. In addition, specific symptoms and signs, such as: headache, neck pain, peripheral neuropathy, syncope, tinnitus, vertigo, ataxia, pinprick test, and Romberg sign were noted. The extent of tonsillar herniation and the presence, location, and extent of syrinxes were also noted.

Overall, sixty-seven percent scored 13 or higher, meaning a very good outcome, while 29% scored 9-12, and 4% scored 8 or less (Table 1). In order to identify specific predictors of outcome, the researchers – after utilizing some statistical tests – decided to use a cut-off score of 11 to denote a better or worse outcome. Using these criteria, 82% scored 11 or above for a good outcome, while 18% scored less than 11 for what was called a worse outcome. It is interesting to note that this lines up with the general finding that about 80% of patients experience a significant improvement in symptoms from surgery.

The team then identified seven statistically significant predictors of poor outcomes and three predictors of positive outcomes (Table 2). The only symptom which predicted a poor outcome by itself was peripheral neuropathy, which encompassed any type of peripheral nerve pain or abnormal sensations (such as tingling in the hands or feet). Patients with peripheral neuropathy were nearly three times as likely to have a poor outcome – meaning a CCOS score of 10 or less – than patients who did not have this symptom. Peripheral neuropathy combined with a Valsalva induced headache or neck pain were also significant predictors of poor outcomes at about the same level. However, patients with peripheral neuropathy and syncope (blacking out) were nearly five times as likely to have poor outcomes.

For positive predictors, the data showed that patients under 18 at the time of surgery were more likely to have good outcomes than adults. This effect was even more pronounced among males with boys under 18 being more than three times as likely to have a positive outcome. While there is a general impression in the Chiari community that children have better outcomes, hard data such as this can have a profound effect on families struggling to make decisions.

In an unusual finding, the data also showed, counter intuitively, that patients with a syrinx were actually more likely to have a positive outcome. Since this goes against the general thinking, and previous research, it could be that this result is a statistical anomaly and highlights the danger of drawing strong conclusions from just one study.

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

Positive and negative predictors for good outcome after decompressive surgery for Chiari malformation type 1 as scored on the Chicago Chiari Outcome Scale. Clinical article. Heiss JD, Suffredini G, Smith R, DeVroom HL, Patronas NJ, Butman JA, Thomas F, Oldfield EH. J Neurosurg Spine. 2010 Dec;13(6):729-42.

The authors stress that they would like to apply the CCOS to a much larger group of patients, both retrospectively across institutions, and in a prospective manner (meaning that patients would be followed using the CCOS scale from the start). If the CCOS proves to be valuable and becomes widely adopted, it may prove to be the spark in structured, sound, Chiari research that the community has been desperately waiting for.

Table 1
Total CCOS Outcome Scores
(167 Patients)

| | |
|------|-----|
| >13 | 67% |
| 9-12 | 29% |
| 4-8 | 4% |

Table 2
Predictors of Poor Outcomes

| Signs/Symptoms | x More Likely Poor Outcome |
|----------------------------------|----------------------------|
| Peripheral Neuropathy | 2.91 |
| Peripheral Neuro & Headache | 2.85 |
| Peripheral Neuro & Neck Pain | 2.49 |
| Peripheral Neuro & Syncope | 4.64 |
| Pinprick Loss & Romberg | 4.06 |
| Pinprick Loss & Paresis | 3.18 |
| Peripheral Neuro & Pinprick Loss | 3.12 |

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