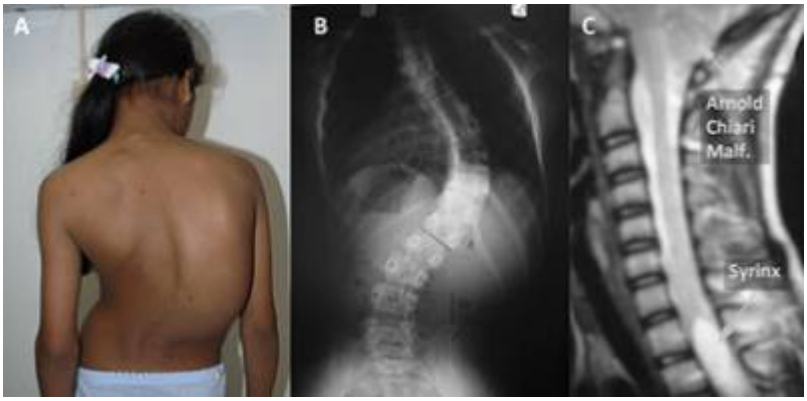
Chiari related scoliosis



Spinal deformity is an important clinical manifestation of Chiari type 1 deformity and syringomyelia.

Epidemiology

The prevalence of scoliosis in patients with Chiari Malformation and syringomyelia (CIM+SM) approaches 80% in some studies [1) 2) 3) 4) 5)].

Risk factors

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 [6) 7) 8) 9) 10) 11)].

Syrinx characteristics, but not tonsil position, were related to the presence of scoliosis in patients with CM-I, and there was an independent association of syrinx length and holocord syrinx with scoliosis. Further study is needed to evaluate the nature of the relationship between syrinx and scoliosis in patients with CM-I [12)].

Diagnosis

A challenge for physicians who see children with scoliosis is deciding when an MRI is warranted to look for neurological problems such as Chiari. Since scoliosis is not uncommon among adolescents, and because only a small percentage of those cases are actually related to Chiari, ordering an MRI for every child with scoliosis is not practical. In several studies, researchers have tried to find unique characteristics of Chiari related scoliosis which can alert doctors to when an MRI should be performed. Based on this work, some doctors recommend that Chiari should be checked for if there are any neurological signs and/or severe curves. Others have tried to focus on curve patterns that aren't typically seen, for example certain types of double curves.

Outcome

The safety posterior spinal fusion and deformity correction in CIM+SM remains controversial and the outcomes are not well described [13) 14) 15) 16) 17) 18)].

Up to half of patients require spinal fusion despite neurosurgical intervention and nonoperative management [19) 20) 21) 22) 23)].

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 [24)].

Case series

A large multicenter retrospective and prospective registry of pediatric patients with CM-I (tonsils ≥ 5 mm below the foramen magnum) and syrinx (≥ 3 mm in axial width) was reviewed for clinical and radiological characteristics of CM-I, syrinx, and scoliosis (coronal curve ≥ 10°).

Based on available imaging of patients with CM-I and syrinx, 260 of 825 patients (31%) had a clear diagnosis of scoliosis based on radiographs or coronal MRI. Forty-nine patients (5.9%) did not have scoliosis, and in 516 (63%) patients, a clear determination of the presence or absence of scoliosis could not be made. Comparison of patients with and those without a definite scoliosis diagnosis indicated that scoliosis was associated with wider syrinxes (8.7 vs 6.3 mm, OR 1.25, $p < 0.001$), longer syrinxes (10.3 vs 6.2 levels, OR 1.18, $p < 0.001$), syrinxes with their rostral extent located in the cervical spine (94% vs 80%, OR 3.91, $p = 0.001$), and holocord syrinxes (50% vs 16%, OR 5.61, $p < 0.001$). Multivariable regression analysis revealed syrinx length and the presence of holocord syrinx to be independent predictors of scoliosis in this patient cohort. Scoliosis was not associated with sex, age at CM-I diagnosis, tonsil position, pB-C2 distance (measured perpendicular distance from the ventral dura to a line drawn from the basion to the posterior-inferior aspect of C2), clivoaxial angle, or frontal-occipital horn ratio. Average curve magnitude was 29.9°, and 37.7% of patients had a left thoracic curve. Older age at CM-I or syrinx diagnosis ($p < 0.0001$) was associated with greater curve magnitude whereas there was no association between syrinx dimensions and curve magnitude.

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2018

Chotai et al. conducted a retrospective review at a single tertiary center for children undergoing Posterior fossa decompression (PFD) with untreated scoliosis, and identified 17 patients with complete follow-up data and imaging.

Overall, scoliosis improved in 7 (41.2%) patients, worsened in 9 (52.9%), and remained unchanged in 1 (5.9%) after PFD (mean follow-up of 7.8 ± 4.1 months). We found that 3 of the 8 (38%) children with

early-onset scoliosis eventually needed scoliosis corrective surgery, which was needed in 7 of the 9 (78%) patients with adolescent-onset scoliosis. In addition, only 1 patient (17%) with a preoperative scoliosis curve <35 degrees and 9 patients (82%) with a curve ≥35 degrees required surgery for scoliosis correction despite PFD (p = 0.018).

In certain patients, PFD for CM-I may lead to improvement or stabilization of scoliosis [26].

2017

Previous reports have addressed the short-term response of patients with Chiari-related scoliosis (CRS) to suboccipital decompression and duraplasty (SODD); however, the long-term behavior of the curve has not been well defined.

Ravindra et al. undertook a longitudinal study of a cohort of patients who underwent SODD for CRS to determine whether there are factors related to Chiari malformation (CM) that predict long-term scoliotic curve behavior and need for deformity correction. METHODS The authors retrospectively reviewed cases in which patients underwent SODD for CRS during a 14-year period at a single center. Clinical (age, sex, and associated disorders/syndromes) and radiographic (CM type, tonsillar descent, pBC2 line, clival-axial angle [CXA], syrinx length and level, and initial Cobb angle) information was evaluated to identify associations with the primary outcome: delayed thoracolumbar fusion for progressive scoliosis. RESULTS Twenty-eight patients were identified, but 4 were lost to follow-up and 1 underwent fusion within a year. Among the remaining 23 patients, 11 required fusion surgery at an average of 88.3 ± 15.4 months after SODD, including 7 (30%) who needed fusion more than 5 years after SODD. On univariate analysis, a lower CXA (131.5° ± 4.8° vs 146.5° ± 4.6°, p = 0.034), pBC2 > 9 mm (64% vs 25%, p = 0.06), and higher initial Cobb angle (35.1° ± 3.6° vs 22.8° ± 4.0°, p = 0.035) were associated with the need for thoracolumbar fusion. Multivariable modeling revealed that lower CXA was independently associated with a need for delayed thoracolumbar fusion (OR 1.12, p = 0.0128).

This investigation demonstrates the long-term outcome and natural history of CRS after SODD. The durability of the effect of SODD on CRS and curve behavior is poor, with late curve progression occurring in 30% of patients. Factors associated with CRS progression include an initial pBC2 > 9 mm, lower CXA, and higher Cobb angle. Lower CXA was an independent predictor of delayed thoracolumbar fusion. Further study is necessary on a larger cohort of patients to fully elucidate this relationship [27].

2016

Mackel et al. conducted a multicenter retrospective review of 44 patients, aged 18 years or younger, diagnosed with Chiari I malformation and scoliosis who underwent posterior fossa decompression from 2000 to 2010. The outcome of interest was the need for spinal fusion after decompression. RESULTS Overall, 18 patients (40%) underwent posterior fossa decompression alone, and 26 patients (60%) required a spinal fusion after the decompression. The mean Cobb angle at presentation and the proportion of patients with curves > 35° differed between the decompression-only and fusion cohorts (30.7° ± 11.8° vs 52.1° ± 26.3°, p = 0.002; 5 of 18 vs 17 of 26, p = 0.031). An odds ratio of 1.0625 favoring a need for fusion was established for each 1° of increase in Cobb angle (p = 0.012, OR 1.0625, 95% CI 1.0135-1.1138). Among the 14 patients older than 10 years of age with a primary Cobb angle exceeding 35°, 13 (93%) ultimately required fusion. Patients with at least 1 year of follow-

up whose curves progressed more 10° after decompression were younger than those without curve progression (6.1 ± 3.0 years vs 13.7 ± 3.2 years, p = 0.001, Mann-Whitney U-test). Left apical thoracic curves constituted a higher proportion of curves in the decompression-only group (8 of 16 vs 1 of 21, p = 0.002). CONCLUSIONS The need for fusion after posterior fossa decompression reflected the curve severity at clinical presentation. Patients presenting with curves measuring > 35°, as well as those greater than 10 years of age, may be at greater risk for requiring fusion after posterior fossa decompression, while patients less than 10 years of age may require routine monitoring for curve progression. Left apical thoracic curves may have a better response to Chiari malformation decompression [28].

2015

Strahle et al. sought to determine if there is an independent association between CM-I and scoliosis when controlling for syrinx status.

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

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

A syrinx was independently associated with scoliosis in a large pediatric population undergoing MRI. CM-I was not independently associated with scoliosis when controlling for age, sex, and syrinx status. Because CM-I is not independently associated with scoliosis, scoliosis should not necessarily be considered a symptom of low cerebellar tonsil position in patients without a syrinx [29].

2013

A retrospective study was conducted on 22 patients with CMS who received brace treatment of scoliosis after PFD. Forty-four age- and sex-matched patients with idiopathic scoliosis (IS) who were treated with bracing served as the control group. The bracing outcome was considered a failure if the curve worsened 6° or more; otherwise, the treatment was considered to be successful.

The age and Risser sign were similar between patients with CMS and IS at brace initiation. The initial curve magnitude of patients with CMS (mean, 32.9° ± 6.3°; range, 20°-45°) was marginally significantly larger than that of patients with IS (mean, 29.6° ± 6.4°; range, 20°-45°). Until the final follow-up, a 6° or more worsening of the major curve occurred in 8 patients with CMS (36%) and in 15 patients with IS (34%). Overall, 7 patients with CMS (32%) and 13 patients with IS (30%) underwent spinal fusion surgery. No significant differences were observed between the 2 groups in the surgery rates or the bracing success rates (P > 0.05). In patients with CMS, neither the performance of syringosubarachnoid shunting nor the extent of tonsillar descent correlated with the bracing

outcomes, whereas a double major curve pattern was found to be predictive for the failure of bracing.

Brace treatment subsequent to PFD is effective in preventing curve progression for 64% of patients with CMS, which is comparable with the rate that is observed in patients with IS. Double major curve pattern may be a risk factor in predicting treatment failure in patients with CMS [30].

Case reports

Tanaka et al. report the result of an 8-year follow-up of a 13-year-old girl with severe scoliosis associated with Chiari malformation and a large syringomyelia. The patient presented at the hospital at the age of 13 with a 68° scoliosis. Magnetic resonance imaging showed Chiari malformation and a large syringomyelia. Neurosurgical treatment involved foramen magnum decompression and partial C1 laminectomy, but the scoliosis still progressed.

They present the first case report of a rare course of scoliosis in a patient with CM-I and a large syringomyelia [31].

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