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RESEARCH

Predictors of syrinx presentation and outcomes in pediatric Chiari malformation type I: a single institution experience of 218 consecutive syrinx patients

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Abstract

Purpose Chiari I malformation (CM-I) in pediatric patients can impose substantial neurologic and functional impairment. Additionally, the presence of syrinx is often a harbinger of clinical compromise, but little attention has been devoted to identifying features associated with syrinx development and the clinical impact of syrinx resolution. Therefore, this study aims to identify clinical and radiographic variables associated with preoperative syrinx presence and postoperative syrinx reduction in pediatric patients with CM-I and determine the relationship between postoperative syrinx reduction and clinical symptom improvement.

Methods The authors performed a retrospective analysis of 435 consecutive pediatric patients who underwent surgical treatment of CM-I from 2001 to 2021 at a single tertiary pediatric medical center. All patients underwent pre- and postoperative MRI, and clinical and radiographic variables were recorded and subject to inferential analysis.

Results Syrinx at presentation was independently associated with symptoms of spinal cord dysfunction at presentation (OR 2.17 (95% CI 1.05–4.48); $p=0.036$), scoliosis (OR 5.33 (2.34–10.86); $p=0.001$), and greater pB-C2 (posterior basion to C2 distance) measurement length (OR 1.14 (95% CI 1.01–1.30); $p=0.040$). Syrinx at presentation was inversely associated with tussive headaches at presentation (OR 0.27 (95% CI 0.16–0.47); $p=0.001$) and cranial nerve deficits at presentation (OR 0.49 (95% CI 0.26–0.92); $p=0.025$). Postoperatively, patients with radiographic evidence of syrinx improvement had greater rates of symptom improvement (93.1% vs 82.1%; $p=0.049$), better CCOS scores (15.4 vs 14.2; $p=0.001$), and decreased rates of readmission (6.0% vs 25.0%, $p=0.002$) and reoperation (0.5% vs 35.7%; $p=0.001$). The difference in syrinx resolution was similar but not statistically significant (10.3% vs 16.7%; $p=0.251$). AO joint anomaly (OR 0.20, 95% CI 0.04–0.95; $p=0.026$) and foramen magnum diameter (OR 1.12, 95% CI 1.00–1.25; $p=0.049$) were the only independent predictors of syrinx improvement, and surgical technique was the only predictor for syrinx resolution (OR 2.44, 95% CI 1.08–5.50; $p=0.031$). Patients that underwent tonsil reduction surgery whose syrinx improved had a wider foramen magnum diameter than those whose did not improve (34.3 vs 31.7; $p=0.028$).

Conclusions Radiographic syrinx improvement is associated with greater rates of symptom improvement and less readmissions and reoperations for CM-I. AO joint anomalies and narrower foramen magnums were independent risk factors for the lack of syrinx improvement. These novel insights will help guide preoperative patient counseling, pre- and intraoperative surgical decision-making, and postoperative clinical prognostication in the treatment of pediatric CM-I.

Keywords Chiari I · Syringomyelia · Syrinx · Outcomes

Introduction

Chiari I malformation (CM-I) is defined according to the following criteria: (1) greater than 5 mm of caudal descent of the cerebellar tonsils below the foramen magnum or (2) less than 5 mm tonsillar descent below the foramen magnum with aberrant cerebrospinal fluid (CSF) flow dynamics on

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cine magnetic resonance imaging (MRI) [1]. Despite this definition's ubiquity in clinical application, the degree of tonsil descent has not consistently correlated with clinical presentation, prognosis [2–8], or disease severity [9]. As a result, many alternative radiographic parameters have been proposed as predictors of clinical presentation and outcomes.

Clivoaxial angle (CXA) and foramen magnum (FM) diameter have varying evidence to support their use [3, 10–13], and newer measurements such as obex position, posterior basion to C2 distance (pB-C2), and occipital condyle-C2 sagittal vertebral alignment (C-C2SVA) have demonstrated associations with clinical symptom severity and need for occipitocervical fusion or ventral decompression [14–20]. Similarly, preoperative syrinx size ≥ 6 mm is the only neuroimaging parameter included in the Chiari Severity Index as a predictor of postoperative improvement [9], but little attention has been devoted to clarifying the factors associated with the presence of syrinx or how treatment-related changes in syrinx size correlate with symptom severity [3, 13]. Another common limitation is the small sample sizes of the currently available evidence relating postoperative syrinx resolution and symptom improvement [3].

To address this knowledge gap, our study provides a large, single institution series detailing (1) clinical and radiographic predictors of syrinx presence in the preoperative setting and (2) the association of postoperative syrinx improvement and clinical symptoms after surgical treatment of CM-I. Outcomes were compared between two thresholds of syrinx improvement, as there remains uncertainty on the targeted syrinx improvement required to minimize long-term neurological decline and thus justify surgical intervention.

Methods

Study design and data collection

The study was conducted in compliance with the Health Insurance Portability and Accountability Act (HIPAA) and received institutional review board (IRB) approval. All patients younger than 18 years who underwent surgery for Chiari I malformation between May 2001 and May 2021 were retrospectively reviewed. Patients were excluded from the study if they were diagnosed with craniofacial dysostosis or skeletal dysplasia or if they had less than 3 months of postoperative follow-up. All eligible patients were evaluated with preoperative and postoperative MRI, and demographic, clinical, procedural, and radiographic variables were recorded. Preoperative presentation symptoms included occipital tussive (Chiari) headaches, headaches atypical for CM-I (non-Chiari), vomiting, cranial nerve dysfunction (ocular palsies, choking, gagging, snoring, dysphagia, or breathing issues), cerebellar dysfunction (ataxia, clumsiness, nystagmus, vertigo, or coordination problems), and spinal cord dysfunction (abnormal reflexes, toe-walking, enuresis, paresthesia, or weakness).

The following preoperative radiographic parameters were measured: the presence of syrinx, scoliosis, medullary kink, hydrocephalus, maximal anterior–posterior diameter of syrinx, syrinx length by number of vertebral body levels, foramen magnum diameter (anterior–posterior), tonsil length below the foramen magnum, CXA, retroversion of the dens, and pB-C2. Anomaly of the atlantooccipital joint was defined as abnormal structural apposition on MRI. Postoperative syrinx measurements were performed using the patient's most recent follow-up MRI. Postoperative syrinx improvement was defined as a decrease in anterior–posterior maximal syrinx diameter compared to the preoperative syrinx diameter, while syrinx resolution was defined as greater than 50% reduction in either syrinx length or anterior–posterior diameter. When evaluating the presence of syrinx, we did not distinguish between syringomyelia and hydromyelia (dilation of the central canal of spinal cord). While the developmental etiology of these entities is distinct, clinical and neurologic sequelae are often very similar.

All surgeries were performed at a single tertiary pediatric medical center. Most patients had clinical symptoms that led to their diagnosis of CM-1, but a subset of patients were incidentally found to have CM-1 with syrinx, such as after presenting for a trauma, and were operated on to prevent expansion or neurologic decline related to their syrinx. All patients underwent suboccipital craniectomy (SOC) with duraplasty. Intradural work was performed according to surgeon preference and was classified into two groups: decompression without tonsil reduction (with or without lysis of arachnoid adhesions) or decompression with tonsil reduction (tonsillar coagulation or subpial tonsillar resection). Postoperative symptom improvement was defined as the reduction or improvement of preoperative Chiari-related symptoms, as expressed by the patient or parents in postoperative follow-up visits. Postoperative outcome was also quantified by the Chicago Chiari Outcome Score [21]. Noted complications include aseptic meningitis, pseudomeningocele, and CSF leak, along with readmission and reoperation rates.

Statistical analysis

Statistical analyses were performed using STATA Statistical Software (version 16, StataCorp). Descriptive statistics were used to determine the frequency, mean, median, standard deviation, interquartile range, or 95% confidence interval. Quantitative variables were first assessed for normal distribution using the Shapiro–Wilk's test. Pearson chi-square was performed to compare the difference in frequencies between categorical variables. Differences in means of normally distributed variables were compared using Student's *t*

Table 1 Relationships between preoperative syrinx and clinical variables

	Total	Syrinx present	Syrinx absent	*p*-value
Overall population: *n* (%)	435	218 (50.1%)	217 (49.9%)	
Demographic				
Age (years): mean (95% CI)	8.4 (7.9–8.9)	9.7 (9.1–10.3)	7.0 (6.4–7.7)	**0.001**
Sex (male): *n* (%)	216 (49.4%)	101 (45.9%)	115 (53.0%)	0.138
Symptoms: n (%)				
Chiari headache	186 (42.6%)	65 (29.6%)	121 (55.8%)	**0.001**
Non-Chiari headache	40 (9.2%)	22 (10.0%)	18 (8.3%)	0.537
Vomit	24 (5.5%)	7 (3.2%)	17 (7.8%)	**0.033**
Cranial nerve	126 (28.8%)	44 (20.0%)	82 (37.8%)	**0.001**
Cerebellar	37 (8.5%)	16 (7.3%)	21 (9.7%)	0.367
Brainstem/spinal cord	99 (22.7%)	61 (27.7%)	38 (17.5%)	**0.011**
None (incidental)	44 (10.1%)	41 (18.6%)	3 (1.4%)	**0.001**

P values in bold reached statistical significance, i.e., < .05

test. Variables with non-normal distributions were compared using the Mann–Whitney *U* test. Univariate and multivariate logistic regression were used to determine the independent association of multiple risk factors with categorical outcome variables. Statistical significance was set at $\alpha = 0.05$.

Results

Presence of syrinx and symptomatology

Of the 435 patients in total, 22 (50.1%) had a preoperative syrinx (Table 1). Patients with syrinx at presentation were older (9.7 years vs 7.0; $p = 0.001$) and were less likely to endorse Chiari-type headaches (29.6% vs 55.8%; $p = 0.001$) or vomiting (3.2% vs 7.8%; $p = 0.033$). There was no difference in sex (45.9% male vs 53.0% female; $p = 0.138$) and the reported rates of non-Chiari headaches (10.0% vs 8.3%; $p = 0.537$). Additionally, patients with syrinx were more likely to present with symptoms of spinal cord dysfunction (27.7% vs 17.5%; $p = 0.011$) but less likely to present with cranial nerve symptoms (20.0% vs 37.8%; $p = 0.001$). Cerebellar symptoms were not related to preoperative syrinx presence (7.3% vs 9.7%; $p = 0.367$). Forty-one (18.6%) of the syrinx patients presented after incidentally identifying Chiari pathology on imaging, whereas only 3 (1.4%) non-syrinx patients presented in this manner ($p = 0.001$). Twenty-two patients presented incidentally without symptoms or scoliosis. There was no difference in average follow-up (31.1 vs 34.5 months; $p = 0.231$).

Tables 1 and 2 summarize the clinical and radiographic parameters associated with preoperative syrinx presence. Notably, patients with syrinx had higher rates of scoliosis (31.7% vs 5.9%; $p = 0.001$) and hydrocephalus (8.2% vs 3.7%; $p = 0.043$), but rates of retroflexed odontoid (40.0% vs 30.0%; $p = 0.059$) and medullary kink (10.0% vs 13.8%; $p = 0.217$) were not significantly different. Patients with syrinx had greater pB-C2 distance (7.2 vs 6.5mm; $p = 0.007$) and foramen magnum diameter (33.9 vs 32.9mm; $p = 0.022$), but there was no significant difference in cerebellar tonsil length or CXA.

Table 2 Association of preoperative radiographic factors with syrinx at presentation

	Total	Syrinx	No syrinx	*p*-value
Scoliosis	66 (19.8%)	57 (31.7%)	9 (5.9%)	**0.001**
Dens retroflexion	141 (35.6%)	82 (40.0%)	59 (30.9%)	0.059
Hydrocephalus	26 (6.0%)	18 (8.2%)	8 (3.7%)	**0.043**
Medullary kink	52 (11.9%)	22 (10.0%)	30 (13.8%)	0.217
CXA: Degrees (95% CI)	134.7 (133.2–136.1)	133.8 (131.8–135.7)	135.8 (133.5–138.0)	0.185
pB-C2: mm (IQR)	6.9 (5.8–8.1)	7.2 (6.0–8.3)	6.5 (5.6–7.5)	**0.007**
Tonsil length: mm (IQR)	13.0 (9.5–16.7)	12.8 (9.5–13.7)	13.3 (9.4–18.0)	0.640
Foramen magnum diameter: mm (95% CI)	33.4 (33.0–33.9)	33.9 (33.3–34.5)	32.9 (32.4–33.5)	**0.022**

P values in bold reached statistical significance, i.e., < .05

CXA clivoaxial angle, *pB-C2* basion to posterior C2 length

Table 3 Multivariate regression of preclinical factors associated with the presence of syrinx

	Presence of syrinx	
	OR (95% CI)	*p*-value
Age	1.13 (1.06–1.21)	**0.001**
Chiari headache	0.20 (0.11–0.36)	**0.001**
Cranial nerve	0.55 (0.29–1.05)	0.069
Brainstem/spinal cord	1.80 (0.85–3.83)	0.126
Vomit	1.17 (0.35–3.94)	0.801
Scoliosis	4.09 (1.76–9.50)	**0.001**
Hydrocephalus	3.41 (0.98–11.86)	0.054
pB-C2	1.12 (0.96–1.30)	0.160
Foramen magnum diameter	0.98 (0.91–1.06)	0.615

P values in bold reached statistical significance, i.e., < .05

pB-C2 basion to posterior C2 length

Determining which factors were independently associated with syrinx (Table 3), multivariate logistic regression analysis revealed that scoliosis (OR 4.09; 95% CI 1.76–9.50; $p=0.001$) and age (OR 1.13; 95% CI 1.06–1.21; $p=0.001$) were predictors of preoperative syrinx presence, while Chiari headaches (OR 0.20; 95% CI 0.11–0.36; $p=0.001$) was associated with the absence of preoperative syrinx. Vomiting (OR 1.17; 95% CI 0.35–3.94; $p=0.801$), brainstem/spinal cord dysfunction (OR 1.80; 95% CI 0.85–3.83; $p=0.126$), cranial nerve symptoms (OR 0.55; 95% CI 0.29–1.05; $p=0.069$), hydrocephalus (OR 3.41; 95% CI 0.98–11.86; $p=0.054$), pB-C2 (OR 1.12; 95% CI 0.96–1.30; $p=0.160$), and foramen magnum diameter (OR 0.98; 95% CI 0.91–1.06; $p=0.615$) were not independent risk factors.

Impact of syrinx improvement on postoperative outcomes

Table 4 compares clinical variables and postoperative outcomes with syrinx improvement status. Notably, postoperative improvement in syrinx diameter was significantly associated with patient-reported postop symptom improvement (93.1% vs 81.5%, $p=0.038$), higher CCOS (15.4 vs 14.2; $p=0.001$), and lower rates of readmission (6.0% vs 26.1%; $p=0.001$) and reoperation (0.5% vs 37.0%, $p=0.001$). There was also less follow-up for those whose syrinx improved (29.4 months vs 45.7; $p=0.005$). The incidence of aseptic meningitis, pseudomeningocele, and CSF leak were not associated with postoperative syrinx outcomes. Syrinx resolution followed similar patterns, apart from there was no difference in symptom improvement (93.3% vs 87.0%; $p=0.147$) despite a similar difference in CCOS values (15.4 vs 14.7; $p=0.001$) as well as follow-up length (29.4 months vs 37.3; $p=0.077$). There was also no difference in age or sex between syrinx response status groups.

Table 4 Association of radiographic syrinx improvement with clinical factors

	Syrinx improvement	No syrinx improvement	*p*-value	Syrinx resolution	No syrinx resolution	*p*-value
Overall	191 (87.6%)	27 (12.4%)		165 (75.7%)	53 (24.3%)	
Follow-up (months): mean (95% CI)	29.4 (26.0–32.7)	45.7 (26.3–65.2)	**0.005**	29.4 (25.7–33.2)	37.3 (26.9–47.7)	0.077
Demographic						
Age (years): mean (95% CI)	9.8 (9.1–10.4)	8.9 (6.8–10.9)	0.320	9.9 (9.2–10.5)	9.0 (7.7–10.2)	0.203
Sex (male): *n* (%)	89 (46.6%)	10 (37.0%)	0.350	75 (45.7%)	10 (44.4%)	0.869
Symptom outcome						
Improved: *n* (%)	178 (93.2%)	22 (81.5%)	**0.038**	153 (93.3%)	47 (87.0%)	0.147
CCOS: mean (95% CI)	15.4 (15.2–15.5)	14.2 (13.3–15.0)	**0.001**	15.4 (15.2–15.5)	14.7 (14.2–15.2)	**0.001**
Complications: n (%)						
Readmission	9 (6.0%)	6 (26.1%)	**0.001**	7 (5.6%)	8 (16.3%)	**0.023**
Aseptic meningitis	5 (3.3%)	0 (0.0%)	0.386	4 (3.2%)	1 (2.1%)	0.695
Pseudomeningocele	3 (1.6%)	1 (3.7%)	0.440	3 (1.8%)	1 (1.9%)	0.991
CSF leak	3 (1.6%)	0 (0.0%)	0.512	3 (1.8%)	0 (0.0%)	0.317
Reoperation	1 (0.5%)	10 (37.0%)	**0.001**	1 (0.6%)	10 (18.5%)	**0.001**

P values in bold reached statistical significance, i.e., < .05

CCOS Chicago Chiari Outcome Scale, *CSF* cerebrospinal fluid

Table 5 Association of preoperative radiographic variables with syrinx and symptom outcomes

	Syrinx improvement	No syrinx improvement	*p*-value	Syrinx resolution	No syrinx resolution	*p*-value
CXA: Degrees (95% CI)	134.2 (132.1–136.7)	130.5 (124.0–137.0)	0.227	133.8 (131.5–136.0)	133.6 (129.3–137.8)	0.941
pB-C2: mm (IQR)	7.3 (6.9–7.6)	7.2 (6.3–7.9)	0.832	7.3 (5.9–8.3)	7.2 (6.0–8.2)	0.754
Tonsil length: mm (IQR)	13.7 (12.6–14.9)	12.8 (10.0–15.6)	0.564	12.9 (9.7–15.6)	11.9 (8.8–17.8)	0.496
Foramen magnum diameter: mm (95% CI)	34.2 (33.5–34.8)	32.2 (29.6–34.7)	**0.041**	34.2 (33.5–34.8)	33.3 (31.9–34.6)	0.214
Basilar invagination: *n* (%)	15 (9.9%)	3 (13.6%)	0.588	14 (11.1%)	4 (8.3%)	0.591
Atlantooccipital joint anomaly: *n* (%)	15 (9.9%)	6 (27.3%)	**0.019**	13 (10.3%)	8 (16.7%)	0.251
Dens retroflexion: *n* (%)	69 (38.6%)	12 (50.0%)	0.282	60 (39.5%)	21 (41.2%)	0.830
Scoliosis: *n* (%)	51 (32.7%)	5 (22.7%)	0.346	46 (35.4%)	10 (20.8%)	0.064

P values in bold reached statistical significance, i.e., < .05

CXA clivoaxial angle, *pB-C2* basion to posterior C2 length

Other radiographic associations with postoperative outcomes

Table 5 summarizes the relationships between preoperative radiographic parameters and postoperative syrinx improvement and symptom improvement. The factors to demonstrate a significant association with syrinx improvement were atlantooccipital (AO) joint anomaly (9.9% vs 27.3%; $p=0.019$) and foramen magnum width (34.2 vs 32.2 mm; $p=0.041$). The difference in syrinx resolution was similar but not statistically significant for both variables (10.3% vs 16.7%; $p=0.251$ and 34.2 vs 33.3 mm; $p=0.214$). Controlling for radiographic variables (Table 6), AO joint anomaly (OR 0.20, 95% CI 0.04–0.95; $p=0.026$) and foramen magnum diameter (OR 1.12, 95% CI 1.00–1.25; $p=0.049$) were both independent predictors of syrinx improvement, and surgical technique was the only predictor for syrinx resolution (OR 2.44, 95% CI 1.08–5.50; $p=0.031$). When patients were stratified by surgical technique and syrinx improvement status (Table 7), there was a significant difference in the foramen magnum diameter between patients whose syrinx improved versus did not improve that underwent tonsil reduction surgery (34.3 vs 31.7; $p=0.028$) but not in patients who underwent non-tonsil reducing surgery (33.2 vs 33.3; $p=0.959$).

Discussion

The pathophysiology of syrinx is complex and multifactorial. It is both influenced by pressure differentials secondary to obstruction of CSF flow at the cervicomedullary junction as well as disturbances in transcellular fluid dynamics in the spinal cord vasculature [22]. Despite progress in our understanding of syrinx pathophysiology, the predictive and prognostic values of radiographic markers for CM-I and syringomyelia remain inconsistent [4, 9]. While multiple studies have attempted to identify associations between such factors with syrinx presence or improvement [5, 6, 8, 13, 23], our study of a large cohort of consecutively treated patients uncovers novel relationships between radiographic variables and syrinx incidence and postoperative improvement.

Preoperatively, our study found multiple associations between symptomatology and syrinx presence. The relationship between syrinx and scoliosis or older age has been described previously [8, 13, 24], but it is curious that age

Table 6 Multivariate regression for syrinx improvement

	Syrinx improvement		Syrinx resolution	
	OR (95% CI)	*p*-value	OR (95% CI)	*p*-value
Surgery group (tonsil reduction)	2.35 (0.85–6.54)	0.100	2.44 (1.08–5.50)	**0.031**
Basilar invagination	2.75 (0.36–20.86)	0.330	4.14 (0.66–25.86)	0.128
Atlantooccipital joint anomaly	0.20 (0.04–0.95)	**0.043**	0.30 (0.07–1.34)	0.114
Foramen magnum diameter	1.12 (1.00–1.25)	**0.049**	1.03 (0.95–1.12)	0.450
Dens retroflexion	0.76 (0.27–2.17)	0.613	0.90 (0.42–1.93)	0.790

P values in bold reached statistical significance, i.e., < .05

Table 7 Effect of foramen magnum diameter on surgical technique efficacy

	Non-tonsil reduction	Tonsil reduction	*p*-value
Foramen magnum diameter: mm (95% CI)			
Improved syrinx	33.2 (31.1–35.2)	34.3 (33.7–34.9)	0.192
Unimproved syrinx	33.3 (30.1–36.5)	31.7 (27.8–35.6)	0.523
***p*-value**	0.959	**0.028**	

P values in bold reached statistical significance, i.e., < .05

remained an independent risk factor for syrinx after multivariate regression. It is possible that there is a change in CSF dynamics over the course of central nervous system development, increasing the risk of syringogenesis, but that has yet to be described. The observation between syrinx presence and symptoms of spinal cord dysfunction aligns with previous smaller studies [25–27]. It is likely that these symptoms arise from centrifugal compression of the spinal cord by an expansile syrinx or from disruption of neuronal pathways by syrinx dissection through spinal cord parenchyma [28–30].

In contrast, patients without syrinx were more likely to present with Chiari-type headaches, vomiting, and cranial nerve symptoms, even after controlling for other clinical factors at presentation. Similarly, Godzik et al. found that 27% of patients without syrinx presented with headaches compared to only 11% of patients with syrinx, but this difference was not statistically significant [25]. Their study, however, did not differentiate between Chiari- and non-Chiari headaches, and their sample size was limited to 36 patients. A potential explanation for this phenomenon is that patients without syrinx are more likely to present with other Chiari symptoms such as the trademark occipital tussive headache. It also may be that CM-I patients with occipital headaches are more likely to seek medical attention and get diagnosed earlier than patients without these symptoms, allowing time for syrinx to develop in the latter. Nevertheless, the authors acknowledge the heterogeneous complexity of syrinx pathogenesis and that there are likely a variety of mechanisms in how syringomyelia leads to its symptomatology. Of note, we had 22 patients that were discovered incidentally; they presented with syrinx but no symptoms, including no scoliosis.

Radiographically, greater pB-C2 length and wider foramen magnum diameter both associated with preoperative syrinx presence while tonsil length and clivoaxial angle (CXA) did not, which contrasts previous reports. A recent systematic review only found associations between pB-C2 and syrinx improvement, but not the presence, whereas tonsil length, clivus gradient, and scoliosis curvature greater than 20° were all associated with syrinx presence [3]. Additionally, Haller et al. found that obex position and tonsil position were independently associated with syrinx presence, while basilar invagination, occipitalization of the atlas, pB-C2 distance, and clival canal angle were not [6]. Despite mixed conclusions in the literature, the relationship between pB-C2 and syrinx presence may be explained by the alteration of fluid pressure dynamics and obstruction of CSF flow at the cervicomedullary junction. The pB-C2 value quantifies the degree of ventral brainstem impingement at the cervicomedullary junction, contributing to CSF flow obstruction that may drive syrinx development [16].

Postoperatively, patients with syrinx improvement had increased rates of symptom improvement, higher CCOS scores, and decreased rates of complications and readmissions. Current literature on the association between syrinx improvement and symptom improvement is mixed. In a study of 20 children, Furtado et al. found no significant association between symptomatic improvement and changes in syrinx size or presence [31]. Conversely, Hale et al. demonstrated that syrinx resolution was associated with improvement in headaches and neck pain (91% vs 77%) [24]. They also found age to be a significant predictor, which we did not. Neither of these studies, however, reported CCOS or complication rates, where we found lower readmission and reoperation rates but no difference in aseptic meningitis, pseudomeningocele, or CSF leak among syrinx-improved patients. Interestingly, we found a small but significant difference, which has not been described previously, in the rate of symptom improvement between patients who experienced no syrinx improvement and those who experienced no resolution. This may indicate that any decrease in syrinx size, and thus centrifugal pressure on the spinal cord, may alleviate related symptoms, but given the similar number of reoperations, it underscores an important point for syrinx patients in that surgery serves a dual purpose by also preventing long-term neurologic decline.

The radiographic predictors of syrinx improvement in this cohort were AO joint anomaly and foramen magnum width. When performing multivariate regression, AO joint anomaly and foramen magnum diameter were the only independent predictors of syrinx improvement, while surgical technique involving tonsillar reduction was the only one for syrinx resolution. Greater foramen magnum diameter was associated with increased likelihood of syrinx improvement, which alludes to the importance of adequate decompression in alleviating the aberrant CSF dynamics that lead to syringogenesis. To the authors' knowledge, the description of AO joint anomaly as a risk factor for syrinx persistence is a novel finding and requires further study on how to best treat these patients.

Limitations and future directions

The current study is not without limitations. Retrospective studies inherently incorporate selection bias in data collection and analysis, and not all possible radiographic parameters relevant to CM-I were measured (including obex position) [6, 19, 20]. Additionally, the study population had a preponderance of patients with syrinx compared to the general Chiari population, alluding to a population bias of patients that receive treatment at a large academic medical center. Future studies would benefit from further quantification of syrinx improvement, including a threshold value for postoperative syrinx size or percentage reduction in syrinx size that best predict clinical improvement or prevention of future decline. Deriving these proposed thresholds would likely require validation in a larger, prospective cohort of patients in a multicenter study.

Conclusion

The current study reports clinical and radiographic variables associated with preoperative syrinx presence and postoperative syrinx improvement. We observed that syrinx improvement or resolution after surgery is associated with greater CCOS and decreased complications. Moreover, surgical technique with tonsil reduction was associated with improved syrinx outcomes, and foramen magnum diameter and AO joint anomaly were the only two independent predictors of syrinx improvement and unimprovement, respectively. These insights will help guide preoperative patient counseling, surgical approach considerations, and postoperative prognostication when treating CM-I with syrinx.

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Declarations

Ethical approval The study was approved under the institutional ethical review board (IRB #STU-082018-025).

Conflict of interest The authors declare no competing interests.

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