HHS Public Access

Author manuscript

Eur Spine J. Author manuscript; available in PMC 2017 February 01.

Published in final edited form as:

Eur Spine J. 2016 February ; 25(2): 619–626. doi:10.1007/s00586-015-4011-1.

Comparison of Spinal Deformity in Children with Chiari I Malformation with and without Syringomyelia: Matched Cohort Study

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Abstract

Purpose—Describe curve patterns in patients with Chiari Malformation I (CIM) without syringomyelia, and compare to patients with Chiari Malformation with syringomyelia (CIM+SM).

Methods—Review of medical records from 2000 to 2013 at a single institution was performed to identify CIM patients with scoliosis. Patients with CIM were matched (1:1) by age and gender to CIM+SM. Radiographic curve patterns, MRI-based craniovertebral junction parameters, and associated neurological signs were compared between the two cohorts.

Results—Eighteen patients with CIM-associated scoliosis in the absence of syringomyelia were identified; 14 (78%) were female, with mean age of 11.5±4.5 years. Mean tonsillar descent was 9.9±4.1 mm in the CIM group and 9.1±3.0mm in the CIM+SM group (p=0.57). Average syrinx diameter in the CIM+SM group was 9.0±2.7mm. CIM patients demonstrated less severe scoliotic curves (32.1° vs. 46.1°, p = 0.04), despite comparable thoracic kyphosis (43.7° vs. 49.6°, p = 0.85). Two (11%) patients with CIM demonstrated thoracic apex left deformities compared to 9/18 (50%) in the CIM+SM cohort (p = 0.01). Neurological abnormalities were only observed in the group with syringomyelia (6/18, or 33%; p = 0.007).

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Conflict of Interest: This project was supported by the Clinical and Translational Science Award (CTSA) program of the National Center for Advancing Translational Sciences (NCATS) of the National Institutes of Health (NIH) under Award Number TL1 TR000449. Additional funding was also provided by the NIH T35 NHLBI Training Grant, under Grant Number 5 T35 HL007815. Washington University, Department of Orthopaedic Surgery – Spine Service received grant monies from Axial Biotech, DePuy Synthes Spine, an NIH grant (2010–2015), and AOSpine, SRS and Norton Healthcare, Louisville, KY (Scoli-RISK-1 study), philanthropic research funding from the Fox Family Foundation (Prospective Pediatric Spinal Deformity study), fellowship funding from AOSpine North America (funds/fellow year). Dr. Lenke shares numerous patents with Medtronic (unpaid). He is a consultant for DePuy Synthes Spine, K2M, Medtronic (monies donated to a charitable foundation). He receives substantial royalties from Medtronic and modest royalties from Quality Medical Publishing. Dr. Lenke also receives or has received reimbursement related to meetings/courses from AOSpine, BroadWater, DePuy Synthes Spine, K2M, Medtronic, Scoliosis Research Society, Seattle Science Foundation, Stryker Spine, The Spinal Research Foundation. Dr. Limbrick has technology licensed to Allied Minds, Inc. for an unrelated project.

Conclusion—In the largest series specifically evaluating CIM and scoliosis, we found that these patients appear to present with fewer atypical curve features, with less severe scoliotic curves, fewer apex left curves, and fewer related neurological abnormalities than CIM+SM. Notably, equivalent thoracic kyphosis was observed in both groups. Future studies are needed to better understand pathogenesis of spinal deformity in CIM with and without SM.

Keywords

Chiari malformation; syringomyelia; scoliosis; kyphosis; spinal deformity

INTRODUCTION

Chiari I malformation (CIM) with syringomyelia (CIM+SM) is associated with scoliosis, with 14–50% of patients demonstrating clinically significant spinal deformity. [1–3] Atypical curve patterns associated with CIM+SM-related deformities, including apex left curves, thoracic hyperkyphosis, early onset, and rapid progression suggest an underlying process that is distinct from idiopathic scoliosis. [4, 5] Some have suggested that the pathogenesis involves the destruction of motor neurons by the expanding syrinx, leading to weakness of paraspinal muscles and progressive loss of spinal alignment. [6]

Scoliosis exists in CIM patients in the absence of syringomyelia (CIM alone), with a prevalence ranging from 8–30%. [7, 8] Several authors have suggested that the absence of syrinx in these patients implies an alternate mechanism for the etiology of scoliosis in CIM alone, [9, 10] while others argue that this rate may reflect a basal level of scoliosis unrelated to CIM. [1] This subgroup of patients remains largely unstudied and has been reported primarily in the context of case-reports and small observational studies, and there is a particular dearth of data regarding the differences in presentation in CIM compared with CIM+SM.

This matched cohort study focuses on this key subgroup of patients who presented with CIM and scoliosis without syringomyelia, comparing them to patients with the more common presentation of CIM+SM-associated scoliosis. Using clinical, radiographic, and MRI data, we (1) describe curve patterns in cohort of patients with scoliosis in the presence of CIM alone, and (2) compare the initial curve patterns and physical findings with age/gender matched cohorts of CIM+SM-associated scoliosis patients.

METHODS

Patient Population

After Institutional Review Board approval, patients who had been evaluated for CIM and scoliosis between 2000 and 2013 at a single institution were identified using ICD-9 codes (754.2, 737, 741, 348.4) and manual review of an institutional database. Inclusion criteria for the search were age ≤ 18 years, diagnosis of CIM (tonsillar position ≥ 5 mm below McCrae's line), and scoliosis with major Cobb angle >10 degrees. Exclusion criteria included: previous Chiari decompression; missing MRI or x-ray studies; spinal deformity related to tumor, infection, arachnoiditis, or previous trauma to the spinal cord; history of

myelomeningocele, tethered cord, or other dysraphism; subjects with connective tissue diseases or genetic syndromes. For the purposes of this study, syringomyelia was defined as cavitation of the spinal cord measuring ≥ 2 mm in the anterior-posterior dimension on axial MR imaging. Confirmation of cavitation was performed using both T1- and T2 sequences; T2 sequences alone were not sufficient to document syrinx presence.

We identified 18 patients (18/542, 3.3%) with spinal deformity and CIM alone (no syringomyelia). Patients with scoliosis and CIM alone were matched one-to-one with patient with scoliosis and CIM+SM. Matching characteristics included age (within 1 year) and gender. In the event of multiple matches, a random number generator was used to select the appropriate match. Patient data were extracted from the medical records and reviewed for demographic information, presenting symptoms, neurological abnormalities, medical co-morbidities, all available radiological studies, and history of subsequent surgical procedures.

Radiographic Review

MRIs of the spine obtained *prior to Chiari decompression* were reviewed for both groups. The following parameters were recorded: (1) tonsillar position, measured from McRae's Line to the caudal-most tip of the cerebellar tonsil; (2) tonsillar herniation classified by the grading scheme described by Ono et al., designating Grade I as a tonsil descending beyond the foramen magnum without reaching the C1 arch, Grade II with a tonsil descending to the C1 arch, and Grade III with a tonsil descending beyond the C1 arch;[2] (3) symmetry of tonsillar ectopia, determined as the ratio of the distance of tonsillar descent between the left-sided tonsil and right-sided tonsil in the coronal plane ratio (ratio <0.9 or >1.10 was defined as asymmetric); [11] (4) posterior basion to C2 distance (pB-C2: distance measured from the B-C2 line, which is drawn from the basion to the infero-posterior C2 body);[11] (5) clivus-canal angle (angle formed by Wackenheim's clivus baseline and the posterior C2 vertebral body line). [8, 12] In the CIM+SM group, syrinx size was expressed as the maximum anteroposterior diameter (mm) of the syrinx cavity. Syrinx length, measured in vertebral segments traversed, was also recorded.

Standard radiographic measurements of spinal deformity were recorded on standing posteroanterior x-rays obtained prior to Chiari decompression or spinal fusion. Curve magnitude was measured using the standard Cobb method. [13] The apex of the curve (left or right) was defined as the direction of the convexity according to convention. Sagittal alignment of the spine and pelvis were assessed using: (1) pelvic incidence;[14, 15] (2) sacral slope[14, 15] (3) sagittal vertical axis;[14, 15] (4) lumbar lordosis;[14, 15] (5) thoracic kyphosis (measured from T2 to T12). [14, 15] Based on previously reported data, thoracic kyphosis was categorized as: hypokyphosis (<20), normal (20–50), and hyperkyphosis (>50). [16] Where available, change in major curve magnitude (progression/stabilization/improvement) following posterior fossa decompression was determined after a minimum of 12-month followup.

Statistical Analysis

Data were reported using mean and standard deviation. Analysis of continuous and dichotomous variables was conducted using the independent-samples t-test and Fisher's

exact test, respectively. Statistical significance was set at a threshold of $p < 0.05$. Statistical analysis was performed using SPSS v.22 (SPSS, Inc, Chicago, IL).

RESULTS

Patient Population

Eighteen CIM patients were matched to eighteen CIM+SM patients based on age and gender. Mean age was 11.5 ± 4.5 years (CIM: 11.5; CIM+SM: 11.6, p = 0.94), and 22% (8/36) of the patients were male (CIM: 22%; CIM+SM: 22%, p = 1.0). Table 1 demonstrates a comparison of demographic and clinical characteristics of the two groups of patients. The majority of patients in both groups were initially referred for management of associated scoliosis (CIM: 61%; CIM+SM: 83%, p=0.136). On physical examination, the CIM+SM group demonstrated a higher rate of neurological abnormalities (CIM: 0%; CIM+SM: 33%, p = 0.007), including upper extremity paresthesias (N=3), asymmetric reflexes (N=2), and lower extremity weakness (N=1).

MRI Imaging

On MR imaging, CIM patients demonstrated a mean tonsillar descent of 9.9 mm ± 4.1, compared to 9.1 mm ± 3.0 in the CIM+SM cohort (p=0.50). There was no statistically significant difference between two cohorts in the prevalence of Grade I tonsillar ectopia (61% vs. 44%, p=0.195), No significant differences were observed in craniovertebral junction parameters between CIM and CIM+SM cohorts (Table 2). All patients with a measurable clival canal angle had CXA >125°; 50% had CXA <150° (CIM: 46% vs. CIM +SM: 54%, p = 1.0). The CIM+SM group had mean syrinx size 9.0 mm ± 2.7, with mean syrinx length of 13 ± 5 vertebral levels.

Radiographic Imaging

Radiographic data are reported in Table 3. Mean coronal Cobb angle was greater in the CIM +SM cohort than CIM at presentation (CIM: 32.1° ± 22.4°; CIM+SM: 46.1° ± 15.7°, p=0.04). The CIM+SM cohort had a higher frequency of apex left deformities than CIM (CIM: 11%; CIM+SM: 50%, p = 0.011). No difference was observed between the two groups in regards to thoracic kyphosis (CIM: 43.7° ± 24.9°; CIM+SM: 49.6° ± 20.2°, p=0.85), nor SVA (CIM: −12.6 mm ± 39.6; CIM+SM: −21.5 mm ± 15.0 p=0.433). Overall, 14 patients (39%) demonstrated thoracic kyphosis greater than 50 degrees; 6/18 (33%) in CIM and 8/18 (44%) in CIM+SM (p=0.733).

Across both groups, kyphosis was associated with the extent of tonsillar herniation (Spearman's rho= −0.483, p = 0.009). No association was found between the degree of kyphosis and age (p = 0.943), gender (p = 1.00), BMI (p = 0.868), presence of neck pain (p=0.681), or size of coronal deformity (p = 0.544). Curve direction was not associated with tonsil laterality (p=0.136), and degree of scoliosis was unrelated to distance of tonsillar herniation (p=0.851). In the CIM+SM cohort, syrinx size did not correlate with either kyphosis (p = 0.815) or scoliotic deformity (p = 0.929).

Surgical Details

In the CIM group, 7/18 (39%) underwent posterior fossa decompression (PFD) with C1 laminectomy at our institution; nine patients (9/18, 50%) underwent scoliosis surgery. Five patients were followed >6 months after Chiari decompression and prior to scoliosis surgery (mean followup = 22 months), with 3/5 (60%) demonstrating curve progression (average 26°) and 2/5 (40%) demonstrating stabilization. In the CIM+SM cohort, 15/18 (83%) patients underwent PFD; fourteen (14/18, 78%) underwent scoliosis surgery. Twelve patients were followed >6 months (mean followup= 17 months), with 2/12 (17%) demonstrating curve improvement (average 26°), 5/12 (42%) stabilization, and 5/12 (42%) progression (average 32°). The difference in progression rates between CIM+SM and CIM (42% vs. 60%) was not significant (p=0.49).

DISCUSSION

The prevalence of scoliosis in the overall population approaches 4%. [17] In patients with CIM, the risk of scoliosis is more than fourfold. [8] Although the etiology of spinal deformity in this population is unclear, the close relationship between spinal deformity, CIM, and syringomyelia is well recognized. Huebert and MacKinnon postulated that in patients with syringomyelia, spinal deformity arises from asymmetric damage of the motor neurons innervating the paraxial musculature. [6] Experimentally-induced syringomyelia in an animal model demonstrated damage to the anterior and posterior horn cells, implicating damage to the motor neurons innervating the paraxial musculature. [18] More recently, a meta-analysis by Hwang et al. found a 50% rate of improvement in scoliotic curves in patients with surgical reduction of syringomyelia. [18, 19]

However, there are reports of spinal deformity in CIM patients without syringomyelia. In a large observational study, Strahle et al. found that nearly 5% of patients had a diagnosis of spinal deformity without evidence of syringomyelia. [8] Some authors have suggested that the presence of spinal deformity in the absence of syringomyelia may argue against the theoretical mechanism of scoliosis development, or simply represent an alternate or independent pathogenesis. In 2003, Brockmeyer et al. [9] suggested that syringomyelia may not be critical for the development of scoliosis. Tubbs et al. [10] recognized that CIM without syrinx may represent a presyrinx state, or that brainstem herniation, particularly asymmetric herniation, may contribute to paraspinal weakness and loss of spinal alignment. Thus, the relationship of CIM and spinal deformity in the absence of SM is unclear, and it remains possible that the deformity observed in CIM reflects the prevalence in the population as a whole. [20]

Investigating the differences between spinal deformity presentation in CIM patients with and without syringomyelia may help shed light on the pathogenesis of deformity in this population. There is a limited amount of published literature on the rare combination of CIM alone and spinal deformity. In a review of our own institution, we found eighteen CIM patients and compared them to a matched cohort of patients with CIM+SM-associated scoliosis. We found that CIM patients demonstrated some atypical radiographic features of scoliosis, although not to the extent present with CIM+SM. Surprisingly, we did not appreciate major differences between the two groups in terms of the degree of tonsillar

descent or commonly used craniovertebral junction parameters, such as clival canal angle and pB-C2.

In 2006, Tubbs et al. [10] reported a case of a 16 year-old female with Chiari I Malformation and mild levoscoliosis without syringomyelia, identifying 24 patients in past literature. In a review of the literature, we identified 134 patients (6%) with CIM-associated scoliosis across 2,131 Chiari I Malformation patients reported in the context of large observational cohort studies. Based on available information, we found that patients with CIM and scoliosis were predominantly female (73%; 37/51), presenting with moderate, right-facing scoliotic curves (67%; 4/6), and fewer neurological abnormalities on examination (8%; 1/13). As the current study includes gender as a matching criterion, this precludes the direct comparison of gender as a contributing risk factor for spinal deformity in the CIM cohort; however, given the predominance of females in the CIM cohort (78%) this may indicate a distinct role of gender in this population. Likewise, the lack of neurological abnormalities in CIM patients is consistent with our findings, suggesting that perhaps CIM+SM patients are more likely to present with neurological deficits, presumably due to the underlying syringomyelia.

In comparison with adolescent idiopathic scoliosis (AIS), where apex right curves are typical, CIM+SM patients demonstrate apex left curves in as many as 42.5% of patients. [4] One potential explanation for the larger number of left-facing curves is that symmetric tonsillar herniation or asymmetric syringomyelia may predispose to a more equal distribution of curve morphologies. Recently, Zhu et al. [21] reported an 85% correlation of tonsillar asymmetry and syrinx asymmetry with curve convexity, suggesting that this atypical feature of the scoliosis may be closely associated with the underlying neurological pathology. The fact that in the current study, both Chiari groups demonstrate a high frequency of apex left curves suggests that the pathogenesis may be distinct from AIS. Further, it is possible that our observation of more left-facing deformities in CIM+SM may simply reflect random variation within a small sample.

Hyperkyphosis in a scoliosis patient may be suggestive of an underlying neurological etiology. [22] Qiu et al. [4] described a 28% prevalence of hyperkyphosis in 87 CIM+SM patients. Recently Zhu et al. [15] reported a 21% prevalence of hyperkyphosis among Chiari Malformation patients with left-facing curves, compared to 7% prevalence in right-facing curves. While 13 patients in the latter study did not demonstrate syrinx, the authors did not differentiate on the basis of syrinx presence.

In 2007, Batzdorf described the frequency of kyphosis following suboccipital decompression and laminectomy in Chiari patients. [23] In their study, 4% of patients initially presented with kyphosis, and 14% demonstrated thoracic or cervical kyphosis following re-operation. [23] The authors attributed this to a “double-hit” phenomenon, in which the already jeopardized paraspinal muscle nerve supply was compromised by the expanding syrinx cavity, and exacerbated by the additional soft-tissue damage from the laminectomy procedure. Batzdorf also noted that gravity may play a significant role in the development of kyphosis in the presence of existing spinal instability, particularly in the context of high thoracic syrinxes. [23] Whether sagittal deformity in CIM without

syringomyelia reflects similar processes contributing to the scoliosis, or secondarily to another process such as brainstem compression remains unclear. However, given the atypical presentation of the CIM curves it is possible that these do not simply reflect the basal level of scoliosis, but rather represent deformity of neurological origin.

The role of surgical decompression in the treatment of scoliosis in CIM patients without syrinx is undefined. Most authors would argue for surgical decompression in the presence of syrinx; however the etiology, and therefore causative factor, of deformity in patients without syrinx is not known. Studies have demonstrated the benefit of decompression for curve stabilization when syrinx in present, [18, 19] yet in many instances the deformity continues to progress. In our study 83% of CIM+SM patients underwent PFD compared to only 39% of CIM; interestingly, 42% of CIM+SM patients demonstrated curve progression compared to 60% of CIM patients however this was not significant. In general, at our institution, those patients with small syrinx or those with small curve are not thought to benefit from PFD if asymptomatic; they undergo closer follow-up, with close conversation between orthopedic and neurosurgical teams. Moving into the future, it will be critical to approach management of this rare and interesting disease via a prospective, multicenter approach.

Major limitations of the current study are the retrospective design and small sample size. With a limited cohort size, we are likely underpowered to detect significant differences between groups. In order to address this issue, we chose to compare the cohort of patients identified with a matched cohort chosen from an existing database of patients with Chiari Malformation and syringomyelia. However, due to the rarity of scoliosis in the context of Chiari Malformation without syringomyelia, we believe the sample size cannot be adequately increased without multi-institutional cooperation. Future studies are needed to evaluate the pathogenesis of spinal deformity, as well as the impact of Chiari decompression on curve progression between the two related manifestations of CIM.

Despite its limitations, the current study is the largest dedicated series of CIM associated scoliosis without syringomyelia in the literature. In comparison to the traditionally syringomyelia-associated scoliosis in Chiari malformation, these patients demonstrated similar extent of hyperkyphosis, with less degree of scoliotic curve progression, and fewer neurological signs. This suggests that both groups may develop neurogenic scoliosis related to the underlying Chiari malformation, without discernable differences at the craniovertebral junction.

CONCLUSION

In the largest cohort of this patient population to be reported to date, we report a 3.3% incidence of scoliosis in CIM patients without syringomyelia. Despite similar degree of tonsillar herniation, patients with Chiari I Malformation and scoliosis in the presence of syringomyelia (CIM+SM) were more likely to demonstrate atypical features than patients with CIM-associated scoliosis in the absence of syringomyelia (CIM). We also observed that a higher rate of patients in the CIM+SM group ultimately underwent a surgical intervention - either posterior fossa decompression with C1 laminectomy, scoliosis surgery, or both - as compared to the CIM group.

Acknowledgments

We would like to acknowledge the generosity of Sam and Betsy Reeves and their support of the Park-Reeves Syringomyelia Research Consortium as well as Mateo Dalla Fontana and the O'Keefe family. We would also like to thank Mike Lehmkuhl for providing assistance with project coordination.

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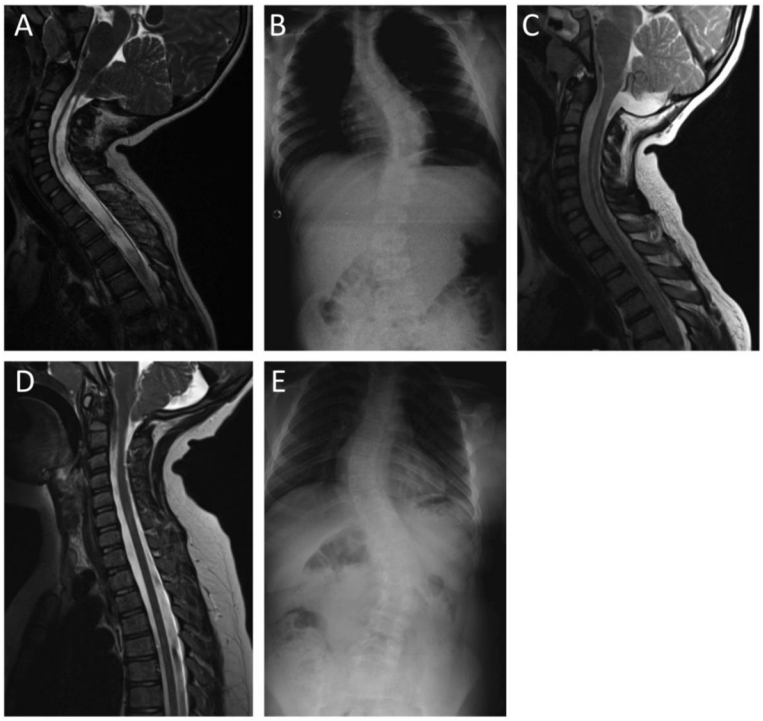

Figure 1.

Magnetic Resonance (MR) imaging and radiographs of two patients with Chiari I Malformation with (A) and without syringomyelia (D). Sagittal T2-weighted MR image of 7-year-old male with 12mm of tonsillar herniation and a large, 12mm wide syrinx (A). Anteroposterior standing radiograph of patient with syringomyelia, demonstrating a 50-degree left-facing thoracolumbar deformity, with curve apex at T10 level (B). Postoperative changes of 2 years after decompression, with interval narrowing of the syrinx extending from C2 to T9 (C). Patient proceeded to fusion (not pictured). Sagittal T2-weigthed MR image of 7-year-old male with 13 mm of tonsillar herniation without evidence of syringomyelia (D). Anteroposterior standing radiograph of patient without syringomyelia, demonstrating a 32-degree right-facing thoracic deformity, with curve apex at the T9 level (E). Patient also proceeded to fusion (not pictured).

Table 1

Demographic and Clinical Characteristics of Chiari I Malformation Patients with Scoliosis

	Syrinx (N = 18)	No Syrinx (N = 18)	P-Value*
Age	11.5 ± 4.4	11.6 ± 4.7	0.94
No. Males (%)	4 (22%)	4 (22%)	1
BMI	21.7 ± 8.1	23.4 ± 5.2	0.55
Presenting Symptom			
Scoliosis (%)	15 (83%)	11 (61%)	0.136
Headache (%)	2 (11%)	5 (27%)	0.206
Paresthesia (%)	1(5%)	0 (0%)	0.311
Neurological Exam			
Neurological Exam Finding (%)	6 (33%)	0	0.007
Sensory Deficit (%)	3 (17%)	0	
Abnormal Reflexes (%)	2 (11%)	0	
Motor Deficit (%)	1 (6%)	0	

BMI indicates body mass index

*Fisher's exact test used for categorical variables, independent-sample t test for continuous variables

Table 2

MRI Characteristics of Chiari I Malformation patients with Scoliosis

	Syrinx (N=18)	No Syrinx (N=18)	P-Value*
Tonsillar Herniation (mm)	9.1 ± 3.0	9.9 ± 4.1	0.570
Grade I	8 (44%)	11 (61%)	0.195
Grade II	8 (44%)	3 (17%)	
Grade III	2 (11%)	3 (17%)	
Tonsillar Symmetry**			
Symmetric	6 (46%)	6 (43%)	0.690
Asymmetric: R dominant	2 (15%)	4 (29%)	
Asymmetric: L dominant	5 (38%)	4 (29%)	
Odontoid pb-C2 (mm)	4.2 ± 1.6	3.9 ± 1.9	0.624
CXA	150° ± 13.8°	151° ± 12.0°	0.741
Syrinx Diameter (mm)	9.0 ± 2.7	--	
Syrinx Length (levels)	13 ± 5	--	

CXA indicates clival canal angle

* Fisher's exact test used for categorical variables, independent-samples t test for continuous variables

** Not all patients had available coronal imaging for determination of tonsillar dominance. Sum does not equal

Table 3

Curve Characteristics

	Syrinx (N=18)	No Syrinx (N=18)	P-Value*
Coronal			
Cobb Angle	46.1° ± 15.7°	32.1° ± 22.4°	0.009
No. Left Facing Curves	9 (50%)	2 (11%)	0.011
No. Thoracolumbar Curves	9 (50%)	5 (28%)	0.171
Coronal Balance (mm)	13.2	12.3	0.95
Sagittal			
Thoracic Kyphosis	49.6° ± 20.2°	43.7° ± 24.9°	0.850
Pelvic Incidence	42.1° ± 15.1°	44.4° ± 13.4°	0.860
Sacral Slope	37.1° ± 9.8°	34.3° ± 6.9°	0.472
Pelvic Tilt	8.3° ± 9.0°	10.1° ± 10.1°	0.678
SVA (mm)	−21.5 ± 15.0	−12.6 ± 39.6	0.433

SVA indicates sagittal vertical alignment