Relationship of syrinx size and tonsillar descent to spinal deformity in Chiari malformation Type I with associated syringomyelia

Clinical article

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Object. Chiari malformation Type I (CM-I) is a developmental abnormality often associated with a spinal syrinx. Patients with syringomyelia are known to have an increased risk of scoliosis, yet the influence of specific radiographically demonstrated features on the prevalence of scoliosis remains unclear. The primary objective of the present study was to investigate the relationship of maximum syrinx diameter and tonsillar descent to the presence of scoliosis in patients with CM-I-associated syringomyelia. A secondary objective was to explore the role of craniocervical junction (CVJ) characteristics as additional risk factors for scoliosis.

Methods. The authors conducted a retrospective review of pediatric patients evaluated for CM-I with syringomyelia at a single institution in the period from 2000 to 2012. Syrinx morphology and CVJ parameters were evaluated with MRI, whereas the presence of scoliosis was determined using standard radiographic criteria. Multiple logistic regression was used to analyze radiological features that were independently associated with scoliosis.

Results. Ninety-two patients with CM-I and syringomyelia were identified. The mean age was 10.5 ± 5 years. Thirty-five (38%) of 92 patients had spine deformity; 23 (66%) of these 35 patients were referred primarily for deformity, and 12 (34%) were diagnosed with deformity during workup for other symptoms. Multiple regression analysis revealed maximum syrinx diameter > 6 mm (OR 12.1, 95% CI 3.63–40.57, $p < 0.001$) and moderate (5–12 mm) rather than severe (> 12 mm) tonsillar herniation (OR 7.64, 95% CI 2.3–25.31, $p = 0.001$) as significant predictors of spine deformity when controlling for age, sex, and syrinx location.

Conclusions. The current study further elucidates the association between CM-I and spinal deformity by defining specific radiographic characteristics associated with the presence of scoliosis. Specifically, patients presenting with larger maximum syrinx diameters (> 6 mm) have an increased risk of scoliosis.
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KEY WORDS • Chiari malformation • syringomyelia • scoliosis

CHIARI malformation Type I (CM-I) is a developmental abnormality of the craniocervical junction (CVJ), often associated with spinal cord abnormalities such as syringomyelia and scoliosis.^{10,33} The rate of scoliosis in pediatric patients with CM-I has been reported to be as high as 80% in those with concurrent syringomyelia.^{6,10,16,20,31} Some authors have speculated that the etiology of scoliosis in such patients is related to the effect of the expanding spinal cord syrinx on the function of medially located motor neurons; the resulting imbalance in the paraspinal musculature is thought to predispose the individual to spinal deformity.^{11,13,18} Others

have questioned the validity of this model and suggested tonsillar compression of the cervicomedullary junction as an alternate etiology of scoliosis in patients with CM-I.⁴

Despite the uncertain etiology, scoliosis in the context of syringomyelia and CM-I is frequently treated with decompression of the CVJ to stabilize the spinal deformity.^{7,8,15} Prior studies have demonstrated the importance of early neurosurgical intervention in patients with scoliosis to avoid or delay curve progression.^{6,7,16} While this approach is widely practiced, no guidelines are in place to identify the patients at risk for concurrent spinal deformity. Although authors have examined radiological features of CM-I-related scoliosis, none have reported the differences in the radiological features of syringomyelia and the CVJ between patients with and those without scoliosis.

We hypothesized that a larger maximum syrinx size

Abbreviations used in this paper: AP = anteroposterior; CM-I = Chiari malformation Type I; CVJ = craniocervical junction; CXA = clivus canal angle.

Syrinx size and scoliosis in Chiari malformation Type I

portends a greater risk of spinal deformity. In the current study, we investigated the relationship between maximum syrinx diameter and the presence of scoliosis in patients with CM-I and syringomyelia. A secondary aim was to explore the role of CVJ characteristics as additional risk factors for scoliosis.

Methods

Patient Population

After obtaining institutional review board approval from Washington University School of Medicine and St. Louis Shriner's Hospital, where patients were co-managed, we queried a pediatric neurosurgical database for patients evaluated for CM-I with syringomyelia in the period between 2000 and 2012. Criteria for inclusion in the search were an age of 18 years or younger, diagnosis of CM-I (tonsillar descent beyond the foramen magnum ≥ 5 mm), and maximum anteroposterior (AP) diameter of syringomyelia ≥ 2 mm. Exclusion criteria were previous decompression or spinal fusion; missing preoperative MRI or radiographic studies; idiopathic syringomyelia or syringomyelia related to tumor, infection, arachnoiditis, or previous trauma to the spinal cord; history of myelomeningocele, tethered cord, or other dysraphism; and connective tissue diseases or genetic syndromes. We reviewed demographic data, presenting symptoms, neurological deficits, and medical comorbidities, as well as pre- and postoperative radiological studies.

Imaging Data

Two different investigators, including a board-certified radiologist, assessed features of the posterior cranial fossa and the CVJ by using brain or cervical spine MRI. They measured: 1) tonsillar herniation, as defined by the distance between the tip of the cerebellar tonsil perpendicular to McRae's line (basion-opisthion line); 2) position of the obex relative to the foramen magnum (quantified by the perpendicular distance between the obex and a line drawn from the basion perpendicular to the central canal);^{35,36} 3) CM 1.5, if present (defined as descent of the obex below the foramen magnum);^{34,36} 4) pb-C2, perpendicular distance between the ventral dura to the line that joins the basion to the posterior portion of the axis body inferior endplate;⁹ 5) odontoid inclination (angle between the dens and the posterior wall of the C-2 vertebral body);³⁷ 6) clivus canal angle (CXA; determined by the angle between the Wackenheim's clivus line and the posterior wall of the C-2 vertebral body);^{28,31} 7) basilar invagination, if present;²⁷ and 8) medullary kinking, if present.²⁷

The location and morphology of the syrinx were evaluated using both T1- and T2-weighted MRI. Sagittal images of the spine and brain were used to determine the rostral and caudal extent of the syrinx, recorded as the number of vertebral segments traversed (as described by Ono et al.²⁰). Axial images were used to determine maximal AP diameter of the syrinx (in mm). Results of CSF flow studies were obtained when available.

The location and severity of spinal deformity were recorded using the standard Cobb technique with available imaging.³¹ Scoliosis was defined as a Cobb angle $> 10^\circ$.¹⁴ Patients without available imaging records were ex-

cluded from analysis, despite prior description of clinically significant scoliosis ($> 10^\circ$) in the medical records.

Statistical Analysis

Univariate statistical analysis was performed to identify the unadjusted association of basic demographic and radiological covariates with the presence of spinal deformity. Proportions were analyzed using Fisher's exact test, and continuous data were analyzed using univariate logistic regression. Tonsillar herniation and CXA were categorized using a cutoff of 1 SD from the mean. Collinearity between radiological covariates was examined using Pearson's correlation coefficients. Those covariates satisfying the statistical entry criteria ($p < 0.2$) on univariate analysis were included in the logistic regression model. Multiple logistic regression analysis was performed to assess independent risk factors for spinal deformity while adjusting for age and sex. The Hosmer-Lemeshow test was used to check goodness-of-fit of the model, and the predictive value of the multivariate model was expressed using the c-statistic. Statistical significance was established using a cutoff of $p < 0.05$. Data were analyzed using SPSS version 19.0 software (IBM).

Results

Patient Characteristics

Basic demographic data and chief complaints of the 92 patients (51 females and 41 males) with CM-I and syringomyelia are presented in Table 1. The overall mean age at syrinx diagnosis was 10 ± 5 years.

Radiological Features of Study Population

Syringomyelia was present in all patients in the study, with a mean maximal AP syrinx diameter of 5.9 ± 3.9 mm and syrinx length of 10 ± 5 vertebral levels. The syrinx crossed the cervicothoracic junction in 52 (56%) of 92 patients, was isolated to the thoracic or lumbar spine in 12 (13%), and spanned the length of the spinal cord in 6 (6%). On preoperative T1-weighted MRI, the mean cerebellar tonsillar herniation was 14.4 ± 7.2 mm, mean obex descent was 10.2 ± 5.0 mm, mean pb-C2 was 3.9 ± 1.8 mm, mean odontoid inclination angle was $16^\circ \pm 8^\circ$, and mean CXA was $155^\circ \pm 11^\circ$. Basilar invagination was identified in 3 patients and medullary kinking in 18 (20%) of 92. There was no correlation between tonsillar descent (Spearman's $\rho = -0.152$, $p = 0.149$) or CXA (Spearman's $\rho = -0.159$, $p = 0.131$) and maximum syrinx diameter. Syrinx size between the 2 groups of tonsillar descent (> 12 mm, 5–12 mm) was comparable (5.7 vs 6.1 , $p = 0.818$).

Cerebrospinal fluid flow studies were performed in 33 patients (36%); 7 (21%) of these patients demonstrated normal flow, 12 (36%) demonstrated posterior flow restriction only, and 14 (42%) demonstrated both anterior and posterior flow restriction. Tonsillar pulsation was observed in 14 patients (42%). Flow restriction was more often observed in patients with severe tonsillar herniation than in those with moderate herniation (100% vs 50%, $p = 0.001$); however, no difference existed in flow restriction between those with and those without scoliosis (100% vs 76%, $p = 0.55$). Syrinx

TABLE 1: Basic demographic and clinical characteristics of 92 patients with CM-I and syringomyelia*

Parameter	w/o Scoliosis (%)	w/ Scoliosis (%)	p Value
no. of patients	57	35	
mean age at syrinx diagnosis in yrs	10 ± 5.3	9.6 ± 3.8	0.696
no. of males	27 (47)	14 (40)	0.490
mean BMI	18 ± 6.5	19.2 ± 6.5	0.498
chief complaint			
scoliosis	0 (0)	23 (66)	—
neurological symptoms	33 (58)	17 (48)	—
headache	38 (67)	9 (26)	—
combination of symptoms	19 (33)	14 (40)	—

* Values represent the means ± standard deviation unless indicated otherwise. BMI = body mass index; — = not applicable.

size was unrelated to extent of flow restriction ($p = 0.26$) and presence of tonsillar pulsation ($p = 0.572$). These data were excluded from multivariate analysis because of the limited number of patients with available CSF flow studies.

Radiographic Features of Scoliosis

Formal preoperative spine radiographs (standing AP and lateral long cassette radiographs) were available in 35 (100%) of 35 patients. Thirty-five (38%) of 92 patients had spine deformity; 14 were male and 21 were female ($p = 0.472$). Spinal deformity had been previously diagnosed in 23 (66%) of the 35 and was diagnosed during workup for presenting symptoms in 12 (34%). Preoperative radiographic records were available for review in all patients with scoliosis (Table 2). The mean major coronal Cobb angle was $30.4^\circ \pm 15^\circ$; 18 curves (51%) were apex left. The mean thoracic kyphosis was $47^\circ \pm 15^\circ$, with 32 (91%) of 35 patients demonstrating a sagittal curve $> 30^\circ$. Patients diagnosed with scoliosis prior to referral to our institution demonstrated a larger Cobb angle (35° vs 22° , $p = 0.017$).

Of the 35 patients with spinal deformity, 15 (43%) ultimately required definitive spinal fusion within 24 months of posterior fossa decompression. Surgeries were offered in cases in which the coronal deformity had progressed to more than 50° , as measured using the traditional Cobb technique. Before reaching this radiographic measure, all patients were nonoperatively treated with various techniques, according to physician and patient preferences. Patients requiring spinal reconstruction demonstrated greater initial coronal deformity (40° vs 25° , $p = 0.003$) and greater sagittal deformity (55° vs 42° , $p = 0.027$). An initial coronal Cobb angle $> 30^\circ$ ($p = 0.014$) and kyphosis $> 45^\circ$ ($p = 0.03$) corresponded to a higher likelihood of scoliosis surgery. Age at decompression ($p = 0.786$), sex ($p = 0.176$), body mass index ($p = 0.779$), preoperative maximal syrinx diameter ($p = 0.152$), and tonsillar herniation ($p = 0.426$) were not associated with scoliosis surgery.

Radiological Features of Scoliosis

Univariate analysis of radiological risk factors associated with scoliosis are presented in Table 3. The presence of scoliosis was significantly associated with a larger maximum AP syrinx diameter (OR 1.5, 95% CI 1.3–1.8,

$p < 0.001$); a syrinx diameter > 6 mm corresponded to a higher rate of scoliosis (OR 6.80, 95% CI 2.64–17.52, $p < 0.001$). The relationship between syrinx size and scoliosis is demonstrated in Fig. 1. A cervicothoracic syrinx was more common in patients with spinal deformity (83% vs 40%, $p = 0.002$). The mean tonsillar herniation was 11.7 ± 5.7 mm in patients with scoliosis, as compared with 16.2 ± 7.6 mm in those without scoliosis (OR 0.898, 95% CI 0.83–0.97, $p = 0.004$). Patients with tonsillar herniation of 5–12 mm were more likely to demonstrate scoliosis (OR 3.38, 95% CI 1.40–8.16, $p = 0.007$), despite no difference in syrinx size ($p = 0.818$). No difference existed in the CXA ($p = 0.912$), pb-C2 ($p = 0.598$), odontoid inclination ($p = 0.547$), or presence of CM 1.5 ($p = 0.92$) between patients with and without scoliosis.

Following multivariate logistic regression analysis, maximum syrinx diameter > 6 mm (OR 12.1, 95% CI

TABLE 2: Radiographic features of scoliosis in 35 patients with CM-I and syringomyelia*

Parameter	Value
mean age in yrs	10 ± 4
no. of males (%)	14 (40)
mean BMI	21 ± 8
coronal	
mean major Cobb angle	30.4 ± 15°
no. of convex lt curves (%)	18 (51)
apex: IQR	T8–11
mean proximal thoracic Cobb angle	14 ± 11°
mean middle thoracic Cobb angle	27 ± 17°
mean thoracolumbar Cobb angle	17 ± 11°
mean coronal balance in mm	10 ± 9
sagittal	
mean major Cobb angle†	47 ± 15°
mean T2–5 Cobb angle	15 ± 7°
mean T5–12 Cobb angle	31 ± 19°
mean T12–S1 Cobb angle	55 ± 17°
apex: IQR	T6–8

* IQR = interquartile range.

† Ninety-one percent of patients had sagittal Cobb angle $> 30^\circ$.

Syrinx size and scoliosis in Chiari malformation Type I

TABLE 3: Univariate analysis of radiological features associated with scoliosis in patients with CM-I and syringomyelia

Parameter	w/o Scoliosis (%)	w/ Scoliosis (%)	p Value (2-sided)	OR (95% CI)
no. of patients	57	35		
CVJ				
mean tonsillar herniation in mm	16.2 ± 7.6	11.7 ± 5.7	0.004	0.898 (0.832–0.969)
no. w/ tonsillar herniation of 5–12 mm	19 (33)	22 (63)	0.007	3.385 (1.405–8.155)
no. w/ CM 1.5	17 (30)	11 (31)	0.920	1.07 (0.433–2.68)
mean obex descent in mm	11 ± 5	9.1 ± 4.7	0.084	1.084 (0.989–1.187)
no. w/ obex <11 mm	28 (49)	12 (34)	0.144	0.552 (0.218–1.249)
mean odontoid pb-C2 in mm	3.8 ± 1.9	4 ± 1.6	0.598	1.067 (0.842–1.351)
mean dens inclination	15.9 ± 7.8°	14.9 ± 7.7°	0.547	0.983 (0.930–1.039)
mean CXA	155.5 ± 10.8°	155.8 ± 11.4°	0.912	1.002 (0.964–1.042)
no. w/ CXA >150°	37 (65)	22 (63)	0.755	1.151 (0.447–2.777)
no. w/ basilar invagination	2 (4)	1 (3)	0.865	1.236 (0.108–14.16)
no. w/ medullary kinking	13 (23)	5 (14)	0.321	1.773 (0.572–5.493)
syrinx morphology				
mean syrinx diameter in mm	4.1 ± 3	8.9 ± 3.4	<0.001	1.537 (1.285–1.839)
no. w/ syrinx diameter >6 mm	17 (30)	26 (74)	<0.001	6.797 (2.637–17.521)
no. w/ syrinx diameter >7 mm	10 (18)	24 (68)	<0.001	10.255 (3.82–27.53)
mean syrinx length (no. of levels)	8.1 ± 5	13.8 ± 4	<0.001	1.264 (1.136–1.406)
syrinx location				
no. cervical	21 (37)	1 (3)	—	—
no. cervicothoracic	23 (40)	29 (83)	0.002	26.5 (3.31–211.8)
no. thoracolumbar	9 (16)	3 (9)	0.111	7 (0.639–76.708)
no. holocord	4 (7)	2 (6)	0.079	10.5 (0.758–145.35)
CSF flow studies				
no. w/ flow study performed	29 (51)	4 (11)	<0.001	—
no. w/ flow restriction	22 (76)	4 (100)	0.555	—
no. w/ tonsillar pulsation	11 (38)	3 (75)	0.279	—

3.63–40.57, $p < 0.001$) and moderate tonsillar herniation (5–12 mm; OR 7.64, 95% CI 2.3–25.31, $p = 0.001$) remained independently associated with the presence of spine deformity when adjusted for age, sex, and syrinx location (Table 4). No significant interactions were found. The model was stable and robust with a Hosmer-Lemeshow chi-square of 0.28 ($p = 0.869$) and c-statistic of 0.807 ($p < 0.001$).

Discussion

We have shown a strong association of the radiological features of syringomyelia and the CVJ with scoliosis in 92 children with CM-I and syringomyelia. Specifically, a larger maximum syrinx diameter and moderate (5–12 mm) rather than severe (> 12 mm) tonsillar ectopia were independently associated with a higher frequency of scoliosis. No association was observed between syrinx location and the presence of scoliosis when adjusted for syrinx size. While the severity of the spinal deformity did not correlate with the diameter of the syrinx, patients with more severe initial deformity (scoliosis > 30° or kyphosis > 45°) were more likely to require surgery for deformity correction despite decompression.

Previous authors have recognized the association among CM-I, scoliosis, and syringomyelia.^{6,20,36} The atypical patterns of CM-I-related deformities, such as left thoracic apices and hyperkyphosis, rapid progression, and early onset, suggest an etiology unique from that of adolescent idiopathic scoliosis.^{24,38} Both syringomyelia and hindbrain compression have been hypothesized in the pathogenesis of CM-I-related scoliosis; however, the role of each as a risk factor remains poorly defined.^{3,21,25,26} Although previous studies have recognized that CM-I patients with syringomyelia are more likely to demonstrate scoliosis,^{16,20,36} risk factors for clinically significant spinal deformity and the need for corrective spinal surgery are still debated.^{20,21,24}

Both Ozerdemoglu et al.²¹ and Qiu et al.²⁴ have explored the relationship between syrinx morphology and spinal deformity characteristics. In a series of 112 pediatric patients with scoliosis, Ozerdemoglu and colleagues reported a range of syrinx diameters (2–18 mm) with no correlation to curve magnitude.²¹ Recently, Qiu et al.²⁴ found no association between syrinx length or ratio of syrinx/cord diameter and curve severity in 83 patients. Similar to the study by Yeom et al.,³⁹ the current study revealed no association between syrinx characteristics and

Fig. 1. A scatterplot of scoliosis frequency against maximal AP syrnix diameter. Patients with a larger maximal syrnix diameter have scoliosis at a higher frequency. A maximum AP syrnix diameter > 6 mm indicates a higher prevalence of scoliosis.

coronal Cobb angle. Only Ono et al.²⁰ noted a correlation between scoliosis severity (12.1° vs 37.7°) and syrnix length (13.4 vs 15.9 levels) in an adult population with CM-I. Yet, in a study by Krieger et al.,¹⁶ all 79 patients with CM-I and scoliosis demonstrated a syrnix width > 6 mm. Recognizing the strong association between syrnix parameters and scoliosis, these authors implicated syrnix size in spinal growth. The preliminary association between greater maximum syrnix diameter and scoliosis in the current study builds on and significantly strengthens this theory. As we have shown, patients with a syrnix diameter > 6 mm demonstrate a significantly higher rate of scoliosis (OR 12.1). This finding also suggests that the mechanisms involved in scoliosis development may be distinct from those involved in curve progression, as syrnix size does not appear to be associated with the magnitude of coronal deformity.

Some authors describe a spatial component in the relationship between syringomyelia and scoliosis in CM-I.²¹ Ozerdemoglu et al.²¹ found that patients with a caudally located maximal syrnix diameter demonstrated distal major curve apices. The cervicothoracic junction was most often affected by a syrnix, with the largest syrnix area in the C5–7 region. Attenello et al.¹ found that the majority of patients with scoliosis demonstrated cervical (90%) and thoracic (95%) syringes. Tokunaga et al.³² described a syrnix at the cervicothoracic junction in 96% (22 of 23) of patients with scoliosis. The mean syrnix/spinal cord ratio in their study was 49%, with maximum dilation occurring most often at the C-6 level (17 of 23 cases). The current study showed a higher rate of scoliosis among cervicothoracic and holocord syringes on univariate analysis; however, when ad-

justed for syrnix length and maximum diameter, syrnix location did not confer an additional risk for scoliosis. This may suggest the susceptibility of the cervicothoracic junction to syrnix expansion and highlights the strength of the association between syrnix size and scoliosis.²¹

The prevalence of scoliosis in CM-I without syringomyelia has been reported in 8%–30% of patients.^{19,31} Some authors suggest that this prevalence implicates tonsillar herniation as the underlying etiology of CM-I-related scoliosis, while others argue that this rate may reflect a basal level of scoliosis unrelated to CM-I.¹⁶ Although this unique patient population was not included in the current study design, we intend to investigate it in a future study.

Few studies have shown a direct association between CVJ parameters and scoliosis in CM-I patients with or without syringomyelia. Only Ono et al.²⁰ reported a relation between levels of tonsillar descent and spinal deformity characteristics; they documented 70% concordance between the laterality of cerebellar tonsil asymmetry and the direction of major curve convexity. To our knowledge, no previous study has found moderate tonsillar ectopia (5–12 mm) to be associated with scoliosis in patients with CM-I and syringomyelia. Attenello et al.¹ found that 90% of patients with CM-I and scoliosis demonstrated “moderate ectopia,” which was defined as tonsillar descent between C-1 and C-2. Similar to the value noted in the current study, Tokunaga et al.³² reported a mean tonsillar herniation of 9.7 mm in 23 patients with CM-I-related scoliosis. Other studies have also found a high proportion of patients with moderate descent. Qiu et al.²⁴ showed that 44% of tonsils descended to the region between the foramen magnum and the C-1 level in 87 patients with scoliosis. They found no correlation between tonsillar herniation and curve severity. In a study of CM-I-related scoliosis, Krieger et al.¹⁶ found that 84% of patients had tonsils above the C-2 level.

It is surprising that patients with only moderate tonsillar ectopia (5–12 mm) have a greater risk of clinically significant scoliosis, whereas patients with more severe tonsillar descent (> 12 mm) demonstrate a lower prevalence of scoliosis. While no difference in syrnix size exists between the 2 groups, several studies have reported a similarly perplexing association between moderate ectopia and the presence of syringomyelia in CM-I.^{23,30,37} In light of this observation, our findings may reflect a complex process at the CVJ.¹⁷ Several authors have suggested that the degree of obex descent at the CVJ could influence the spinal axis and the development of syringomyelia.²⁹ Yet, we observed no association between obex descent and the presence of CM 1.5 with scoliosis. Although dynamic imaging of CSF flow in patients with CM has helped shape our understanding of the pathophysiology of syringomyelia,⁵ we were unable to include these studies in the multivariate analysis

TABLE 4: Independent risk factors for scoliosis in patients with CM-I and syringomyelia*

Parameter	OR	95% CI	p Value
max syrnix diameter >6 mm	12.1	3.63–40.57	<0.001
tonsillar herniation of 5–12 mm	7.64	2.3–25.31	0.001

* Hosmer-Lemeshow chi-square = 0.28, p = 0.869.

Syrinx size and scoliosis in Chiari malformation Type I

because of the small number of patients with complete testing. Likely, there is a dynamic component to the development of scoliosis that warrants further investigation using dynamic imaging.

Ultimately, the goal of decompression in CM-I patients with scoliosis is to stabilize curve progression and thereby prevent the need for complex spinal reconstruction. Although the natural history of scoliosis in CM-I remains unclear, a recent meta-analysis by Hwang et al.¹² suggests the benefit of decompression and syrinx reduction in improving scoliotic curve progression. But despite adequate posterior fossa decompression, the rate of scoliosis surgery ranges between 15% and 50%, comparable with the 43% described in the current study.^{2,6,16,25} Past authors have described an older age, level of spinal deformity, extent of syrinx resolution, and degree of initial scoliosis as risk factors for curve progression and spinal fusion.^{1,2,6,12,22,25} In the largest study to date, Krieger et al.¹⁶ noted a higher rate of surgery in patients with an initial Cobb angle > 25°, with no association with preoperative syrinx size or extent of syrinx resolution. Similarly, we observed that patients with scoliosis > 30° as well as those with kyphosis > 50° were significantly more likely to require spinal reconstruction within the follow-up period. However, the decision to perform spinal reconstruction involves multiple variables, among which are patient and physician preferences. While it is likely that early intervention plays a role in avoiding surgery, additional studies are needed to adequately determine the optimal management of spinal health in these complex cases.

A central limitation of our study is its retrospective, cross-sectional design. The data and their interpretation are limited by completeness of the medical records and consistency between preoperative evaluations, particularly the availability of standing plain films of the spine. A number of patients did not seek orthopedic care at our institution, thus limiting our ability to capture longitudinal data and examine the temporal relationship among syringomyelia, scoliosis development, and scoliosis surgery. Importantly, our study population included only those patients with syringomyelia ≥ 2 mm on MRI evaluation; we hope to investigate patients with CM-I–related scoliosis without syringomyelia in the future. Note also that this study is subject to the bias of a tertiary care referral center, as the composition of our study population represents only those patients referred to our clinic. To capture the true incidence of spinal deformity in patients with CM-I and better understand risk factors of progression and functional outcome, future research in this field must come in the form of a prospective, multicenter effort such as that of the Park-Reeves Syringomyelia Research Consortium.

No study has examined radiological features of the CVJ and syringomyelia as potential risk factors for the presence of scoliosis in CM-I with syringomyelia. We have shown that coexistent scoliosis is more likely with a larger maximum syrinx diameter and moderate tonsillar herniation (5–12 mm). While the results of this study have limited clinical implications, the findings presented herein suggest that dilation of the spinal cord by syringomyelia, as well as compression at the cervicomedullary junction, appears to influence spinal growth in CM-I.

Prospective studies conducted through the Park-Reeves Syringomyelia Research Consortium are directed toward further elucidating these complex relationships.

Conclusions

Results from this retrospective study demonstrate a strong association between maximum syrinx size and the presence of spinal deformity in patients with CM-I and syringomyelia. This finding represents an early but important component in the investigation of potential common mechanisms involved in the pathogenesis of these complex conditions.

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Disclosure

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