

# Functional Outcome for Chiari Malformation Using a Novel Scoring System

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## Abstract

**Background** The authors report functional outcomes comparing various surgical techniques, using a Chiari-specific assessment tool. They also intend to externally validate the performance of the CCOS by comparing with gestalt outcome.

**Methods** Total cohort comprised 73 surgically treated patients, and patients were divided into two groups: patients who were operated upon at the authors' institute and those who were evaluated at their institute but underwent surgery elsewhere due to various reasons. Functional outcome was evaluated on the basis of the Chicago Chiari outcome scale (CCOS) and gestalt outcome scale. Mean duration of follow-up was  $10.23 \pm 5.8$  months.

**Results** In the authors' cohort of 73 patients, 76.70% ( $n = 56$ ) were improved, 23.30% ( $n = 17$ ) were unchanged, and none of them deteriorated. The median CCOS was  $14 \pm 1.34$  (range: 11–16). There was no statistical difference in outcome between the different operative groups (foramen magnum decompression, duraplasty, tonsillar resection "other"). The CCOS value of 14 has excellent sensitivity (0.95) and good specificity (0.746) for identifying patients with good gestalt outcome.

**Conclusion** The authors found a clear correlation between higher CCOS score and gestalt outcome. There was no statistical difference in outcome between the different operative groups.

## Keywords

- ▶ Chiari malformation
- ▶ syrinx
- ▶ functional outcome
- ▶ Chicago Chiari outcome scale
- ▶ posterior fossa decompression

## Introduction

Chiari malformation I (CMI) is a fairly common anomaly of the hind brain that was first described over a century ago by Hans Chiari during postmortem examination.<sup>1</sup> Patients usually require surgery for long-term presenting symptoms. Surgical decompression is highly recommended in symptomatic patients with cerebrospinal fluid (CSF) flow obstruction.<sup>2–5</sup> There is little consensus on the surgical technique used for decompression, although it is commonly achieved via a suboccipital craniectomy and C1/C2 laminectomy, with or without dural splitting, duraplasty, tonsillar coagulation and resection, arachnoid lysis, and posterior fossa reconstruction,

with the ultimate goal being an increase in retrocerebellar space and improvement in CSF flow.

Surgical outcome can be quantified subjectively by noting improvement in various symptoms, objectively by clinical examination and outcome scales. Assessment of patient outcomes is often intricate by lack of standardized outcome measures specific to CM-I. To address the deficiency of reporting of validated surgical outcomes, Aliaga et al<sup>6</sup> developed the Chicago Chiari outcome scale (CCOS), which was the only CM-I-specific standardized outcome scale that correlated with well-accepted measures of outcome according to a recent meta-analysis.<sup>7</sup> We report functional outcomes comparing various surgical techniques, using a Chiari-specific

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assessment tool. We also intend to externally validate the performance of the CCOS by comparing with gestalt outcome.

## Patients and Methods

Total cohort comprised 73 surgically treated patients, and patients were divided into two groups: patients who were operated upon at our institute and those who were evaluated at our institute but underwent surgery elsewhere due to various reasons (► Fig. 1). These patients were in regular follow-up after surgery. The operative details of elsewhere operated patients were not analyzed.

Forty patients with CMI and syrinx who underwent surgery at our institute between August 2013 and October 2015 were evaluated. The diagnosis of CMI was made using sagittal T1-weighted magnetic resonance imaging (MRI) studies of the brain and was defined as tonsillar herniation of at least 5 mm below the level of the foramen magnum. Patients undergoing decompression were followed regularly, and outcomes were evaluated based on the last available follow-up. Mean duration of follow-up was  $10.23 \pm 5.8$  months.

## Clinical Outcome Measures

All patients were evaluated by one reviewer using both the gestalt outcome and CCOS. Our primary interest was the overall functional outcome.

Each patient was placed into a gestalt outcome group of “improved,” “unchanged,” or “worse” (I/U/W). The 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. Slight or no change in symptoms that did not affect patients’ quality of life or worsening of their clinical status 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.

The primary clinical outcome measure was the composite CCOS score. The CCOS is a standardized, recently validated tool for evaluating clinical outcome in CM1 patients. Patients were also evaluated for presence of other symptoms and for temporal changes in their symptomatology for pain

symptoms, nonpain symptoms, functionality, and complications, with a total possible score of 16.

## Results

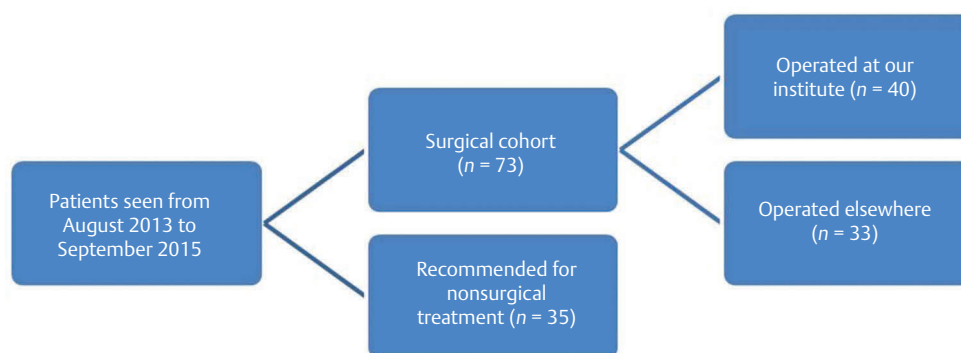
The mean age at surgery was 34 years ( $34.50 \pm 13.82$ , range: 4–60 years); 13% ( $n = 10/73$ ) were younger than 18 years and 43% ( $n = 32/73$ ) were female. Most patients presented with multiple symptoms, including headache (52.10%), neck pain (61.60%), paresthesias (45.2%), motor symptoms (56.20%), scoliosis (13.70%), bulbar symptoms (6.8%), gait disturbances (35.60%), and bladder disturbances (6.80%) (► Table 1). The mean duration of symptoms was 21 months ( $21.57 \pm 20.47$ ).

## Surgical Procedure

Of the 40 patients operated upon at our institute, 28 (70%) underwent foramen magnum decompression with augmentative duraplasty, 5 (12.5%) underwent foramen magnum decompression alone, and 2 (5%) underwent foramen magnum decompression with duraplasty and tonsillar resection/coagulation. One patient underwent foramen magnum decompression with duraplasty with C1–C2 posterior fusion. Two patients underwent foramen magnum decompression with C1–C2 posterior fusion for craniovertebral instability.

**Table 1** Symptoms at initial presentation

Symptoms	Percentage (%)
Headache	52.1
Neck pain	61.6
Paraesthesia	45.2
Motor symptoms	56.2
Numbness	32.9
Wasting	38.4
Sleep disturbance	19.2
Visual disturbance	2.7
Tinnitus	1.4
Bulbar symptoms	6.8
Gait disturbance	35.6
Bladder disturbances	6.8
Scoliosis	13.7



**Fig. 1** Flow diagram showing recruitment process for this study.

Two patients underwent only ventriculoperitoneal shunt for associated hydrocephalus.

### Complications

Surgical complications were assessed throughout patient's postoperative course, from the time of surgical decompression to the last clinic visit. Complications were experienced by four patients after the initial procedure. Three patients experienced CSF-related complications after surgery. One patient had CSF leak from operative site wound, and one developed supratentorial subdural hygromas following decompressive surgery that was managed conservatively. One patient had CSF leak following ventriculoperitoneal (VP) shunt that required shunt removal, and one patient had discharge from wound site that was managed conservatively. None of them had an irreversible injury after surgery.

### Outcome According to Gestalt and Chicago Chiari Outcome Scales

In our cohort of 73 patients, 58 (79.50%) had improved on gestalt scale whereas 15 (20.50%) remained unchanged.

In our cohort of 73 patients, 76.70% ( $n = 56$ ) were improved, 23.30% ( $n = 17$ ) were unchanged, and none of them deteriorated on CCOS. The median CCOS was  $14 \pm 1.34$  (range: 11–16). When grouping patients into CCOS score ranges, 56 (76.70%) patients achieved a score between 13 and 16 and labeled as improved; 17 (23.30%) patients achieved a CCOS score between 9 and 12 and labeled as unimproved.

The aptitude of the CCOS to differentiate patients with good gestalt outcome was plotted by means of a receiver operating characteristic (ROC) curve. The area under the ROC curve was 0.815, signifying excellent prediction of gestalt

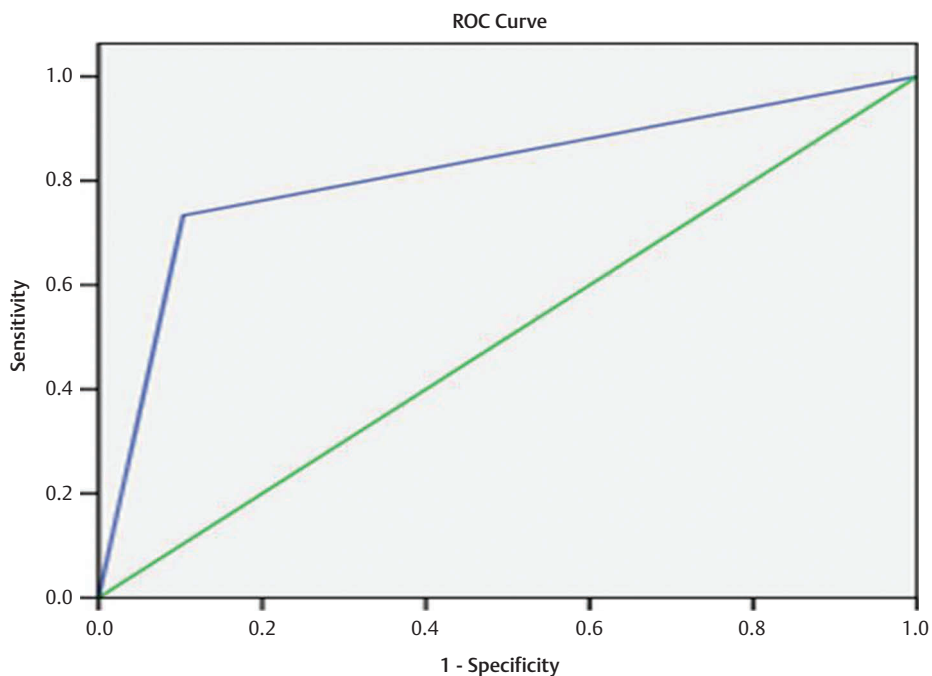
outcome. Sensitivities and specificities for the CCOS were also calculated to determine the optimal CCOS value to use as a cutoff score (► Fig. 2). These results showed that a CCOS value of 14 has excellent sensitivity (0.95) and good specificity (0.746) for identifying patients with good gestalt outcome (► Fig. 2).

### Outcome According to Chicago Chiari Outcome Scale Based on Surgical Technique

Operative details were available only for 40 patients operated at our center. Five patients underwent only bony decompression leaving the dura intact. Posterior fossa decompression (PFD) without opening of the dura improved patients' condition in 80% ( $n = 4$ ) of the cases, whereas PFD with duraplasty and with tonsillar manipulation improved the initial symptomatology in 85.7% ( $n = 24$ ) and 100% ( $n = 2$ ), respectively. One (33.3%) patient who underwent posterior fusion along with decompression reported improvement; 20% ( $n = 1$ ) of patients who underwent bony decompression alone were unchanged whereas in group of patients who underwent duraplasty with bony decompression, 14.3% ( $n = 4$ ) did not benefit from the procedure (► Table 2). Moreover, on chi-square test difference failed to reach statistical significance in improvement among various groups ( $p = 0.143$ ).

### Discussion

Using a novel scoring system, the CCOS, we were able to quantify the functional outcome in a cohort of CMI patients. Aliaga et al have recently introduced a scoring system (CCOS) for assessing outcomes in CMI patients and compared their scoring system with the Gestalt outcome group classifying



**Fig. 2** Graph showing the receiver operating characteristic (ROC) for prediction of CCOS using gestalt outcome scale. The ROC had an area under the curve of 0.815 ( $p < 0.001$ ).

**Table 2** Outcome according to CCOS based on surgical technique

Operative technique	Improved (CCOS 13–16)	Unchanged (CCOS 9–12)
Foramen magnum decompression ( <i>n</i> = 5)	4 (80%)	1 (20%)
Duraplasty ( <i>n</i> = 28)	24 (85.7%)	4 (14.3%)
Tonsillar resection/coagulation ( <i>n</i> = 2)	2 (100%)	0
Shunt ( <i>n</i> = 2)	2 (100%)	0
Posterior fusion ( <i>n</i> = 3)	1 (33.3%)	2 (66.7%)

Abbreviation: CCOS, Chicago Chiari outcome scale.

patients as improved, unchanged, or worse and showed that the scores of their novel scoring system correlated with the Gestalt outcome grouping. They reported that “improved” patients generally scored between 13 and 16, “unchanged” patients between 9 and 12 whereas “worse” patients scored between 4 and 8.<sup>6</sup> Hekman et al used the CCOS to determine positive and negative predictors of good outcome and observed that sensory deficits correlated with lower CCOS scores.<sup>8</sup>

The operative approach remains controversial. Several studies have compared osseous decompression with and without dural opening. Erdogan et al reported similar clinical outcomes between duraplasty and non-duraplasty groups.<sup>9</sup> Durham et al in a meta-analysis concluded no overall significant difference between osseous decompression with and without duraplasty and noted higher recurrence rate when the dura was not opened but a higher rate of complication when it was.<sup>10</sup>

In this study, PFD without opening of the dura improved patient's condition in 80% of the cases, whereas PFD with duraplasty and with tonsillar manipulation improved the initial symptomatology in 85.7% and 100%, respectively. We failed to find statistical significance in improvement among various groups ( $p = 0.143$ , chi-square test) and concluded no overall significant difference between osseous decompression with and without duraplasty; however, the low number of patients treated with PFD limits the statistical power of our study.

When the dura was opened, the surgical complications included one case of CSF leaks—one case of subdural hygroma that subsequently resolved with conservative treatment only and one case of superficial wound infections. When the dura was not opened, none of our patients had a complication. Hayhurst et al reported overall 30% complication rate in the dural opening group compared with 12.5% in the bony decompression group.<sup>11</sup> Alfieri et al reported 2.4% of CSF leak rate, of that 1% required VP shunt. Erdogan et al reported 11% CSF leak rate, all of which were in the duraplasty group, and interestingly, hospital stay in the non-duraplasty group was on average 5.4 days compared with 14.2 days in the duraplasty groups. Overall complication rate in our surgical cohort A was 10%. We conclude that extradural decompression without duraplasty might be associated with lesser complication rates with similar outcome.

## Conclusion

Using a novel Chiari-specific scoring tool, we found improved functional outcome in almost 76.70% of our cohort. In the statistical assessment of the CCOS, we found a clear correlation between higher CCOS score and gestalt outcome. Further refinement and increased use of this validated, disease-specific outcome scale should be encouraged, and it will enable easier comparison of treatment strategies. Surgical decompression without duraplasty provides the benefits of surgical decompression while avoiding the complications of intradural techniques. There was no statistical difference in outcome between the different operative groups (foramen magnum decompression, duraplasty, tonsillar resection “other”), but our study highlights the need of performing randomized controlled trial studies on surgical techniques in CMI patients.

### Conflict of Interest

All the authors certify that they have no affiliations with or involvement in any organization or entity with any financial interest (such as honoraria; educational grants; participation in speakers' bureaus; membership, employment, consultancies, stock ownership, or other equity interest; and expert testimony or patent-licensing arrangements), or nonfinancial interest (such as personal or professional relationships, affiliations, knowledge, or beliefs) in the subject matter or materials discussed in this manuscript.

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### Ethical Approval

For this type of study formal consent is not required.

### Informed Consent

Informed consent was obtained from all individual participants included in this study.

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