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Original Article

A Prospective Study of CSF Flow Dynamics Across Foramen Magnum in Adult Chiari Malformation/Syringomyelia Complex and its Clinical Correlation with Outcomes after Surgery

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Abstract

Introduction Chiari I malformation refers to cerebellar tonsillar descent below the foramen magnum and is frequently associated with syringomyelia. Prior cerebrospinal fluid (CSF) flow studies correlated the clinical severity of these lesions with general flow velocity or bulk flow at the foramen magnum; however, these techniques have not assessed the effect on surgical outcomes. The study aims to present clinical and radiological factors and CSF flow parameters (pre- and postoperative) that affect the surgical outcome.

Materials and Methods The institutional ethics committee approved the study. We collected the prospective clinical data, including pre- and postoperative symptoms. Functional grades were determined along with the change in clinical improvement based on clinical examination notes, the change in functional grade was calculated, and the radiologic data were analyzed according to the degree of clinical improvement. The surgical procedure included suboccipital bony decompression with duroplasty. Patients were followed up at 1 month, 3 months, and 1 year.

Results There were a total of 25 patients. The mean age of the patients was 45.52 ± 13.37 years, with 40% being males and 60% being females. After the surgery, there was a significant increase in the anterior and posterior CSF flows at the foramen magnum. Most had a resolution in the headache and sensory symptoms, while the lower cranial nerve and motor symptoms had a minor resolution at follow-up. Age, sex, and headache do not correlate with the outcome or syrinx improvement. Motor power

Keywords
- Chiari malformation
- syrinx
- CSF flow dynamics
- Craniovertebral junction (CVJ) anomalies
- foramen magnum decompression
- syringomyelia

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in the lower limb (preintervention) and syrinx shape (preintervention) were significantly associated with the variable “anterior flow at the foramen magnum” (preintervention; $p < 0.05$). Average flow, cervicomedullary angle (postintervention), sensory symptom progression, and CSF flow change anterior to the foramen magnum were significantly associated with the variable “anterior flow at the foramen magnum” (postintervention; $p < 0.05$).

Conclusion Even after surgery, persistent foramen magnum CSF outflow obstruction has a robust negative correlation with the outcome. The Chiari outcome predictability index has shown a significant correlation with patient outcomes and can be used to inform patients about the expected outcome. The results of the present study will be helpful in stratifying patients according to their desired outcomes.

Introduction

Chiari malformation is a condition in which brain tissue extends into the spinal canal. It occurs when the posterior fossa is uncommonly tiny. Chiari I malformation refers to cerebellar tonsillar descent below the foramen magnum.[1–3] The malformation is a progressive disease with varying symptoms, ranging from headache to myelopathic features to the involvement of lower cranial nerves.[4,5]

Chiari I malformation is frequently associated with syringomyelia (cystic cavity in the spinal cord).[4,6,7]

The current treatment for Chiari I malformation aims to establish cerebrospinal fluid (CSF) flow at the level of the foramen magnum or address the atlantoaxial joint view of instability.[8,9] These surgeries aim to halt and reverse the progression of syringomyelia and improve the quality of life.[4,10,11] However, the outcome of surgery varies among patients.[10–12]

By conducting CSF flow studies, investigators have attempted to correlate the clinical severity of the disease with CSF flow velocity or bulk flow at the foramen magnum; however, these techniques have not allowed consistent prediction of symptomatology, explanation of the presence of syringomyelia, or the assessment of the hydrodynamic characteristics of decompression. There are multiple studies on the surgical outcome but very few on predictors of outcome.[10,11,13]

Aims and Objectives

- To study the various clinico-radiological factors affecting the outcome in Chiari malformation patients undergoing surgery.
- To study the CSF flow dynamics across the foramen magnum in patients with Chiari malformation before and after the foramen magnum decompression (FMD) surgery.
- We correlate the CSF flow dynamics across the foramen magnum with the preoperative and postoperative clinical status.

Materials and Methods

Study Design

This is a hospital-based prospective cohort study.

Study Population

Patients aged 18 to 70 years with Chiari I malformation who presented between July 2019 and January 2021 and consented to participate were included in the study.

Inclusion Criteria

- Age between 18 and 70 years.
- Symptomatic patients with Chiari I malformation with or without syringomyelia.
- Patients with Chiari 0 malformation and syringomyelia.
- Patients with Chiari II malformation without brainstem descent.

Exclusion Criteria

- Chiari II, III, or IV malformations.
- Patients with tonsillar descent who had no symptoms and syringomyelia.
- Syringomyelia related to tumors.
- Posttraumatic syringomyelia.
- Syringomyelia associated with spinal dysraphism.
- Craniovertebral junction complex bony anomaly causing cervicomedullary junction (CMJ) compression.

Methodology

We collected prospective clinical data, including pre- and postoperative symptom resolution, determined the functional grades and change in clinical improvement based on clinical examination, calculated the shift in functional grade, and analyzed the radiologic data according to the degree of clinical improvement.

Neurologic Assessment

Clinical details were obtained from the medical records. The functional grade was determined using the functional grading system from Noudel et al.[14]

Postoperatively, the patients were followed up in the clinic for between 5 and 40 days (mean = 16.27 days), with continual follow-up at several-month intervals. Clinical improvement was determined by a clinician who assessed only the clinical symptoms but was blinded to the pre- or postoperative radiologic results. Based on their last documented visit, patients were categorized as having no partial or complete resolution of their symptoms.

Radiologic Studies

Magnetic Resonance Imaging Protocol

All patients underwent pre- and postoperative magnetic resonance imaging (MRI), including axial and sagittal T1- and T2-weighted fast spin-echo sequences, sagittal and axial cardiac gated phase contrast cine mode images, sagittal cardiac gated cine true fast imaging with steady-state precession (true FISP), and sagittal high-spatial-resolution constructive interference in steady-state (CISS) sequences. The midsagittal craniocervical junction region phase contrast cine-mode images provided a quasi-real-time dynamic assessment of the CSF flow characteristics. The sequence derived signal contrast between flowing and stationary nuclei by sensitizing the phase of the transverse magnetization to the velocity of motion and enabled measurement of flow velocity and flow pulsation magnitude or simply a qualitative assessment of CSF flow. The CSF velocity was defined as the peak systolic velocity in the transverse plane through the foramen of magnum, measured using the AW VolumeShare 5 software (GE Medical Systems). Heavily T2-weighted MR CISS sequences provided exquisite delineation of the parenchyma–CSF interfaces and detailed assessment of the cerebellar morphology.

Chiari I malformation was diagnosed using sagittal T1-weighted MRI brain studies and was defined as tonsillar herniation of at least 5 mm below the level of the foramen magnum.

Narrowing of the CSF spaces anterior and posterior to the spinal cord was assessed on T2-weighted imaging. Postoperative improvement in CSF flow was demonstrated by comparing T2 signal on pre- and postoperative images and signaling in the posterior fossa and around the spinal cord. Phase-contrast cine flow imaging was compared pre- and postoperatively and assessed on all patients postoperatively to confirm adequate CSF flow. The postoperative MRI with cine was obtained approximately 3 months after surgery.

Surgical Technique

A standard suboccipital craniectomy of the width of the foramen magnum and approximately the same height was performed to ensure a good decompression of the cerebellar hemispheres, brainstem, and midline structures. For patients with cerebellar tonsil herniation below the C1, a posterior arch removal was also performed to decompress the cervical spinal cord. The dura was opened in a **Y**-shaped manner and reflected radially. The intradural contents were explored. Any adhesions observed were lysed, thus restoring the egress of CSF from the ventricle and across the CMJ. An augmented lax duraplasty was performed in a watertight fashion, which were closed in layers.

Postoperative Care and Follow-Up

All the patients were closely monitored in the intensive care unit (ICU) for 24 hours postoperatively for cardiovascular, respiratory, and neurologic changes. On postoperative day 1, patients were fully mobilized with a soft collar and were discharged as per the institute's protocols of postoperative management, which is usually after removal of stitches on the seventh postoperative day. Patients returned to the clinic at 3 months, 6 months, and 1 year with an MRI of the craniovertebral junction and cine MRI flow study. Postoperative complications were divided into two groups:

- Early complications (up to 30 days):
  CSF leak and/or pseudomeningocele.
  Meningitis: infective and chemical.
  Hematoma.
- Late complications (after 30 days):
  New or enlarging syrinx.
  Obstruction of CSF flows across the CMJ due to scarring.
  Cerebellar ptosis.

Statistical Analysis

A univariate analysis was used to compare groups, and $p < 0.05$ was considered significant. Statistical analyses were performed using EPI Info 7. Descriptive statistics measures, mean and standard deviation, were used to summarize the numerical data and count and percentage for summarizing the nominal data. The study period was 18 months. All the patients meeting the inclusion criteria were enrolled for the study. A comparison of categorical data was made using an unpaired *t*-test. The continuous data were compared using Student's *t*-test/Mann–Whitney *U* test, as applicable.

Ethical Issues

Ethical clearance has been taken from the Institute's Human Ethics Committee for conducting this study.

Results

A total of 25 patients were included in the study. The mean age of the patients was 45.52 ± 13.37 years, with 40% being males and 60% females. More than half of the patients had occipital headaches at presentation, while one-fourth had no headache, and the rest had holocranial headaches. The headache was severe in 40% of the patients. There were no motor signs in three patients, while eight patients presented with motor involvement in the form of weakness, atrophy, and spasticity, and one patient each had weakness or atrophy. Forty percent of the patients had lower cranial nerve involvement in dysarthria or dysphagia. The sensory symptoms included paresthesias in 14 patients, dysesthesias in 10 patients, and posterior column dysfunction in 1 patient. The details of the demographic profile and clinical presentation are given in ►**Table 1**.

The radiological parameters were recorded using the details mentioned in the "Methodology" section. The mean pB-C2 distance (a line is drawn from the basion to the inferoposterior aspect of C2; pB-C2 is then measured as the perpendicular distance running through the odontoid tip to the line drawn previously as shown by the horizontal *arrow* in ►**Supplementary Fig. S1**, available in the online version) before intervention was 5.66 ± 1.85, which changed to 5.18 ± 1.75 after surgical intervention. Similarly, the position of the obex changed from a mean of 1.85 ± 0.76 to 1.78 ± 0.76 after surgery. There was a significant increase in the anterior flow at the foramen magnum with the surgical intervention, with a mean of 0.22 ± 0.10 beats/min before intervention to 5.00 ± 1.80 beats/min postintervention. Similarly, there was an increase in the posterior flow at the foramen magnum with intervention (1.01 ± 1.41 to 4.09 ± 1.22 beats/min). The details of the effect of surgical intervention are given in ►**Table 2**.

Twenty-one patients had complete resolution of headache after surgical intervention, while 4 had an improvement or maintained the status quo. None of the patients had worsened headaches after surgery. Sensory symptoms were resolved in 88% of the patients. The most minor resolution of symptoms was seen for the lower cranial nerve outcomes (48%), followed by motor symptoms (44%). Even if these symptoms were not resolved, the majority showed improvement or status quo for the symptoms, while there was no worsening.

All patients had tonsillar descent below McRae's line, with 84% having peg-like morphology and 16% having wedge morphology of the tonsils. In 60% of the patients, the descent was still below the C1 level; in the rest, the descent was to the C1 posterior arch level. Six patients had no syrinx, while 15 had a fusiform shape, and 4 had a tubular shape of the syrinx. The details of the pre- and postsurgery patterns of sensorimotor involvement, tonsillar descent, and syrinx morphology are presented in ►**Supplementary** –►**Tables S1–S3** (available in the online version). The summary of improvement in the functional status parameters of the patients is shown in ►**Supplementary Fig. S2** (available in the online version).

"CSF Flow Change Anterior" and "Tonsillar Descent (Postintervention)"

There was no significant difference between the various groups regarding the distribution of tonsillar descent (postintervention; $\chi^2 = 1.604$, $p = 0.527$). The strength of association between the two variables was low (Cramer's V) = 0.25 (low association); (bias-corrected Cramer's V) = 0.15 (low association; ►**Supplementary Table S4**, available in the online version).

"CSF Flow Change Anterior" and "Syrinx (Postintervention)"

There was no significant difference between the various groups regarding the distribution of syrinx (postintervention; $\chi^2 = 1.103$, $p = 0.402$; ►**Supplementary Table S5**, available in the online version).

Motor power in the lower limb (preintervention) and syrinx shape (preintervention) were significantly associated with the variable "anterior flow at the foramen magnum (preintervention)" ($p < 0.05$).

Average flow, cervicomedullary angle (postintervention), sensory symptoms progression, and CSF flow change anterior to the foramen magnum were significantly associated with the variable "anterior flow at the foramen magnum (postintervention)" ($p < 0.05$).

Discussion

Patients who present to the hospital with Chiari malformations are often in a dilemma about their quality of life after surgery. This gets further accentuated when the pathology, unclear natural history of the disease, and possible surgical outcomes are explained. This apprehension is more evident in patients with concomitant syringomyelia. During this study, by studying various preoperative clinical and radiological factors, we have attempted to develop a clinical tool to help predict the surgical outcomes in these patients.

Age

Chiari malformations are a disease affecting all ages, from children to older adults. The minimum age of patients in our study was 25 years, and the maximum was 75 years. The mean age at presentation in the adult population was 45.5 years. Furtado et al,[15] in their study, have not found any significant association between age and improvement in outcome. In our study, age does not seem to correlate statistically with the patient's outcome or syringomyelia improvement ($p = 0.051$).

Sex

Most of the studies in the literature have reported a female preponderance in Chiari I malformation and syringomyelia.[1,16–18] The female-to-male ratio in our study was 1.5:1. We found no statistically significant relationship between gender with the outcome and syringomyelia improvement, which was consistent with concurrent literature.[1,15] Compared to males, a more substantial percentage of a better outcome was observed in female patients, which may be due to the female preponderance of the disease.

Symptoms and Signs

Symptoms and signs in our study were clubbed into five groups: headache, sensory symptoms, motor symptoms, lower cranial nerve symptoms, and functional status.

Headache

Headache was the most common specific symptom in the study. In our research, 72% of patients had headaches as presenting symptoms. In 64.0% of the population, headaches radiated to the neck region.

Concurrent literature had reported improved outcomes of headaches following surgery.[1,19,20] In our study, 84% of the

Table 1 Demographic profile and clinical presentation

Basic details	Mean ± SD, Median (IQR), Min–Max, Frequency (%)
Age (y)	45.52 ± 13.37 || 45.00 (37.00–52.00) || 25.00 - 75.00
Gender	
Male	10 (40.0%)
Female	15 (60.0%)
Height (m)	1.63 ± 0.08 || 1.62 (1.58-1.66) || 1.48 - 1.78
Weight (kg)	68.12 ± 12.90 || 65.00 (58.00-78.00) || 45.00 - 90.00
Handedness	
Right	20 (80.0%)
Left	5 (20.0%)
Headache	
No headache	7 (28.0%)
Occipital	14 (56.0%)
Holocranial	4 (16.0%)
Radiation of headache (yes)	16 (64.0%)
Aggravating factors—headache	
None	8 (32.0%)
Laughing/coughing	9 (36.0%)
Movement	4 (16.0%)
Position change	4 (16.0%)
Severity of headache	
No	7 (28.0%)
Mild	1 (4.0%)
Moderate	7 (28.0%)
Severe	10 (40.0%)
Motor symptoms	
None	3 (12.0%)
Weakness	1 (4.0%)
Weakness, atrophy	1 (4.0%)
Weakness, atrophy, spasticity	8 (32.0%)
Weakness, atrophy, spasticity, gait changes	10 (40.0%)
Weakness, spasticity	2 (8.0%)
Vision	
Normal	12 (48.0%)
Blurred	5 (20.0%)
Nystagmus	7 (28.0%)
Diplopia	1 (4.0%)

(*Continued*)

Table 1 (*Continued*)

Basic details	Mean ± SD, Median (IQR), Min–Max, Frequency (%)
Motor score	4.00 ± 0.58 || 4.00 (4.00–4.00) || 3.00–5.00
Functionality	
No	0 (0.0%)
Mild impairment	2 (8.0%)
Moderate impairment	14 (56.0%)
Severe impairment	7 (28.0%)
Unable to work	2 (8.0%)
Lower cranial involvement	
None	15 (60.0%)
Dysarthria	5 (20.0%)
Dysphagia, dysarthria	4 (16.0%)
Dysphagia, other, dysarthria	1 (4.0%)
Sensory symptoms (yes)	25 (100.0%)
Sensory deficit	
None	0 (0.0%)
Paresthesia	14 (56.0%)
Dysesthesia	10 (40.0%)
Posterior column dysfunction	1 (4.0%)
Involvement of hypoesthesia	
Upper limb, trunk	2 (8.0%)
Right upper limb, trunk	7 (28.0%)
Left upper limb, trunk	9 (36.0%)
Right upper limb, right lower limb, trunk–anterior, trunk–posterior	7 (28.0%)

patients had a resolution of pain. There was a partial resolution in 16% of patients, or it remained the same.

The headache correlated with syrinx length below McRae's line (mm) and CSF flow changes in the anterior aspect. Headache was absent in patients with Chiari 0 malformation, which may point toward tonsillar herniation being the cause of the headache.[21]

Sensory Symptoms, Motor Symptoms, and Preoperative Functional Status

The motor symptoms were present in 80% of the study population, and sensory symptoms were present in all cases.

Most researchers have defined postoperative functional improvement or deterioration using preoperative sensory and motor symptoms.[22] Greenberg et al observed that patients with myelopathic features had poor outcomes compared to other patient groups.[20] Tisell et al[23] observed a

Table 2 Effects of surgical intervention on radiological parameters at cervicomedullary (CM) junction

CM junctions	Mean ± SD (preintervention)	Mean ± SD (postintervention)	Median (IQR), preintervention	Median (IQR), postintervention	Min–Max (preintervention)	Min–Max (postintervention)
PB–C2 distance (preintervention)	5.66 ± 1.85	5.18 ± 1.75	6.00 (4.60–7.00)	5.30 (4.00–6.30)	2.0–9.0	1.8–8.3
Obex position (preintervention)	1.85 ± 0.76	1.78 ± 0.76	1.60 (1.30–2.10)	1.60 (1.30–2.30)	0.7–3.7	0.1–4.3
Cervicomedullary angle (preintervention)	142.44 ± 15.07	148.96 ± 12.21	142.00 (135.00–158.00)	150.00 (140.00–158.00)	112.0–165.0	130.0–170.0
Pre-/post-PB C2 change	1.57 ± 1.43		1.20 (0.40–2.40)		0.1–5.1	
Pre-/postobex change	−0.07 ± 0.95		0.00 (−0.63 to 0.30)		−2.1 to 2.6	
CM angle change	6.52 ± 16.85		8.00 (−6.00 to 15.00)		−28 to 53	
Anterior flow at the foramen magnum (preintervention), beats/min	0.22 ± 0.10	5.00 ± 1.80	0.22 (0.14–0.29)	5.20 (4.30–6.60)	0.0–2.1	0.1–7.4
Posterior flow at the foramen magnum (preintervention), beats/min	1.01 ± 1.41	4.09 ± 1.22	1.06 (5.00–2.07)	3.80 (3.20–4.70)	0.0–4.9	2.2–6.8

Abbreviations: IQR, interquartile range; Max, maximum; Min, minimum; SD, standard deviation.

significant improvement in patients with paresis and sensory dysfunction after surgery.

We have found that the presence of motor or sensory features had a significant negative correlation with the functional outcome score ($p < 0.002$).

Furtado et al[15] have studied the preoperative functional status of the patient and their improvement but have not commented on the correlation of preoperative functional status with postoperative functional status.

Of note, we found that the presence of sensory symptom had a significant positive correlation with the presence of syringomyelia ($p = 0.001$). This can be explained by the involvement of the spinothalamic tracts during the evolution of the syrinx.

Lower Cranial Nerve Symptoms

Milhorat et al found 52% involvement of the lower cranial nerves.[4] Aitken et al found 4% involvement of the lower cranial nerves.[19] In our study, 40% had symptoms of lower cranial nerve symptoms. These patients had a negative correlation with the outcome score. One of the reasons for this could be that lower cranial nerve deficit might present at a later stage and represent a more severe form of the disease.[24–26]

Duration of the First Symptom Less Than 9 Months

The median duration of the appearance of the first symptom to presentation in our study was 9 months. The patient who presented earlier than 9 months had a positive correlation with outcome. This is consistent with the existing literature, which indicates that a shorter symptom duration correlates with better outcome.[27,28] However, we did not find any significant correlation between symptom duration and syrinx resolution.

Preoperative Radiological Factors

Tonsillar Descent and Syringomyelia

The natural history of syringomyelia is variable with the symptoms and syrinx ranging from progression to spontaneous resolution.[29–31] Also, there are reports where syringomyelia has remained stable for many years.[32,33] As syringomyelia associated with Chiari 0 malformation tends to remain stable, it is likely that tonsillar herniation plays a role in syrinx progression by causing CSF flow obstruction at the foramen magnum.[34–37] In Chiari 0 malformation, CSF flow abnormality is not as severe because the foramen magnum is not crowded and hence tends to remain stable.

In the present study, all the patients with tonsillar herniation and syringomyelia underwent surgery. Of 19 patients with Chiari I malformation and syringomyelia, the syrinx resolved in 9 (45%) patients and remained stable in 10 (55%) patients. Patients with tonsillar herniation had greater syrinx resolution postsurgery ($p = 0.003$). This is probably because tonsillar herniation is the main reason for foramen magnum crowding in Chiari I patients, which gets relieved by surgical decompression.

Effect of Syringomyelia on Outcome

There are contrasting studies on the effect of syringomyelia on outcomes of Chiari I malformation. Some believe that the presence of syringomyelia is associated with poor outcome,[37–39] while some have reported a favorable effect on outcome.[40] Also, there are reports of patient with asymptomatic syringomyelia.[34–37] In our study, 19 patients had syringomyelia and 1 was asymptomatic. It could be possible if syringomyelia might be secondary to some other pathology[41,42] and does not become symptomatic unless it is progressive.[35,37,43]

Syringomyelia regression has a positive correlation with outcome ($p < 0.001$). In our study, 95% of patients in whom the syrinx improved had a good outcome. None of the patients with syrinx progression had a good outcome.

Extent of Syringomyelia

Imae concluded that there was no significant relationship between the degree of tonsillar herniation and the size, length, and position of the syrinx cavity.[44] The most common extent of the syrinx was cervicodorsal, which is similar to those found by other authors.[45] Nishizawa et al analyzed nine unoperated patients with syringomyelia and concluded that the longitudinal extent of syringomyelia based on MRI does not have a prognostic value.[46] Nakamura et al[47] found the extent and width of syringomyelia to be significant determinants of clinical course in asymptomatic patients. They recommended surgical treatment for syringomyelia that extends over four or more vertebrae. We found a negative correlation between the extent of syrinx and the outcome score ($p = 0.022$). We also noted a correlation of the diameter of the syrinx with the extent of the syrinx ($p = 0.035$). Those patient with a larger syrinx might have irreversible damage to the cord during its progression and may not improve totally even after resolution of the syrinx.

We have not found any correlation between the longitudinal extent of syringomyelia and the outcome or syringomyelia improvement. Wetjen et al also found no relation between the extent of syringomyelia and syrinx narrowing.[17]

Syrinx Diameter Less Than 6 mm

Greenberg et al noted a significant negative association between the presence of a large syrinx ≥6 mm and postoperative improvement.[20] Furtado et al found that change in cord diameter, not syrinx diameter affects the outcome.[15] In our study, we found a significant positive correlation between the outcome and a syrinx with maximum diameter ≤6 mm.

In all, 95% patients with a syrinx diameter less than 6 mm had a good outcome as compared to 72.7% patients with a syrinx diameter greater than 6 mm. A diameter of 6 mm may represent the cutoff where most patients begin to experience symptoms and cord damage; however, it may not be a rule. In our case series, there were patients with a syrinx diameter of 6 and 8 mm who did not undergo surgery and remained stable. However, none of the patients with a syrinx diameter less than 6 had a poor outcome.

Shape of Syrinx

In syringomyelia, enlargement can be either fusiform or tubular, depending on the length of the syringomyelic cavity or cavities.[48] There are views that these tubular cavities represent hydromyelia.[49] However, the author also notes that hydromyelia seen today might be the underlying predisposition for developing a syringomyelia if an adequate impulse takes place.[49] We have found that a syringomyelia with a tubular shape tends to be more stable ($p < 0.001$), whereas a fusiform-shaped syringomyelia is more likely to be unstable. In our study, 54% of tubular syrinxes remained stable postsurgery; however, only 7% of fusiform syrinxes remained stable; they either improved or worsened.

Management and Postoperative Follow-Up

The outcome scores of patients undergoing FMD alone or with duroplasty or added C1 arch excision were similar when compared to the whole cohort.

Ellenbogen et al[50] reviewed 65 patients with Chiari I malformation and compared them with 20 normal patients. They found that, as compared with normal individuals, there was a dramatic difference between the durations of caudal CSF flow across the brainstem at the foramen magnum in Chiari I patients. They noted that successful treatment of Chiari I patients requires reestablishing CSF pathways by a surgical procedure appropriate for the presumed origin and nature of the obstruction.[50] They added that creating an enlarged posterior fossa is beneficial and can be addressed in many creative ways. However, they cautioned against removing too much bone due to the potential consequence of cerebellar sag.[50,51]

Chiari Outcome Predictability Score

Based on the factors found to have significant correlation with outcome and syrinx improvement, we created an outcome predictive index, Chiari outcome predictability index (COPI). More than 90% of patients with COPI > 0 have chances of good outcome as compared to 66% in other groups. This tool can help stratify patients into three groups as shown in ►**Table 3**, and prognosticate the expected outcome.

Limitations

The median clinical follow-up in our study was close to 2 years. Our results are, therefore, only applicable to short-term outcomes of these patients. We reviewed cases spanning a 10-year period and included all patients with Chiari malformations we could contact and have access to their radiological imaging and/or report. The inclusion of retrospective patient data along with prospective cohort is a limitation of our study.

Further prospective studies with larger sample size will ascertain the usefulness of COPI.

This study does not predict the type of surgery that may be appropriate for the patient. Factors that can help in deciding the type of surgical intervention required for the patients can be further studied. In addition, other limitations

Table 3 Effect of surgery on outcome of symptoms

Progression	Resolved/no new symptom	Improved and unimpaired	Unchanged/ refractory	Worse
Headache outcome	21 (84.0%)	4 (16.0%)	0 (0.0%)	0 (0.0%)
Sensory symptom outcome	22 (88.0%)	3 (12.0%)	0 (0.0%)	0 (0.0%)
Motor symptom outcome	11 (44.0%)	14 (56.0%)	0 (0.0%)	0 (0.0%)
Lower cranial nerve outcome	12 (48.0%)	13 (52.0%)	0 (0.0%)	0 (0.0%)
Complications outcome	23 (92.0%)	2 (8.0%)	0 (0.0%)	0 (0.0%)
Functional outcome	15 (60.0%)	8 (32.0%)	1 (4.0%)	1 (4.0%)

include the low number of cases on the whole, very low number of cases included in each subgroup for analysis, unclear kappa for the observers demarcating the variables for each variant suggesting possibility of ambiguity of the interpretation of the variables, selection biases during the surgery as for the techniques, amount or advantages of releasing the adhesions, and the amount of hemorrhage in the field of surgery.

In addition, some studies have shown an association between the quantitatively measured tonsillar descent and the maximum dimeter of the syrinx and its impact on resolution posttonsillar reduction surgery. We have not performed the quantitative analysis on the tonsillar descent and its impact on the syrinx diameter. Performing a region of interest (ROI) analysis on the syrinx scoring system and quantitative tonsillar descent in future prospective studies would add strength to the existing literature.

Conclusion

Clinically, the functional status of the patient is the most important predictor of postoperative outcome. The presence of motor, sensory, or lower cranial nerve symptoms is associated with poor outcome. The duration of the first symptom at the time of presentation has a significant correlation with syrinx improvement. We have found an appreciable but not significant correlation between the duration of the first symptoms and the outcome score.

Radiologically, postoperative, syringomyelia progression was associated with poor outcomes. Persistent foramen magnum CSF outflow obstruction, even after surgery, has a strong negative correlation with the outcome. The COPI has shown a significant correlation with patient outcome and can be used to inform patients about the expected result.

Authors' Contributions

M.P. was responsible for data collection, design, analysis, and manuscript writing. A.S. was responsible for hypothesis, supervision, study design, data analysis, presentation of results, and reviewing the manuscript. P.C. was involved in supervision, manuscript writing, and administrative support. S.R. was involved in administrative support, clinical details, surgical details, and reviewing the manuscript. R.M. was involved in manuscript writing, reviewing the manuscript, and design. A.A. was involved in administrative support, reviewing the manuscript, and supervision. S.N. was involved in supervision, data analysis, reviewing the manuscript, and administrative support.

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Conflict of Interest

None declared.

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