Published in final edited form as:

Spine J. 2015 September 1; 15(9): 2002–2008. doi:10.1016/j.spinee.2015.04.048.

Risks and Outcomes of Spinal Deformity Surgery in Chiari Malformation with Syringomyelia versus Adolescent Idiopathic Scoliosis

Jakub Godzik, MD¹, Terrence F. Holekamp, MD, PhD², David D. Limbrick, MD, PhD^{2,3}, Lawrence G. Lenke, MD¹, TS Park, MD^{2,3}, Wilson Z. Ray, MD², Keith Bridwell, MD¹, and Michael P. Kelly, MD^{1,†}

¹Department of Orthopaedic Surgery, Washington University School of Medicine, St. Louis, MO

²Department of Neurological Surgery, Washington University School of Medicine, St. Louis, MO

³St. Louis Children's Hospital, St. Louis, MO

Abstract

Background Context—Chiari Malformation, Type 1, with syringomyelia (CIM+SM) is often associated with spinal deformity. The safety of scoliosis surgery this population is controversial, and has never been directly compared with adolescent idiopathic scoliosis (AIS). Purpose: Compare the safety and subjective outcomes of spinal deformity surgery between patients with Chiari Malformation I associated scoliosis and a matched Adolescent Idiopathic Scoliosis cohort.

Study Design—Retrospective matched cohort analysis

Patient Sample—Patients with CIM+SM and treated with spinal fusion for spinal deformity were identified in the surgical records of a single institution and were matched, 1:1, with AIS patients undergoing spinal fusion at the same institution.

Outcome Measures—Neurological monitoring data quality and integrity, radiographic parameters, Scoliosis Research Society-22 Scores.

Methods—A clinical database was reviewed for patients undergoing spinal reconstruction for CIM+SM associated spinal deformity at our institution from 2000 to 2012. Thirty-six CIM+SM patients were identified and matched to an AIS cohort (1:1) based on age, gender, major curve magnitude, fusion length, and revision status. Demographics, deformity morphology, surgical details, neuromonitoring data, and pre- and postoperative SRS-22 scores were recorded at a minimum 2-year followup. Changes in SRS-22 scores were compared within and between groups. Complications and neurological monitoring data issues were compared between groups.

[†]Corresponding Author and Address: Michael P. Kelly, MD, Assistant Professor, Department of Orthopaedic Surgery, Washington University School of Medicine, 660 South Euclid Avenue, Campus Box 8233, St Louis, Missouri 63110, kellymi@wudosis.wustl.edu, Tel: (314) 747-2535; Fax: (314) 747-2599.

Publisher's Disclaimer: This is a PDF file of an unedited manuscript that has been accepted for publication. As a service to our customers we are providing this early version of the manuscript. The manuscript will undergo copyediting, typesetting, and review of the resulting proof before it is published in its final citable form. Please note that during the production process errors may be discovered which could affect the content, and all legal disclaimers that apply to the journal pertain.

Results—Mean age was 14.5 ± 5 years (CIM+SM: 14.6 ± 5 ; AIS: 14.4 ± 5), and 42% of patients were male. Preoperative mean major coronal Cobb measured $58^\circ \pm 25$ vs. $57^\circ \pm 17$ ($p=0.84$) with mean kyphosis $52^\circ \pm 17$ vs. $41^\circ \pm 20$ ($p=0.018$). An average of 10.4 ± 2.6 vertebral levels were fused (10.4 ± 2.8 vs. 10.4 ± 2.3 , $p=0.928$). No differences existed in surgical approach ($p=0.336$), estimated blood loss ($680\text{mL} \pm 720$ vs. 660 ± 310 , $p=0.845$), or duration of surgery ($6.0 \text{ hours} \pm 2.2$ vs. 5.6 ± 2 , $p=0.434$). Complication rate was comparable between the two groups (33% vs. 14%, $p=0.052$). CIM+SM experienced more neurological complications (11% vs. 0%, $p=0.04$) and neuromonitoring difficulties (28% vs. 3%, $p=0.007$) than the AIS cohort. Mean curve correction was comparable at 2 years (58% CIM+SM vs. 64% AIS, $p=0.2$). At follow-up, both CIM+SM and AIS groups demonstrated improved cumulative SRS-22 outcome subscores (CIM+SM: +0.4, $p=0.027$; AIS: +0.3, $p<0.001$). No difference in outcome subscores existed between CIM+SM and AIS groups.

Conclusions—While CIM+SM patients undergoing spine reconstruction can expect similar deformity corrections and outcomes scores to AIS patients, they also experience higher rates of neuromonitoring difficulties and neurological complications related to surgery. Surgeons should be prepared for these difficulties, particularly in children with larger syrinx size.

INTRODUCTION

Chiari Type I Malformation, a developmental abnormality of the hindbrain, is frequently associated with syringomyelia and scoliosis.[1, 2] The prevalence of scoliosis in patients with Chiari Malformation and syringomyelia (CIM+SM) approaches 80% in some studies, with up to half of patients requiring spinal fusion despite neurosurgical intervention and nonoperative management.[1, 3-6] Previous authors have suggested that risk factors for curve progression and spinal fusion include older age, the location of spinal deformity, extent of syrinx resolution, and degree of initial scoliosis.[4, 7-11]

The safety posterior spinal fusion and deformity correction in CIM+SM remains controversial and the outcomes are not well described.[4, 5, 12-15] While early reports and series found a high rate of complication associated with spinal deformity surgery in CIM+SM patients, recent studies have reported few instances of neurological deficit.[3, 13, 16] Although most surgeons would advocate decompression of the Chiari I malformation and syringomyelia to promote curve resolution and reduce the risk of neurologic complication, some authors have suggested otherwise.[6, 14, 17] To date, no study has directly compared the safety and effectiveness of spinal fusion in CIM+SM with adolescent idiopathic scoliosis (AIS).

In the current study, we sought to compare the safety and subjective outcomes of spinal deformity surgery between patients with CIM+SM associated scoliosis and a matched AIS cohort. Our hypothesis was that patients with CIM+SM associated spinal deformity would experience higher rates of neurological deficits with equivalent outcomes at a minimum of two years followup.

MATERIAL AND METHODS

Patient Population

After Institutional Review Board approval, patients who had undergone spinal deformity surgery for either CIM+SM associated spinal deformity or Adolescent Idiopathic Scoliosis (AIS) between 2000 and 2012 were identified in an operative database at a single institution. We identified forty-one patients who underwent instrumented fusion between 2000 and 2012, of whom thirty-six (87%) had a minimum of 2-year follow-up. All CIM+SM patients were matched one-to-one with adolescent idiopathic scoliosis (AIS) patients undergoing instrumented fusion. Matching characteristics included: age (within 1 year), gender, major curve magnitude (Cobb angle within 10°), fusion length, and revision status. Those patients with kyphosing scoliosis were matched on the Cobb angle magnitude of both sagittal and coronal curves. In the event of multiple matches, a random number generator was used to select the appropriate match.

Patient data were extracted from the medical records, surgical history was reviewed for outcomes and complications of past neurosurgical procedures. Surgical data for the index surgery included surgical approach (anterior, posterior, combined), staging (single, staged), levels of instrumentation, estimated blood loss (EBL), operative time, and use of posterior column osteotomies (PCO). Complications within the follow-up period were categorized as major or minor as per Glassman et al.;[18] new postoperative neurological deficits were noted as neurological complications.

Intraoperative Monitoring

Intraoperative monitoring data were collected from an institutional neurological monitoring database. Monitoring modalities included somatosensory-evoked potentials (SSEP), motor-evoked potentials (NMEP), descending neurogenic evoked potentials (DNEP), triggered electromyography (T-EMG), and spontaneous electromyography (S-EMG). Absence of baseline signal or an intraoperative loss of signal below threshold were categorized as neuromonitoring difficulty.

Radiographic Review

Preoperative and postoperative (2-year) radiographic studies were reviewed by 2 co-authors blinded to group assignment (CIM+SM vs. AIS). Standard radiographic measurements were taken, including Cobb angle for major coronal curve, coronal alignment, thoracic kyphosis, sagittal vertical axis alignment by C7 plumb line, lumbar lordosis, sacral slope, pelvic incidence, and pelvic tilt. [19, 20]

Magnetic resonance (MR) imaging of the spine and craniocervical junction prior to spinal fusion was reviewed; when available, imaging prior to posterior fossa decompression (PFD) was also reviewed. The syrinx size was expressed as the maximum anteroposterior diameter (mm) of the syrinx cavity and the number of vertebral levels traversed.

Outcomes

Where available, health-related quality-of-life measures as obtained by the Scoliosis Research Society Questionnaire (SRS-22, -24, -29, or -30), were reviewed at preoperative and at latest postoperative evaluation.

Analysis

Data were reported using mean and standard deviation. Analysis of continuous and dichotomous variables was conducted using the independent t-test and chi-square test, respectively. Statistical analysis of paired data was performed using Wilcoxon signed rank test of non-parametric data. Statistical significance was set at a threshold of $p < 0.05$. Statistical analysis was performed using SPSS v.21 (SPSS, Inc., Chicago, IL).

RESULTS

Patients

Thirty-six CIM+SM patients were matched to thirty-six AIS patients with a minimum of two-year follow-up. Mean age was 14.4 ± 5.5 years (CIM+SM: 14.6 ± 5 ; AIS: 14.4 ± 5 , $p = 0.84$), and 42% (30/72) of patients were male. Table 1 demonstrates a comparison of initial clinical and radiographic presentation between the CIM+SM and AIS cohorts. Preoperative coronal deformities were similar. The CIM+SM group had significantly more thoracic kyphosis ($52^\circ \pm 17$ vs. $41^\circ \pm 20$, $p = 0.018$), without a significant difference in preoperative sagittal alignment (C7 Plumb: $2.5\text{mm} \pm 34$ vs. $-9\text{mm} \pm 40$, $p = 0.248$). There were no significant differences between groups BMI, number of instrumented levels, surgical approach, number of posterior column osteotomies, and mean follow-up. There were significant differences between the two groups in preoperative neurological abnormalities (12/36 or 33% in CIM+SM vs. 2/36 or 6% in AIS, $p = 0.003$), and preoperative SRS-22 scores in the function subdomain (3.5 for CIM+SM vs. 4.2 for AIS, $p = 0.02$). Neurological abnormalities in CIM+SM included long-tract signs (N=4), paresthesia (N=3), asymmetric reflexes (N=2), lower extremity weakness (N=1), and incontinence (N=1); abnormalities in the AIS cohort included minor lower extremity weakness (N=2).

Syringomyelia: Neurosurgical Management

Ninety-two percent (33/36) of patients with CIM+SM had prior neurosurgical intervention for syringomyelia; 32/26 (89%) had posterior fossa decompression (PFD) prior to spinal deformity surgery; 1 patient had undergone syringopleural shunting (direct decompression of syringomyelia). The mean age at syrinx diagnosis was 9.84 ± 4.2 years; mean age at PFD was 10.3 ± 4.5 years, with a mean delay of 3 ± 3.4 years before spinal surgery. Mean pre-PFD tonsillar herniation was $11.5 \pm 0.5\text{mm}$, average maximum AP syrinx diameter $7.9 \pm 3\text{mm}$, and syrinx length 14 ± 4 vertebral levels. After decompression, syrinx width decreased an average of 52% in diameter (9.0 mm to 4.3mm, $p < 0.001$), with 68% of patients demonstrating a syrinx reduction more than 50% in maximum diameter. Syrinx length decreased on average by two vertebral levels to 11.4 ± 5 vertebral levels. Prior decompression or reduction in syrinx diameter was unrelated to presence of neurological deficits at the time of scoliosis surgery.

Twenty-two patients had available AP/lateral spine radiographs prior to neurosurgical decompression for comparison, with an average pre-PFD major Cobb angle of $44 \pm 17.7^\circ$. All spinal deformities were initially treated non-operatively with a variety of techniques, according to physician and patient preferences. Following decompression, the major curve progressed in magnitude in 14 (63%) patients, to an average of $57 \pm 18.4^\circ$ (30%, $p < 0.001$) within a period of 28 ± 22 months. Change in syrinx size was not associated with extent of curve progression ($p = 0.686$). The mean syrinx diameter immediately prior to spinal deformity surgery of the entire CIM+SM cohort was 4.4 ± 2.4 mm. Surgical indications included significant curve progression following neurosurgical decompression and curve size greater than 50 degrees using traditional Cobb technique.

Scoliosis Surgery: Surgical Details

All patients underwent fusion with instrumentation. Fifty patients (50/72, 69%) underwent PSF, seventeen patients (17/72, 24%) underwent staged ASF and PSF procedures, and five patients (5/72, 7%) underwent ASF alone with an average fusion length of 10 ± 3 levels (Table 3). There were no significant differences between the two groups in surgical approach ($p = 0.213$), proportion of patients undergoing thoracoplasty (2/36 or 6% for CIM+SM vs. 2/36 or 6% for AIS, $p = 1.0$), nor number of instrumented levels (10.4 ± 3 levels for CIM+SM vs. 10.4 ± 2 levels for AIS, $p = 0.928$). Operative times were not significantly different between groups (6 ± 2.2 hours for CIM+SM vs. 5.6 ± 2 hours for AIS, $p = 0.434$) nor was total estimated blood loss (680 ± 720 mL for CIM+SM vs. 660 ± 310 mL for AIS, $p = 0.845$).

Complications

The overall rate of complications in the CIM+SM group was 33% (12/36), compared to 14% (5/26) in the AIS cohort ($p = 0.052$) (Table 2). The CIM+SM cohort experienced more neurological complications in the perioperative period (4/36 or 11% for CIM+SM vs. 0/36 or 0% for AIS, $p = 0.04$): 2 cases of transient lower extremity single nerve root deficits (<6 months); 1 case of permanent nerve root deficit (L4/L5); 1 spinal cord deficit (ASIA C) requiring reoperation and prolonged length of stay. Patients experiencing new neurological deficits were more likely to have undergone PCOs (50% vs. 10%, $p = 0.027$), with greater extent of kyphosis correction (49% vs. 12%, $p = 0.054$; 27 degrees vs 8 degrees, $p = 0.049$). Patients sustaining neurological deficits also experienced greater EBL (1800 ± 1400 mL vs. 500 ± 400 mL, $p < 0.001$) and longer surgical times (8.6 ± 2.3 vs. 5.6 ± 1.9 hours, $p = 0.027$). Development of new neurological deficit was not associated with patient age ($p = 0.235$), history of chiari decompression ($p = 0.466$), duration between decompression and scoliosis surgery ($p = 0.33$), extent of syrinx reduction ($p = 0.395$), preoperative syrinx diameter ($p = 0.794$), or presence of preoperative neurological deficit ($p = 1.0$).

There were four reoperations in the CIM+SM cohort (4/36 or 11%). All reoperation were late (>3 months after surgery) for pseudoarthrosis (N=3) and progressive deformity (N=1). In contrast, only one patient in the AIS cohort required reoperation at 10 years after initial fusion for coronal and sagittal curve decompensation (1/36 or 3%, $p = 0.164$).

Neurological Monitoring

Neuromonitoring difficulties (lack of baseline or loss of signal) were more frequent in the CIM+SM cohort (CIM+SM: 10/36(28%), AIS:1/34(3%), $p = 0.007$). The CIM+SM group had an 11% (4/36) rate of false positives compared to 4% in the AIS cohort ($p = 0.358$). Neuromonitoring difficulties were associated with larger syrinx diameter (10 mm vs. 7 mm, $p = 0.013$) and greater syrinx length (17 vs. 13 levels, $p = 0.006$). CIM+SM patients that did not undergo PFD prior to spinal correction did not have a higher rate of neuromonitoring difficulties ($p = 0.293$). Patients experiencing neuromonitoring difficulties demonstrated a 38% rate of neurologic deficit compared to 2% in those without neuromonitoring difficulty ($p = 0.01$).

Radiographic Results

Radiographic results are reported in Table 2. Mean curve correction was comparable between the CIM+SM and AIS group at 2 years (58% CIM+SM vs. 64% AIS, $p = 0.20$). For the CIM+SM group, the mean preoperative coronal Cobb decreased from 58 degrees to 24 degrees at 2-year follow up (mean 58% correction, $p = 0.001$); for the AIS group, the mean preoperative coronal Cobb decreased from 57 degrees to 22 degrees (mean 64% correction, $p < 0.001$). Mean thoracic kyphosis decreased from 52 degrees preoperatively to 41 degrees in the CIM+SM group and was unchanged in the AIS group ($p = 0.015$). No difference was observed between postoperative C7 coronal alignment (12.5 mm CIM+SM vs. 10.5 mm AIS, $p = 0.405$) or between postoperative C7 sagittal alignment (-6.7 mm CIM+SM vs. -7.1 mm AIS, $p = 0.972$).

Outcomes

Patient outcomes scores were available in 22/36 (61%) of the CIM+SM cohort and 23/36 (63%) of the AIS cohort (Table 4). Both groups experienced improved SRS scores at final follow-up (+0.4 for CIM+SM, $p = 0.027$; +0.3 for AIS, $p < 0.001$). The AIS group had greatest benefit in the self-image (+0.74, $p < 0.001$) and satisfaction subdomains (+0.76, $p = 0.001$), while the CIM+SM group had greatest improvement in self-image (+0.65, $p = 0.013$) and function (+0.6, $p = 0.021$). When directly compared, the CIM+SM group had greater improvement than AIS in the function domain (+0.6 vs. 0, $p = 0.05$), satisfaction (+0.9 vs. +0.8, $p = 0.026$). In the CIM+SM cohort, postoperative sagittal alignment was moderately correlated with function ($r = 0.47$, $p = 0.025$), pain ($r = 0.47$, $p = 0.025$), and cumulative SRS-22 ($r = 0.43$, $p = 0.043$) at last follow-up. Neither postoperative Cobb angle ($r = -0.39$, $p = 0.07$) nor postoperative kyphosis ($r = -0.37$, $p = 0.084$) were correlated with improvement in SRS-22 outcome scores.

DISCUSSION

The purpose of this study was to compare the safety and outcomes of spinal deformity surgery in Chiari Malformation I with associated syringomyelia (CIM+SM) to patients with Adolescent Idiopathic Scoliosis (AIS). We found a higher rate of neurological deficits (4/36), despite prior Chiari decompression and concurrent neuromonitoring. Difficulties obtaining stable, baseline monitoring data were more common in the CIM+SM group as well. (CIM+SM: 28% (10/36), AIS:3% (1/34), $p = 0.007$). These higher rates of monitoring

difficulties were associated with larger syrinx diameter, but unrelated to physical exam findings. CIM+SM and AIS patients demonstrated similarly improved SRS-22 scores; data not previously reported. This reinforces the benefits of surgery in this disease, in the setting of an increased risk of neurological deficit.

Two recent, small series reported no neurological monitoring difficulties and no new neurologic deficits in CIM+SM spinal deformity surgeries.[14, 21] In our cohort, new neurological deficits occurred in 11% (4/36) of the CIM+SM cohort, compared to none in the AIS cohort. This rate is higher than the published rate (1.3%) of new neurological deficits in the Scoliosis Research Society morbidity and mortality database and suggests that Chiari-associated deformities pose a higher risk of new neurological deficits than patients with idiopathic deformities.[22] Xie *et al* reported a series of all vertebral column shortening procedures and suggested that this carried less neurological risk as it takes tension off the spinal cord.[21] In our series, a number of patients were treated with PCOs, which shorten the posterior column but may lengthen the anterior column. This may increase tension within the spinal cord. In conjunction with hydrostatic pressures within the syrinx, lengthening the anterior column may provide an additive risk, suggesting that PCO be avoided in this patient population. This is supported by our finding that more aggressive kyphosis correction was associated with neurological deficits. Reoperation due to curve progression was uncommon, similar to the findings of Bradley *et al*, and careful selection of fusion levels is common to all scoliosis surgeries.[14]

Given the frequency of neurological monitoring difficulties and new neurological deficit, surgeons treating CIM+SM deformities should be prepared for these problems. Patients must be taught rehearsed Stanara wakeup tests and surgeons should be prepared to encounter neuromonitoring data changes during surgery and to make appropriate corrective maneuvers. Future research will help elucidate what the true risk factors for new neurological deficit are. We have found that aggressive correction of kyphosis, via PCO, may lead to a higher risk. Xie *et al* recommended vertebral column resection, perhaps the most dangerous of spine procedures, for management of these deformities. Given the vast difference in approaches, more data are needed to provide quality data for informed decision making on behalf of the patient. The majority of patients in our series experienced progression of the deformity after decompression of the Chiari malformation and the potential benefit of decompression needs further study.

The study is limited by the retrospective design, small sample size, and is likely underpowered to detect differences in rare events. As with any retrospective study, data are limited by the accuracy and availability of medical records. Our reported complication rates are higher than previous retrospective studies investigating this pathology, however. Matching was performed using a computer algorithm in an attempt to limit selection bias and allowing for randomization of controls in the event of multiple matches. In order to maximize the number of patients included in this study, matching on basis of radiographic criteria was limited to major curve magnitude; as a result of not matching across two planes, the CIM+SM group demonstrated slightly greater average preoperative kyphosis (52° vs. 41°). However, we must acknowledge that the difference in neurological deficits is influenced by this difference and not by the intraspinal pathology. Given the small

difference in average kyphosis, we feel this is an unlikely situation. Lastly, a number of patients had missing preoperative quality-of-life data, limiting the conclusions regarding outcomes data and we may overestimate the true benefits of surgery. As spinal deformities and associated CIM remain relatively uncommon, we require a large number of patients to reach more definitive conclusions regarding the true safety profile and patient outcomes following spinal surgery. Future research in this regard must come in the form of a prospective, multicenter effort to adequately power comparisons and capture accurate data points.

CONCLUSION

Chiari Malformation patients undergoing spinal deformity surgery demonstrate improvement in subjective outcome scores comparable to a cohort of idiopathic spinal deformity patients after a minimum 2-year follow-up. However, surgical management of spinal deformity in patients with underlying Chiari Malformation and syringomyelia carries a higher risk of new neurological deficit, despite adequate neurosurgical decompression and intraoperative neuromonitoring. The association of kyphosis correction and posterior column osteotomy with neurological deficits may warrant consideration and less aggressive correction of preoperative kyphosis.

References

1. Tubbs R, Beckman J, Naftel R, Chern J. Institutional experience with 500 cases of surgically treated pediatric Chiari malformation Type I. *J Neurosurg Pediatr.* 2011; 7:248–56. [PubMed: 21361762]
2. Hankinson T, Tubbs RS, Wellons JC. Duraplasty or not? An evidence-based review of the pediatric Chiari I malformation. *Childs Nerv Syst.* 2011; 27:35–40. [PubMed: 20890606]
3. Yeom JS, Lee C-K, Park K-W, et al. Scoliosis associated with syringomyelia: analysis of MRI and curve progression. *Eur Spine J.* 2007; 16:1629–35. [PubMed: 17701226]
4. Eule JM, Erickson Ma, O'Brien MF, Handler M. Chiari I malformation associated with syringomyelia and scoliosis: a twenty-year review of surgical and nonsurgical treatment in a pediatric population. *Spine.* 2002; 27:1451–5. [PubMed: 12131745]
5. Phillips LH, Blanco JS, Sussman MD. Direct spinal stimulation for intraoperative monitoring during scoliosis surgery. *Muscle & nerve.* 1995; 18:319–25. [PubMed: 7870110]
6. Farley, Fa; Puryear, A.; Hall, JM.; Muraszko, K. Curve progression in scoliosis associated with Chiari I malformation following suboccipital decompression. *Journal of spinal disorders & techniques.* 2002; 15:410–4. [PubMed: 12394666]
7. Attenello FJ, McGirt MJ, Garcés-Ambrossi GL, Chaichana KL, Carson B, Jallo GI. Suboccipital decompression for Chiari I malformation: outcome comparison of duraplasty with expanded polytetrafluoroethylene dural substitute versus pericranial autograft. *Childs Nerv Syst.* 2009; 25:183–90. [PubMed: 18769929]
8. Bhangoo R, Sgouros S. Scoliosis in children with Chiari I-related syringomyelia. *Childs Nerv Syst.* 2006; 22:1154–7. [PubMed: 16541292]
9. Hwang SW, Samdani AF, Jea A, et al. Outcomes of Chiari I-associated scoliosis after intervention: a meta-analysis of the pediatric literature. *Childs Nerv Syst.* 2012
10. Ozerdemoglu, Ra; Transfeldt, EE.; Denis, F. Value of treating primary causes of syrinx in scoliosis associated with syringomyelia. *Spine.* 2003; 28:806–14. [PubMed: 12698125]
11. Sengupta DK, Dorgan J, Findlay GF. Can hindbrain decompression for syringomyelia lead to regression of scoliosis? *Eur Spine J.* 2000; 9:198–201. [PubMed: 10905436]
12. Huebert HT, MacKinnon WB. Syringomyelia and scoliosis. *The Journal of bone and joint surgery British volume.* 1969; 51:338–43. [PubMed: 5770414]

13. Zhang H-Q, Deng A, Liu S-H, et al. Adult thoracolumbar or lumbar scoliosis with Chiari malformation and syringomyelia: a retrospective study of correction and fusion strategies. *Archives of orthopaedic and trauma surgery*. 2011; 131:475–80. [PubMed: 20632021]
14. Bradley LJ, Ratahi ED, Crawford Ha, Barnes MJ. The outcomes of scoliosis surgery in patients with syringomyelia. *Spine*. 2007; 32:2327–33. [PubMed: 17906574]
15. Ferguson RL, Devine J, Stasikelis P, Caskey P, Allen BL. Outcomes in Surgical Treatment of “Idiopathic-Like” Scoliosis Associated With Syringomyelia. *Journal of spinal disorders & techniques*. 2002; 15:301–6. [PubMed: 12177546]
16. Xie J, Wang Y, Zhao Z, et al. One-stage and posterior approach for correction of moderate to severe scoliosis in adolescents associated with Chiari I malformation: is a prior suboccipital decompression always necessary? *Eur Spine J*. 2011; 20:1106–13. [PubMed: 21399931]
17. Charray O, Koop S, Winter R, Lonstein J, Denis F, Bailey W. Syringomyelia and scoliosis: a review of twenty-five pediatric patients. *J Pediatr Orthop*. 1994; 14:309–17. [PubMed: 8006160]
18. Glassman S, Berven S, Bridwell K, Horton W, Dimar J. Correlation of Radiographic Parameters and Clinical Symptoms in Adult Scoliosis. *Spine (Phila Pa 1976)*. 2005; 30:682–8. [PubMed: 15770185]
19. Ames C, Smith J, Scheer J, et al. Impact of spinopelvic alignment on decision making in deformity surgery in adults. *J Neurosurg Spine*. 2012; 16:547–64. [PubMed: 22443546]
20. Zhu Z, Sha S, Liu Z, et al. Sagittal spinopelvic alignment in adolescent thoracic scoliosis secondary to Chiari I malformation: a comparison between the left and the right curves. *Eur Spine J*. 2013
21. Xie J-M, Zhang Y, Wang Y-S, et al. The risk factors of neurologic deficits of one-stage posterior vertebral column resection for patients with severe and rigid spinal deformities. *Eur Spine J*. 2013
22. Hamilton DK, Smith JS, Sansur Ca, et al. Rates of new neurological deficit associated with spine surgery based on 108,419 procedures: a report of the scoliosis research society morbidity and mortality committee. *Spine*. 2011; 36:1218–28. [PubMed: 21217448]

Table 1

Comparison of Initial Clinical and Radiographic Data: CIM+SM and AIS Cohorts

	CIM+SM	AIS	p-value [*]
Demographic Data			
Mean Age $\pm$ SD (yrs)	14.5 $\pm$ 5	14.4 $\pm$ 5	0.84
No. Females (%)	21 (58%)	21 (58%)	1
BMI (kg/m ²)	22.8	20.4	0.1
Preoperative Neurological Abnormality	12 (33%)	2 (6%)	<0.001
Preoperative Radiographic Data			
Coronal Cobb Angle $\pm$ SD	58 $\pm$ 25°	57 $\pm$ 17°	0.84
No. Apex Left Deformities	10 (28%)	5 (14%)	0.146
Thoracic Kyphosis $\pm$ SD	52 $\pm$ 17°	41 $\pm$ 20°	0.018
Sagittal C7 Plumb $\pm$ SD (mm)	2.5 $\pm$ 34	-9 $\pm$ 40	0.248

* Independent t-test for continuous variables; Chi-Square test for categorical variables Chiari Malformation with Syringomyelia = CIM+SM; Adolescent Idiopathic Scoliosis = AIS

Table 2

Complications by Group

	CIM+SM	AIS
Complication		
Major		
Nerve Root Deficit	3	0
Spinal Cord Deficit	1	0
Minor		
Excessive Bleeding	2	1
Pneumothorax	1	3
Superficial Infection	1	0
Pseudarthrosis	3	0
Instrumentation Failure	1	1
Total	12	5

Table 3

Clinical and Radiographic Results: CIM+SM and AIS

	CIM+SM	AIS	p-value [*]
Surgical Approach			
Posterior (%)	23 (64%)	27 (75%)	0.336
Anterior (%)	4 (11%)	1 (3%)	
Combined (%)	9 (25%)	8 (22%)	
Instrumented Levels (±SD)	10.4±2.8	10.4±2.3	0.928
Osteotomy (%)	5 (14%)	5 (14%)	1.00
Operative Time (±SD, hrs)	6.0±2.2	5.6±2.0	0.434
Estimated Blood Loss (±SD, mL)	680±720	660±310	0.845
Postoperative Radiographic Data			
Coronal Cobb Angle ± SD	24 ± 13°	22 ± 13°	0.445
Percent Correction	58%	64%	0.2 ^{**}
Thoracic Kyphosis ± SD	41 ± 15°	38 ± 17°	0.396
Percent Correction	17%	0%	0.091
Sagittal C7 Plumb ± SD (mm)	-6.7±53	-7.1±36	0.972
Total Complications (%)	12 (36%)	5 (14%)	0.052
Neurological Complications	4(11%)	0(0%)	0.04
Reoperations	4(11%)	1(3%)	0.164

* Independent t-test for continuous variables; Chi-square test for categorical variables

** Mann-Whitney Test

Chiari Malformation with Syringomyelia = CIM+SM; Adolescent Idiopathic Scoliosis = AIS

Table 4

Scoliosis Research Society-22 Scores.

	CIM+SM			AIS		
	Preoperative	Postoperative	p-value*	Preoperative	Postoperative	p-value**
Pain	3.9	3.8	0.571	4.2	4.3	0.753
Self-Image	3.6	4.2	0.013	3.5	4.2	0.826
Function	3.6	4.2	0.021	4.2	4.2	0.05
Satisfaction	3.2	4.1	0.067	3.7	4.5	0.001
Mental	4.1	4.4	0.125	4.1	4.3	0.866
Cumulative	3.7	4.1	0.027	4.0	4.3	<0.001

* Wilcoxon Signed Rank test;

** Wilcoxon Signed Rank test between groups