Chiari I Malformations and Scoliosis: The Importance of Syringomyelia

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Glossary

CM-I-Chiari I Malformation

SM-Syringomyelia

CM-I + SM-Chiari I Malformation with Concomitant Syringomyelia

CM-I-Only-Chiari I Malformation without Concomitant Syringomyelia

PFD-Posterior Fossa Decompression

EOS-Early-onset-Scoliosis

AIS-Adolescent-onset-Scoliosis

Abstract

Purpose- The association between Chiari I Malformation (CM-I) and scoliosis is well recognized, but it is unclear if surgical decompression of Chiari malformations alters scoliosis progression long-term in patients with associated syringomyelia (SM). The purpose of this study is to assess the effect of neurosurgical Chiari decompression on the natural history of scoliosis in CM-I patients with SM and CM-I patients without SM.

Methods- Retrospectively, patients diagnosed with scoliosis (Cobb>10°), CM I, and SM over a 15-year period were identified. Patients with congenital, syndromic or concomitant causes of neuromuscular curves were excluded. Clinical and major curve characteristics were recorded at time of scoliosis diagnosis, before decompression and at last follow-up.

Results
CM-I+ SM-Seventy-eight (25M, 53F) patients with a mean age of 9.3 years (1-19) at scoliosis diagnosis were included. Median follow-up was 5 years (range, 2-17). Forty-five patients were <10 years of age and 33 patients were ≥ 10 years of age at scoliosis diagnosis and neurosurgical decompression. Thirty of 78 (41%) patients presented with atypical curves as defined by criteria put forth by Spiegel et al. 67 (86%) patients underwent decompression, of which 26 (39%) improved, 18 (27%) stabilized and 23 (34%) progressed with 13 of the 23(19%) requiring deformity correction surgery. The average scaled SM size decreased from 91.3±67.21 at presentation to 23.6±22.20 scaled units at follow-up (P<0.001). Multivariate analysis determined a one-unit reduction in syrinx scale increased the odds of curve improvement by 1% (OR=1.01; 95% CI = 1.00-1.02; p=0.01) and decreased the odds of fusion by 3% (OR=1.03; 95% CI=1.00-1.05; p=0.03). For each additional unit of syrinx scale at presentation, the odds of fusion increased by 3% (OR=1.03; 95% CI=1.00-1.05; p=0.03).

CM-I Only-Fifty-one (17M, 34F) patients with a mean age of 10 years (range,1-18) at scoliosis diagnosis were included. Median follow-up was 3.6 years (IQR,2.0 to 6.6). More than half of the cohort (29/51, 57%) was treated with bracing **(Table 5)**. Curve magnitude progressed an average of 4.5 degrees (95% CI = 0.5 to 8.4 degrees) for the cohort from baseline to most-recent follow-up. Multivariable analysis determined that there was no association between change in curve magnitude and patient age, initial curve, chiari size, or curve shape (all p>0.05).

Conclusions- Improvement in SM scale size at follow-up is associated with spinal deformity improvement following CM-I decompression.

Significance-SM improvement following decompression is associated with spinal deformity improvement at follow-up.

Level of Evidence: 3

Introduction

While the majority of patients with scoliosis has idiopathic scoliosis without an underlying cause, intraspinal pathology as the cause of scoliosis must be ruled out in certain patients. Although intraspinal pathology is commonly found in association with scoliosis, little is known about the management and evolution of scoliosis in this patient population (1-5),(6). Chairi-1 malformations (CM-I) are the most common intraspinal anomaly found in patients under 18 (5). Most CM-I are found incidentally in otherwise healthy individuals (5). Scoliosis is present in up to 20% of patients with CM-I and up to 88% of patients who have CM-I and concomitant syringomyelia (SM) (7, 8).

SM is thought to mediate scoliosis development in CM-I patients with concomitant SM (9-14). CM-I is the most common cause of SM with the concomitance rate reported to be 50% to 70%.(15, 16) Two proposed mechanisms for SM formation in CM-I patients exist, including the water-hammer theory and the one-way valve theory. The water-hammer theory proposes that arterial pulses from the choroid plexus transmitting CSF down through an abnormal fourth ventricle mediate SM development.(9) The one-way valve theory postulates unequal pressure generated with the Valsalva maneuver causes increased pressure in the spinal cord and SM development. (9) MRI studies have shown many CM-I patients have aberrant communication between the ventricles but neither theory has been proven.(9, 11) Recent biochemical and histologic data suggest that enlarging SM causes asymmetrical injury to the anterior horn of the spinal cord, (10, 13, 14) leading to denervation and weakness of the paraspinal muscles, causing scoliosis development.(10, 13, 14) However, this does not explain scoliosis development in CM-I patients without SM (17, 18), leading many to suggest that asymmetric compression of the cervicomedullary junction by the cerebellar tonsils may cause direct spinal cord injury and scoliosis in the absence of SM.(17-21) Recent studies have found a high rate of atypical curves in CM-I patients without SM, suggesting a non-idiopathic etiology.(17, 18, 21) Research on the management and natural history of scoliosis in this population is scant. All prior series examining CM-I and scoliosis are small and the majority of patients have concomitant SM.[(14, 22-28)] While research on CM-I patients with SM suggest Posterior Fossa Decompression (PFD) can lead to curve improvement,[(14, 22-28)] little is known about the natural history of scoliosis in CM-I patients without concomitant SM.(14, 17, 18, 21, 29, 30) Even with this paucity of research, most neurosurgeons recommend PFD as an intervention for curve management in CM-I patients without SM.

While the relationship between CM-I, SM and scoliosis has been well documented, the potential benefits of neurosurgical decompression are not well known.[(14, 22-28)] While several series have shown that patients may experience spinal deformity improvement following decompression, many patients in these series also experienced deformity progression.(22, 24, 26, 31, 32) Many of these series were small, and contain patients with and without SM. These studies also had limited follow-up, leading some to postulate that the benefits of decompression on curve progression may be temporary.[(14, 22-28)] As such, the long-term benefit of decompression in CM-I+SM patients remains unclear. To the authors' knowledge, this is the first scoliosis study comparing CM-I+SM patients treated with and without neurosurgical decompression. The authors also believe this to be the largest long-term follow-up study examining CM-I patients with concomitant SM. As such, we seek to describe the presentation and natural history of scoliosis progression in CM-I patients with concomitant SM undergoing decompression. We also seek to characterize the factors associated with deformity progression and fusion at last follow-up in this population.

Through a separate study, we also sought to examine the natural history of scoliosis progression in CM-I patients without concomitant SM (CM-I only) undergoing decompression and treated non-operatively. To the authors knowledge, this serves as the first study describing the natural history of scoliosis and the benefit of decompression on curve decompression in this population.

Materials and Methods

Study: CM-I patients with concomitant SM

Institution Review board approval was obtained prior to beginning this study. Electronic medical records were examined to identify all patients diagnosed with scoliosis, CM-I and SM from 2000 to 2015. Patients with concomitant causes of neuromuscular, congenital and syndromic curves were excluded. Patients who underwent deformity correction surgery before decompression were also excluded. Scoliosis was defined as a coronal deformity greater than or equal to 10° using the Cobb method. Thirty-nine patients were found to have less than 2 years of radiographic follow-up and were excluded. Overall, 78 patients were identified that met these inclusion criteria. Preoperative and postoperative medical records were reviewed for all patients. The presenting complaints, demographic data, neurological exam findings, secondary treatments (bracing, casting) and deformity correction details were collected for all patients. All patients treated with decompression underwent primary sub-occipital decompression with C1 laminectomy. Six patients underwent fourth ventricle to subarachnoid space shunting at the time of decompression.

Scoliosis Evaluation

Direction, magnitude, and location of major and minor curves were recorded at presentation, before decompression, and at last follow-up. Early onset scoliosis (EOS) was defined as scoliosis diagnosed and treated with PFD <10 years of age. Adolescent-onset scoliosis (AOS) was defined as scoliosis diagnosed and treated with PFD ≥10 years of age. Single major and double major curves were documented. The classification system put forth by Spiegel et al. was used to define atypical curves.(33) Factors associated with deformity progression were examined trichotomously for all analyses. Trichotomous grouping was follows: Cobb improvement by greater than 5°, stabilization (a change in Cobb of 5° or less), and deformity progression of more than 5°. Grouping was based on change in major curve between presentation and last follow-up.

Syringomyelia Evaluation

Syringomyelia diagnosis was confirmed with magnetic resonance imaging (MRI) imaging for all patients. Syringomyelia was defined as a central cyst on T2-weighted MRI. Patients were all followed with serial imaging. SM length was defined as the number of vertebral segments spanned.(12) SM width was defined as the maximal syrinx width in millimeters on sagittal MRI. Scaled SM area was defined as the syrinx width in millimeters multiplied by its length in vertebral angles. SM dimensioned were measured before decompression and at follow-up for all patients, regardless of decompression.

Statistical Analysis

Patient and condition characteristics were summarized for all patients. Continuous characteristics were summarized by mean and standard deviation (SD) or by median and interquartile range (IQR; 25th to 75th percentile), as appropriate and categorical characteristics were summarized by frequency and percent.

Patient and condition characteristics, including patient age, sex, initial major curve size, curve shape, kyphosis, syrinx size, and whether the patient was treated with bracing were analyzed for potential effect on change in major curve and change in syrinx size using multinomial logistic regression. Outcomes were trichotomized as described above. Sub-group analysis was conducted for subjects who underwent decompression. Multinomial logistic regression was used to assess the effects of age, initial curve, curve shape and bracing on curve outcome after decompression. Odds ratios along with 95% confidence intervals were estimated for significant effects. All tests were two-sided and p-values less than 0.05 were considered significant

Study: CM-I Patients without Concomitant SM

Institution Review board approval was obtained prior to beginning this study. Electronic medical records were examined to identify all patients diagnosed with scoliosis and CM-I from 2000 to 2015. Patients with SM were excluded. Patients with concomitant causes of neuromuscular, congenital and syndromic curves were also excluded. Patients who underwent deformity correction surgery before decompression were also excluded. The Cobb method was used to measure major and minor curves characteristics at scoliosis diagnosis, before decompression and at last follow-up. Scoliosis was defined as a coronal deformity greater than or equal to 10° in the coronal plain. Forty-six patients were found to have less than 2 years of radiographic follow-up and were excluded. Overall, 51 patients were identified that met these inclusion criteria.

Preoperative medical record and postoperative were reviewed for all patients. The chief complaints, demographic data, neurological exam findings, and secondary treatments (bracing) and deformity correction details were collected for all patients. All patients treated with decompression underwent primary sub-occipital decompression with C1 laminectomy and no patients underwent fourth ventricle to subarachnoid space shunting.

Scoliosis Evaluation

The Cobb method was used to measure major and minor curves at presentation, before decompression, and at last follow-up. Direction and location of major and minor curves were recorded. Patients with single major and double major curves were noted. The classification system put forth my Spiegel et al. was used to define atypical curves.(33) Deformity progression were examined in a continuous manner for regression analysis and by group for all other analysis. Deformity improvement was defined as Cobb angle improvement greater than 5°, stabilization was defined as a change in Cobb of 5° or less, and deformity progression was defined as Cobb progression of more than 5°. Patients were grouped based on curves at last follow-up. Early onset scoliosis (EOS) was defined as scoliosis diagnosed and decompression <10 years of age. Adolescent idiopathic Scoliosis(AIS) was defined as scoliosis diagnosed and decompression ≥10 years of age.

Statistical Analysis

Patient and condition characteristics were summarized for all patients. Continuous characteristics were summarized by mean and standard deviation (SD) or by median and interquartile range (IQR; 25th to 75th percentile), as appropriate and categorical characteristics were summarized by frequency and percent.

Patient and condition characteristics, including patient age, sex, initial major curve size, curve shape, kyphosis, and whether the patient was treated with bracing, were analyzed for potential effect on change in major curve and change in syrinx size using multinomial logistic regression. Outcomes were trichotomized as described above. Subgroup analysis was conducted for subjects who underwent decompression. Multinomial logistic regression was used to assess the effects of age, initial curve, curve shape and bracing on curve outcome after decompression. Odds ratios along with 95% confidence intervals were estimated for significant effects. All tests were two-sided and p-values less than 0.05 were considered significant.

Results

Study: CM-I patients with Concomitant SM

Seventy-eight patients (32% male) met the inclusion criteria for this study. The average age (±SD) at scoliosis diagnosis was 9.3±3.7 (range, 1-19) years **(Table 1).** Median follow-up was 5 (IQR, 3-8) (range, 2-17) years. Thirty-two (42%) patients had EOS and 44 (58%) were AOS patients.

Seventy-six presented with a chief complaint of scoliosis and were found to have CM-I and SM through subsequent evaluation. Two patients (2.5%) presented to neurology: 1 with clonus and 1 with chronic headaches and were subsequently found to have CM-I, SM and scoliosis. Through subsequent neurological work-up, 10 patients who presented with scoliosis as their chief complaint were found to have a history of chronic headaches, 9 were found to have back pain and 3 were found to have upper extremity numbness through there neurological exam. Overall, 29% of patients were found to have an abnormal neurologic exam.

Major and Atypical Curve Patterns

Major curve patterns at presentation included: 17 (22%) right thoracic curves, 23 (30%) right thoracolumbar curves, 1 (1%) right lumbar curve, 12 (15%) left thoracic curves, 22 (28%) left thoracolumbar curves, and 3 (4%) left lumbar curves. Sixty (77%) patients had single major curves and 18 (24%) had double major curves. Thirty of 78 (38%) patients presented with atypical curves as defined by criteria put forth by Spiegel et al.(33) Curves with a left thoracic component were identified in 21 patients (27%), including single thoracic (13, 15%) and left thoracic/right thoracolumbar (8, 9%). Patterns with an upper thoracic component (double thoracic and triple curves) were observed in 11 (14%) patients.

Syringomyelia and Curve progression

Overall, 67 (86%) patients underwent neurosurgical decompression at a mean age 9.3±3.52 years. Of the 67 patients who underwent decompression, 27 (40%) patients improved, 18(27%) patients stabilized and 22 (33%) patients progressed with 13 (19%) requiring deformity correction surgery **(Table 4)**. Multinomial regression determined that there was no significant association between patient age, major curve location, or bracing treatment with curve progression or fusion **(Table 4)**.

Following decompression, the average SM width was reduced from 7.3±3.72 *mm* (mean ± SD) at presentation to 2.7±1.62 *mm* at follow-up (p<0.001). The average SM length decreased from 11.0±4.68 vertebral levels to 7.7±4.46 levels (p<0.001). The average scaled SM size decreased from 91.3±4.68 at presentation to 23.5±4.42 scaled units at follow-up (p<0.001) **(Table 2).** Multivariate analysis determined that that for each 1-unit decrease in syrinx scale size at follow-up, the odds of curve improvement increased by 1% (OR=1.01; 95% CI = 1.00-1.02; p=0.01) **(Table 3).** Five of sixty-seven (7%) patients had complete SM resolution after neurosurgical decompression. Five patients did not have an appreciable decrease in syrinx size after primary decompression and required a secondary decompression. Two patients were treated with decompression alone and three patients were treated with decompression and fourth ventricle to subarachnoid shunting. All patients had SM improvement following final decompression. Overall, patients experiencing an 83% reduction in scaled syrinx size following decompression **(Table 2)**.

Overall, twenty-three (29%) patients underwent fusion at follow-up. Multivariate analysis demonstrated that the odds of fusion increased by 3% (OR=1.03; 95% CI=1.00-1.05; p=0.03) for each additional unit of syrinx scale at presentation. Furthermore, the odds of fusion decreased by 3% (OR=1.03; 95% CI = 1.0-1.06); p=0.02) for each l-unit decrease in scale size **(Table 3).**

Patients < 10 at scoliosis diagnosis were compared to patients ≥ 10 at the time of scoliosis diagnosis. Patients <10 where found to have SM that spanned 3 more vertebral levels (p=0.04) at presentation. However, no difference was found in SM improvement following decompression between these groups (p>0.05).

Additional Characteristics and Curve Progression

Multivariable analysis determined that for patients with an initial curve ≥ 40 °, the odds of curve improvement at follow-up decreased by 74% (OR=0.26; 95% CI = 0.08-0.98; p=0.03); with 22/50 (44%) of decompressed patients with curve <40° experiencing curve progression compared to 5/17 (29%) of patients with curves ≥40°. Multivariable analysis determined that for patients with an initial curves ≥40°, the odds of fusion increased 4 times (OR=4.3; 95% CI = 1.31 to 14.11; p=0.02), with 22% (11/50) of decompressed patients with curve <40° requiring deformity correction surgery compared to 53% (9/17) of patients with curves ≥40 °.

Multivariable analysis of decompressed patients found patients with single major curves were more like to improve in curve magnitude compared to double major curves (OR=7.8; 95% CI = 1.1-58.4; p=0.04). Twenty-three of 52(44%) of patients with single major curves experienced improvement compared to 2/14 (14%) of patients with double major curves. Seven of 14(50%) of patients with double major curves went on to require deformity correction surgery compared to 12/52 (23%) of patients with single major curves (Table 4). Multivariable analysis of decompressed patients also found that subjects with a larger CM-I at presentation were more likely to experience stabilization in curve magnitude (OR=1.24; 95% CI=1.02-1.5; p=0.03). No association was detected between Chiari size at presentation and the likelihood of improved curve magnitude (p=0.25).

Patients <10 at the time of scoliosis diagnosis and decompression were more likely to experience spinal deformity improvement following decompression. Nineteen of 40(48%) patients diagnosed at <10 years of age showed improvement at follow-up compared to 7/27 (26%) of patients diagnosed at ≥ 10 years of age, but these differences were not significant (p=0.22) (Table 4).

Eleven patients in this study (14%) did not undergo decompression and were treated with non-operative management. Of patients who did not undergo decompression, 1 (9%) improved, 6 (55%) stabilized and 4 (36%) progressed. Of the 67 patients who underwent decompression, 26 (39%) patients improved, 18 (27%) patients stabilized and 23 (34%) patients progressed.

Study: CM-I Patients without Concomitant SM

Fifty-one patients met the inclusion criteria for this study. Seventeen males (33%) and thirty-four females (67%) were included. The average age (±SD) at scoliosis diagnosis was 10.2±4.29 (range 1-18) years. Average follow-up was 3.6 (IQR 2-6.6) years. Twenty-two (43%) patients were EOS. Twenty-eight (55%) patients were AOS. Overall, 11(22%) patients underwent PFD. The average Cobb angle at presentation was 30.7°±16.24°. Average Cobb angle at follow-up was 35.1°±19.60°. Fifteen (29%) patients went on to require deformity correction surgery.

Thirty-nine patients presented with a chief complaint of scoliosis and were found to have CM-I through subsequent evaluation. Twelve (24%) presented to neurology, including 8 with headaches, 2 with back pain, 1 with seizures, 1 for toe-walking and one with nausea; these patients were subsequently found to have CM-I and scoliosis. Through subsequent neurological work-up, 4 patients who presented with scoliosis as their chief complaint were found to have a history of chronic headaches and 2 were found to have back pain through their neurological exam. Overall, 35% of patients were found to have an abnormal neurologic exam.

Major and Atypical Curve Patterns

Major curve and minor curve patterns were recorded for all patients. Major curve patterns were as follows: eleven (22%) presented with right thoracic curves, fifteen (29%) with right thoracolumbar curves, none (0%) with a right lumbar curve, six (12%) with left thoracic curves, eight (16%) with left thoracolumbar curves, and eleven (22%) with left lumbar curves. Thirty-one (61%) of patients had single major curves and 20(39%) of patients had double major curves. (**Table 5**).

Atypical curves were defined using the criteria put forth by Spiegel et al. Eighteen of 51(35%) patients fell into this category. Curves with a left thoracic component were identified in ten patients (20%), including single thoracic (4, 8%) and left thoracic/right thoracolumbar (4; 8%) curves. Patterns with an upper thoracic component (6 double thoracic and 2 triple curves) were observed in 8 (16%) patients.

Change in Curve

Curve magnitude progressed an average of 4.5 degrees (95% CI=0.5 to 8.4 degrees) from baseline to most-recent follow-up in this cohort **(Table 6)**. Only 16 patients (31%; 95% CI=19.5 to 46%) showed improvement in curve magnitude at follow-up. Multivariable analysis determined that there was no association between change in curve magnitude and patient age, initial curve, Chiari size, curve shape, or bracing treatment (all $p>0.05$).

Fusion

Nearly one-third of the cohort (15/51, 29%) underwent on to fusion at follow-up **(Table 6)**. Multivariable analysis determined that there was no association between the likelihood of fusion and patient age, initial curve, Chiari size, and curve shape (all $p>0.05$).

Decompression and Bracing

Eleven (22%; 95% CI=12 to 36%) patients were treated with decompression. Subjects with a single major curve were more likely to be treated with decompression compared to those with a double major curve. There were no other associations between patient or curve characteristics and decompression intervention **(Table 7)**. There were also no differences detected in outcomes given decompression treatment (all p>0.05; **Table 7**). Patients treated with bracing were more likely to improve and less likely to experience curve progress, independent of decompression status **(Table 8).**

Discussion

CM-I is the most common intraspinal anomaly found in children.(5) CM-I is defined as caudal ectopia of one or both of the cerebellar tonsil 5mm or more below the foramen magnum. (34) A recent population based study of 14,118 found that 3.8% of people under 18 have CM-I malformations.(5) CM-I is the most common condition leading to SM development with the concomitance rate between SM and CM-I reported to be 50% to 76%.(15, 16) Recent studies have suggested that the direct impaction of the cerebellar tonsils can cause scoliosis in CM-I patients without SM.(17, 19) The leading theory of scoliosis development in CM-I patients suggests that the expanding SM causes asymmetric injury to the anterior horn of the spinal cord.(10, 13, 14) This injury subsequently causes asymmetric denervation and weakness to the spinal muscles, causing coronal deformity to develop. Recent biochemical and histologic studies support this theory and have found that decompression surgery improves innervation of spinal muscles,(13) suggesting a mechanism for the spinal deformity improvement and stabilization following decompression.

Decompression in CM-I patients with concomitant Syringomyelia

Most neurosurgeons recommend decompression of CM-I in the setting of SM with the goal of reversing or preventing the neurological sequelae associated with SM.(29, 35) The benefit of decompression with regards to to spinal deformity progression is less clear.(7) Prior studies examining decompression in CM-I patients have found rates of scoliosis improvement of 18% to 38%.(22, 25, 26, 31, 36) As the length of follow-up in these studies is short, many have questioned the long-term benefit of PFD in CM-I +SM patients. [14, 22-28] In the current CM-I+SM study, 9% of patients who did not undergo decompression experienced spontaneous spinal deformity improvement, compared to 40% of patients who underwent decompression. Spontaneous Cobb improvement is uncommon in the idiopathic population and these results suggest a benefit of decompression in CM+SM patients. (7)

Curve progression following decompression in CM-I +SM patients is common, making it difficult to assess patient prognosis in this population. Prior studies have found rates of deformity progression from 36% to 89%.(22, 25, 26, 31, 36, 37) In the current study 34% of patients experienced spinal deformity progression with 13 (19%) requiring deformity correction surgery at long-term follow-up.

Importance of Syringomyelia improvement Post-PFD in CM-I patients with concomitant Syringomyelia

While the relationship between SM and denervation of paraspinal muscles has been studied in vitro (10, 13, 14), little is known about the relationship between SM characteristics and scoliosis prognosis. SM improvement following decompression has been observed in 65% to 93% of patients. (31, 32, 36, 38, 39) While some studies have observed curve improvement in patients with SM, many studies have reported curve progression in the setting of SM improvement. (22, 26, 31, 32, 36, 39-41) Prior studies have not found an association between

SM characteristics and initial curve magnitude or the risk of progression.(22, 26, 36, 39-41) In this study, syrinx improvement was associated with spinal deformity improvement at follow-up. As a decrease in syrinx width and length were both associated with curve improvement, a scaled measurement for SM area was developed. This study found patients with larger SM at presentation were more likely to require deformity correction at follow-up. A one-unit reduction in scale SM area size at follow-up was associated with a 1% increase in the odds of curve improvement and a 3% decrease in the odds of fusion, with the average patient experiencing a 70-unit reduction in SM size at follow-up. In this study, 7.5% of patients had no SM improvement following primary decompression. These patients all underwent secondary decompression and all experienced subsequent SM improvement, with 80% experiencing deformity improvement. Krieger et al. found a similar rate of reoperation, with 8.5% of patients requiring reoperation. Preoperative SM length, SM width, SM area, Cobb angle, CM-I size, and age were not found to influence the likelihood of decompression failure, suggesting serial imaging is needed for all patients to assess SM regression in the post-surgical period.(4)

Prevalence of Atypical Curves In CM-I Patients with concomitant Syringomyelia

Significant work has been done to classify typical and atypical curves in CM-I patients. Spiegal et al. found that curve patterns present in less than 2% of the idiopathic population are common in CM-I patients.(33) In this study, 38% of patients presented with curves defined as atypical by Spiegel et al. (33) This supports the idea that these curves suggest intraspinal pathology.(17, 42) Prior studies have found similar rates of atypical curves ranging from 43.7% in Qiu et al. to 51.2% in Spiegel et al.(12, 33) Most studies have not detailed how curve patterns impact curve progression.(8, 26, 37, 43) In this study, single-major curves were more likely to improve when compared with double major curves. Flynn et al. found that eight of nine double major curves experienced progression following decompression in a study of 15 patients. (25) Zhu found double major curves were present in 47% of progressors compared to 11% of non-progressors.(44) Attenello et al. suggested thoracolumbar curves were more likely to progress.(31) None of these associations were observed in this study. In the current study patients with left thoracolumbar major curves were more likely to improve when compared to other major curve patterns. More than half of the 27 patients who improved at follow-up presented with left thoracolumbar curves. Senguta et al. observed curve improvement was more common in patients with left thoracic curves, with 75% of these patients avoiding deformity correction surgery.(22) No association between fusion or progression and left thoracic curves was found in this study.

Importance of Curve Magnitude in CM-I Patients with Concomitant Syringomyelia

Authors have suggested that curves greater than 30 to 40° are more likely to progress or require fusion. Tubbs et al. found no improvement in patients with curves > 40°.(16) Nagib found scoliosis improvement in 6 of 6 patients with curves <30° and stabilization in 4 of 4 patients with curves ≥ 30°.(45) Ghanem et al. found that 5 of 5 patients who presented with curves ≥ 40° required fusion.(23) In this study, patients with Cobb angles over 40° had a lower odds of improvement and a greater odds of fusion at follow-up. After decompression. 59% of these patients experiencing progression compared to 26% of patients with curves ≤ 40°. Consistent with findings by Brockmeyer et al. and Smith et al., some patients with curves > 40° improved, but these patients should be considered exceptions rather than the rule.(7, 26) In this study, 53% of these patients with curves> 40° went on to require deformity correction surgery compared to 20% of patients who presented with curves ≤ 40°. Although patients with curves > 40° are more likely progress and require deformity correction surgery, decompression should always be carried out before deformity correction surgery to reduce the risk of neurological deficits after surgery.(7)

Importance of Age in CM-I Patients with Concomitant Syringomyelia

Prior studies have suggested that younger patients are more likely to experience spinal deformity improvement. Muhonen et al. reported scoliosis improvement in 3 of 3 curves in patients <10 despite one curve exceeding 40°.(27) Flynn et al. and Brockmeyer et al. found that 70% (7/10), and 91% (10/11) of patients <10 avoided fusion respectively.(25, 26) No association between age curve progression or fusion requirement was found in this study. When curve improvement and stabilization are used to define surgical success, 68% of patients <10 years of age experienced surgical success following decompression compared to 63% of patients ≥10 years of age. While little has been written about the benefit of decompression in patients ≥ 10, these results are promising and suggest that decompression may alter deformity progression in patients ≥10.

Importance of Kyphosis in CM-I Patients with Concomitant Syringomyelia

Flynn et al. observed that half of progressors had >50° of kyphosis compared to 1 of 7 nonprogressers.(25) Zhu et al. found no association with curve progression and kyphosis in a study of 54 CM-I+SM patients who underwent PFD.(44) In this study, we found no association begween kyphosis and risk of progression or fusion.

Importance of Bracing Post-PFD in CM-I Patients with Concomitant Syringomyelia

The benefit of bracing following decompression is heavily debated. The benefit of bracing following PFD remains controversial. Zhu et al. found bracing to be a statistically significant predictor of curve improvement in a study of 54 patients.(44) Sha et al. found 24% (8 of 33) of patients who underwent bracing required deformity correction surgery compared to 43% (13 of 21) of those without bracing.(13) Other studies have suggested that the scoliosis correction is not maintained in this population after bracing is concluded. In this study, no association between bracing and Cobb angle progression was found in patients who underwent decompression.

Establishing a Minimal Syringomyelia Size that Warrants Decompression in CM-I Patients with Concomitant Syringomyelia

As all prior studies examining the value of decompression have consisted exclusively of patients who underwent decompression, it has been difficult to assess the degree to which decompression alters the natural history of scoliosis in this population. Eleven patients in this study (14%) did not undergo decompression and were treated with non-operative management. Ten of these patients were treated non-operatively due to the small size of their SM. One patient was treated non-operatively against medical advice due to parental request. Only 9% of these patients experienced curve improvement at last follow-up, compared to 39% of patients treated with decompression. The non-decompressed group also had a fusion rate of 36% compared to 19% of decompressed patients. While our study finding that large patients with larger SM are more likely to progress to fusion is consistent with the mainstay recommendations by neurosurgery, the progression data in our cohort who did not undergo decompression make it difficult to ascertain which patients should be treated non-operatively.

Scoliosis in CM-I Patients without Concomitant Syringomyelia

While the mechanism of scoliosis in CM-I patients remains unclear, most theories have suggested that scoliosis in CMI patients is mediated by syingomeylia.(9-11, 13, 14) Biochemical and in vitro studies have found that syringomyelia induces the denervation of paraspinal muscles in CMI patients, subsequently causing scoliosis via asymmetrical muscular injury and weakness.(14, 46) However, syringomyelia does not completely explain scoliosis in CMI patients as scoliosis is relatively common in CMI patient without concomitant syringomyelia.(27, 37, 39) The paucity of research on this population of CMI patients makes the management of scoliosis in these patients unclear.(7)

There remains significant conflicting evidence surrounding the influence of tonsillar ectopia on the development of scoliosis. In reviewing the abnormal position of cerebellar tonsils in 203 scoliosis patients, Sun et al. found a significant correlation between tonsillar ectopia and uncommon thoracic curves.(18) In a study of 42 patients with CMI, Tubbs et al. found that the unilateral clinical and physical exam findings corresponded to the asymmetric tonsillar herniation. (47) Additionally Zhu et al. found that the dominant side of the tonsillar ectopia correlated with the convex side of the scoliosis curve in up to 88.5% of CM-I patients. (42) Additionally, Cheng et al. found tonsillar ectopia to be more common in AIS patients compared to age-matched controls (30), leading many to believe that direct compression of the spinal cord by tonsillar ectopia is involved in the pathogenesis of scoliosis in CM-I patients.(17, 30) While, these results imply tonsillar ectopia may play a role in the development of scoliosis, the natural history of scoliosis in CM-I patients without SM has not been examined. Additionally, the benefit of decompression and bracing has not been examined on this population. This represents the first study examining the natural history of scoliosis in CM-I patients without SM. It is also the first study examining the benefit of decompression and bracing in CM-I patients without SM.

Prevalence of Abnormal Neurological Findings at the Time of Presentation in CM-I Patients

without Concomitant Syringomyelia

In the current study, 35% of patient were found to have abnormal neurological findings at the time of presentation. The most common presenting symptom was headaches, which were found in 20% of patients. Prior studies including CM-I patients with concomitant syringomyelia patients also found headaches to be the most common neurologic finding in CM-I patients.(48, 49) While prior studies on CM-I Patients with concomitant syringomyelia found many CM-I patients with motor weakness and upper motor neuron signs on exam, including hyperreflexia and clonus, no patients in this study presented with weakness, hyperreflexia or clonus, suggesting motor weakness and upper motor neuron signs may be a symptom of syringomyelia instead of CM-I**.** The findings by Alzate et al. also support this theory as 0% (0/32) of their CMI presented with motor deficits compared to 24% (8/34) of their CMI + SM patients. (35, 48, 49) In this study, patients with significant tonsillar herniation were more likely to present with neurologic findings when compared to patients with less significant tonsillar ectopia, suggesting tonsillar herniation is the cause of neurologic findings in these patients. Prior studies on CM-I patients have found that most CM-I-only patient experience resolution of neurologic sequelae following decompression. (35)

Prevalence of atypical curves at initial presentation in CM-I Patients without concomitant Syringomyelia

In this study, 18 of 51 (35%) patients presented with atypical curves based on the criteria put forth by Spiegal et al.(33) Zhu et al. found atypical curves in 26 of 26 CM-I-only patients with scoliosis, arguing in favor of a non-idiopathic etiology of scoliosis in these patients.(17) Although the benefit of PFD in this population is unknown, the majority of neurosurgeons report that they perform PFD on CM-I patients without concomitant syringomyelia to prevent curve progression. (29) While recent data has suggested tonsillar ectopia can cause scoliosis independent of SM, to the authors knowledge this is the first study examining curve progression in CM-I-only patients with scoliosis.(13, 14, 17, 30) In this study, no association was found between decompression and curve improvement. Overall, 0%(0/11) patients treated with decompression experienced curve improvement at follow-up compared to 27%(11/40) of patients treated non-operatively. Much lower than the 18% to 38% rate of curve improvement was seen in CM-I Patients with concomitant Syringomyelia.(22, 25, 26, 31, 36) Additionally, there was no association between the degree of tonsillar ectopia and initial curve magnitude or curve progression. Together, these results suggest different principles guide scoliosis development in CM-I+SM patients compared to patients CM-I-only patients. The findings by Strahle et al. of an independent association with SM and Scoliosis without any association between CM-I and scoliosis when controlling for syrinx status in a cohort of 14,118 patients support this theory.[(5)] Some reports have suggested there is an independent relationship between low cerebellar tonsil position and scoliosis,(18-21, 30, 50) but most of these cases are drawn from larger case series that include a majority of patients with concomitant SM. The increased incidence of MRI screening in recent years has shown us that cerebellar tonsil position is distributed in an approximately normal distribution pattern and tonsillar ectopia is not rare, even in healthy individuals.(21, 51) As low-lying cerebellar tonsils and scoliosis are common, it is not surprising that many individuals with both conditions are identified. Consistent with prior studies, this study failed to demonstrate an association between low tonsil position and curve severity.(18, 21)

Bracing in CM-I Patients without concomitant Syringomyelia

Despite decades of research, the role of bracing in preventing curve progression in CMI patients remains controversial.(52) Prior studies in CMI patients with SM have found curve progression to be almost universal in patients braced without PFD.(7, 53) These patients are thought to progress despite bracing because the SM, which represents the primary cause of their curve, has not been addressed. However, studies have found that bracing may be beneficial in CMI patients with SM post-PFD.(27, 44, 52, 53) In contrast with prior studies, CM-I patients without SM treated with bracing in this study were less likely to experience curve progression regardless of PFD. Overall, 33% (10/30) of the patients treated with bracing experienced curve progression at follow-up compared to 57% (12/21) of the patients not treated with bracing. Additionally, 33% (10/30) of patients treated with bracing experienced curve improvement at follow-up compared to 5% (1/21) of patients treated without bracing. As curve progression is almost universal in CM-I patients with concomitant SM treated with bracing without PFD, these results suggest that curve progression in CM-I only patients is governed by different principles than in patients with SM.(7, 53)

Factors Associated with Curve Progression at Follow-up in CM-I Patients without Concomitant Syringomyelia

Similar to the findings by Flynn et al, double major curves were more likely to progress at follow-up compared to single major curves, with 60%(12/20) of double major curves progressing at follow-up compared to 33% (10/31) of patients with single major curves.(25) Zhu et al. found a similar trend in CM-I patients with Scoliosis.(44) While studies conducted in CMI patients with SM have found age, kyphosis, and initial curve magnitude to be predictors of curve progression, no such trends were found in this study.(25, 44)

Conclusion

CM-I Patients with concomitant Syringomyelia

SM improvement following decompression surgery is associated with a greater odds of curve improvement in patients with CM-I+SM. SM improvement following decompression surgery is also associated with a decreased odds of deformity correction at follow-up. Patients with curves >40° and double major curves are less likely to experience deformity correction at follow-up. No association between age and curve improvement was found, suggesting EOS and AIS patients can benefit from decompression. While our study found that patients with larger SM are more likely to undergo deformity correction at follow-up, the high rate of progression in the patients with smaller SM who did not undergo decompression makes it difficult to ascertain which patients should be treated non-operatively.

CM-I Patients without Concomitant Syringomyelia

As decompression surgery carries associated risks, it is important to understand the benefit of this surgery in different populations of CM-I patients.(54) The results of this study suggest that scoliosis in CM-I patients without SM is driven by a different process than scoliosis in patients with concomitant SM. The results of the current study do not suggest decompression is beneficial in altering curve progression in CM-I patients without SM. Additionally, in the current study many patients had favorable outcomes with bracing independent of decompression, suggesting this population can benefit significantly from non-operative management alone.

Limitations

CM-I Patients with concomitant Syringomyelia

Limitations of the study include the retrospective nature of the study, and reliance on electronic medical record documentation with incomplete data (both kyphosis and length of time in brace were not documented for all patients). The group undergoing decompression was small and demonstrated diverse SM characteristics. This study was carried out at a tertiary care center where the same neurosurgeons and orthopedic surgeons operate using the same practice guidelines and the patient population may not be generalizable.

CM-I Patients without concomitant Syringomyelia

This study has limitation inherit to being a retrospective study. The group undergoing decompression was small. Kyphosis data were incomplete, and we were not able to assess this variable for all patients. All patients were treated by the same neurosurgeons and orthopedic surgeons using the same practice guidelines.

Suggestions for Future Work

Collectively, the results of these two studies will likely have significant impact on patient management and CM-I research in the short-term and long term. Our study on CM+SM patients represents the largest and first long-term follow-up study on scoliosis in this population. It is the first study to show that effects of PFD are long-lasting and carry into adulthood. It is the first to show a significant association between SM improvement and curve improvement. It is also the first study to show that a large syrinx at presentation is associated with an increased odds of fusion at follow-up. Additionally, we developed a new measurement modality for SM size through this study that can help predict likelihood of curve improvement and risk of fusion at follow-up.

It has been proposed that in CM-I patients scoliosis is caused by expanding SM.(13-15, 22, 24, 31, 32, 36) The results of this clinical study support the theory that SM size and SM improvement were associated with curve progression and improvement. Additionally, the degree of CM-I ectopia was not associated with curve progression, arguing strongly against the theory of direct compression.(18, 47, 54) Currently, biochemical and histologic date suggest that expanding SM causes injury to anterior horn cells of the spinal cord.(13, 55-57)Data suggest that this damage leads to denervation of skeletal muscle. (13, 55-57) Recent studies examining the expression of embryonic γ-subunit and abnormal spread of acetylcholine receptors at the neuromuscular junction support this theory, as acetylcholine receptors are upregulated in injured skeletal muscle cells.(14) Innervation is essential for the maintenance and survival of skeletal myofibrils. (13, 55-57) In the absence of innervation, myofibrils become prone to apoptosis, as anti-apoptotic protein Bcl-2 is downregulated in these myofibrils and pro-apoptotic protein Bax is upregulated. (13, 55-57) This increase in Bax-to-bcl-2 ratio leads to increased cytochrome C leakage into the cytosol, leading to apoptosis. (13, 55-57) Studies have suggested that after SM decompression, the bax-to-bcl-2 ratio in these cells may improve and lead to reinnervation and improved myofibril fitness.(13) Previously, many have argued against this theory, citing no association with SM improvement and curve improvement. While this current study suggests SM is associated with curve improvement, it also provides a framework for future investigation into SM-induced scoliosis research. Given that there are overwhelming data suggesting that decompression is beneficial for preventing neurologic sequelae, even CM-I+ SM patients with small SM are treated with PFD. This makes comparative studies difficult. As such, it has been difficult to determine which patient subsets require decompression vs. non-operative conservative management. As PFD carries many associated risks, it is important to know which patient populations many benefit from decompression.(54)

With this framework for SM size and improvement, one can now study the association between SM size and biochemical surrogates for muscle injury such as Bax and Bcl-2. From these studies we should be able to determine the size at which SM is likely to cause spinal cord injury and denervation. These studies would also allow us to better understand the relationship between SM width, SM length, SM area, and SM curve progression and neurologic sequelae.

Additionally, this new measurement modality can be used to better determine surgical success in CM-I patients treated with PFD. The possibility that SM improvement after PFD serves as an important predictor of scoliosis prognosis raises many management question. The most important question may be whether all patients should be reimaged to assess the success of the PFD and how to define PFD failure. Prior studies and the results of this study suggest that maximal surgical benefit occurs at about 6 months after PFD.(58, 59) However, results from this study and others suggest that maximal curve improvement can occur many months after this time point. In this study, 7.5% of patients had no SM improvement following primary decompression. These patients all underwent secondary decompression and all experienced subsequent SM improvement with 80% experiencing deformity improvement. Krieger et al. found a similar rate of reoperation, with 8.5% of patients requiring reoperation, with such a high rate of reoperation that we believe that reimaging at 6-months in all patients is indicated.(8) However, while the management is clear in reimaged patients who have no appreciable change in SM size, more studies are necessary to guide the management of patients with partial SM improvement. Finally, we believe this measurement modality can be used to compare PFD procedures, with the outcome being SM improvement rate and percentage.

We believe that the results of our CM-I only study will have significant short-term and long-term impact. As the first study on curve progression in this population, we believe the most significant effect on management in the short-term is to inform neurosurgeons and orthopedic surgeons that CM-I PFD in the absence of SM many not influence curve progression. Arguments put forth regarding CM-I as a cause of scoliosis in the absence of SM revolve around tonsillar ectopia causing direct, spinal cord injury through a similar mechanism as SM.(17, 18) The results of this study argue against this theory. Firstly, patients with SM often present with upper motor neuron symptoms including clonus and hyperreflexia, as upper motor neurons are injured by the expanding SM. Additionally, patients with SM often present with motor difficulty and hypertonia or hypertonia. These are also signs of spinal cord injury. In this study and in findings by Alzate et al., no CM-I-only patients presented with these symptoms, arguing against this theory.(35) Additionally, in this study patients who underwent decompression were less likely to experience curve improvement compared to patients who did not undergo decompression, arguing against CM-I as the cause of scoliosis in this population. Additionally, patients with SM treated with bracing alone have been found to have poor outcomes because spinal injury, which is thought to be the cause of scoliosis in these patients, is not addressed.(7, 53) In this study, many CM-I-only patients had positive outcomes with bracing alone, arguing against the theory of direct spinal and muscular injury caused by tonsillar ectopia. Finally, consistent with prior studies, this study demonstrated no association between tonsillar ectopia and curve severity at presentation, further arguing against this theory.(18, 21) To definitively rule out this theory, biochemical studies should be performed to assess for muscle dennervation in this population. However, we believe that the results of this study suggest that PFD for curve management in CM-I-only patients is relatively contraindicated. Additionally, the results of this study suggest that bracing in these patients may prevent curve progression and lower the need for deformity correction surgery at follow-up.

Summary

CM-I with Concomitant SM

Improvement in SM scale size after decompression is associated with spinal deformity improvement at follow-up.

Improvement in SM scale size after decompression is associated a decreased risk of spinal deformity progression.

Patients with larger SM at presentation are more likely to require fusion at follow-up.

CM-I patients with single major curves are more likely to improve when compared to patients with double major curves.

Curves >40 degrees were likely were more likely to progress and require fusion.

Curves in EOS and AIS patients experienced a benefit following decompression.

CM-I without Concomitant SM

Degree of tonsillar ectopia correlates with the risk of neurologic deficits at presentation.

Degree of tonsillar ectopia does not correlate with curve severity at presentation or curve progression at follow-up.

No association between decompression and curve progression at follow-up was observed.

Patients treated with bracing were more likely to experience curve improvement and less likely to experience curve progression, independent of PFD status.

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Table 1. Patient and condition characteristics (N=78).		
Characteristic	**Freq.**	**(%)**
Sex (*% male*)	25	(32%)
Age at scoliosis diagnosis (*mean* ± *SD; N=76*)	9.3	± 3.68
Age group		
Age <10 years	33	(42%)
Age ≥ 10 years	45	(58%)
Age at procedure (*years; mean* ± *SD*)	9.9	± 4.18
Chiari size (*mean* ± *SD*)	11.2	± 4.23
Decompression	67	(86%)
Age at Decompression (*years; mean* ± *SD*)	9.3	± 3.52
Curve type		
Right thoracic	17	(22%)
Right thoracolumbar	23	(30%)
Right lumbar	1	(1%)
Left thoracic	12	(15%)
Left thoracolumbar	22	(28%)
Left lumbar	3	(4%)
Curve shape		
Single major curve	60	(77%)
Double major curve	19	(24%)
Initial curve magnitude (*degrees; mean* ± *SD*)	34.4	± 10.91
Initial Chiari size (*mm; mean* ± *SD*)	11.2	± 4.23
Initial Syrinx size		
Length (*mm; mean* ± *SD*)	141.9	± 73.15
Width (*mm; mean* ± *SD*)	7.3	± 3.72
Levels (*mean* ± *SD*)	11	± 5
Scale (*width*levels*)	91.3	± 67.21

*The number in parentheses (*N*=) represents the number of patients with available data for the given characteristic.

Table 2. Curve magnitude and syrinx dimensions over time (N=78).

	Curve Magnitude		Syrinx Dimensions					
			Width		Levels		Scale	
Time point	**Mean**	**± SD**	**Mean**	**± SD**	**Mean**	**± SD**	**Mean**	**± SD**
Presentation	34.4	± 10.91	7.3	± 3.70	11	± 6	91.3	± 4.68
Most-recent followup	38	± 23.36	2.7	± 1.66	8	± 4	23.5	± 4.42
Change from pres to followup	3.6	± 20.52	-4.7	± 3.67	-4	± 4	-69.8	± 3.97
Percent improved (*freq. (%)*)	35	(45%)	63	(80%)	48	(64%)	65	(83%)

Table 3. Results of multivariable logistic regression analysis for improvement in curve magnitude and fusion.

Improvement in curve magnitude (35/78 = 45%)			
Factors	**OR**	**95% CI**	**P**
Initial curve >40 degrees	0.26	[0.08-0.98]	0.03
Change in syrinx scale	1.01	[1.00-1.02]	0.01

Fusion (23/78 = 29%)			
Factors	**OR**	**95% CI**	**P**
Initial curve >40 degrees	4.3	[1.31-14.11]	0.02
Initial syrinx scale	1.03	[1.00-1.05]	0.03
change in syrinx scale	0.97	[0.94-0.99]	0.02

Table 4. Characteristics by outcome group for decompression subjects only (N=67).

Variable	Progressed (n=22) Freq.	(%)	Stabilized (n=18) Freq.	(%)	Improved (n=27) Freq.	(%)	P*
Change in curve (*mean* ± *SD*)	24.9	± 17.43	0.8	± 3.37	-14.5	± 7.91	
Patient/condition characteristics							
Sex (*% male*)	8	(36%)	5	(28%)	8	(30%)	
Age at scoliosis diagnosis (*mean* ± *SD*)	8.4	± 3.19	10.5	± 3.01	7.8	± 3.51	
Age group (*% ≥ 10 years*)	13	(59%)	8	(44%)	19	(70%)	0.22, 0.82
Chiari size (*mean* ± *SD*)	11.2	± 4.37	13.3	± 3.89	11.3	± 4.03	0.03, 0.25
Initial kyphosis (*mean* ± *SD*)	35.8	± 8.69	34.4	± 6.80	34.4	± 5.41	
Initial curve (*mean* ± *SD*)	38.2	± 11.87	34.4	± 10.00	33.3	± 8.90	
Initial curve < 40 degrees	12	(55%)	16	(89%)	22	(82%)	0.06, 0.26
Curve characteristics							
Right thoracic	7	(32%)	3	(17%)	4	(15%)	
Right thoracolumbar	6	(27%)	9	(50%)	5	(19%)	
Right lumbar	0	(0%)	1	(6%)	0	(0%)	
Left thoracic	4	(18%)	4	(22%)	3	(11%)	
Left thoracolumbar	4	(18%)	1	(6%)	14	(52%)	
Left lumbar	1	(5%)	0	(0%)	1	(4%)	
Curve shape							
Single major curve	14	(64%)	14	(78%)	24	(92%)	0.20, 0.04
Double major curve	8	(36%)	4	(22%)	2	(8%)	
Initial Syrinx size							
Length (*mean* ± *SD*)	144.7	± 62.42	155.5	± 86.60	160.5	± 63.97	
Width (*mean* ± *SD*)	7.8	± 3.40	7	± 3.39	9	± 3.14	
Levels (*mean* ± *SD*)	11.6	± 4.33	11.3	± 4.25	12.6	± 4.29	
Scale (*width*levels*)	98.6	± 63.93	83.9	± 54.12	121.6	± 67.15	
Treatment characteristics							
Bracing	14	(64%)	12	(67%)	16	(59%)	0.55, 0.33
Casting	1	(5%)	0	(0%)	1	(4%)	

*P-values are based on a multivariable multinomial logistic regression model. The first value is associated with the comparison between the odds of stabilized outcome versus progressed outcome and the second value is associated with the comparison between the odds of improved outcome versus progressed outcome.

Table 5. Patient and condition characteristics (N=51).		
Characteristic	**Freq.**	**(%)**
Sex (*% male*)	17	(33%)
Age at scoliosis diagnosis (*years; mean ± SD; N=76*)	10	± 4.3
Age group		
Age <10 years	29	(57%)
Age >= 10 years	22	(43%)
Age at procedure (*years; mean ± SD*)	10	± 4.4
Decompression	11	(22%)
Age at Decompression (*years; mean ± SD*)	8	± 5.0
Curve type		
Right thoracic	11	(22%)
Right thoracolumbar	15	(29%)
Right lumbar	0	(0%)
Left thoracic	6	(12%)
Left thoracolumbar	8	(16%)
Left lumbar	11	(22%)
Curve shape		
Single major curve	31	(61%)
Double major curve	20	(39%)
Initial curve magnitude (*degrees; mean ± SD*)	31	± 16.2
Initial Chiari size (*mm; mean ± SD*)	9	± 3.6
Bracing	29	(57%)
Casting	1	(2%)

Table 6. Outcomes (N=51).		
Characteristic	**Mean**	**± SD**
Curve magnitude		
Presentation	30.8	± 16.16
Most-recent followup	35.3	± 19.95
Change (presentation to followup)	4.5	± 13.97
Percent improved (presentation to followup)	16	(31%)
Fusion (%; 95% CI)	29%	(17.9 to 44.0%)

SD, standard deviation; CI, confidence interval

Table 7. Patient, condition, treatment and outcome characteristics by decompression.

Characteristic	Decompression (n=11) Freq.	(%)	No decompression (n=40) Freq.	(%)	P
Patient/condition characteristics					
Sex *(% male)*	6	(55%)	11	(28%)	0.15
Age at scoliosis diagnosis (*years; mean ± SD*)	9	± 5.01	10	± 4.1	--
Initial kyphosis (*degrees; mean ± SD*)	36	± 16.6	36	± 9.8	1.00
Initial curve magnitude (*degrees; mean ± SD*)	24	± 13.6	33	± 16.5	0.10
Curve characteristics					
Right thoracic	2	(18%)	9	(23%)	0.86
Right thoracolumbar	5	(46%)	10	(25%)	
Right lumbar	0	(0%)	0	(0%)	
Left thoracic	1	(9%)	5	(13%)	
Left thoracolumbar	1	(9%)	7	(18%)	
Left lumbar	2	(18%)	9	(23%)	
Curve shape					
Single major curve	10	(91%)	21	(53%)	0.03
Double major curve	1	(9%)	19	(48%)	
Initial Chiari size (*mm; mean ± SD*)	10.5	± 5.34	8.6	± 3.01	0.29
Treatment characteristics					
Bracing	5	(46%)	24	(60%)	0.50
Casting	1	(9%)	0	(0%)	0.22
Outcomes					
Fusion	2	(18%)	13	(33%)	0.36
Curve magnitude at followup (*degrees; mean ± SD*)	31	± 17.1	37	± 20.7	0.42
Change in curve magnitude (*degrees; mean ± SD*)	7	± 13.4	4	± 14.2	0.55
Change in curve type	2	(18%)	14	(35%)	0.30

SD, standard deviation

Table 8. Characteristics by outcome group (N=51).

	Progressed (n=22)		Stabilized (n=18)		Improved (n=11)	
Variable	**Freq.**	**(%)**	**Freq.**	**(%)**	**Freq.**	**(%)**
Change in curve (*mean* ± *SD*)	16.2	± 10.34	0.9	± 2.76	-13.2	± 8.26
Patient characteristics						
Sex (*% male*)	7	(32%)	6	(33%)	4	(36%)
Age at scoliosis diagnosis (*mean* ± *SD*)	10.2	± 3.59	11	± 5.20	7.8	± 3.43
Age ≥ 10 years	8	(36%)	5	(28%)	8	(73%)
Chiari size (*mean* ± *SD*)	8.1	± 2.83	10.1	± 4.47	8.9	± 3.36
Initial kyphosis (*mean* ± *SD*)	31.5	± 7.67	38.1	± 13.87	40	± 9.77
Initial curve (*mean* ± *SD*)	29.8	± 16.89	32.2	± 18.07	30.4	± 12.06
Curve > 40	17	(77%)	13	(72%)	9	(82%)
Curve characteristics						
Right thoracic	9	(41%)	1	(6%)	1	(9%)
Right thoracolumbar	2	(9%)	10	(56%)	3	(27%)
Right lumbar	0	(0%)	0	(0%)	0	(0%)
Left thoracic	3	(14%)	1	(6%)	2	(18%)
Left thoracolumbar	3	(14%)	2	(11%)	3	(27%)
Left lumbar	5	(23%)	4	(22%)	2	(18%)
Curve shape						
Single major curve	10	(46%)	13	(72%)	8	(73%)
Double major curve	12	(55%)	5	(28%)	3	(27%)
Treatment characteristics						
Decompression	3	(14%)	8	(44%)	0	(0%)
Bracing	10	(46%)	9	(50%)	10	(91%)
Casting	0	(0%)	1	(6%)	0	(0%)