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A Novel Scoring System for Assessing Chiari Malformation Type I Treatment Outcomes

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Abstract

BACKGROUND: Outcome assessment for the management of Chiari malformation type 1 is difficult because of the lack of a reliable and specific surgical outcome assessment scale. Such a scale could reliably correlate postoperative outcomes with preoperative symptoms.

OBJECTIVE: We developed a novel scoring system and applied it retrospectively to 146 patients treated at our institution in order to create and verify a simple and quantifiable assessment of Chiari outcomes.

METHODS: The Chicago Chiari Outcome Scale (CCOS) uses 4 postoperative outcome categories (pain, nonpain symptoms, functionality, and complications) graded 1 to 4 for a total possible score of 16. As a comparison with current Chiari outcome methodology, each patient was also placed into a gestalt outcome group of “improved,” “unchanged,” or “worse” (I/U/W). Patients were stratified by CCOS scores and by I/U/W group.

RESULTS: Stratifying patients by total CCOS scores showed that patients who achieved CCOS scores between 13 and 16 were predominantly in the I/U/W improved group (n = 101, 69%); scores between 9 and 12 were predominantly I/U/W unchanged (n = 39, 27%), and scores between 4 and 8 were I/U/W worse (n = 6, 4%). Symptom subscore results provided insight into the specifics of the overall outcome in addition to the more quantitative nature of the 16-point scale.

CONCLUSION: We describe a CCOS that assigns higher scores to patients judged improved by gestalt I/U/W ratings and lower scores to those who were unchanged or worse while defining outcome in 4 specific subcategories. As such, this CCOS should allow for a more unified and quantifiable outcome assessment after Chiari surgery.

Keywords

Chiari malformation type 1; Outcome assessment; Posterior fossa decompression; Scoring system

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Disclosure

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Outcome assessment after surgery for Chiari malformation type 1 (CM1) is difficult because of the lack of a reliable and unified scoring system. The constellation of presenting symptoms of CM1 can be quite variable.^{1–4} However, there are several strong indications for posterior fossa decompression (PFD), such as drop attacks, dysphagia with aspiration, apnea, and the presence of a syrinx.^{4–8} Beyond these indications, other symptoms and signs may or may not warrant intervention. The estimation of whether those interventions will result in benefit is often based on previous outcomes, and that relationship is not always clear.^{4,6,9–12} We developed a simple and quantifiable outcome scoring system, the Chicago Chiari Outcome Scale (CCOS), and applied it retrospectively to 146 patients treated at our institution. Previous reports use outcome labels such as “improved,” “unchanged,” or “worse,” or some variant of this scheme.^{13–19} The CCOS is multifaceted, and a general gestalt description does not fully address the various nuances in outcome. Patients who present with many Chiari symptoms may only experience relief from some of them after intervention.^{2,20} The CCOS uses 4 postoperative categories to assess outcomes: pain symptoms, nonpain symptoms, functionality (ability to attend work or school), and complications. Here, we present the CCOS and how it can be applied to a cohort of patients in comparison with the traditional gestalt assessments of “improved,” “unchanged,” or “worse” outcome. Finally, we discuss general implications for the use of this novel Chiari outcome scoring system.

PATIENTS AND METHODS

Charts of Chiari patients were reviewed under an institutional review board-approved protocol at our institution. Five separate raters blinded to each other’s ratings performed chart review assessments on a consecutive series of 246 first-time surgically treated Chiari patients. Of this group, 35 had their initial Chiari surgeries at outside institutions, and 211 were first-time surgical Chiari patients, initially treated at our institution. All of these 211 patients underwent suboccipital craniectomy, C-1 laminectomy, and expansile duraplasty with autologous pericranium. These 35 patients were excluded, because factors related to a previous Chiari surgery was not included in this initial presentation of the CCOS. Because it is possible for a variety of factors to impact the resolution of Chiari syndrome, including surgical technique and potential sequelae from the surgery itself, it was felt appropriate to address those issues in a revised version of the CCOS for reoperative patients.^{6–11,13,15,16} Twenty-eight of the 211 patients had limited follow-up at our institution. Of these remaining 183 first-time surgical patients, 146 had data notations to populate all of the data fields for preoperative symptoms as well as postoperative outcome. Ninety-six patients were female and 50 were male. Patients’ mean age at diagnosis was 20 ± 16.7 years (range, 1–55). The mean time from onset of symptoms to diagnosis of CM1 was 33.5 ± 42.9 months (range, <1 month to 20 years). The mean time from diagnosis to PFD was 7.7 ± 17.8 months (range, <1–132).

Patients’ primary symptoms were documented, along with the presence or absence of symptoms and signs specific to the Chiari syndrome. Four postoperative categories were assessed in each patient: pain symptoms, nonpain symptoms, functionality, and surgical complications. Each category was graded from 1 to 4, using higher numbers to represent better outcomes (Figure 1). The improvement of pain and nonpain symptoms was

determined by comparing the patients' pre-operative state with their postoperative state at the last clinic visit. The majority (>95%) of chart notes were written and/or reviewed and signed by the senior author. The remaining notes were authored by 2 other faculty members.

For children unable to fully express themselves to the examiners, subjective symptomatic histories were obtained from parental or caretaker interviews—both before and after surgery. Although formally a different approach than in an older child or adult, the basis of assessing outcome remains the symptomatic change reported within an individual based on pre- and postoperative symptoms and signs.

Pain symptoms commonly included tussive headache, neck and shoulder pain, and dysesthesia in the upper extremities. If symptoms that were present before PFD completely resolved at the last clinic visit, patients received a score of 4. This meant that pain symptoms were completely absent. New Chiari-related pain symptoms that arose post-operatively were considered at the last clinic visit and handled similarly; patients were assigned a score of 4 only if both preoperative and postoperative symptoms resolved. New postoperative pain symptoms that were not Chiari-related were not considered when applying the CCOS. These included pain symptoms caused by unrelated diagnoses or head and neck trauma unrelated to patients' Chiari surgery. If the pain symptoms persisted but improved, meaning a decrease in frequency, intensity, and/or duration, with or without the use of medication, patients received a score of 3. This score was also assigned when patients had preoperative symptoms that resolved, but postoperative symptoms that persisted, albeit improved and/or managed with medication. Likewise, if new postoperative pain symptoms arose and resolved, but preoperative pain symptoms improved without full resolution, patients received a score of 3. When either old preoperative or new postoperative pain symptoms were unchanged and refractory to medication, patients received a score of 2, regardless of the improvement of other symptoms. When either old preoperative or new postoperative symptoms increased in frequency, intensity, and/or duration, patients received a score of 1.

Nonpain symptoms commonly included dysphagia, ataxia, vertigo, muscle weakness, sensory loss, tinnitus, paresthesias, and drop attacks, as well as signs, such as presence of syrinx or other neurological signs. A grading strategy similar to that used for pain symptoms was used when assigning a score of either 4 or 1 for nonpain symptoms. If nonpain symptoms decreased in frequency, intensity, and/or duration, and did not cause impairments, patients received a score of 3. When either old preoperative or new Chiari-related postoperative nonpain symptoms persisted in an unchanged manner, or improved but left the patient impaired, patients received a score of 2.

Functionality, meaning the patients' ability to attend work, school, or usual daily responsibilities, was also assessed at the last clinic visit. When patients were able to fully attend their typical daily responsibilities in their usual manner, they were assigned a score of 4. Impairment, whether due to pain or nonpain symptoms, that allowed greater than 50% attendance of daily responsibilities led to a score of 3. When patients' impairment allowed only less than 50% attendance of daily responsibilities, they received a score of 2. If patients were unable to attend to their daily responsibilities at all, they received a score of 1.

Surgical complications were assessed throughout patients' postoperative course, from the time of surgical decompression to the last clinic visit. They included typical surgical complications such as wound infection, chemical meningitis, and CSF leak, all of which tend to be more random in occurrence. Also included are complications that tend to be less random and more specific to Chiari patients, such as the need for posterior fossa reexploration for fourth ventricular outflow obstruction due to scarring and pseudotumor cerebri, possibly leading to shunt placement, shunt infection, and/or shunt malfunction. Patients who experienced none of these or any other surgical complications received a score of 4. Transient complications that resolved in an unprotracted manner, such as minor wound infections, led to a score of 3. Similarly, increased intracranial pressure that resolved after a single lumbar puncture and lowering of CSF pressure also led to a score of 3. Persistent complications that were well controlled led to a score of 2. For example, patients who had persistent pseudotumor cerebri that required medical and/or surgical intervention to improve, such as repeated lumbar punctures or shunt placement, received a score of 2. Recurring shunt malfunction and/or infection led to a score of 2 if patients were well between shunt surgeries. Separate from the issue of pseudotumor cerebri, recurrence of Chiari symptoms that necessitated posterior fossa reexploration and had subsequent resolution of symptoms led to a score of 2. Patients with persistent complications that were poorly controlled received a score of 1. When pseudotumor cerebri persisted and did not improve despite medical and/or surgical intervention, patients received a score of 1. Similarly, if posterior fossa reexploration did not relieve recurrent Chiari symptoms, patients received a score of 1. Persistent shunt malfunction and/or infection that did not respond to surgical intervention led to a score of 1. This meant that patients experienced consistent worsening between shunt surgeries.

Each patient was also placed into a gestalt outcome group of "improved," "unchanged," or "worse" (I/U/W) by the rating team. The determination of patient outcome was based on overall change of factors that affected quality of life from their preoperative baseline. Partial to full resolution of symptoms leading to improvement in quality of life placed a patient in the "improved" outcome group. However, resolution of preoperative symptoms had to be weighed against new postoperative symptoms and complications such as pseudotumor cerebri. Recurrence of symptoms such that quality of life was similar to preoperative baseline placed a patient in the "unchanged" outcome group. Worsening of overall quality of life, whether from increasing severity of symptoms or recurring complications after PFD, placed a patient in the "worse" outcome group. We used the gestalt and inherently unclear nature of this grading system for comparison with CCOS scores and to relate to Chiari treatment outcomes literature. Patients were stratified by their total Chiari scores to show the distribution of I/U/W outcomes based on total score. Three groups of score ranges were defined before that analysis of the scores or their relationship to the I/U/W scores were investigated: 4 to 8, 9 to 12, and 13 to 16 to correspond with the I/U/W categories. The percentages of I/U/W patients within each of these score ranges were determined. Patients were also stratified by their outcome group, I/U/W, and then median Chiari scores (with ranges) were found for each of the 4 categories within each group as well as median total outcome scores (with ranges). The CCOS is a nonlinear scale, similar to the Glasgow Coma Scale. Reports in the literature have shown the application of median and ranges to cohorts

of patients analyzed with a nonlinear scale, such as the Glasgow Coma Scale.^{21,22} A one-way analysis of variance was used to compare the total scores between the 3 outcome groups, I/U/W; a *P* value of < .05 was considered statistically significant.

Interrater reliability of the retrospective chart analysis was examined. Sixty charts were randomly chosen such that the distribution of I/U/W within the 60 charts matched the initial distribution of the 146 patients. Thirty of the 60 charts were chosen at random. These 30 charts were reevaluated by all 5 researchers in the same manner as the initial retrospective chart analysis. The results of these 30 chart analyses were examined to find the variation in both the total CCOS score and the gestalt outcome group assigned. The Fleiss *k*-statistic was used to assess interrater agreement of gestalt group assignment.

RESULTS

The mean time from initial surgery to the last clinic visit was 29.3 ± 29.8 months (range, 1–142). Although Chiari patients usually show improvement after surgery, there are those whose symptoms are recalcitrant and follow-up of any length is adequate for evaluation and scoring. Of those whose symptoms resolve and then recur, the recurrence of symptoms is generally 6 to 12 months after their initial surgery.^{3,13,16} Given that trend, we believe that patients with follow-up of at least 1 year can be compared. In our cohort of 146 patients, 99 patients had their last clinic visit at our institution 1 year or more after surgery. Thirty-two of the remaining 47 patients had their last clinic visit less than 1 year after surgery at our institution, but informed the senior author or his staff of their status via local follow-up 1 year or more after surgery. Of the total 146 patients, 101 patients (69.2%) had improved outcome, 39 patients (26.7%) had unchanged outcome, and 6 patients (4.1%) had worse outcome. Stratifying patients by total scores showed that patients who were labeled as improved after PFD predominantly achieved outcome scores between 13 and 16 (Figure 2). Similarly, patients who were labeled as unchanged predominantly achieved scores between 9 and 12, and patients who were labeled as worse achieved scores between 4 and 8. When grouping patients into score ranges, 94 patients achieved a score between 13 and 16. Of those, 93 were graded as improved, 1 was unchanged, and none were worse. Forty-five patients achieved scores between 9 and 12. Of those, 8 were graded as improved, 36 were unchanged, and 1 was worse. Finally, 7 patients achieved scores between 4 and 8. In this range, none were graded as improved, 2 were unchanged, and 5 were worse (Table).

Figures 3, 4, and 5 show the distribution of scores within each of the 4 postoperative categories, median category scores (with ranges), and median total outcome score (with ranges) when patients were stratified by I/U/W outcome group. The improved patients had a median total score of 14 (range, 10–16), with median individual category scores of 3 (range, 3–4), 3 (range, 2–4), 4 (range, 2–4), and 4 (range, 2–4) (pain symptoms, nonpain symptoms, functionality, complications). The unchanged patients had a median total score of 10 (range, 7–13) with median individual category scores of 2 (range, 2–4), 2 (range, 1–4), 3 (range, 1–4), and 2 (range, 1–4). Patients with worse outcome had a median total score of 7.5 (range, 4–9) with median individual category scores of 2 (range, 1–2), 2 (range, 1–2), 2.5 (range, 1–3), and 1.5 (range, 1–2). One-way analysis of variance showed that the differences in total

scores between all 3 outcome groups, I/U/W, were statistically significant ($F = 169.69$, $F\text{-crit} = 3.06$, $P < .001$).

The examination of interrater reliability showed an absolute average agreement of 88% with regard to which gestalt outcome group, I/U/W, a patient was placed into ($\kappa = 0.57$). The main source of discrepancy was found between patients being placed into either the “unchanged” or “worse” outcome group. When solely analyzing the “improved” outcome category, there was an absolute average agreement of 93% ($\kappa = 0.68$). Each of the 30 charts used for the interrater reliability analysis received Chiari score assessments by the 5 researchers, and standard deviations were found for each of the 30 total outcome scores. The mean score deviation amongst the 5 researchers for total outcome scores was 1.56 ± 0.74 (range, 0–2.86), on the 16-point scale.

DISCUSSION

Chiari malformation type 1 presents in a variety of ways and the guidelines for surgical intervention are not always clear. The presence of certain signs and symptoms are strong indicators for surgical intervention.^{4–8} However, PFD is sometimes performed in the absence of those strong indicators when the relative indications, many of which are sensory in nature such as intractable head pain, are such that possible improvement is believed to outweigh the surgical risks. We describe an outcome-scoring system (the CCOS) that quantifies specific aspects of outcomes following PFD in order to better predict how well surgery will treat a patient with a specific constellation of presenting complaints. Over large populations of patients, CCOS grading would gauge which specific Chiari syndrome presentation before PFD leads to which specific outcome, because not all symptom constellations respond equally to surgery.^{2,9,15,20}

Our CCOS includes both preoperative symptoms and new postoperative symptoms in its assessments. This creates the possibility of confusion or inconsistency when assessing these 2 entities within 1 category. To reduce this possibility, the CCOS has a high threshold for improvement: to receive a high outcome score, all symptoms must improve, without any symptoms worsening (see Patients and Methods). Conversely, a low threshold was used for assessing deterioration: lower scores are given when only 1 symptom (either preoperative or new postoperative) worsens, despite improvement of other symptoms.

The CCOS uses 4 postoperative categories of 4 points each that sum to a total score. The scores were compared with traditional gestalt assessments of “improved,” “unchanged,” and “worse” outcome. We found reliable correlation between the 3 gestalt outcome groups of I/U/W and individual scores assigned in the 4 postoperative categories. Those who improved mostly scored 4’s and 3’s. Those who were unchanged tended to score 3’s and 2’s and those who were worse scored 2’s and 1’s. We also found reliable correlation between the patients’ outcome group of I/U/W and their total scores. The majority of patients who improved after PFD achieved total outcome scores between 13 and 16. A minimum score of 13 serves as a useful cutoff for a general label of “improved.” Twenty-eight patients received a score of 13 (27 improved, 1 unchanged). Of those, 21 patients (all improved) had one of their category scores as a 4, and all others as a 3. This would indicate that the patient had a good outcome

in at least 3 categories and an excellent outcome in 1 category. The remaining 7 patients' scores included two 4's, one 3, and one 2. Because one of the categories scored a 2, at least 1 assessment category was unchanged despite the other categorical assessments showing improvement and resolution. This combination of scores still serves as a useful cutoff for a general label of "improved." No patient received the combination of scores that included three 4's and a 1 (to sum to 13). This was highly unlikely, because it meant one aspect had to drastically worsen in the face of all other aspects completely resolving.

However, despite most of the CCOS total scores coinciding well with the I/U/W scoring, there were outliers. These were patients who were assigned scores higher or lower than would be expected from their I/U/W scores. Patients whose I/U/W score was improved but who had scores of 10, 11, or 12 tended to be in poorer condition before PFD. This situation led to lower CCOS scores as the final outcome, which was "improved" over their preoperative baseline in the I/U/W scale, but could not accumulate more than 12 points. Similarly, patients who were rated I/U/W "unchanged" and scored 7 or 8 and presented with very severe symptoms before PFD were in fact unchanged, but were unable to accumulate more than 8 points based on preoperative symptomatology. These observations point to a nuance of the CCOS in that overall patient outcome score in the CCOS is weighted more toward absolute outcome rather than outcome relative to preoperative symptoms. The merit of this aspect of the CCOS is that absolute outcome (as opposed to relative outcome) can be correlated with preoperative severity of disease—a comparison that the I/U/W scoring cannot support. One patient was I/U/W "unchanged" and scored 13 on the CCOS. Before PFD, this patient had mild pain symptoms and very few nonpain symptoms. The nonpain symptoms largely resolved, but there was a full return of pain symptoms. In comparison with other patients who were I/U/W unchanged, this patient was in relatively better condition before PFD and was able to accumulate more CCOS points than most of the other unchanged patients. The utility of the CCOS can be seen here because this type of patient will have a high scoring outcome with or without PFD and begs the question of whether PFD is needed at all.

One purpose of the scale is to more clearly quantify outcomes so that judgments can be made about conditions before PFD and treatment. Previous reports have also sought to find predictors of outcome based on preoperative symptoms, signs, and associated diagnoses.^{3,23} Higher scores should reflect better outcomes in order for those numbers to be meaningful, although that reflects only absolute outcome. Because the scale grades an absolute level of function (which, in theory, can be graded preoperatively) in 3 of the 4 subscores, global preoperative function can be used as a predictor for CCOS score. Although higher scores can still be associated with an unchanged outcome and lower scores can still be associated with an improved outcome, this trend was in the minority and reflects the small number of patients with extremely good or extremely poor preoperative state. The dominant trend in the application of the CCOS is that higher scores reflect better outcomes and lower scores reflect worse outcomes, as seen in Figure 1.

We found that CCOS score groupings (4–9, 8–12, 13–16) from approximately 8% of the patients did not correlate with the corresponding I/U/W outcome group. These patients can be examined to find trends or reasons for the discrepancy between CCOS score and I/U/W

outcome, as described above. The CCOS score is weighted toward absolute outcome in the function and complication subscores rather than outcome relative to preoperative state (as in the entire I/U/W scale) and insight can be gained from this detail. Examination of the 4 separate outcome subscores in an individual patient illuminates preoperative symptoms that do or do not respond to PFD, such as in the patients who were in the I/U/W unchanged category yet scored above 12 or below 9 in the CCOS because of relative changes in combination with absolute outcome events. In this sense, the CCOS does describe specific preoperative conditions that lead to specific types of postoperative outcomes, as it is intended to do.

There was a high degree of agreement between the 5 raters with regard to which gestalt outcome group, I/U/W, a patient was placed into. Rating of outcome groups is assumed to be somewhat more subjective and influenced by the interpretation of the rater. Degrees of change over time are condensed down to a single category. Despite the subjective nature of this assessment method, there was an absolute average interrater agreement of 93% with regard to patients being placed into the “improved” outcome group. There was a lower degree of agreement with regard to the “unchanged” and “worse” outcome groups, suggesting that it is easier to recognize when patients’ overall condition improves. This is reassuring because the majority of patients tend to be labeled “improved” after Chiari surgery, both in our experience and in the analyses done by other groups.^{13–19} However, the degree or manner of improvement remains unspecified and continues to be a limitation with this style of I/U/W assessment.

We would like to define a system that aids us in determining which Chiari patients are most likely to benefit from decompressive surgery, and be able to define the details of the benefit provided. We found reliable correlation between the gestalt outcome groups and CCOS scores, which suggests that the CCOS may not be more useful than the gestalt outcome groups for “overall” assessment of outcome. However, extra detail that the CCOS subcategories provide further validates the overall assessment and allows outcome prediction to include specific categories of postoperative symptoms and function. This particular strength of the scoring system allows not only an overall assessment, but also the assignment of predictive value to a preoperative presentation that correlates with specific individual postoperative symptoms—allowing enhanced ability to prepare patients for possible outcome scenarios.

It is difficult to translate the subjective experience of pain into truly objective measures. The Brief Pain Inventory has been validated in various studies in patients with cancer and in patients with chronic nonmalignant pain. Although the Brief Pain Inventory can reliably track changes in pain over time, the instrument is 8 pages in length and would be rather unwieldy to use in combination with the rest of the CCOS categories. We recognize that the CCOS relies on some subjective determination of pain and nonpain symptoms. However, we believe that a compromise between a user-friendly outcome scale and objective measures of a subjective experience is most useful. This compromise provides some objective qualifiers to the assessment of pain and nonpain symptoms, yet keeps the outcome scale easily accessible for the purpose of correlating preoperative state to postoperative outcome.

The CCOS was applied in a retrospective manner and will be validated with a prospective study in the future. A standardized set of questions for the 4 categories of the CCOS will be beneficial for such a prospective study. It is difficult, however, to precisely determine what these questions should include and exclude; this will be more completely addressed in the future prospective study. The following are an initial set of questions that reflect how the CCOS was designed to be used; these are illustrated in Figure 6.

For the Pain and Nonpain Categories

(1) “Is the patient completely without any Chiari-related pain/non-pain symptoms?” If yes, the score is 4. If no: (2a) “Are the patient’s preoperative Chiari-related pain/nonpain symptoms improved or well managed medically?” and/or (2b) “Are any new postoperative Chiari-related pain/nonpain symptoms managed so as not to interfere with the patient’s activity?” If yes to both questions 2a and 2b, the score is 3. If no to either question: (3a) “Are the patient’s preoperative Chiari-related pain/nonpain symptoms unchanged, and/or (3b) are they worse or are there any new postoperative pain/nonpain symptoms refractory to medical management?” If yes to 3a and no to 3b, the score is 2; if no to 3a and yes to 3b, the score is 1, because the patient is worse than before the surgery.

For the Functionality Category

(1) “Does the patient experience no interference in their daily activities of living from their Chiari-related symptoms?” If yes, the score is 4. If no: (2) “Is the patient able to attend work or school the majority (>50%) of the time?” If yes, the score is 3. If no: (3) “Is the patient able to attend but his/her attendance is significantly impaired (<50% of the time) by his/her Chiari-related symptoms?” If yes, the score is 2. If no: (4) “Is the patient unable to attend work or school due to Chiari symptoms?” If yes, the score is 1.

For the Complications Category

(1) “Did the patient experience no complications after surgery (ie, wound infection, CSF leak, chemical meningitis, increased CSF pressure, deep vein thrombosis, pulmonary embolism, etc.)?” If yes, the score is 4. If no: (2) “Did the patient experience transient complications after surgery that have since resolved (ie, wound infection, CSF leak, etc.)?” If yes, the score is 3. If no: (3) “Does the patient experience a persistent complication that can be managed medically or surgically (eg, anticonvulsants for persistent new-onset seizures, successful shunt placement for persistent elevated CSF pressure, etc.)?” If yes, the score is 2. If no: “Are there persistent and poorly controlled postoperative complications (ie, unstable angina refractory to medical management after perioperative myocardial infarction, etc.)?” If yes, the score is 1.

It should be noted that the nonpain score of the CCOS focuses on symptoms. Therefore, postoperative radiological findings were not factored into the CCOS system. However, a syrinx is a special, often symptom-related “sign” that is intimately tied to Chiari syndrome. It is common to see a syrinx become smaller after surgery but still persist despite symptom resolution, and such an outcome is important to any Chiari outcome scale. Thus, we propose that a preoperatively symptomatic syrinx that is resolved, unchanged, or unresolved postoperatively will be scored (as if it were a “symptom”) in the nonpain category as a 4 if

the related symptoms resolve, as a 3 if the symptoms are reduced but unresolved, as a 2 if the symptoms are unchanged, and as a 1 if the symptoms were increased. Asymptomatic syrinx will be scored (as if it were a “symptom”) in the nonpain category as a 4 for resolution, a 3 for decrease without resolution, as a 2 for not changing, and as a 1 for enlarging. If a syrinx enlarges after surgery, the nonpain category will always be scored as a 1.

Our initial survey of patient charts found 211 first-time surgical Chiari patients. Of these, 28 had limited follow-up at our institution, and 37 did not have enough information in their chart notes to populate all 4 CCOS categories. This introduces a potential source of bias. Because information needed to define a score was not included in the clinic notes of 37 of the 183 eligible patients, it may have been that these specific CCOS categories were much less of a salient issue for a patient and thus not explored during the visit and not documented. If that category was indeed unremarkable, it would likely have led to either no change or an improvement in overall score.

The outcome assessments made by the CCOS are more detailed than those previously described. Pillay et al²⁴ described a classification system in 1991 using MRI. However, the actual outcomes assessment was based on a system described by Levy et al²⁵. This system placed patients into 5 continuous categories purely based on motor function, both in the upper and lower extremity. The range in the lower extremity was described as being from normal unassisted walking to complete plegia. Patients were considered improved/worse if they crossed from 1 motor category to another after surgical intervention. Pillay applied this system to 35 patients, initially separating them based on the presence or absence of a syrinx. Whereas this assessment system has the benefit of placing patients into strict categories, the degree of change is not described. It is not known across how many motor function levels an “improved” patient may have moved. The final level of function is also not known; the “improved” group may be a mix of both high-and low-functioning patients. The CCOS clarifies these uncertainties by assigning each patient a final and absolute outcome level based on the 4 postoperative categories.

Bindal et al²⁶ described a classification and outcomes assessment system in 1995, applied to 22 symptomatic patients that underwent surgical treatment for Chiari syndrome. The classification system categorized patients by the presence or absence of a syrinx, and whether that syrinx was symptomatic. The outcomes assessment system assigned points based on the number of symptoms and signs of each patient. Each patient was then given a single, summed preoperative score and improvement in symptoms was represented by a reduction in the total score. A disadvantage of this system is the inability to compare the degree of change between patients, because the change in score for each patient functions on a relative scale specific to each patient. Patients could have had largely varying degrees of change in score, depending on the initial preoperative score. Our scoring system allows more direct comparison from 1 patient’s score to another. One patient’s degree of improvement is compared with another on the same absolute scale. Also, the system described by Bindal et al combines the various symptoms each patient has into 1 score. In doing so, some of the detail is lost. The scoring system we describe here is able to supply more detail and address changes regarding specific categories following PFD.

The pain symptoms category is separated from nonpain symptoms, because some patient groups may be more likely to improve in 1 category than another, even if there is overall improvement in both categories. Similarly, symptoms from 1 category may be more likely to recur than from another category.³ Providing a functionality category gives a more complete picture of patients' ability to fulfill societal requirements such as work and school. For example, it may be that patients are able to still attend work or school, despite a clinical assessment of unchanged or mildly worsening symptoms. The results of most case series only report changes in typical Chiari symptoms. Mueller and Oro²⁷ provide one of the very few assessments of patients' self-perception of their functional capacity and quality of life before and after PFD. Although their study did not aim to report changes in typical Chiari symptoms, the CCOS combines these various outcome measures into one unified assessment tool. Finally, the complications category takes into account the complexity of risk from surgical sequelae when patients do not have absolute indications for PFD.^{4,16} It is important to note that that complications category includes both typical surgical complications, which are more likely to be random, and complications more specific to Chiari patients and PFD, which are less likely to be random. The latter category includes late scarring of fourth ventricular outflow pathways and elevated CSF pressures (pseudotumor cerebri, which has been observed after Chiari surgery).^{28,29} It may be that mild symptoms in fact do not warrant PFD enough to outweigh the risk of surgical complications.^{3,4,16,30}

Limitations of using simple outcome labels, such as the I/U/W, are the subjective nature of the evaluation and absence of uniform outcome measures.^{7,31} In previous assessments, the degree of improvement needed in order to label a patient as "improved" is not always clear. The issue is further confused if several markers (quality of life, clinical change, absence of complications) are grouped under one heading, such as "improved" or "not improved."^{13–19,23,32–36} A patient may improve in one aspect but worsen in another by differing degrees, and may still have either an overall improved or unchanged outcome. The CCOS aims to lessen this subjectivity by assigning a number based on defined levels in well-separated categories. The present study is retrospective in nature. Several items within the study will be improved when we conduct prospective work in the near future. For example, we can implement the use of a preoperative assessment to compare with the postoperative CCOS. Also, during patient clinic visits we can actively acquire information to fill in the CCOS categories allowing all patients to be assessed by the CCOS. Such prospective data collection may also reduce variability in interpreting subjective reporting of symptomatology. Finally, application of the CCOS several times to the same patient at predefined time intervals after PFD also affords the ability to characterize change over time. Ultimately, it aims to steer away from the sole use of nomenclature such as "improved," "unchanged," and "worse." The scoring system aims to provide a more quantifiable method amenable to statistical analyses.

CONCLUSION

The CCOS, applied retrospectively to a group of Chiari surgical patients, assigns higher scores to those who were improved after PFD and proportionately lower scores to those who were unchanged or worse. This system allows for a more unified outcome assessment system with specific assessment of subscores that can be used in other studies examining

recurrence of specific types of symptoms and prediction models. Further application of the CCOS will refine its use and improve its ability to assess operative Chiari outcomes.

ABBREVIATIONS:

CCOS	Chicago Chiari Outcome Scale
CM1	Chiari malformation type 1
I/U/W	“improved,” “unchanged,” or “worse”
PFD	posterior fossa decompression

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Chicago Chiari Outcome Scale				
<u>Pain</u>	<u>Non-pain</u>	<u>Functionality</u>	<u>Complications</u>	<u>Total Score</u>
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

FIGURE 1.
The Chicago Chiari Outcome Scale.

FIGURE 2.

Chiari patient outcomes stratified by total CCOS score to show the distribution of “improved,” “unchanged,” and “worse” outcome group. CCOS, Chicago Chiari Outcome Scale.

FIGURE 3.

Median scores, ranges, and percentage of distribution of scores within the 4 postoperative categories of the Chicago Chiari Outcome Scale for 101 Chiari patients (69.2%) with improved outcome. Sx, symptoms.

FIGURE 4.

Median scores, ranges, and percentage of distribution of scores within the 4 postoperative categories of the Chicago Chiari Outcome Scale for 39 Chiari patients (26.7%) with unchanged outcome. Sx, symptoms.

FIGURE 5.

Median scores, ranges, and percentage of distribution of scores within the 4 postoperative categories of the Chicago Chiari Outcome Scale for 6 Chiari patients (4.1%) with worse outcome. Sx, symptoms.

FIGURE 6.

Flowchart illustrating an initial set of questions that reflect how the CCOS was designed to be used. CCOS, Chicago Chiari Outcome Scale.

TABLE.Chiari Patient Outcomes Categorized by CCOS Score Ranges and Outcome Groups^a

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

^aCCOS, Chicago Chiari Outcome Scale.