

Intraspinal Anomalies and Their Interplay with Spinal Deformity in Early Onset Scoliosis

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Abstract: Numerous articles have reported the presence of intraspinal anomalies in children with early onset scoliosis (EOS) and have found varying effect of neurosurgical intervention on the risk of spinal deformity progression. However, no review to date has performed a detailed analysis of the implications of intraspinal anomalies on scoliotic development and the implications of neurosurgical intervention on curve progression in children with EOS. This article reviewed the relevant information as it pertains to children < 10 years of age with scoliosis associated with tethered cord, syringomyelia, and Arnold-Chiari (Chiari) malformations. The type of spinal cord anomaly and its effect on scoliosis development and progression is reviewed as well as the identification of risk factors for curve progression following neurosurgical intervention.

Key Concepts:

- Intraspinal anomalies have an increased incidence in children and may influence the initiation and development of early onset scoliosis (EOS).
- Tethered cord release in EOS children may not provide significant benefit to prevent curve progression.
- Younger age with smaller presented coronal curve deformity is associated with a higher rate of curve resolution following posterior fossa decompression in Chiari malformations.
- Multicenter, prospective studies are needed to better understand associated spinal deformity behavior following neurosurgical intervention.

Introduction

Early onset scoliosis (EOS) has a well-documented association with intraspinal anomalies consisting of abnormalities of the spinal cord or brainstem to include Arnold-Chiari malformations, syringomyelia, and tethered cord.^{1,11,29,63} The incidence of associated

intraspinal anomalies in presumed idiopathic EOS has been reported to range from 5-21%^{11,14,24,29,44,45} with a generally accepted recommendation for neural axis magnetic resonance imaging (MRI) for children presenting with scoliosis before the age of 10 years to determine the presence of pathology and ascertain the

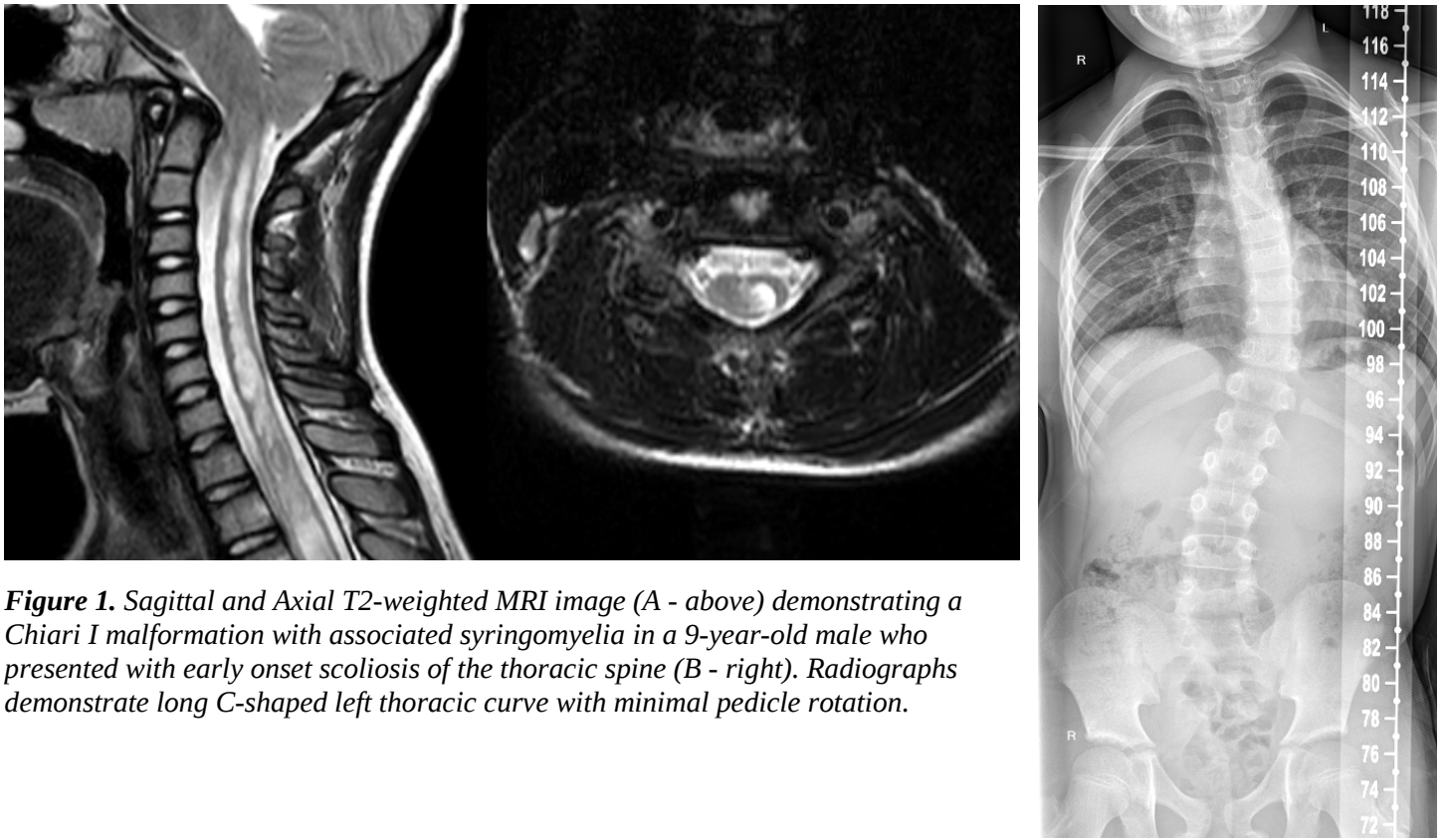


Figure 1. Sagittal and Axial T2-weighted MRI image (A - above) demonstrating a Chiari I malformation with associated syringomyelia in a 9-year-old male who presented with early onset scoliosis of the thoracic spine (B - right). Radiographs demonstrate long C-shaped left thoracic curve with minimal pedicle rotation.

need for neurosurgical intervention. Not only do children with EOS and concomitant intraspinal anomalies have a higher incidence of neuromonitoring changes with higher rates of neurologic complications,⁴¹ but there are also reports of curve resolution following neurosurgical intervention.²⁷ Several case series have reported the outcomes of neurosurgical intervention in children with EOS associated with various intraspinal anomalies, identifying several factors associated with curve progression and resolution.^{3,5-8,12,15,17,18,28,31,34,39,43,47,49,57}

This review summarizes the current literature detailing the implications of intraspinal anomalies on scoliotic development and the implications of neurosurgical intervention on curve progression in EOS. In particular, we aim to describe the relevant information in regard to tethered cord, syringomyelia, and Arnold-Chiari (Chiari) malformations in the treatment of EOS.

Epidemiology of Intraspinal Anomalies in EOS

The most common intraspinal anomalies encountered in EOS consist of syringomyelia, Chiari malformations, and tethered cord. The overall incidence of the various types of intraspinal anomalies range from 5-21% in children with EOS.^{11,14,24,29,44,45} However, the true incidence of intraspinal anomalies may not be known due to age selection bias in the reported literature which provides an arbitrary distinction for age at diagnosis to distinguish EOS and adolescent scoliosis.

Reviewing the literature, isolated syringomyelia account for 13.8-28.5% of intraspinal abnormalities, combined syringomyelia and Chiari malformation 19-62.5% (Figure 1), isolated Chiari malformation 0-45.7%, and tethered cord 0-43%.^{29,44,45,63} A less frequently seen condition in EOS is diastematomyelia, also referred to as split cord malformation, with an incidence ranging from 0-6.4%, with an apparent geographic predilection in

Southeast Asia.^{44,63} Heemskerk et al.²⁵ performed a systematic review of neural axis anomalies in patients of all ages with presumed idiopathic scoliosis, identifying an overall prevalence of 11.4%, with syringomyelia (3.7%), Chiari malformation (3%), and combined Chiari malformation with syringomyelia (2.5%) being the most common diagnoses. Risk factors for intraspinal anomalies included EOS, male gender, atypical curve patterns (i.e., left thoracic), increased thoracic kyphosis, and abnormal neurologic findings.

Clinical and Radiographic Evaluation for Suspected Intraspinal Anomalies

Identifying concerning factors for an underlying intraspinal anomaly in EOS is vital to ensure early identification and appropriate treatment; however, this can be challenging due to patient limitations and an inability to describe or articulate associated neurologic symptoms, especially in the infantile cohort. Dobbs et al.¹¹ suggested that the presence of a scoliotic magnitude ≥ 20 degrees in infantile patients, defined as 3 years or younger, should raise concern for an intraspinal anomaly. Additionally, the authors recommended obtaining a neural axis MRI in all infantile scoliosis patients due to the high rate (80%) of neurosurgical intervention in children with otherwise normal physical examinations. A retrospective review by Pahys et al.⁴⁴ supported these recommendations, despite a lower overall incidence of intraspinal anomalies (13%). Gupta et al.²⁴ expanded the age range for recommending MRI analysis to include juvenile patients, defined as age < 10 years, presenting with coronal deformity magnitudes > 20-degrees.

There are patient characteristics and physical examination findings that should raise concern for an associated intraspinal anomaly. The presence of EOS in a male patient has been associated with a higher rate of intraspinal anomalies.^{2,20,40,59,63} Children with tethered cord often present with pain, heel cord contractures, acute gait abnormalities, lower extremity weakness, and bowel/urologic symptoms.^{7,13,35} Neurologic examination

is important and abnormal superficial abdominal reflexes are commonly associated with syringomyelia^{40,62} and warrant MRI investigation.^{1,2,23,26,40,62}

A syringomyelia, however, is frequently associated with a Chiari malformation and together are more commonly encountered in children <3 years,^{29,44,45,63} with oropharyngeal dysfunction representing a common presenting symptom.^{1,23} For children aged 3-5 years, presenting symptoms include oropharyngeal dysfunction, in addition to headaches, and headache or neck pain worsened with Valsalva.^{1,23} Additional findings in Chiari malformations include extremity weakness,²³ abnormal deep tendon reflexes,²³ nystagmus,²⁶ and vocal cord dysfunction.²³ In patients aged 3-10 years, the presence of a concomitant syringomyelia is also associated with a higher rate of abnormal neurologic findings on examination.^{1,23}

An atypical scoliotic deformity pattern, to include a left thoracic curve, has been reported as highly suggestive for an intraspinal anomaly with an incidence between 27-50%.^{2,19,29,46,59,63,68} Wu et al.⁵⁹ reviewed 68 patients between 7-24 years of age with presumed idiopathic scoliosis and left thoracic deformities. An intraspinal anomaly was identified in 54% of patients and was more frequent in children with greater magnitudes of deformity. Severe scoliotic deformity, defined as a coronal magnitude >45 degrees despite immaturity, has been accepted as a significant risk factor for an underlying intraspinal anomaly. Morcuende et al.³⁸ reported severe deformities to have a 32% probability of positive MRI findings when seen in isolation, increasing to 86% when there was an associated neurologic abnormality. Additional atypical curve characteristics, including rapid progression (>1 degree/month), headache, or back pain had only a 3% probability of an abnormal MRI when severe deformity and abnormal neurologic findings were excluded.

In children with Chiari I malformations, an associated syringomyelia is predictive of concomitant scoliotic deformity, carrying an Odd's ratio of 9.08.⁵⁴ Zhu et

al.⁶⁵ reported that Chiari I malformations with syringomyelia had a similar radiographic appearance to cases of isolated syringomyelia in the sagittal and coronal planes. Godzik et al.¹⁹ performed a matched cohort study of spinal deformity patients with Chiari I malformations, with and without syringomyelia, finding that in the absence of an associated syringomyelia, there were significantly less atypical curve patterns with less severe deformity magnitudes than in the combined Chiari I/syringomyelia cohort. Although this data suggests that syringomyelia may be an important driver of the spinal deformity, other authors have identified atypical patterns in upwards of 50% of children with isolated Chiari I malformations.⁶⁸

Sagittal plane deformity also requires particular attention in EOS. Whereas idiopathic scoliosis is commonly associated with apical lordotic alignment, increased thoracic kyphosis is a risk factor for syringomyelia with and without Chiari malformations.^{4,33,46,52,58} Loder et al.³³ extended this sagittal plane analysis proximally to include the cervical spine, recommending clinicians maintain a strong suspicion for an underlying Chiari malformation, with or without syringomyelia, for children presenting with thoracic kyphosis > 40 degrees and cervical lordosis >0 degrees.

Influence of Intraspinal Anomaly on Spinal Deformity

The interplay between intraspinal anomalies and the initiation/propagation of spinal deformity is complex and varies based upon the underlying spinal dysraphism. In isolated tethered cord, the true prevalence of scoliosis is not known, but estimated at 31%,⁷ and similarly, the pathophysiology of scoliosis development remains unclear despite several proposed hypotheses.^{7,16,61} Conversely, the development of scoliosis in children with Chiari malformations has been amply investigated and can be influenced by cerebellar tonsillar displacement, extent of descent, syringomyelia deviation, diameter of syringomyelia, and morphology.

The position of the cerebellar tonsils in Chiari malformations and its association with curve convexity is controversial. Zhu et al.⁶⁸ reported a 78% concordance between the dominant side of asymmetrically displaced cerebellar tonsils and the thoracic convexity. Additional studies have supported this relationship^{9,60,66} while others have challenged this.^{54,55} The extent of cerebellar descent is also associated with the severity of spinal deformity.^{9,21} Godzik et al.²¹ found that moderate tonsillar displacement (5-12 mm) was an independent predictor of spinal deformity rather than severe tonsillar descent (>12 mm) when controlling for age, gender, and syringomyelia location.

Syringomyelia properties have also been implicated in the development of spinal deformity. Strahle et al.⁵⁴ reviewed over 14,000 cervical and brain MRI studies in children <18 years old, and the presence of an associated syringomyelia was independently predictive of concomitant scoliosis, in addition to female gender and older patient age. Other studies have also reported significant relationships for associated syringomyelia properties.^{9,60,66} The laterality of dominant syringomyelia deviation has been associated with curve convexity.^{9,21,60,65,66} Chiari I-associated syringomyelia are unique in comparison to idiopathic or tethered cord associated syringomyelia in that they are significantly wider and more likely to have cranial extension to the cervical spine.⁵³ Data from the Park-Reeves Syringomyelia Research Consortium identified that syringomyelia width and distal extent, in addition to holocord morphology—a syringomyelia that involves the entire spinal cord—are associated with spinal deformity and are independent predictors of underlying Chiari I malformations.⁵⁵

The pathophysiology of a Chiari lesion leading to scoliosis development may be due to alterations in cerebrospinal fluid (CSF) mechanics and flow characteristics.^{30,45} Alterations in CSF flow may be causative of syringomyelia development in this setting,

due to resultant venous and capillary dilation distal to the obstruction resulting in a mechanical stress on the spinal cord that impairs the blood-spinal cord barrier allowing for ultrafiltration and the accumulation of protein-poor fluid collection.³² Additionally, CSF flow alterations have been identified as a potential initiator for the development of spinal deformity.⁶⁴ However, further research is needed to investigate and better elucidate the underlying mechanisms of spinal deformity development and propagation in children with Chiari I malformations.

Effect of Neurosurgical Intervention on Spinal Deformity

Tethered Cord

A preponderance of data is available on the effect of tethered cord release in patients when the tether is associated with myelodysplasia.^{7,10,36,48} McLone et al.³⁶ reported the effect of detethering on spinal deformity progression in myelomeningocele (Figure 2), finding deformity stabilization or improvement in 63% of children at 2-7 years follow-up. Factors that influence the outcomes of deformity progression including patient age, skeletal maturity, level of the myelodysplastic lesion, and presenting coronal curve magnitude.^{2,38,51,54,55}

An evidence-based review of detethering in myelomeningocele patients concluded there was insufficient data to make definitive conclusions regarding the causative effect of a tethered cord on scoliosis development and whether detethering decreases the risk of deformity progression.¹² Goldstein et al.²² reported a multicenter, multidisciplinary retrospective review of myelomeningocele patients and the authors recommended against prophylactic detethering in asymptomatic patients with myelomeningocele.

For idiopathic EOS patients with tethered cords, there is less available literature to guide surgeons on risk factors for spinal deformity progression as it is hard to control for confounding variables like large curve magnitude

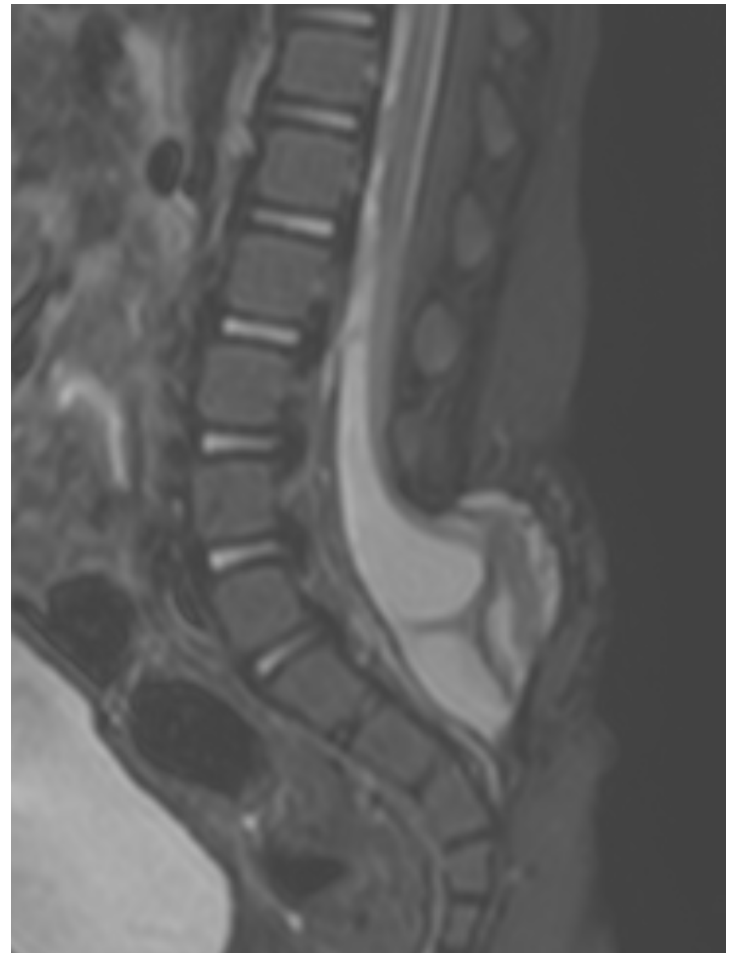


Figure 2. Sagittal T2-weighted MRI image demonstrating a myelomeningocele following closure with tethered cord

and immaturity.⁷ In the setting of an idiopathic tethered cord and progressive scoliosis, some surgeons recommend simultaneous release prior to correction. Mehta et al.³⁷ demonstrated that concurrent detethering and deformity correction can be pursued with less cumulative operating room time, less blood loss, a shorter hospital length of stay, with a lower incidence of durotomy, wound infection, and recurrent tethering.

Chiari Malformation

Neurosurgical intervention for Chiari malformations largely consists of posterior fossa decompression (PFD) which has a variable effect on spinal deformity

First Author	Publication Year	Patients with EOS (N)	Average Age (yrs.)	Average Follow-Up (Months)	% Curve Progression	Risk Factors for Progression
Farley ¹⁵	2002	5	8.7	34.7	80.00%	None listed
Flynn ¹⁷	2004	9	6.02	77.67	33.30%	Older age at presentation (11.4 vs 6.5 years); Larger preoperative Cobb angle (45 deg vs 28 deg); Double curve patterns (100% vs 14.3%); Axial rotation >2+ (75% vs 0%); Kyphosis > 50 deg (50% vs 14.3%)
Brockmeyer ⁵	2003	11	4.54	27.4	9.10%	Age > 10 years (older patients excluded from data collection); Preoperative Cobb > 40 degrees
Sengupta ⁴⁹	2000	8	8.5	36	37.50%	Age > 10 years (older patients excluded from data collection); Curve pattern other than left thoracic
Ozerdemoglu ⁴²	2003	6	6.2	91	12.50%	Age > 10 years (older patients excluded from data collection)
Charry ⁶	1994	14	7.5	33.7	42.80%	Age > 10 years (older patients excluded from data collection)
Attenello ³	2008	14	7.36	39	42.80%	Failure of syringomyelia to resolve (4-fold risk); Thoracolumbar scoliotic apex (5-fold risk)
Kontio ³⁰	2002	6	6.3	60.3	50.00%	None listed
Ghanem ¹⁸	1997	4	7.8	53.4	100.00%	Preoperative Cobb > 40 degrees
Eule ¹²	2002	19	8.67	62	73.70%	Age >8 years at time of decompression
Chotai ⁸	2018	8	6.4	8.5	25%	Age > 10 years (older patients excluded from data collection); Preoperative Cobb > 35 degrees
Ravindra ⁴⁷	2018	23	8.1	63.2	47.80%	Larger Preoperative Cobb (35 deg vs 22.8 deg); lower clival-axial angle (131.5 deg vs 146.5 deg), pBC2 line > 9 mm
Mackel ³⁴	2016	18 of 44	10.4	72	60%	Preoperative Cobb > 35 degrees (Odds Ratio 4.9); Age > 10 years with Cobb > 35 degrees increased Odds Ratio to 17 for progression to spinal fusion
Jankowski ²⁸	2016	4 of 15	7	58.5	53.30%	Preoperative Cobb > 30 degrees, Chiari II malformation
Krieger ³¹	2011	41 of 79	NR	35	21 of 79 (25.5%)	Preoperative Cobb > 20 degrees
Muhonen ³⁹	1992	3 of 11	5.5	35	0%	Age > 10 years (older patients excluded from data collection)
Verhofste ⁵⁷	2020	38 of 65	8.9	82.8	27%%	Larger Preoperative Cobb (43 deg vs 34 deg); Less Cobb improvement 1 year following Neurosurgical decompression; Double and Triple major Curves; Age > 10 years

Table 1. A summary of articles reporting the outcomes of posterior fossa decompression in children with Chiari malformations and associated early onset scoliosis (EOS). Provided values include only children < 10 years of age from within the respective studies.

*Shaded area indicates articles not included by Hwang et al.²⁹

progression. Hwang et al.²⁷ performed a meta-analysis including 12 clinical articles on children age <19 years undergoing neurosurgical intervention for Chiari I malformations with associated scoliosis. Overall, 37% demonstrated deformity magnitude improvement, 18% maintained stable curve magnitudes, and 45% progressed. In the absence of syringomyelia, scoliosis does occur in Chiari malformations but is less common. Tubbs et al.⁵⁶ identified 49 children, all adolescents, with this condition and reported varied responses of spinal deformity to PFD.

A summary of the available studies reporting on PFD and scoliosis progression for children with EOS and Chiari malformation is provided in Table 1. Age > 10 years and severity of deformity at presentation is associated with a higher risk of deformity progression and has been reported by several authors.^{5,6,8,34,42,49,57,67} The exact age cutoff remains under debate but many authors use an age of 10 as a threshold for progression. The degree of curve magnitude at presentation that is likely to lead to progression with a Chiari is also controversial, varying threshold values for coronal plane deformity being reported as risk factors for progression, including 20 degrees,³¹ 30 degrees,²⁸ 35 degrees,³⁴ 40 degrees,^{5,18} and 44.5 degrees.⁶⁷ In addition to age and the magnitude of spinal deformity, the radiographic improvement in the syrinx following neurosurgical intervention can also serve as a risk factor for progression. A syringomyelia that fails to resolve following decompression has been cited as a risk factor for deformity progression.^{4,64}

Scoliosis Characteristics

Characteristics of the spinal deformity and anatomic differences have also been implicated in the risk for deformity progression. Specific curve patterns have been associated with higher rates of deformity progression including double major curve^{17,67} and patterns other than left thoracic.⁴⁹ Flynn et al.¹⁷ identified axial rotation above 2+, as assessed by the

Nash-Moe classification, as associated with a 75% rate of deformity progression as compared to 0% in children with $\leq 2+$ axial rotation as well as sagittal plane deformity, defined as a thoracic kyphosis >50 degrees, as an additional risk factor.

What has not been widely discussed is the role that bracing can play in preventing progression. Zhu et al.⁶⁷ reported that in their series of 25 children undergoing PFD, those who were indicated for bracing demonstrated a significantly lower rate of deformity progression. Sha et al.⁵⁰ reviewed 21 children who underwent observation following PFD compared against 33 patients who were braced. Deformity progression, defined as >5-degree increase in magnitude, occurred in significantly more of the patients in the observational cohort (62% vs 30%). Additionally, when compared with children with idiopathic scoliosis, bracing was equally effective in preventing spinal deformity progression.⁵¹ More prospective research, however, is needed to better understand the importance of bracing in preventing deformity progression.

Summary

Intraspinal anomalies are relatively common in children with EOS and require prompt evaluation. Current evidence suggests that characteristics of the intraspinal pathology may influence the development of spinal deformity. Neurosurgical intervention can influence the natural history of spinal deformity in children with intraspinal pathology, especially with regard to Chiari malformations. Positive prognostic variables for age vary based upon the underlying pathology, but children with smaller deformity magnitudes and younger age at presentation are associated with higher rates of deformity stabilization or improvement following intervention. However, the available literature is quite disparate in spinal deformity progression rates and highlights the need for prospective, multicenter studies to gain a more concise understanding of the true risk for deformity progression.

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