The association between Chiari malformation Type I, spinal syrinx, and scoliosis

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OBJECT Chiari malformation Type I (CM-I) is often found in patients with scoliosis. Most previous reports of CM-I and scoliosis have focused on patients with CM-I and a spinal syrinx. The relationship between CM-I and scoliosis in the absence of a syrinx has never been defined clearly. The authors sought to determine if there is an independent association between CM-I and scoliosis when controlling for syrinx status.

METHODS The medical records of 14,118 consecutive patients aged ≤ 18 years who underwent brain or cervical spine MRI at a single institution in an 11-year span were reviewed to identify patients with CM-I, scoliosis, and/or syrinx. The relationship between CM-I and scoliosis was analyzed by using multivariate regression analysis and controlling for age, sex, CM-I status, and syrinx status.

RESULTS In this cohort, 509 patients had CM-I, 1740 patients had scoliosis, and 243 patients had a spinal syrinx. The presence of CM-I, the presence of syrinx, older age, and female sex were each significantly associated with scoliosis in the univariate analysis. In the multivariate regression analysis, older age (OR 1.02 [95% CI 1.01–1.03]; $p < 0.0001$), female sex (OR 1.71 [95% CI 1.54–1.90]; $p < 0.0001$), and syrinx (OR 9.08 [95% CI 6.82–12.10]; $p < 0.0001$) were each independently associated with scoliosis. CM-I was not independently associated with scoliosis when controlling for these other variables (OR 0.99 [95% CI 0.79–1.29]; $p = 0.9$).

CONCLUSIONS A syrinx was independently associated with scoliosis in a large pediatric population undergoing MRI. CM-I was not independently associated with scoliosis when controlling for age, sex, and syrinx status. Because CM-I is not independently associated with scoliosis, scoliosis should not necessarily be considered a symptom of low cerebellar tonsil position in patients without a syrinx.

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KEY WORDS Chiari malformation Type I; scoliosis; spinal syrinx

CHIARI malformation Type I (CM-I) is often associated with a spinal syrinx.^{26,33} Patients who have both CM-I and a syrinx are more likely to undergo surgery than those with CM-I alone.²⁸ For this reason, surgical series tend to overestimate the true prevalence of syringes associated with CM-I.^{24,33} When all patients with CM-I discovered on imaging are considered without selecting for those who are symptomatic or undergoing treatment, a syrinx is found in a smaller, but still substantial, percentage of those with CM-I.³⁰ Spinal syrinx is associated with scoliosis in some individuals,^{3,18,19,36} perhaps as a result of asymmetrical injury to the spinal cord from an expanding cyst.¹⁶

Although most researchers agree that CM-I can cause a spinal syrinx and that a spinal syrinx can cause scoliosis,^{3,5,6,11,12,15,20} the association of CM-I and scoliosis in the absence of a syrinx has never been defined properly and remains controversial. Some researchers have speculated that asymmetrical compression of the cervicomedullary junction by the cerebellar tonsils can result in scoliosis even in the absence of a spinal syrinx.^{5,6,34,37} Nevertheless, there is scant existing evidence for such a causal relationship between CM-I and scoliosis in the absence of a syrinx. Arguing in favor of such a relationship are several case reports of patients with CM-I and scoliosis in the absence of a syrinx.^{13,22,34} However, given the high preva-

ABBREVIATIONS CM-I = Chiari malformation Type I; OR = odds ratio.

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lence of both CM-I^{23,30} and scoliosis,^{7,35} both conditions in an individual can occur by chance in many instances. Therefore, the existence of reports of individual patients or even small series of patients with both findings does not prove a causative relationship. In addition, there are case series in which CM-I as a cause of scoliosis has been examined.^{11,20,34} Unfortunately, a majority of these patients also have a spinal syrinx, making it impossible to draw any conclusion on the relationship between CM-I and scoliosis in the absence of a syrinx. To examine this relationship, we analyzed a large cohort of children who, over an 11-year span, underwent brain or cervical spine MRI. We then performed multivariate regression analysis to determine if there was an independent relationship between CM-I and scoliosis when we controlled for syrinx status.

Methods

After approval by the University of Michigan Institutional Review Board, we examined the medical records of 14,118 consecutive children aged ≤ 18 years who underwent brain or cervical spine MRI at the University of Michigan in an 11-year span. Electronic records, including radiology reports, surgical reports, all office consultation notes, and hospital chart entries, were reviewed by using the Electronic Medical Record Search Engine (EMERSE)¹⁴ to identify patients with CM-I, syrinx, and/or scoliosis by searching for the key words “tonsillar ectopia,” “tonsillar herniation,” “tonsillar descent,” “tonsil,” “syrinx,” “syringomyelia,” “hydromyelia,” “Chiari,” or “scoliosis” in any part of the electronic medical record. All imaging records and all other medical records for patients selected in this way were reviewed to confirm the diagnoses.

Each patient was assigned to a category on the basis of his or her syrinx status, CM-I status, and scoliosis status. For the purpose of this analysis, CM-I was defined on imaging as a cerebellar tonsil position at least 5 mm below the foramen magnum.^{1,4,29,30} A spinal syrinx was defined as a spinal cord cyst (hypointense signal on T1-weighted images and hyperintense signal on T2-weighted images) of at least 3 mm wide seen on an axial image.³⁰ Scoliosis was defined as at least a 10° Cobb angle seen on a radiograph. Although radiographs were available for most of the patients with a diagnosis of scoliosis, those without radiographs but with a clear description and diagnosis of scoliosis in the medical record were also included in the scoliosis category.

There were 554 patients excluded from analysis. Patients were excluded if they had Chiari malformation Type II or III or any risk factors for low cerebellar tonsil position other than idiopathic CM-I, including mass effect from tumor, hydrocephalus, intracranial cysts, cerebral edema, or craniosynostosis. Patients who received surgical treatment for CM-I before their first MRI at our institution were excluded. Patients with a syrinx and Chiari malformation Type 0, intramedullary spinal cord tumor, or open myelomeningocele were also excluded. The final analysis included 13,564 patients.

Statistical significance calculations were obtained using the chi-square test, the t-test, ANOVA, and multivariate

regression analysis. Multivariate regression analysis for scoliosis was performed while taking into account CM-I status, syrinx status, sex, and age. Raw odds ratios (ORs) were produced using 2 × 2 tables and chi-square testing. Data were analyzed using SAS 9.3 (SAS Institute, Inc.) and StatPlus (AnalystSoft, Inc.) software. For the multivariate regression analysis, we analyzed CM-I as a categorical variable rather than a continuous variable; patients with a cerebellar tonsil position of ≥ 5 mm below the foramen magnum were analyzed together. Scoliosis and syrinxes were also analyzed as categorical variables for multivariate regression analysis.

Results

In this cohort of 13,564 patients, 52% were male, and the mean age at the time of MRI was 7.7 years. Scoliosis was present in 1740 patients (12.8%), 509 patients (3.8%) had a cerebellar tonsil position of ≥ 5 mm below the foramen magnum, and 243 (1.8%) had a syrinx of ≥ 3 mm in maximal diameter. Of the 1740 patients with scoliosis, 114 (6.6%) had CM-I, 137 (7.9%) had a syrinx, and 72 (4.1%) had both CM-I and a syrinx.

Of the 114 patients with both CM-I and scoliosis, 72 had a syrinx and 42 did not (Table 1). The mean age at the time of the scoliosis diagnosis was 9.5 years (Fig. 1). There was no difference in the mean ages at diagnosis between those with and those without a syrinx. There was a similar sex distribution for those with a syrinx (69.4% female) and those without a syrinx (69.0% female). The patients with both CM-I and a syrinx had a lower cerebellar tonsil position (mean 12.9 mm) than those with CM-I and no syrinx (mean 9.1 mm; $p < 0.001$). Patients with a syrinx were more likely to have a curve of $> 20^\circ$ (70.8%) than those without a syrinx (45.2%; $p < 0.01$). The locations of the curve with the largest Cobb angle for the group as a whole were distributed similarly between the thoracic (51%) and thoracolumbar (46%) regions, with a minority in the lumbar spine (2.6%). Thirteen patients (11.4%) had a left thoracic curve. Neither the presence of a syrinx ($p = 0.9$) nor the syrinx width ($p = 0.3$) was related to the presence of a left thoracic curve. The average greatest curve was 31°. There was no association between cerebellar ton-

TABLE 1. Characteristics of the study patients with CM-I and scoliosis (n = 114)

Characteristic	Syrinx (n = 72)	No Syrinx (n = 42)	p Value
Female (no. [%])	50 (69.4)	29 (69.0)	0.96
Mean age at scoliosis diagnosis (yrs)	9.4	9.7	0.3
Lt thoracic curve (no. [%])	8 (11.1)	5 (11.9)	0.9
Degree of greatest curve (°)	32.1	28.5	0.10
Patients w/ $\leq 20^\circ$ curve (no. [%])	21 (29.2)	23 (54.8)	<0.01
Patients w/ $> 20^\circ$ curve (no. [%])	51 (70.8)	19 (45.2)	<0.01
Mean cerebellar tonsil position (mm)*	12.9	9.1	<0.001

* Below the foramen magnum.

FIG. 1. Bar graph illustrating the number of patients with CM-I stratified according to age at the time of their scoliosis diagnosis.

sil position (analyzed as a continuous variable; $p = 0.7$) or cerebrospinal fluid flow (categorized as normal, decreased anterior/posterior to the foramen magnum, or abnormal tonsillar pulsations) at the foramen magnum ($p = 0.7$) and the degree of the largest curve. There were no associations between syrinx width (analyzed as a continuous variable; $p = 0.4$) or cranial extent of the syrinx ($p = 0.8$) and the degree of the greatest curve. There were no differences in sex, age at CM-I diagnosis, cerebellar tonsil position, or cerebrospinal fluid flow in patients who presented with a curve of $> 20^\circ$ versus those who presented with a curve of $\leq 20^\circ$.

In univariate analysis of the risk factors for scoliosis in the 13,564 patients who underwent MRI, female sex ($p < 0.0001$), presence of a syrinx ($p < 0.0001$), and presence of CM-I ($p < 0.0001$) were each significantly associated with scoliosis. In multivariate regression analysis, female sex ($p < 0.0001$), older age ($p < 0.0001$), and presence of a syrinx ($p < 0.0001$) were independently associated with scoliosis. CM-I was not independently associated with scoliosis when we controlled for these other factors ($p = 0.9$). ORs were calculated for these variables after performing the univariate and multivariate analyses (Table 2). ORs were calculated for female sex (OR 1.71 [95% CI 1.54–1.90]), older age (OR 1.02 [95% CI 1.01–1.03]), and syrinx (OR 9.08 [95% CI 6.82–12.10]) in multivariate analysis. CM-I was not independently associated with scoliosis when we controlled for these other variables (OR 0.99 [95% CI 0.79–1.29]).

Discussion

Patients with both CM-I and scoliosis are frequently referred to neurosurgeons for consideration of Chiari decompression surgery. When a syrinx is also present, the decision to proceed with Chiari decompression is not controversial,²⁸ because the association between CM-I and syrinxes is well established.^{10,26} It is also clear that those patients with both CM-I and a syrinx are at risk for scoliosis.^{11,30,32} Our data confirmed this well-established association between syrinxes and scoliosis. The management of a patient with scoliosis and an otherwise asymptomatic CM-I is not as clear in the absence of an associated syrinx. Our goal was to evaluate this relationship in the absence of a syrinx by analyzing the independent association between CM-I and scoliosis while controlling for syrinx status. ORs from the multivariate analysis indicate that syrinxes, female sex, and age have associations with scoliosis. Because the OR for older age was close to 1, we believe that this finding is unlikely to be clinically significant. Most important is that CM-I was not independently associated with scoliosis in our multivariate analysis.

Some reports have suggested that there is an independent relationship between low cerebellar tonsil position and scoliosis,^{2,8,9,18,25,31} perhaps as a result of dorsal compression of the cervicomedullary junction by the tonsils.^{5,6,37} However, much of the existing evidence that could be used to suggest an independent association for CM-I and scoliosis in the absence of a syrinx is inconclusive. First, there have been several reports of patients with CM-I and sco-

TABLE 2. Odds ratios for scoliosis in the study cohort

Characteristic	Univariate Analysis		Multivariate Analysis	
	p Value	OR (95% CI)	p Value	OR (95% CI)
Female sex	<0.0001	1.76 (1.59–1.95)	<0.0001	1.71 (1.54–1.90)
Syrinx	<0.0001	9.45 (7.29–12.24)	<0.0001	9.08 (6.82–12.10)
CM-I	<0.0001	2.03 (1.64–2.51)	0.94	0.99 (0.79–1.29)

liosis without syringomyelia; however, most of these cases were drawn from larger case series that included a majority of patients with a syrinx and CM-I.^{12,13,22,34} Because both low cerebellar tonsil position and scoliosis are common, it is not surprising that many individuals with both conditions have been identified.^{7,23,30,35} Others have cited an improvement in scoliosis after CM-I decompression in patients without an associated syrinx;^{12,17,34} however, most patients in these series had a syrinx, making it difficult to support any independent effect of CM-I without a syrinx.¹⁷ Also, the natural history of scoliosis is variable, so it is not clear whether a few examples of postsurgical improvement in patients without a syrinx differs from the natural history of this condition seen in other individuals.³⁴ Minor curve improvements are not unusual within groups of patients with scoliosis followed without treatment,²¹ and on the basis of the existing literature, it is difficult to make a case for the therapeutic benefit of Chiari decompression surgery to treat scoliosis in the absence of a syrinx.

Other evidence that has been cited to support the notion that CM-I may cause scoliosis in the absence of a syrinx is the prevalence of CM-I on screening MR images of patients undergoing scoliosis repair.^{18,25} Several studies have noted an association of low cerebellar tonsil position and idiopathic scoliosis.^{2,8,9,18,25,31} Cerebellar tonsil position, however, is distributed in an approximately normal distribution pattern across all age groups, and low tonsil position is not rare, even in healthy individuals.²⁹ Furthermore, some groups have used definitions for tonsil ectopia in patients with scoliosis that fall within the range of normal tonsil position as it is usually defined.^{8,27,31} These reports confirm only that CM-I is found in a small percentage of patients with scoliosis, not that there is an independent association between CM-I and scoliosis. It is notable as well that these studies found no association between low tonsil position and the severity of curve or the presence of an atypical curve.^{2,31}

We analyzed a large number of patients in an MRI database to study the relationship between CM-I and syrinxes. Patients in this cohort were categorized by CM-I, syrinx, and scoliosis status. When we controlled for known risk factors for scoliosis, including presence of a syrinx, female sex, and age, there was no longer an independent association between CM-I and scoliosis. When CM-I is associated with a syrinx, there is an increased risk for scoliosis and, on the basis of our analysis, this increased risk is most likely from the syrinx. In cases of CM-I without a syrinx, no association with scoliosis was found. We conclude that although CM-I is associated with syrinxes and syrinxes are associated with scoliosis, the syrinx is a necessary intermediary in most cases. When CM-I is present without a syrinx, there does not seem to be a significantly increased risk of scoliosis.

There are several limitations to our study. The patients we analyzed are representative of those who underwent MRI at a tertiary referral center and therefore do not represent a true population cohort. The cohort of patients in the study was derived from all individuals who underwent brain or cervical spine imaging in an 11-year span, which ensured that cerebellar tonsil position could be assessed accurately in every case. Because patients with scoliosis

are more likely to be referred for diagnostic imaging, there was a detection bias for scoliosis in this study. For this reason, the number of detected cases of scoliosis in this group was higher than would be expected in the population as a whole. The original imaging interpretation was performed by unblinded observers and, therefore, was subject to observer bias. Finally, the multivariate regression included CM-I status, presence of a syrinx, sex, and age but did not include other potentially important factors such as a history of meningitis, subarachnoid hemorrhage, or bony abnormalities.

We did not find an independent association between CM-I and scoliosis after controlling for syrinx status. The lack of an independent association indicates that CM-I is unlikely to be causative for scoliosis in the absence of a spinal syrinx. Finally, and most important, the lack of an independent association casts some doubt on the existence of any putative therapeutic benefit of CM-I decompression surgery for treating scoliosis alone in the absence of a syrinx or other symptoms.

Conclusions

There is a strong association between syrinxes and scoliosis, but there does not seem to be a significant relationship between CM-I and scoliosis when controlling for the presence of a syrinx.

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Author Contributions

Conception and design: Maher, Strahle. Acquisition of data: all authors. Analysis and interpretation of data: Maher, Strahle, Smith. Drafting the article: Maher, Strahle, Garton, Muraszko. Critically revising the article: Maher, Strahle. Reviewed submitted version of manuscript: all authors. Approved the final version of the manuscript on behalf of all authors: Maher. Study supervision: Maher.

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