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Positive and negative predictors for good outcome after decompressive surgery for Chiari malformation type 1 as scored on the Chicago Chiari Outcome Scale

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Abstract

Objective: Posterior fossa decompression (PFD) is commonly applied as treatment for Chiari malformation type 1 (CM1), an entity which is associated with a variety of presenting symptoms but little data correlating symptoms to surgical outcome. We applied the Chicago Chiari Outcome Scale (CCOS), a novel 16-point tool for evaluating outcome, to a consecutive series of CM1 patients to identify specific factors or symptoms that predispose to a better or worse surgical outcome.

Methods: A series of 167 CM1 patients who underwent initial PFD at our institution (consisting of suboccipital craniectomy, C1 laminectomy, subarachnoid exploration, and expansile autologous pericranial duraplasty) were reviewed. Pre-operative signs, symptoms, and characteristics were recorded, and odds ratios were calculated to identify significant pre-operative factors corresponding to a better or worse outcome on the CCOS.

Results: Sensory deficits and peripheral neuropathy correlated with a lower score on the CCOS. Younger age at the time of surgery and, strikingly, presence of syringomyelia both correlated with a higher CCOS score.

Discussion: Our results identify specific presenting factors that correlated with a better or worse outcome after CM1 decompression. These data also demonstrate that CCOS scoring allows for a rigorous comparison of outcome in different patient populations and between variable operative techniques. 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.

Keywords

Arnold–Chiari malformation type 1; Outcome assessment; Patient; Prognosis; Syringomyelia; Surgery

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Background

Chiari malformation type 1 (CM1) is characterized by herniation of the cerebellar tonsils through the foramen magnum leading to compression of the posterior fossa contents and frequently a pathological obstruction of cerebrospinal fluid flow out of the fourth ventricle.[1,2] Syringomyelia is a frequent associated finding; it is estimated that over 90% of all cases of syringomyelia occur in the presence of CM1.[3] The precise pathophysiology of the symptoms and of the associated syringomyelia remains poorly understood.[1,2,4]

The clinical manifestations of CM1 are diverse. The most common symptoms are occipital headache that occurs or worsens upon the Valsalva maneuver; radiating neck pain; peripheral neuropathy (including dysesthesias and paresthesias); syncope, or 'drop attacks;' tinnitus; dysphagia; cerebellar dysfunction (including ataxia and tremor); vertigo or dizziness; and incontinence.[1]

Though most published data agrees that posterior fossa decompression (PFD) can treat the CM1 syndrome, there remains controversy regarding operative technique and patient selection based on individual risk and benefit.[5] Specifically, why some patients do not experience improvement in symptoms despite demonstration of adequate decompression at the foramen magnum, as evidenced by MRI CINE flow studies[6] or anatomical MR imaging,[7,8] is poorly understood. As pointed out by McGirt and colleagues, symptom recurrence despite adequate decompression may reflect poor patient selection.[8]

We recently introduced a quantitative scale for assessing outcome of Chiari decompression, the Chicago Chiari Outcome Scale (CCOS).[9] In the present study, we applied the CCOS to a group of 167 consecutive patients who have undergone initial PFD for CM1 at our institution in order to identify various demographic and clinical factors associated with CM1 that correlate significantly with improved or worsened outcome after surgery.

Methods and Patients

Patient selection, surgical procedure and follow-up

Charts of 245 consecutive surgical patients who underwent PFD for CM1 over a 12-year period at our institution were reviewed. Of these patients, 210 were first-time PFD patients. All patients underwent suboccipital craniectomy, C1 laminectomy, subarachnoid exploration, and placement of autologous pericranial expansile duraplasty. The senior author (DF) performed over 95% of these operations and the remainder were performed by faculty surgeons trained at our institution, indicating that this patient group underwent uniform application of a specific PFD surgery. All clinic notes at follow-up were generated under the supervision of the operating faculty surgeons.

Of these 210, 29 were lost to follow-up; and 14 lacked sufficient information in their follow up clinic notes to be able to retrospectively apply all four subscores of the CCOS (Table 1). For the remaining 167 patients, common pre-operative signs, symptoms, and demographic information were extracted from the charts.

Patients were divided randomly among five independent raters for assignment of post-operative categories and scores. To determine inter-rater reliability, 30 patients were selected at random and scored by all five raters, as previously described.[9] Surgical outcomes were assessed at the last point of follow-up with the senior author, either by clinic visit or communication with the patient's primary care physician.[9] It has been demonstrated in several series, that Chiari patients frequently will experience some improvement in their symptoms after decompressive surgery, only to have a recurrence 6–12 months after surgery.[8,10,11] Patients were instructed to follow up regularly up to 1 year after surgery, either in our clinic or with their primary care physician; if they were well and stable, they were discharged from follow-up. One hundred and nine patients had their last documented clinic visit at our institution at least 1 year or more after surgery (range: 12–142 months); 39 more had their last documented visit less than 1 year after surgery (range: <1–11 months), but informed the senior author of their status via local follow-up 1 or more years after surgery. The remaining 19 patients were unable to be assessed 1 year or more after surgery (last clinic visit with senior author, range: 2–11 months).

The CCOS consists of four categories, each scored from 1–4: these include pain symptoms, non-pain symptoms, ability to perform daily responsibilities, and surgical complications. The numerical score assigned in each category represents the change from pre-operative baseline to post-operative state at the time of assessment in each category. These four categories are summed to generate a composite score, ranging from 4–16, reflecting the overall outcome of surgery. In the present study, we clarified the initially published scoring criteria:[9] patients who lacked pain symptoms in their initial presentation and who still had no pain symptoms post-operatively received a score of '4' in the pain subcategory. Likewise, patients who lacked non-pain symptoms in their initial presentations and who still had no non-pain symptoms post-operatively received a score of '4' in the non-pain subcategory.

Patient demographics

For the 167 patients analyzed, 37% were male and 63%, female. Fifty-four percent were under the age of 18 at diagnosis, and 53%, at the time of surgery. Thirteen percent had a family history that included CM1 (Table 2).

For each of the 167 patients, we recorded the following signs and symptoms associated with CM1 as present (or absent) immediately before surgery: Headaches associated with the Valsalva maneuver, 44%; neck pain, 34%; peripheral neuropathy (including dysesthesia, paresthesia, and hyperesthesia), 42%; syncope, 16%; tinnitus, 21%; and dysphagia, 17%; cerebellar dysfunction symptoms (ataxia, self-reported gait abnormalities, or tremors), 25%; vertigo or dizziness, 17%; bowel or bladder incontinence, 7% (Table 3); pinprick loss, 44%; positive Romberg sign, 7%; paresis, 18%; and cognitive impairment, 15% (Table 4). Cognitive impairment included developmental delays, memory impairment, and any self-reported changes in cognitive abilities. Eight patients presented with scoliosis (7%). One hundred and ten of the patients had some documentation of the duration of the primary symptom. Eighty five of these had symptoms lasting 1 year or longer (Table 3). Only one patient had concomitant hydrocephalus.

Presence of a syrinx and the degree of tonsillar herniation were determined either directly from MR images or from written reports. Forty-three percent of patients had a documented syrinx pre-operatively (Table 5). The size ranged from less than a single vertebral level to a holocord syrinx, and the location was predominantly in the cervical and thoracic regions. We categorized the degree of herniation by vertebral level, as our patients varied greatly in both age and size. The degree of herniation was able to be evaluated only in 62 of the 167 patients; 43 had pre-operative MRIs accessible to the research staff, and 19 had the degree of herniation (in terms of vertebral level) in either a pre-operative radiology report or clinic note. Of these 62, eight had cerebellar tonsils that did not extend past the foramen magnum; 10 herniated to between the foramen magnum and C1; 28, to C1; 6 between C1 and C2; and 10 to C2 or beyond (Table 5). As for vertebral abnormalities, no patients were treated for basilar invagination or with craniocervical fusion, and none manifested instability post-operatively.

Results

CCOS total scoring distribution

The CCOS total scores ranged from 4–16. For the 167 patients analyzed, 112 scored a 13 or higher on the CCOS; 48, between 9 and 12; and 7, between 4 and 8. We chose to split the scores into only two groups for the purpose of our analysis in assessing ‘better’ versus ‘worse’ outcome. We tested several stratification schemes within the scoring system to determine a cut off for ‘improved’ outcome: 4–12 versus 13–16; 4–11 versus 12–16; 4–10 versus 11–16; and 4–9 versus 10–16. We determined that the cutoff between 4–10 and 11–16 was the most appropriate. Analysis to this level produced the greatest number of significant outcome odds ratios (ORs).

Therefore, those with higher scores (11–16) were deemed to have experienced a good outcome from surgery; those with lower scores (4–10) were deemed to have experienced a poor outcome from surgery. In our 167 patients, 137 (82%) scored an 11 or higher on the CCOS, while 30 (18%) scored a 10 or below. Of those 30 who experienced a ‘poor’ outcome from surgery, we have post-operative CINE flow data for 20. Despite experiencing a recurrence of symptoms, 11 of these had completely normal flow at the foramen magnum, and 7 more had some, albeit reduced, flow. Only 2 of the 20 showed no flow. This further supports the need to identify which factors predispose patients to symptom resolution with surgical intervention, as anatomical decompression is not strictly correlative.

Symptoms, signs, and outcome associations

ORs were generated to identify factors correlative with a better (CCOS 11–16) versus a worse (CCOS 4–10) outcome. ORs were considered to be significant if the 95% confidence interval excluded 1, and if the P value was <0.05. The outcome association of each presenting symptom or sign is shown in Tables 3–5. All factors demonstrating a significant correlation (OR) with outcome are summarized in Table 6. Neither gender nor a history of familial Chiari I correlated significantly with total CCOS score.

Patients presenting with peripheral neuropathy (dysesthesia, parasthesia, or hyperesthesia) were 2.91 times more likely to experience a poor outcome of surgery (1.28–6.61; $P<0.05$). Patients presenting with peripheral neuropathy and Valsalva headache were 2.85 times more likely to experience a poor outcome of surgery (1.22–6.63; $P<0.05$). Patients presenting with peripheral neuropathy and neck pain were 2.49 times more likely to experience a poor outcome of surgery (1.03–6.00; $P<0.05$). Patients presenting with peripheral neuropathy and syncope were 4.64 times more likely to experience a poor outcome of surgery (1.43–15.02; $P<0.05$) (Table 6). No other specific symptom or pairwise combination of symptoms produced significant ORs.

No specific individual sign correlated significantly with outcome, although loss of sensitivity to pinprick was strongly trending towards significance (OR: 2.23; 95% CI: 0.998–5.01; $P=0.0504$). Patients presenting with pinprick loss and a positive Romberg sign were 4.06 times more likely to experience a poor outcome of surgery (1.02–16.15; $P<0.05$), and patients presenting with pinprick loss and paresis were 3.18 times more likely to experience a poor outcome of surgery (1.05–9.56; $P<0.05$). Patients presenting with peripheral neuropathy and pinprick loss were 3.12 times more likely to experience a poor outcome of surgery (1.35–7.19; $P<0.05$) (Table 6). No other specific sign, pairwise combination of signs, or pairwise combination of signs and symptoms produced significant ORs.

There were three factors found to be positive prognostic indicators, i.e. they correlated with a favorable outcome of surgery. Patients who were less than 18 years old at the time of surgery were 2.72 times more likely to have a good outcome than those who were 18 years or older at the time of surgery (1.18–6.26; $P<0.05$) (Table 6). Male patients under the age of 18 years at the time of surgery were even more likely to benefit from surgery (OR: 3.39; 95% CI: 1.12–10.30; $P<0.05$). This is consistent with a variety of reports in the literature that pediatric patients experience better outcomes of decompressive surgery than do adult patients.[12]

Surprisingly, the degree of herniation was not found to correlate significantly with outcome. Perhaps this is due to other anatomical nuances of cerebellar tonsillar anatomy other than length. The most striking finding, however, was that in our series patients with a pre-operative syrinx were 3.94 times more likely to experience a good outcome of surgery (1.52–10.25; $P<0.05$) (Table 6). This directly contradicts the findings of many other studies, which demonstrated that patients with syringomyelia were more likely to experience a poor outcome from surgery.[4,10,13,14] It should be noted that the ORs for a poor outcome for patients without syringomyelia who had peripheral neuropathy trended towards significance (OR: 2.42; 95% CI: 0.95–6.19; $P=0.0644$). Likewise, the OR for a poor outcome for patients without syringomyelia who presented with both pinprick loss and peripheral neuropathy trended towards significance (OR: 2.57; 95% CI: 0.95–6.94; P=0.0623). These trends did not hold for patients with syringomyelia and the respective presenting symptomatology. A prospective study validating these factors as predictive, and allowing for uniform assessment of CCOS scores at set post-surgical time points, will be essential in the future.

Discussion

Decompressive surgery is successful at providing symptom relief for many Chiari patients; however, some patients do not benefit from surgery despite achieving adequate anatomic decompression. Currently, this discrepancy is poorly understood, often making it difficult to advise a patient on whether surgery is the best option for an individual case. Therefore, a determination of which pre-operative characteristics correlate with better outcomes is very helpful. Identifying which subsets of patients for whom the benefits of surgery do and do not outweigh the risks will aid in optimally advising patients about what to expect from surgery. Such data will reduce the incidence of surgical treatment in patients whose outcome would have been better with a more conservative medical management option.

Several attempts to identify which patients respond favorably to decompression have been published, but have largely suffered from small sample size and a poorly defined outcome scoring system.[8,15–17] The most commonly utilized outcome description is that of 'improved', 'unchanged', or 'worse'.[7,8,10–12,14,16,18–20]

These poorly defined categories fail to represent the distinct components of the goals of decompression: to improve symptoms and quality of life with minimal risk. Furthermore, the 'unchanged' category in the conventional outcome scale is ambiguous, i.e. it can include patients who were the same as at baseline as well as patients who experience improvement in some aspects but no change or worsening in others. These shortcomings prompted us to develop a more quantitative system of evaluating post-operative outcome for CM1 decompressive surgery that combines the objective and subjective improvement in the patients' signs and symptoms with the self-perceived benefits to quality of life and the risks of surgery,[9] the CCOS. This has allowed us to identify pre-operative factors that correlate with outcome, based on four discrete categories relevant to the CM1 patient: pain symptoms, non-pain symptoms, ability to perform daily responsibilities, and surgical complications.

To identify pre-operative factors that correlated with outcome, we utilized a binary system (CCOS 4–10 versus 11–16). This proved to be the most significant cutoff statistically, i.e. producing the greatest number of significant ORs for correlation between pre-operative factors and outcome. We believe that this represents resolution of the ambiguity associated with the 'unchanged' category. It also fits with the design of the CCOS, which has a high threshold for improvement and a low threshold for deterioration.

In the present study, peripheral nerve symptoms, either neuropathy (including dysesthesia, paresthesia, and hyperesthesia) or loss of sensitivity to pinprick, correlated with a poor outcome after surgery. The significant pairwise combinations of symptoms also center around these two categories. This agrees with the previous finding that patients with sensory deficits are unlikely to experience improvement.[16] It has previously been argued that sensory or motor deficits and syringomyelia may represent a more advanced stage of the disease.

It is not surprising that age under 18 years at the time of surgery correlated with higher CCOS scores as there are significant differences between the presentation of CM1 in adult and pediatric patients.[10] Children and adults may even vary in response to specific surgical treatments: multiple studies have demonstrated that pediatric patients do better with bone-

only decompression and a conservative craniectomy,[10,11,19,20] though the meta-analysis done by Durham and Fjeld-Olenec, pooling studies from several centers, demonstrated a decreased risk for re-operation when PFD was combined with duraplasty in children.[17] Several studies have demonstrated greater symptom relief in adult patients with syringomyelia when duraplasty is utilized. Some studies have even gone so far as advocating tonsillar resection,[7,14] though there is also objection to such a technique.[11,20,21] It would be possible to better compare these techniques, as well as others such as dura splitting decompression,[22] utilizing the CCOS as a standardized quantitative outcome assessment.

The most striking finding in our study is that patients presenting with syringomyelia were significantly more likely to achieve a better outcome score. This directly contradicts the findings of several other studies,[4,7,10,13,14,23,24] one of which was in a pediatric population only,[10] where the correlation trend between syringomyelia and poor outcome was not statistically significant. It is possible that our technique, which routinely includes duraplasty, provides better symptom relief for patients with syringomyelia, as has been reported previously.[7,23,24]

The other major radiological finding-evaluated degree of tonsillar herniation, did not correlate significantly with outcome. This is not altogether surprising, as the natural history of CM1 is poorly understood. Even after symptoms present, they can spontaneously resolve, [25] though in many, CM1 symptoms slowly progress over long periods of time, with intermittent periods of stability and/or regression.[4,12] Several studies have demonstrated that patients operated earlier in their symptomatic course will respond better.[12,15,16] Many patients with CM1 and significant tonsillar herniation will be diagnosed while asymptomatic with a chance of remaining so over an extended period of time.[1,2,26] Conversely, there is an increasing awareness of a small subset of patients who manifest with a syrinx or other convincing CM1 symptomatology with no discernable herniation.[1,2] It is also widely accepted that the cerebellar tonsils may ascend with age,[27] making the diagnosis of CM1 by tonsillar herniation alone imprecise. Is it possible that other radiological features, such as posterior fossa volume[28] or dynamic cerebrospinal fluid flow imaging,[29] may present a more precise metric for predicting outcome via the CCOS in future studies.

Another possible contributing factor may lie in the fact that our institution has long favored early surgical intervention, with syringomyelia constituting an absolute indication to offer surgery. Many experts believe CM1 to be a progressive disease,[4,12] with patients experiencing better outcomes with early surgical intervention.[8,12,15,16] While the present study does not directly address this question, this is a potentially contributing factor to these unusual results. The most likely contributing factor is that by using a numerical scoring system, we are better able to sort through and detect some form of improvement in the patients that otherwise would have been categorized as ‘unchanged’ in the simple improved/unchanged/worse categorization. This represents the greatest strength of the CCOS, in that it can be used to more thoroughly evaluate patient outcome after decompressive surgery for CM1, allowing for more rigorous comparisons across patient demographics, symptomatology, and a widely variable operative technique.

Analysis of a larger population of patients will be necessary in order to resolve several of the ORs presented here. A larger sample size will also allow examination of a larger group of predictive factors as Dyste *et al.* did to identify the triplet combination of atrophy, ataxia and scoliosis.[16] Another weakness of the study is the inherent bias in retrospective data gathering. Nonetheless, we have identified certain factors as strongly correlating with higher or lower CCOS scores indicating a good or poor overall outcome. These data give quantitative clarity to how patients are likely to benefit from this procedure. In our hands, younger patients and patients presenting with syringomyelia are more likely to experience a good outcome of surgery, and patients presenting with peripheral neuropathy or loss of sensitivity to pinprick are frequently less likely to experience a good outcome from surgery.

We believe that this scale can be applied to determine which sets of patients respond favorably to distinct surgical techniques, resolving lingering questions of surgical intervention versus medical management, as well as which surgical method is most appropriate for which patient. As the scale can easily be applied retrospectively via chart review, it will also facilitate pooling of data for meta-analysis to produce more generalizable results across broader populations of patients. This information has the potential to affect patient selection for surgery by allowing better-informed decisions regarding the likelihood of a good outcome with a specific constellation of presenting symptoms.

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Table 1

Chicago Chiari Outcome Scale scoring criteria[9]

Chicago Chiari Outcome Scale				
Pain	**Nonpain**	**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

Table 2

Patient demographics and pertinent medical history

Characteristic	*N* (incidence %)	CCOS ≥11
Male; female	62; 105 (37%; 63%)	55/62 (89%); 82/105 (78%)
<18 years old at diagnosis	90 (54%)	79/90 (88%)
<18 years old at surgery	89 (53%)	79/89 (89%)
<18 years old at surgery+male	51 (31%)	47/51 (92%)
(+) Familial Chiari 1	21 (13%)	18/21 (86%)

Table 3

Incidence and outcome association of signs evaluated

Symptom	N (incidence %)	CCOS >11
(+) Valsalva headache	73 (44%)	56/73 (78%)
(+) Neck pain	57 (34%)	46/57 (81%)
(+) Peripheral neuropathy	70 (42%)	51/70 (73%)
(+) Syncope	27 (16%)	19/27 (70%)
(+) Tinnitus	35 (21%)	27/35 (77%)
(+) Dysphagia	28 (17%)	28/35 (80%)
(+) Cerebellar symptoms	41 (25%)	37/41 (90%)
(+) vertigo/dizziness	29 (17%)	22/29 (76%)
(+) Incontinence	12 (7%)	10/12 (83%)
(+) Scoliosis	8 (5%)	8/8 (100%)
Primary symptoms lasting >1 year before surgery	85/110 (77%)	67/85 (79%)

Table 4

Incidence and outcome association of symptoms evaluated

Signs	*N* (incidence %)	CCOS ≥11
(+) Pinprick loss	73 (44%)	55/73 (75%)
(+) Romberg sign	12 (7%)	8/12 (67%)
(+) Paresis	30 (18%)	21/30 (70%)
(+) Cognitive impairment	25 (15%)	22/25 (76%)

Table 5

Incidence and outcome association of radiological findings

Radiology										
						N, % (CCOS >/=11, %)				
(+) Syrinx					72 (43%)				66/72 (92%)	
Foramen magnum		FM to C1		At C1		Between C1 and C2		C2 and beyond		
Herniation	8 (13%)	8/8 (100%)	10 (16%)	6/10 (60%)	28 (45%)	21/28 (75%)	6 (10%)	6/6 (100%)	10 (16%)	8/10 (80%)

Table 6

Odds ratios for CCOS <11 (a ‘poor’ outcome) and odds ratios for a CCOS ≥11 (a ‘good’ outcome)

Poor outcome Factor	OR (95% CI) *P* value	Good outcome factor	OR (95% CI) *P* value
Peripheral neuropathy	2.91 (1.28–6.61) 0.0106	<18 years old at surgery	2.72 (1.18–6.26) 0.0184
Peripheral neuropathy + Valsalva headache	2.85 (1.22–6.63) 0.0154	<18 years old at surgery and male	3.39 (1.12–10.30) 0.0310
Peripheral neuropathy + neck pain	2.49 (1.03–6.00) 0.0423	(+) Syrinx	3.94 (1.52–10.25) 0.00489
Peripheral neuropathy + syncope	4.64 (1.43–15.02) 0.0104		
Pinprick loss + (+)Romberg	4.06 (1.02–16.15) 0.0466		
Pinprick loss + paresis	3.18 (1.05–9.56) 0.0400		
Peripheral neuropathy + pinprick loss	3.12 (1.35–7.19) 0.00771		