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CASE SERIES

Chiari I malformations with syringomyelia: long-term results of neurosurgical decompression

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Abstract

Study design Retrospective case series.

Objectives The objective was to assess the long-term outcomes on scoliosis following Chiari-I (CM-I) decompression in patients with CM-I and syringomyelia (SM). A secondary objective was to identify risk factors of scoliosis progression.

Background The association between CM-I with SM and scoliosis is recognized, but it remains unclear if CM-I decompression alters the long-term evolution of scoliosis in patients with associated syringomyelia.

Methods A retrospective review of children with scoliosis, CM-I, and SM during 1997–2015 was performed. Congenital, syndromic, and neuromuscular scoliosis were excluded. Clinical and radiographic characteristics were recorded at presentation, pre-decompression, after 1-year, and latest follow-up. A scale to measure syringomyelia area on MRI was used to evaluate SM changes post-decompression.

Results 65 children with CM-I, SM, and scoliosis and a mean age of 8.9 years (range 0.7–15.8) were identified. Mean follow-up was 6.9 years (range 2.0–20.4). Atypical curves were present in 28 (43%) children. Thirty-eight patients (58%) underwent decompression before 10 years. Syringomyelia size reduced a mean of 70% after decompression ($p < 0.001$). Scoliosis improved in 26 (40%), stabilized in 17 (26%), and progressed in 22 (34%) cases. Early spinal fusion was required in 7 (11%) patients after a mean of 0.5 ± 0.37 years and delayed fusion in 16 (25%) patients after 6.0 ± 3.24 years. The remaining 42 (65%) patients were followed for a median of 6.1 years (range 2.0–12.3) without spine instrumentation or fusion. Fusion patients experienced less improvement in curve magnitude 1-year post-decompression ($p < 0.001$) and had larger curves at presentation (43° vs. 34°; $p = 0.004$).

Conclusions Syringomyelia size decreased by 70% after CM-I decompression and scoliosis stabilized or improved in two-thirds of patients. Greater curve improvement within the first year post-decompression and smaller curves at presentation decreased the risk of spinal fusion. Neurosurgical decompression is recommended in children with CM-I, SM, and scoliosis with the potential to treat all three conditions.

Level of evidence Level IV.

Keywords Chiari I · Syringomyelia · Decompression · Scoliosis · Pediatrics

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Introduction

Intraspinal pathology (ISP) must be ruled out in certain patients with idiopathic scoliosis. ISP is frequently associated with scoliosis, but less is documented about the management and evolution of scoliosis in these patients [1–6]. Chairi-1 malformations (CM-I) are common neuraxis anomalies found in nearly 4% of children under 18 years, with concomitant syringomyelia (SM) and scoliosis found in 88% and 20%, respectively [5, 7, 8].

Syringomyelia in patients with CM-I can be associated with scoliosis [9–14]. Two theories for SM formation exist: (1) water-hammer theory and (2) one-way valve theory. The water-hammer theory proposes that arterial pulses from the choroid plexus transmit cerebrospinal fluid down an abnormal fourth ventricle and mediate SM formation [9]. The one-way valve theory postulates that unequal pressures generated through the Valsalva maneuver causes increased pressure in the spinal cord resulting in SM development [9]. Neither is proven; however, MRI studies have found aberrant communications between the ventricles in CM-I patients [9, 11]. Biochemical and histologic data suggest that enlarging SM causes asymmetrical injury to the anterior horn of the spinal cord that may potentiate scoliosis through denervation and weakness of paraspinal musculature [10, 13, 14].

The relationship between CM-I, SM, and scoliosis is adequately documented, but neurosurgical decompression of CM-I on scoliosis outcome is unknown and mixed results are reported [7, 15–20]. These studies are often limited to small heterogeneous case series with short follow-up and some believe the benefits of decompression are temporary [14, 16–18, 20–24]. A recent series of 23 patients by Ravindra et al. reported poor durability of decompression, with 30% experiencing late curve progression requiring fusion after 5 years [20]. To the authors' knowledge, this is the largest study to date evaluating the long-term outcomes of scoliosis after neurosurgical decompression in children with CM-I and SM.

The main objectives of this study were: (1) to describe the presentation of scoliosis in children with CM-I and SM; and (2) to identify risk factors of scoliosis progression and spinal fusion after decompression.

Methods

IRB approval (IRB-P00023640) was obtained and electronic medical records were reviewed for children with scoliosis, CM-I, and SM during 1997–2015. Neuromuscular, congenital, and syndromic scoliosis and prior spinal deformity surgery were excluded. Scoliosis was defined using the Cobb method: coronal curve $\geq 10°$. Demographics, symptoms, neurologic abnormalities, bracing characteristics, and surgical details were recorded. Neurosurgical posterior fossa decompression (PFD) consisted of primary sub-occipital decompression with C1 laminectomy and Y-shaped duraplasty in all cases. A fourth ventricle-to-subarachnoid space stent was placed during PFD in a minority of cases when severe arachnoid scarring limited fourth ventricle outflow.

Scoliosis evaluation

Direction, magnitude, and location of major curves were evaluated pre-decompression, 1 year, and latest follow-up. The Spiegel et al. classification was used to define atypical curves that included: left thoracic, double thoracic, triple, and long thoracic curves [25]. Factors associated with scoliosis progression were examined trichotomously. Post-decompression groups were created according to the SOSORT criteria: Cobb improvement $> 5°$, stabilization or $\leq 5°$ change in magnitude, and progression $> 5°$.

Non-operative treatment

With the exception of PFD, indications for bracing or casting were similar to the management of idiopathic scoliosis. Children < 2 years with curves $\geq 25°$ were treated with elongation, derotation, and lateral flexion casting ($n = 1$) until bracing was initiated. A Boston-style thoracolumbar sacral orthosis (≥ 18 h/day) or Charleston-type night brace (12 h/night) was prescribed in children ≥ 2 years with residual curves $> 25°$. Bracing was continued until curves stabilized $< 25°$, skeletal maturity was reached (determined by Risser 4 or 5), or surgical intervention was required in progressive curves $\geq 45°$. Due to the long-term follow-up of this study, objective brace-wear parameters were not available and compliance was based on patient and surgeon report. Patients were considered compliant if braces were worn $\geq 2/3$rds the prescribed duration. Skeletally immature patients with stable curves or residual curves $< 25°$ were observed and followed up every 6 months to detect late curve progression.

Chiari I and syringomyelia evaluation

MRI's were performed in cases of early onset scoliosis, atypical curves, or neurologic symptoms and confirmed the diagnosis of CM-I and SM. Chiari I malformations were defined as a caudal descent of the cerebellar tonsils ≥ 5 mm below the foramen magnum. A line was drawn between the inner margins of the foramen magnum, from basion to opisthion, and the distance from this line to the inferior cerebellar tonsils determined CM-I size. A central cyst on T2 MRI confirmed a syringomyelia. Syringomyelia length (vertebral levels spanned) and width (maximum diameter

in millimeters) were measured [12]. Scaled SM size was calculated by multiplying length and width (represented as scaled SM units). Serial MRI's were performed to evaluate SM area pre-and post-decompression.

Statistical analysis

Patient and condition characteristics were summarized using SAS V.9.4 (SAS Inc; Cary, NC, USA). Continuous characteristics were summarized by mean and SD, mean and range, or median and interquartile range (IQR 25th–75th percentile) and categorical characteristics by frequency and percent. Bivariate comparisons were conducted for patient, curve, and treatment characteristics using SOSORT outcome groups described above. Comparisons were conducted using analysis of variance or Chi squared test based on variable type.

Continuous changes in Cobb and SM size were calculated through the difference of pre-and post-decompression measurements: (+) change indicated an increase in size (progression); and (−) changes a decrease (improvement). Multivariable linear modeling was used to analyze the effects of age, sex, Cobb, curve type, CM-I size, SM size, and bracing on change in Cobb. Model selection procedures were implemented to find the most parsimonious model to estimate the effects of patient and curve characteristics on outcomes. Decisions were made using a combination of model fit (based on Akaike's information criterion) and minimal change in effect estimates for significant effects. All tests were two-sided and $p < 0.05$ was considered significant.

Table 1 Patient and condition characteristics ($n = 65$)

Characteristic	Freq.	(%)	Range
Sex (% female)	44	(68%)	
Follow-up duration (years; mean ± SD)	6.9	± 3.9	(2.0–20.4)
Age at scoliosis diagnosis (years; mean ± SD)	8.9	± 3.39	(0.7–15.8)
Age at decompression (years; mean ± SD)	9.1	± 3.45	(1.6–16.7)
Age < 10 years	38	(58%)	
Age ≥ 10 years	27	(42%)	
Curve type			
Right thoracic	13	(20%)	
Right thoracolumbar	19	(29%)	
Right lumbar	1	(2%)	
Left thoracic	11	(17%)	
Left thoracolumbar	19	(29%)	
Left lumbar	2	(3%)	
Curve shape			
Single-major curve	51	(80%)	
Double-major curve	12	(18%)	
Triple-major curve	2	(3%)	
Initial curve magnitude (°; mean ± SD)	36	± 10.4	(14–60)
Initial curve ≥ 40°	20	(31%)	
Curve magnitude for curves ≥ 40° (°; mean ± SD)	49	± 5.2	(43–60)
Initial Chiari size (mm; mean ± SD; $n = 63$)*	11.9	± 4.19	(4.5–21.3)
Initial syrinx size ($n = 64$)*			
Length (mm; mean ± SD)	153.9	± 69.28	(9.4–318.3)
Width (mm; mean ± SD)	8.1	± 3.37	(1.1–15.1)
Levels (mean ± SD)	12	± 4.3	(2–19)
Scale unit size (width × levels; mean ± SD)	104.8	± 64.23	(7.8–246.5)
Post-decompression syrinx size ($n = 64$)*			
Length (mm; mean ± SD)	110.2	± 63.15	(0–230)
Width (mm; mean ± SD)	2.7	± 1.76	(0–8.5)
Levels (mean ± SD)	8	± 4.6	(0–17)
Scale unit size (width×levels; mean ± SD)	27.0	± 23.20	(0–113.7)
% Reduction scale unit size (width × levels; mean ± SD)	70%	± 21.07	(24%–100%)

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

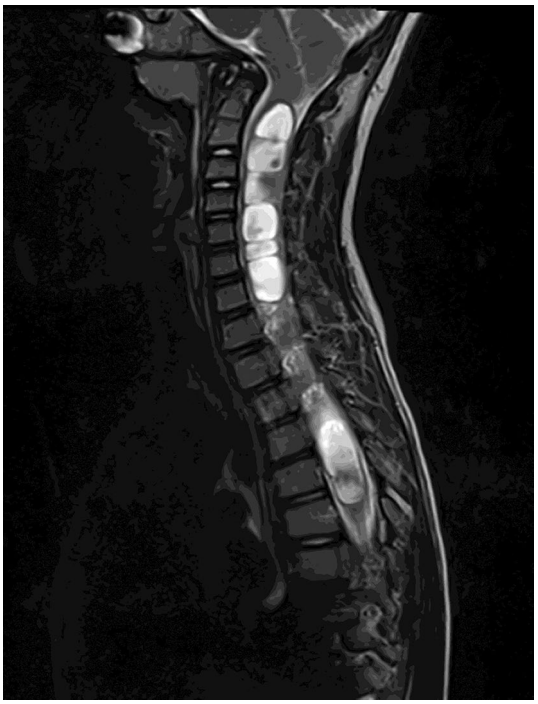

Fig. 1 Case 1: 5.4-year-old girl that presented with scoliosis as the chief complaint. An MRI was performed due to the patient's young age and curve severity. Pre-decompression sagittal T2 MRI demonstrating a Chiari I malformation 11.3 mm below the foramen magnum and a large septated syringomyelia that extends distally 14 levels from the craniocervical junction to below T7. Decompression was performed 2 months after the initial presentation

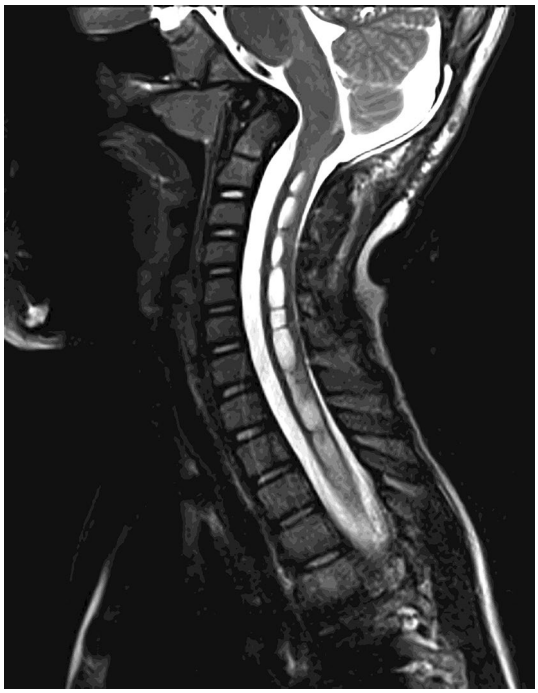

Fig. 2 Case 1 continued: 3-month post-decompression a significant 73% reduction in syringomyelia size is seen on T2 MRI, with adequate spinal cord decompression and cerebrospinal fluid flow surrounding the posterior cerebellum

Results

Patient demographics and presenting symptoms

Sixty-five patients (44 females) with a mean age of 8.9 ± 3.39 years (range 0.7–15.8) at scoliosis diagnosis were identified (Table 1). Mean follow-up was 6.9 years (range 2–20.4). 27 patients (42%) were adolescent-onset (≥ 10 years) and 38 (58%) were early onset scoliosis (< 10 years). 58 patients (89%) presented with scoliosis as the chief complaint, with CM-I and SM identified through MRI evaluation. Five (8%) were diagnosed incidentally. Two (3%) presented with neurologic symptoms: one clonus and another chronic headaches. Of the patients presenting with scoliosis, neurologic history revealed chronic headaches in eight (12%) and upper extremity sensory disturbances in three (5%). Neurologic symptoms were found in a total of 13 patients (20%).

Major and atypical curve patterns

Major curve patterns included: 19 (29%) right thoracolumbar, 19 (29%) left thoracolumbar, 13 (20%) right thoracic, 11 (17%) left thoracic, 2 (3%) left lumbar, and 1 (2%) right lumbar (Table 1). 28 patients (43%) displayed atypical curves defined by Spiegel et al. [25]. 51 (80%) single-major, 12 (18%) double major, and 2 (3%) triple-major curves were seen. Mean major Cobb angle at presentation was 36° ± 10.4° (range 14°–60°) and 20 patients (31%) had curves ≥ 40°. Of the curves > 40°, the mean Cobb was 49° ± 5.2 (range 43°–60°). No correlation was found between Cobb and SM size ($r = 0.10$; 95% CI − 0.15 to 0.34; $p = 0.41$) or CM-I size at presentation ($r = 0.14$; 95% CI − 0.11 to 0.38; $p = 0.26$).

Chiari I malformation

Chiari I malformations were a mean of 11.9 ± 4.2 mm (range 5.0–21.3 mm) below the foramen magnum (Table 1). No correlation was found between Chiari size and SM size ($r = 0.20$; 95% CI − 0.05 to 0.42; $p = 0.12$). Neurosurgical decompression was performed at a mean of 9.1 ± 3.5 years, with 38 (58%) < 10 years. A fourth ventricle to subarachnoid space stent was placed during PFD in six patients (9%).

Table 2 Bivariate comparisons in baseline characteristics across SOSORT outcome groups ($n=65$)

Variable	Progressed Freq. (%)	Stabilized Freq. (%)	Improved Freq. (%)	P*
Number of patients	22 (34%)	17 (26%)	26 (40%)	
Change in curve (°)	24 ± 18.6	2 ± 4.8	− 14 ± 8.2	
Patient/condition characteristics				
Sex (% female)	14 (64%)	12 (71%)	18 (69%)	0.89
Age at decompression (years)	9.6 ± 2.93	10.4 ± 3.22	7.3 ± 3.33	0.005*
Age group (% ≥ 10 years)	12 (55%)	8 (47%)	7 (27%)	
Chiari size (mm)	12.1 ± 4.23	11.9 ± 4.42	11.8 ± 4.17	0.97
Initial kyphosis (°)	39 ± 6.8	32 ± 9.7	34 ± 5.3	0.16
Initial curve (°)	40 ± 10.7	35 ± 12.0	34 ± 8.6	0.15
Initial curve ≥ 40°	11 (50%)	4 (24%)	5 (19%)	0.40
Curve type				< 0.001*
Right thoracic	8 (36%)	2 (12%)	3 (12%)	
Right thoracolumbar	6 (27%)	8 (47%)	5 (19%)	
Right lumbar	0 (0%)	0 (0%)	1 (4%)	
Left thoracic	3 (14%)	6 (35%)	2 (8%)	
Left thoracolumbar	4 (18%)	1 (6%)	14 (54%)	
Left lumbar	1 (5%)	0 (0%)	1 (4%)	
Curve shape				0.02*
Single-major curve	13 (57%)	15 (88%)	23 (92%)	
Double-major curve	8 (35%)	2 (12%)	2 (8%)	
Triple-major curve	2 (9%)	0 (12%)	0 (0%)	
Initial syrinx size				
Length (mm)	159.4 ± 73.95	144.8 ± 73.89	155.1 ± 64.15	0.83
Width (mm)	8.3 ± 3.21	6.3 ± 3.34	9.1 ± 3.18	0.03*
Vertebral levels	12.3 ± 4.33	11.1 ± 4.21	12.3 ± 4.36	0.61
Scale unit size (width × levels)	109.3 ± 62.50	75.9 ± 54.99	120 ± 67.23	0.08
Treatment characteristics				
Postoperative brace	15 (83%)	10 (67%)	19 (83%)	0.54

Data are presented as frequency (%) or mean ± standard deviation

Syringomyelia

Syringomyelia scaled size decreased a mean of 70% after PFD, from 104.8 to 27.0 units (Table 1, Figs. 1, 2). Mean SM width and vertebral levels decreased from 8.1 to 2.7 mm ($p<0.001$) and from 12 to 8 levels ($p<0.001$), respectively. Five patients (7%) did not have an appreciable decrease in SM size and required a second PFD. Of these, two PFD and three PFD with fourth ventricle to subarachnoid shunting were performed. All patients experienced SM improvement following secondary decompression.

Scoliosis outcome

Scoliosis improved in 26 (40%) patients, 17 (26%) stabilized, and 22 (34%) progressed (Table 2, Figs. 3, 4). Differences were detected in SOSORT outcome groups with respect to age at PFD ($p=0.005$), curve type ($p<0.001$), curve shape ($p=0.02$), syrinx width ($p=0.03$), and brace compliance ($p<0.001$) (Table 2). Children were compared based on < 10 or ≥ 10 years. A higher incidence of curve progression was seen in PFD in children ≥ 10 years (55% vs. 27%; $p=0.005$). Outcomes also differed based on curve type. Both (100%) triple-major and 8/12 (67%) double major curves progressed compared to 23/51 (45%) single-major curves ($p=0.02$).

Of these potential risk factors, multivariable analysis determined that change in SM scaled size and change in Cobb within the first year following PFD were the only factors significantly associated with long-term scoliosis outcome (Table 3, Fig. 5). Each additional ten unit decrease in SM scaled size resulted in a 0.8° reduction in curve after PFD ($p=0.03$). Furthermore, after correcting for age and presenting Cobb, each additional 10° improvement in Cobb in the first year post-decompression resulted in a total of 9° reduction in Cobb at latest follow-up ($p<0.001$) (Table 3).

Residual curves ≥ 25° seen post-decompression were braced until curve stabilization, progression ≥ 45° requiring

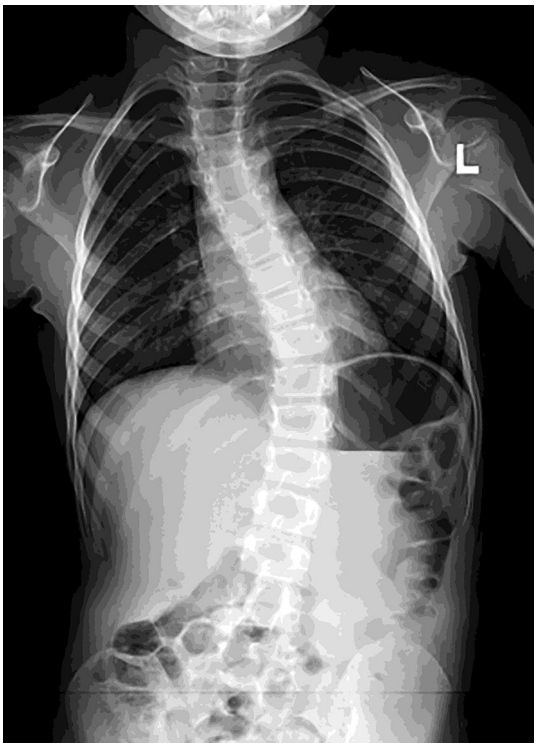

Fig. 3 Case 1 continued: The patient presented with a left thoracolumbar curve measuring 45° as demonstrated on the PA spine X-ray

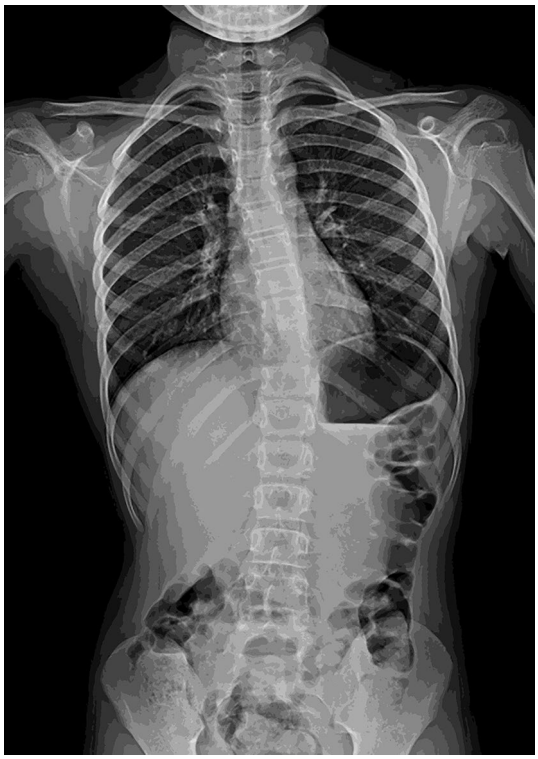

Fig. 4 Case 1 continued: Scoliosis improvement is seen 2 years post-decompression. The Cobb angle has reduced to 18° and corresponds to a 60% curve correction

spinal fusion, or skeletal maturity was reached. A total of 44 patients (68%) were braced. Of these, 41 patients (93%) were prescribed a Boston-style brace and 3 patients (7%) received a Charleston-type brace (Table 4). Twenty-one patients (32%) were not braced after decompression due to residual curves < 25°, progressive curves ≥ 45° requiring immediate spinal fusion, stable curves, or patients approaching skeletal maturity. Bracing details can be found in Table 4.

Spinal fusion

Spinal fusion was performed by the latest follow-up in 23 (35%) patients. Mean follow-up duration of the 42 (65%) non-fusion patients was 6.1 years (range 2.0–12.3). Fifty-two patients (80%) were followed up until skeletal maturity. Seven children (11%) underwent early fusion after PFD at a mean of 0.5 years (IQR 0.3–1.0) and with an average Cobb of 60° (range 41°–75°) (Table 4). The remaining 16 (25%) required delayed fusion after a mean of 6.0 years (IQR 3.5–8.4 years) and Cobb of 63° (range 35°–120°). In cases of late fusion, the Cobb progressed a mean of 22° (range −4° to 63°) between decompression and fusion (Figs. 6, 7, 8, 9). Fusion patients had larger curves at presentation (43° vs. 34°; $p = 0.004$).

No difference in CM-I size (12.0 mm vs. 11.9 mm) was seen in cases with and without fusion. 11 of 20 (55%) patients presenting with curves ≥ 40° progressed after PFD and six required early fusion, but was not significant ($p = 0.40$). 8 of 12 (67%) patients with double-major curves received spinal fusion compared to 13/51 (25%) with single-major curves ($p < 0.001$). Both triple-major curves required fusion.

Discussion

Chiari-I malformations are the most common neuraxis anomalies in children [5]. A recent population based study of 14,118 patients found CM-I in 3.8% of children under 18 years [5]. CM-I is the most common cause of syringomyelia formation with reported incidences between 50 and 76% [26, 27]. The leading theory of scoliosis development postulates that SM expansion causes asymmetric injury to the anterior horn of the spinal cord [10, 13, 14]. This denervation and paraspinal muscle weakness can result in scoliosis. Biochemical and histologic studies support this theory

Table 3 Multivariable results for associations with curve progression

Presentation	Change in curve magnitude								
	Model 1			Model 2			Model 3		
	EE	(95% CI)	*p*	EE	(95% CI)	*p*	EE	(95% CI)	*p*
Sex (% female)	4.16	(−5.5 to 13.8)	0.40						
Age (years; mean ± SD)	−0.38	(−1.8 to 1.0)	0.60	−0.26	(−1.6 to 1.1)	0.72	−0.27	(−1.6 to 1.0)	0.69
Initial curve > 40°	6.24	(−3.7 to 16.1)	0.22	6.29	(−3.6 to 16.2)	0.22	6.38	(−3.3 to 16.0)	0.20
Chiari size (mm)	0.27	(−0.8 to 1.3)	0.62	0.17	(−0.9 to 1.2)	0.75			
Change in syrinx scale (width × level)	0.08	(0.00 to 0.15)	0.04*	0.08	(0.01 to 0.16)	0.03*	0.08	(0.01 to 0.15)	0.03*
Change in curve at 1-year (°)	0.89	(0.40 to 1.37)	< 0.001*	0.87	(0.38 to 1.35)	< 0.001*	0.87	(0.40 to 1.34)	< 0.001*

Each model was adjusted for the covariates with estimated effects listed

SD standard deviation, *CI* confidence interval, *EE* effect estimate

*Significant effect on curve progression and severity at the final follow-up

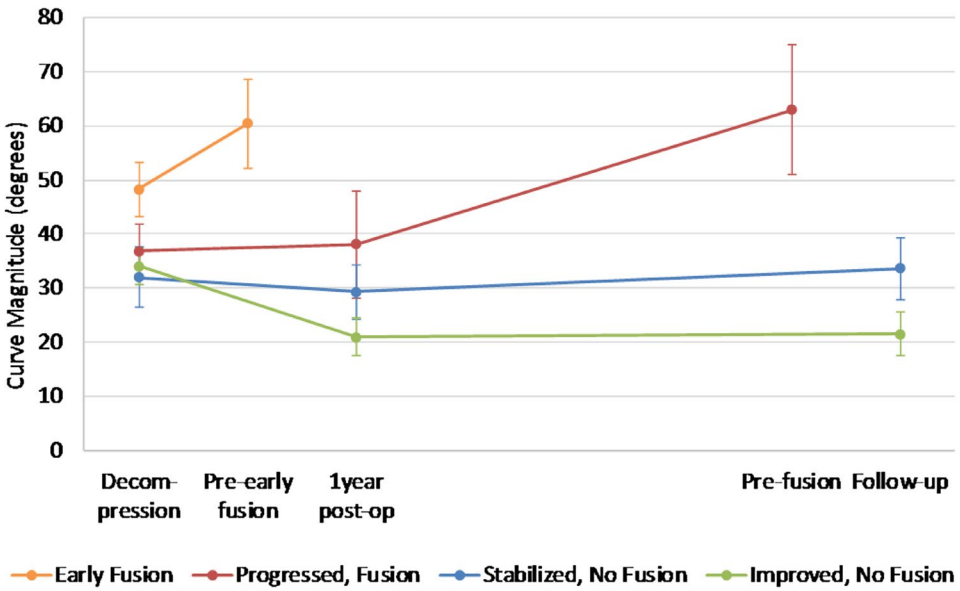

Fig. 5 Change in Cobb angles of various groups after decompression

and report improvement of paraspinal muscle innervation and spinal deformity following PFD [13].

Most authors recommend CM-I decompression in patients with SM to prevent or reverse neurological deterioration [28, 29]. However, the benefits of PFD on spinal deformity are less clear [7]. Prior studies report scoliosis improvement between 18%-38% [15, 16, 18, 20, 22, 30]. The follow-up in these series is limited and many question the long-term benefits of decompression [14, 16–18, 21–24].

While the relationship between SM and paraspinal muscle denervation has been studied in vitro, the association between SM and scoliosis remains unclear [10, 13, 14]. SM improvement following decompression is observed in 65%–93% of patients and is consistent with our results [15, 19, 30–32]. However, many studies report curve progression regardless of SM improvement [15, 16, 18, 19, 25, 30, 32–34]. These studies fail to identify an association between SM characteristics or initial curve magnitude and the risk of progression [16, 18, 30, 32–34]. In this study, a scale was used to quantify SM dimensions to analyze SM changes and scoliosis outcome. We found that SM reduction post-decompression was associated with scoliosis improvement or stabilization in two-thirds of patients. This effect was most pronounced immediately following decompression and patients with larger improvements in Cobb during the first year displayed a lower incidence of delayed spinal fusion.

Curve progression following decompression is not uncommon and prior studies report progression in 30%–89% [15, 16, 18, 20, 22, 30, 35]. In the current study, 22 (35%) progressed, 7 (11%) underwent early fusion, and 16 patients (25%) required delayed spinal fusion after long-term

Table 4 Outcome summary by fusion groups

At decompression	Fusion ($n=23$, 35%)				No fusion ($n=42$, 65%)	
	Early ($n=7$)		Late ($n=16$)			
	Freq. (%)	Range	Freq. (%)	Range	Freq. (%)	Range
Sex (% female)	6 (86%)		11 (69%)		27 (64%)	
Age (years; mean ± SD)	12.3 ± 1.47	(9.9 to 14.5)	8.9 ± 2.70	(5.0 to 14.5)	9.1 ± 3.82	(1.6 to 16.7)
Age ≥ 10 years	6 (86%)		5 (31%)		16 (38%)	
Curve magnitude (°; mean ± SD)	48° ± 6.2	(39° to 60°)	38° ± 10.2	(22° to 57°)	34° ± 9.3	(17° to 57°)
Curve ≥ 40°	6 (86%)		6 (38%)		8 (19%)	
1 year post-decompression	Mean ± SD		Mean ± SD		Mean ± SD	
Change in syrinx scale (width*levels)	− 92 ± 36.1	(− 166 to − 61)	− 71 ± 44.2	(− 138 to − 7)	− 83 ± 64.9	(− 234 to − 5)
Change in curve (°)	18° ± 20.6°	(2° to 62°)	21° ± 12.2°	(− 17° to 33°)	− 10° ± 9.5°	(− 33° to 11°)
Treatment characteristics	Freq. (%)		Freq. (%)		Freq. (%)	
Bracing	NA		13 (81%)		30 (79%)	
Compliance[a]	NA		4 (31%)		25 (83%)	
Curve improvement (°; mean ± SD)	NA		13° ± 5.3°	(5° to 22°)	16° ± 8.7	(1° to 35°)
% Curve correction (mean ± SD)	NA		37% ± 12.7%	(15% to 50%)	46% ± 23.0%	(3% to 90%)
Brace duration (years; mean ± SD)	NA		4.2 ± 2.28	(1.1 to 8.5)	3.4 ± 2.65	(0.8 to 8.4)
Last follow-up or pre-fusion	Mean ± SD		Mean ± SD		Mean ± SD	
Time since decompression (years)	0.5 ± 0.37	(0 to 1.0)	6.0 ± 3.24	(1.9 to 13.8)	6.1 ± 2.78	(2.0 to 12.3)
Curve magnitude (°)	60° ± 11.1°	(41° to 75°)	63° ± 22.9°	(35° to 120°)	34° ± 10.9	(4° to 61°)
Change in curve magnitude (°)			22° ± 17.6°	(− 4° to 63°)	2 ± 5.3	(− 36° to 11°)

SD standard deviation, *NA* not applicable (patients with early fusions were not included in bracing data due to immediate spinal fusion, 1 patient was braced and not included)

[a]Compliance by report was used. Patients that wore the brace > 2/3rds the prescribed duration were considered compliant

follow-up. This is similar to reports by Ravindra et al. who analyzed the long-term durability of decompression on scoliosis [20]. In that series of 23 patients, 7 (30%) required delayed spinal fusion after 5 years and the authors concluded poor durability of PFD on long-term curve control. The mean follow-up in the current series was 6.9 years. However, the IQR of delayed spinal fusion in the current series was 3.5–8.4 years after PFD and our results might underestimate the true incidence of late progression. Case 2 is an interesting example of delayed curve progression after significant initial curve improvement. This case demonstrates the importance of long-term follow-up in these patients, or until skeletal maturity is reached at a minimum.

Different curves are found in patients with CM-I and Spiegel et al. identified atypical curve patterns present in < 2% of idiopathic patients [25]. In this study, 28 (43%) patients presented with atypical curves suggesting an influence of ISP on curve formation [36, 37]. Prior studies also report atypical curves between 44 and 51% [12, 25]. However, most do not comment on scoliosis outcome [8, 16, 35, 38]. In this study, single-major curves improved more often compared to double-major curves and triple-major curves. Flynn et al. found that 8/9 double-major curves experienced progression following PFD [22]. Zhu also reported that double-major curves progressed in 47% compared to 11% without [39]. Senguta et al. observed increased improvement of left thoracic curves, with 75% avoiding spinal fusion [18].

Authors suggest increased progression or fusion rates in those presenting with curves ≥ 30°. Tubbs et al. saw no improvement in curves ≥ 40° [27]. Ghanem et al. also found that 5/5 patients with curves ≥ 40° required fusion [21]. Nagib complemented these studies and reported improvement in 6/6 patients with curves < 30° and stabilization in 4/4 ≥ 30° curves [40]. Other studies observed improvement ≥ 40° curves [7, 16]. This study found that 12/20 patients with curves ≥ 40° required fusion. Of these, six were early fusions and suggests that severe curves are more resilient to SM treatment. Fusion patients also had larger curves at presentation. We believe that decompression should be performed in all children prior to spinal fusion to reduce the risk of perioperative neurological deficits [7]. Furthermore, our results suggest that an increased benefit may be obtained by performing PFD at a younger age in an attempt to minimize initial curve progression.

Other studies also report increased benefits in younger patients. Muhonen et al. found scoliosis improvement in

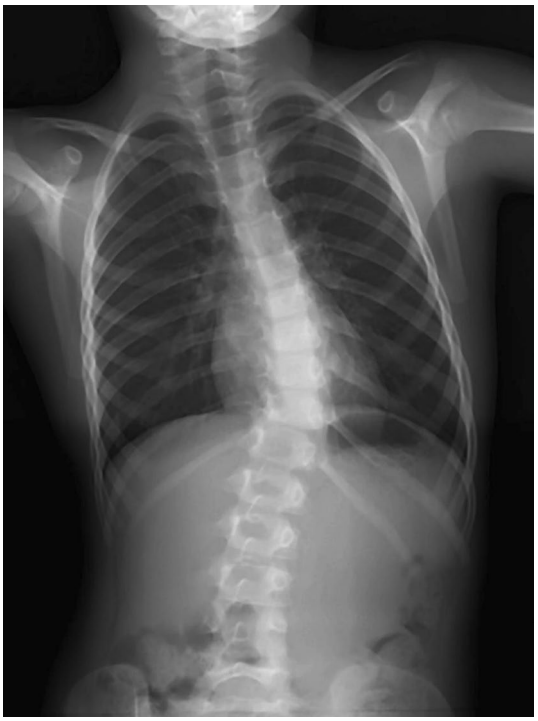

Fig. 6 Case 2: 5.2-year-old girl that presented with scoliosis and back pain. Pre-decompression PA X-ray demonstrating a 36° left thoracolumbar curve. MRI revealed a Chiari I and syringomyelia of 10.8 mm and 188.7 SM scaled units, respectively, and decompression was performed 2 months after the initial visit

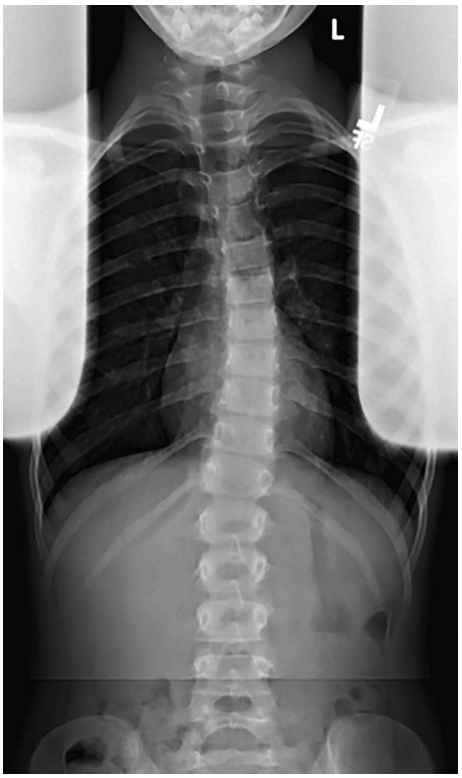

Fig. 7 Case 2 continued: 14-month post-decompression with a successful 90% reduction in SM size. The scoliosis has improved to 16° corresponding to a 56% curve correction

3/3 patients < 10 years, despite one ≥ 40° curve [23]. Flynn et al. and Brockmeyer et al. found that 7/10 (70%) and 10/11 (91%) patients < 10, respectively, avoided fusion [16, 22]. This study also found decreased rates of progression and fusion in younger children. Few authors have evaluated the benefits of decompression in patients ≥ 10. However, our results are promising and decompression may also alter deformity progression in children ≥ 10.

The benefit of bracing has been observed in other studies. Zhu et al. found bracing a predictor of curve improvement in 54 patients [39]. Sha et al. reported that 8/33 (24%) of patients treated with bracing required spinal fusion compared to 13/21 (43%) without [13]. Objective brace compliance monitors were not available during the duration of this study and limits the conclusions that can be drawn from the effect of bracing. However, it is the senior authors' belief that bracing is indicated in patients with residual curves > 25° following decompression and all patients should be followed up until skeletal maturity to detect late curve progression.

The retrospective nature is the largest limitation. Another limitation is the lack of follow-up until skeletal maturity in 13 patients. Late curve progression was seen after a mean of 6 years and studies with shorter follow-up are susceptible to underreporting the true incidence of curve progression and fusion. Another limitation difficult to overcome is the lack of a control group. It is the senior authors' belief that all children should undergo decompression to prevent further progression and neurologic deterioration if an association is suspected between the CM-I and SM. A final limitation is that this study was conducted at a tertiary care center and may not be generalizable.

The biggest merit of this study is that it is the largest series of children with CM-I, SM, and scoliosis that evaluates the long-term outcomes of decompression on scoliosis. A SM scale allowed for a quantifiable method to analyze changes in SM size and correlate PFD to scoliosis outcomes. A reduction in syringomyelia following decompression is associated with improvement or stabilization of scoliosis in two-thirds of patients. The long-term prognosis is determined by the absolute reduction in SM size and the extent of curve improvement during the first year after decompression. Children that underwent decompression at a younger age experienced higher rates of curve improvement. However, scoliosis improvement can occur regardless of age and decompression is recommended in children of all ages.

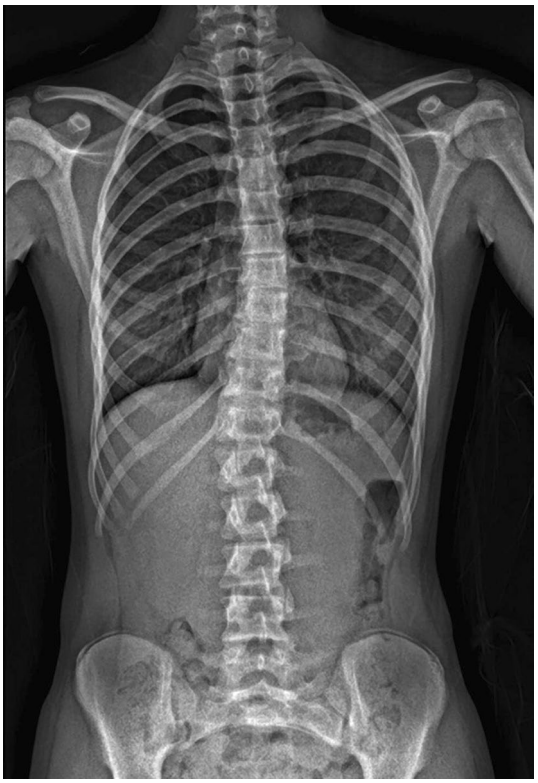

Fig. 8 Case 2 continued: 84-month post-decompression. The patient is now 12 years and the curve has reduced further to 12°. Mild vertebral rotation is seen at the thoracolumbar junction and lumbar spine

Key points

- A mean 70% decrease in syringomyelia scaled size was seen after neurosurgical decompression.
- Scoliosis severity at presentation, smaller reductions in syringomyelia scaled size, less curve improvement in the first year following decompression, and double or triple major curves were risk factors for spinal fusion.
- Spinal fusion was performed more frequently in children that underwent decompression at an older age.
- Neurosurgical decompression is recommended in children of all ages and has potential to improve CM-I, SM, and scoliosis.

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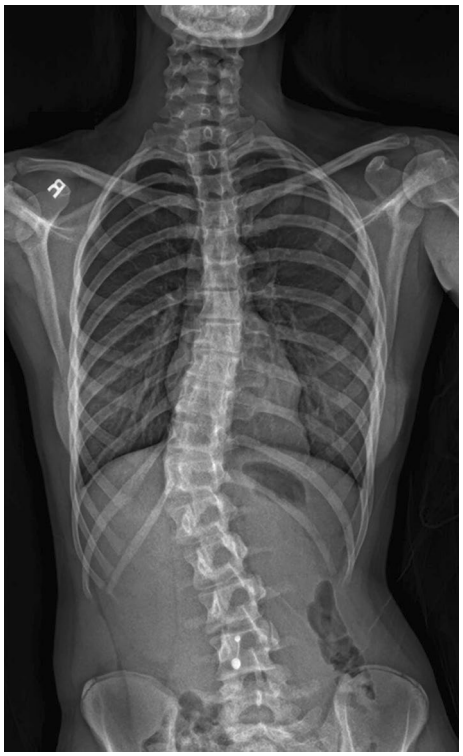

Fig. 9 Case 2 continued: 100-month post-decompression. The patient is now 14 years and a delayed progressive curve of 31° is seen in the opposite direction

Compliance with ethical standards

Ethical approval IRB approval: R00023640-1.

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