Scoliosis and Other Congenital Vertebral Anomalies

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Introduction

Congenital scoliosis, or a lateral curvature of the spine, is driven by vertebral anomalies present at birth. The prevalence of congenital anomalies is estimated at 0.5 to 1/1000 live births (Giampietro et al. 2013) and represents approximately 10% of scoliotic deformities. The development and classification of this type of scoliosis is rooted in either failure of formation or segmentation that typically occurs within the first 6 weeks of embryogenesis (Giampietro et al. 2013). Management strategies rely on a thorough understanding of the type, location, age at presentation, and potential for growth. The true prevalence may be underestimated, as there are some anomalies that remain clinically silent (Giampietro et al. 2013).

Congenital scoliosis occurs more frequently in females compared to males with a ratio of 1.51 to 1 (Basu et al. 2002; Beals et al. 1993; Ghandhari et al. 2015; Rajasekaran et al. 2010; Shahcheraghi and Hobbi 1999; Shen et al. 2013). Multiple congenital curves may occur in up to 25% of cases (Shahcheraghi and Hobbi 1999; Louis et al. 2010; Winter et al. 1968), and compensatory curves often arise in response to the thoracic or lumbar curves (Louis et al. 2010; McMaster and Ohtsuka 1982). These compensatory curves may be minor early in life, but over time may become more clinically significant than the original curvature surrounding the congenital deformity (McMaster

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and Ohtsuka 1982; McMaster and McMaster 2013). Although some malformations may be detected as early as 12–28 weeks in utero, only 25% of malformations are diagnosed within the first year of life (Louis et al. 2010; McMaster and Ohtsuka 1982; McMaster and Singh 1999). Half of patients are diagnosed before 3 years of age (Shahcheraghi and Hobbi 1999; Louis et al. 2010).

Classification and Natural History

Embryologic, genetic, and environmental factors contribute to the etiology of congenital scoliosis. During embryogenesis the ectoderm, endoderm, and mesoderm undergo a specific choreographed sequence that forms the neural tube and paired somites in a head-to-tail fashion. Coordinated signal transduction pathways promote proper segmentation. The foundational basis of our spine and musculature is derived from the transformation of somites into a dermatome, myotome, and sclerotome.

The genetic basis of congenital scoliosis is predominantly sporadic, although it will rarely follow a monogenic inheritance pattern. Congenital spinal deformities have a positive family history in 1–3.4% of cases and may approach up to 8% in smaller case series (Louis et al. 2010; Bollini et al. 2010). Patients with congenital scoliosis may also have a 10–17% family history positive for idiopathic scoliosis, which suggests a genetic susceptibility for spinal deformities (Maisenbacher et al. 2005). Several genetic pathways responsible for proper segmentation as well as somite formation have been implicated in the formation of congenital scoliosis, including inhibitory gradients of FGF/WNT as well as Notch signaling pathways. Certain syndromic cases have also implicated various genetic mutations that follow a Mendelian inheritance pattern that are both autosomal recessive and dominant. Environmental injuries also contribute to congenital malformations. Hypoxic injuries as well as numerous teratogenic causes have been proposed. However, the precise mechanism by which these agents are thought to disrupt spinal development is often unknown (Giampietro et al. 2013).

Congenital scoliosis encompasses patients with vertebral anomalies that are broadly characterized by three subtypes defined by their embryological basis: failure of formation, failure of segmentation, or a combination resulting in a mixed deformity (McMaster and Ohtsuka 1982; Johal et al. 2016; Hedequist and Emans 2007). Vertebral deformities are further described by the spinal region (i.e., cervical, cervicothoracic, thoracic, thoracolumbar, lumbar, lumbosacral, or a combination) and plane (kyphosis vs. scoliosis). Three-dimensional imaging allows broadening of the categories to include deformities of the posterior elements including the pedicles and lamina. Genetic syndromes are also associated with vertebral abnormalities. For the purposes of this review, we will elaborate on nonsyndromic spinal deformities.

Malformations of the vertebral body can result from failures of formation. Anatomical findings may include wedge vertebra, hemivertebra, or butterfly vertebra. Hemivertebra can be distinguished from a wedge vertebra by incorporating an extra spinal segment and is often associated with an extra rib (Beals et al. 1993; McMaster and Singh 1999) (Fig. 1a, b). Hemivertebrae may be further classified into segmented, semisegmented, incarcerated, or unsegmented to reflect synostosis with adjacent vertebrae. Semisegmented vertebrae represent a segment that is fused with the vertebrae above and below without an intervening disc. Incarcerated vertebrae are joined by the pedicles to the adjacent levels, and nonsegmented vertebrae are not separated at either the level above or below. The location of the hemivertebrae may result in a balanced spine if they are contralateral to each other, thus producing a hemimetameric shift. Butterfly vertebrae are often associated with congenital kyphosis (McMaster and Singh 1999).

Abnormal synostosis between vertebrae results from failures of segmentation, including a block vertebra or a unilateral bar. Mixed failures represent a combination of formation and segmentation and have a high rate of progression if unbalanced. More recent classification schemes also include failures of formation or segmentation of the posterior elements. These can be divided by the

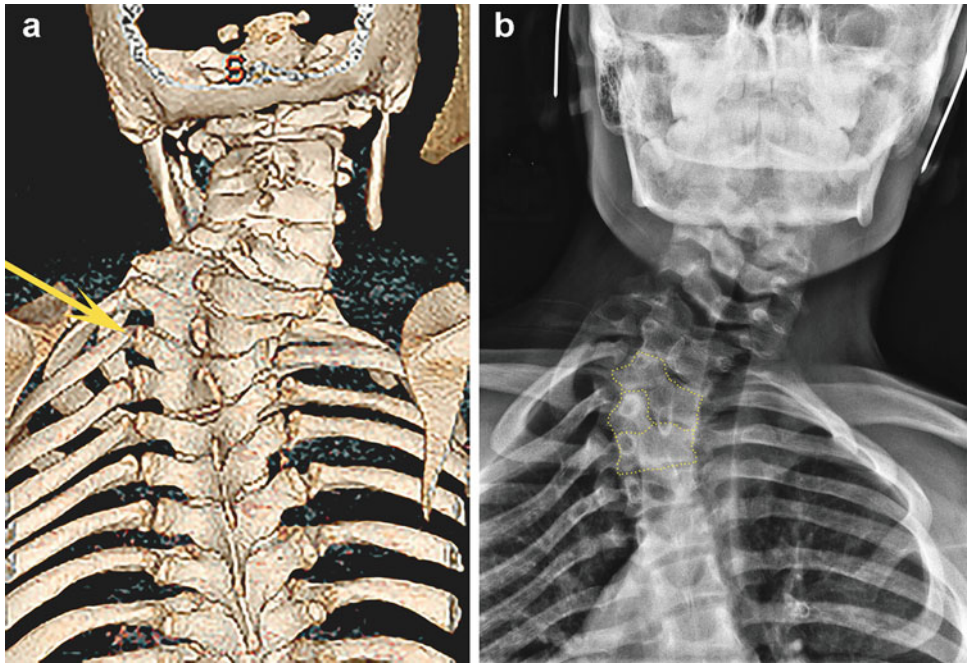

Fig. 1 (**a**) 3D CT reconstruction demonstrating extra rib emanating from the posterior aspect of the hemivertebrae (*yellow arrow*). (**b**) PA x-ray with hemivertebrae outlined

presence of a bipedicle or hemipedicle element reflecting abnormal development of the neural arches (Nakajima et al. 2007). If both anterior and posterior elements share the failure of segmentation or formation, the malformation is in "unison." If the failure is confined to one compartment, it is defined as "discordant." Discordance can be mismatched in that the vertebral body fuses with the opposite side of an adjacent vertebra's posterior element (Nakajima et al. 2007). A difference between the anterior and posterior components of a vertebral level can also be defined as a "mixed complex" (Nakajima et al. 2007).

Patient Evaluation

Clinical presentation of patients with congenital scoliosis varies depending on location, age, and other associated organ system anomalies. Routine studies ordered in newborns for other reasons such as chest radiographs, ultrasound, or MRI may identify congenital anomalies. Treatment strategies will be determined by the patient's age at the time of initial presentation, pattern, and magnitude of the deformity, as well as any associated congenital anomalies.

Initial evaluation includes a detailed history and physical examination. A positive family history of spinal deformity may be noted in up to 20% of patients (Giampietro et al. 2013). Evaluation should also encompass the prenatal and birth periods. Age, height, and weight predict skeletal growth and curve progression and should be noted. Coronal, sagittal, truncal, and pelvic imbalance should also be assessed clinically.

Chest wall abnormalities may occur with congenital scoliosis, and the patient should be examined for deformities, asymmetry, excursion, and inspiratory/expiratory capacity. The rotation and curvature of a thoracic scoliosis may compress the rib cage's height and depth. Thoracic insufficiency syndrome resulting from spinal deformity may occur due to reduced thoracic volumes and functions because of impaired pulmonary growth

in the first 8 years of life (Batra and Ahuja 2008; Muirhead and Conner 1985). Hypoplastic lung development may occur in patients with scoliotic curves approaching 90 degrees or those with a kyphotic contribution (Muirhead and Conner 1985; McMaster et al. 2007). A baseline pulmonary function test should be performed prior to surgical intervention (Muirhead and Conner 1985).

A thorough evaluation of all organ systems should be made, as 30–60% of congenital scoliosis cases have skeletal and nonskeletal abnormalities beyond the skeletal malformation (Beals et al. 1993; Louis et al. 2010; Winter et al. 1968; Maisenbacher et al. 2005; Bernard et al. 1985). In addition, the discovery of one nonskeletal abnormality predicts the presence of multiple (Basu et al. 2002; Beals et al. 1993). Mixed malformations have the highest incidence of associated organ defects, at 70% (Basu et al. 2002), which includes neural, cardiac, genitourinary, or gastrointestinal. Curve location reflects the associated abnormalities; neural and cardiac anomalies are more associated with thoracic curves (Basu et al. 2002; Shen et al. 2013; Winter et al. 1968; Bollini et al. 2010; Bernard et al. 1985), whereas genitourinary anomalies are associated with lumbar or lumbosacral curves (Winter et al. 1968; Bernard et al. 1985; Letts and Bobechko 1974).

Neural axis abnormalities or neurological deficits may present in 4–26% of patients with congenital scoliosis. Of these, 18–71% of patients will have intraspinal abnormalities (Ghandhari et al. 2015; Rajasekaran et al. 2010; McMaster 1984). Patients may also present with neurological abnormalities without intraspinal anomalies in 6–17% of patients, possibly due to cord stretching (Rajasekaran et al. 2010). Neural anomalies may also be associated with anomalies of the rib cage (Basu et al. 2002; Ghandhari et al. 2015; Bollini et al. 2010).

Spinal Imaging

Standard radiographs in both the anteroposterior (AP) and lateral upright dimensions are the basis of diagnosis and follow-up for congenital scoliosis (Batra and Ahuja 2008). These films will identify deformities and measure a curvature's magnitude and progression. If cervical spine anomalies are suspected, cervical spine films (AP and lateral upright images) are indicated. Standard measurements of both the primary and secondary curvatures should assess the coronal, truncal, and sagittal balance. Bending or traction films may be helpful to predict surgical correction (Batra and Ahuja 2008).

Computed tomography (CT) yields greater sensitivity for defining vertebral anatomy in preoperative planning (Hedequist et al. 2004; Newton et al. 2002). The complexity of some underlying deformities as well as better visualization of the posterior elements of the spine has encouraged more routine use of CT imaging (Hedequist et al. 2004; Newton et al. 2002). Curvatures that have a significant kyphotic element also benefit from evaluation with CT (Beals et al. 1993). Reduction of spinal curves may be seen 9–17% in the supine position; therefore, CT is not adequate for serial measurements of the curvature or progression (Yazici et al. 2001). CT carries significant radiation exposure compared with conventional radiography and should not be used for serial follow-up. CT is the preferred modality to visualize chest wall deformities and calculate lung volume, an anatomical measure that correlates with functional pulmonary tests (Kauczor et al. 2002).

Patients with congenital scoliosis should also have screening with magnetic resonance imaging (MRI) given the prevalence of intraspinal anomalies despite a normal neurological exam (Beals et al. 1993). Historically, renal evaluation is performed by ultrasound. However, intravenous pyelogram and renal MRI are alternatives (Batra and Ahuja 2008). Echocardiogram should also evaluate any cardiac dysfunction or anomalies. MR angiography may be an advanced tool to localize vascular abnormalities (Chan and Dormans 2009).

Treatment Options

Observational treatment should be offered to skeletally immature patients with curvatures less than 20 degrees. Serial standing radiographs at 3–6

month intervals can monitor for curve progression (Bernard et al. 1985). Semisegmented and incarcerated hemivertebrae may not require treatment (McMaster and David 1986), and block vertebrae rarely exceed 20 degrees (Letts and Bobechko 1974). Compensatory curves with radiographically documented flexibility or mixed curves composed predominantly of normal vertebrae may respond to bracing therapy. Bracing is not effective in patients without growth potential, and the curvature should be between 25 and 40 degrees (Winter et al. 1968; McMaster and Ohtsuka 1982; Chan and Dormans 2009). Most congenital scoliotic curves are stiff and inflexible or have acute angulations that limit the effectiveness of bracing (Winter et al. 1968; McMaster and Ohtsuka 1982).

Surgical indications are determined based on multiple factors. These include age at diagnosis, location of the curve, and the nature of the underlying vertebral malformation. Over 75% of congenital curves will show progression (Winter et al. 1968; McMaster and Ohtsuka 1982) such that 64–84% of untreated patients develop curves greater than 40 degrees after 10 years of age (Shahcheraghi and Hobbi 1999; Winter et al., 1968; McMaster and Ohtsuka 1982). Spinal curvatures will show greatest growth velocity during periods of significant skeletal growth. The spine undergoes a bimodal growth spurt with the first period during the first 3 years of life and again during puberty (McMaster and Ohtsuka 1982). Spinal deformities that are clinically apparent during the first few years of life have a significant risk of progression given the continued spinal growth until skeletal maturity (Winter et al. 1968).

Curve location can also impact the perception of progression. Thoracic curves, particularly thoracolumbar curves, have the poorest prognosis for progression (Winter et al. 1968; McMaster and Ohtsuka 1982; Letts and Bobechko 1974). Cervicothoracic curves might have a slower progression but become clinically important given the significant aesthetic impact with shoulder imbalance. Lumbar and lumbosacral curves also cause significant decompensation with truncal shift (Winter et al. 1968; McMaster and Ohtsuka 1982). Double curves are often progressive (Winter et al. 1968; McMaster and Ohtsuka 1982), but balanced anomalies may maintain cosmesis and stability (Winter et al. 1968).

The nature of the underlying malformation remains the strongest predictor of outcome (Winter et al. 1968; McMaster and Ohtsuka 1982). All malformations have a possibility of growth, but those with unbalanced growth at the superior and inferior endplates have an aggressive pattern of evolution (Batra and Ahuja 2008). The most severe malformations involve unilateral unsegmented bars with a contralateral hemivertebra. This is then followed by a unilateral unsegmented bar, multiple hemivertebrae, a single hemivertebra, wedge vertebra, and finally a block vertebra (Winter et al. 1968; McMaster and Ohtsuka 1982). Mixed malformations tend to have a slower evolution but are unpredictable in growth and clinical significance (McMaster and Ohtsuka 1982). Concomitant rib abnormalities may affect curve progression, although the impact of this is still controversial (Shahcheraghi and Hobbi 1999; McMaster and McMaster 2013).

Surgical intervention should be considered for curves that exceed 40–50 degrees or those that demonstrate steady progression. In addition, unilateral unsegmented bars, with or without contralateral hemivertebrae, may warrant surgical intervention, especially in children under 5 years of age given the propensity for progression. It is now more broadly accepted that earlier, prophylactic intervention prior to development of a secondary curve is preferred, even in skeletally immature patients.

Patients with intraspinal anomalies or involvement of other organ systems will need full evaluation prior to surgical correction of their congenital curves. Historically, intraspinal anomalies were addressed in a separate procedure prior to scoliosis surgery. However, some anomalies may not need to be addressed if patients are neurologically normal, and if the scoliosis surgical plan involves shortening the spinal column, this may relieve tethering on the spinal cord (Huang et al. 2015; Jalanko et al. 2011). Half of patients with cardiac abnormalities will require treatment of their heart defect (Basu et al. 2002; Letts and Bobechko 1974). Up to one third of patients may

require intervention for their genitourinary anomalies (Basu et al. 2002).

Surgical planning is dictated by the type of anomaly present as well as the patient's age and concern for growth potential. Surgery can be considered in patients under the age of five who have a minimal deformity over a short section of spine with a high propensity for progression. There are few studies that directly compare instrumented versus uninstrumented fusions (Farley et al. 2011). Studies suggest that both procedures present low neurological risk and similar reoperation rates due to fusion failures, curve progression at the point of the deformity, or curve progression of the adjacent levels (Hedequist et al. 2004; Farley et al. 2011; McMaster 1998). Uninstrumented fusions improve the curvature by 4–15 degrees, but the correction is lost over time (Farley et al. 2011; McMaster 1998; Winter et al. 1984). Instrumentation may improve the correction to 11–26 degrees, although nearly 10 degrees of correction will be lost over time. This loss is primarily due to bending of the fusion mass followed by pseudarthrosis (Winter et al. 1984). Instrumentation offers a better initial correction with a potentially lower rate of pseudarthrosis, and so as our implant technology and safety profiles have improved, there has been a general trend towards instrumentation.

Long-segment fusion and instrumentation is generally contraindicated before 8–10 years of age as it can impair lung growth and risk thoracic insufficiency syndrome (Vitale et al. 2008). There is an association between number of segments fused and reduced vital capacity in children undergoing fusion at a younger age (Vitale et al. 2008). Posterior fusions are associated with the development of crankshafting, which is seen in 14–30% of those patients with earlier surgery or larger curvatures (Winter et al. 1984; Kesling et al. 2003).

Patients undergoing corrective surgery for congenital spine abnormalities have the highest risk of neurologic injury, with 2–2.89% of cases reporting neurologic deficits (Reames et al. 2011). This is more than double the risk in surgery for patients with adolescent idiopathic scoliosis. Younger children present technical challenges for neuromonitoring with a propensity for neuromonitoring changes. Intraoperative neuromonitoring includes motor-evoked potentials, somatosensory-evoked potentials, triggered EMG, or neurogenic-evoked potentials (Pastorelli et al. 2011). For any intraoperative neuromonitoring loss, an intraoperative wake-up test may be necessary. Historically, traction therapy was contraindicated, but there may be some utility for gradual curve correction (Rinella et al. 2005). However, in short, rigid, angular, or kyphotic deformities, traction may be less efficacious and carry an increased neurological risk (Rinella et al. 2005).

Convex hemiepiphysiodesis may be considered in patients younger than 5 years with an evolving lumbar or thoracolumbar curve still less than 70 degrees that involves fewer than five segments without excessive kyphosis. This procedure requires concave growth potential and may be considered in fully segmented formation failures. Over time, the discrepancy between the concavity and convexity balances to straighten the spine as the spine continues to growth. Curve correction of 6–20 degrees is seen in 20–77% of cases (Uzumcugil et al. 2004; Winter et al. 1988). There is a wide discrepancy of stabilization of the curvature in 17–70% of cases (Uzumcugil et al. 2004; Winter et al. 1988) and only 0–21% of cases showed progression (Uzumcugil et al. 2004; Winter et al. 1988). While historically this required an anterior-posterior approach, a modified treatment now targets a posterior-only approach coupled with instrumentation or distraction of the concavity (Cheung et al. 2002).

Correction of a severe or inflexible curve may require an angular osteotomy or vertebral column resection. Hybrid techniques combine an apical osteotomy with additional growth modulation procedures harnessing the ability to correct a rigid deformity and allowing for future spinal growth through a growing rod construct (Wang et al. 2012). Hemivertebral excision is typically indicated in the presence of a fully segmented hemivertebral deformity in the thoracolumbar junction, lumbar spine, or lumbosacral spine where the hemivertebrae causes truncal imbalance or compensatory curves (Zhuang et al. 2016).

When compared to a convex hemiepiphysiodesis or in situ fusion, the hemivertebral excision is thought to achieve better curve correction. This comes with an added neurologic risk.

Hemivertebra resection may be achieved through posterior-only, staged anterior-posterior, or simultaneous anterior-posterior approach. Coronal curve correction ranges from 33–71% for anterior-posterior (Bollini et al. 2010; Jalanko et al. 2011) to 54–82% for posterior-only approaches (Jalanko et al. 2011; Cheung et al. 2002). Transient neurological deficits were 0–10% in both approaches, although there are some case series with a significantly higher rate (Jalanko et al. 2011). The posterior-only approach has gained popularity despite difficulty with anatomical visualization of ventral spinal structures as it reduces operative time and hospital stay; it also simplifies the exposure and manipulation of the ventral spinal structures (Jalanko et al. 2011).

Skeletally immature patients with long-segmented, progressive deformity may be treated through a growing construct, either with growing rods (Fig. 2) or an expansion thoracoplasty and vertical expandable prosthetic titanium rib (VEPTR) (Fig. 3). Growing rods help control deformity progression while allowing for spinal and pulmonary growth. This procedure would not be appropriate in progressive curves that lack stable anchor points or in patients with congenital rib fusion requiring a thoracostomy (Wang et al. 2012). VEPTR is indicated in skeletally immature scoliosis cases where congenital rib fusion or absence limits the hemithoracic volume with impending thoracic insufficiency syndrome.

Two techniques exist for growing rod insertion: single rod and dual rod constructs. In the single rod technique, a contoured rod is anchored proximally and distally along the concavity of the spine. With the dual rod technique, a contoured rod is implanted on both the convexity and concavity of the curvature. Both rods are then distracted at approximately 6 month intervals. The dual rod technique offers superior curve correction and has become popularized, although there is concern for bilateral autofusion that may develop over time (Wang et al. 2012; Bess et al. 2010). Anchor points may consist of hooks or claw constructs if pedicle anatomy is inadequate. Pedicle screws offer greater biomechanical strength and fewer complications. Postoperatively a thoracolumbosacral orthosis (TLSO) should be worn for 6 months to help reduce implant failure. The dual rod technique improves Cobb angle in the major curvature by 29–50% and results in 1.2–1.5 cm/y growth potential from T1-S1 with lengthening every 6 months (Wang et al. 2012).

Growing systems have a high rate of complication with curve magnitude being an important risk factor. Approximately, half of patients will experience at least one complication (Bess et al. 2010). The majority of these are implant-related, such as rod breakage or dislodgement (Bess et al. 2010; Watanabe et al. 2013). Minimizing complications may be accomplished by reducing the number of lengthening procedures, implanting at a later age, placing rods submuscularly, and using the dual rod technique (Bess et al. 2010; Watanabe et al. 2013). Noninvasive lengthening techniques have emerged within the last decade to help reduce the total number of procedures. These newer techniques are limited by a short follow-up period of only 1–3 years (Hickey et al. 2014). Magnetic rods also carry a similar concern for autofusion, and whether the magnetic forces can generate enough force to overcome any bony regrowth is unclear.

VEPTR implants have shown preservation of concave and convex growth and improvement of the Cobb angle, lateral deviation of the spine, thoracic height, cervical tilt, and shoulder imbalance (Watanabe et al. 2013). It remains the treatment of choice in patients for whom thoracic insufficiency syndrome is a concern; however, the ultimate impact on pulmonary function is mixed. Improvement in thoracic volume, lung volume, and forced vital capacity are seen (Dede et al. 2014), but age-adjusted lung volume and function do not improve (Dede et al. 2014). In addition, chest wall compliance is halved (Dede et al. 2014). The complication rate approaches 40% over time, with the number of complications increasing in proportion to the number of procedures. The risk profile might exceed that of growing rods (Zivkovic et al. 2014). Complications such as neurologic injury (7% of cases), brachial

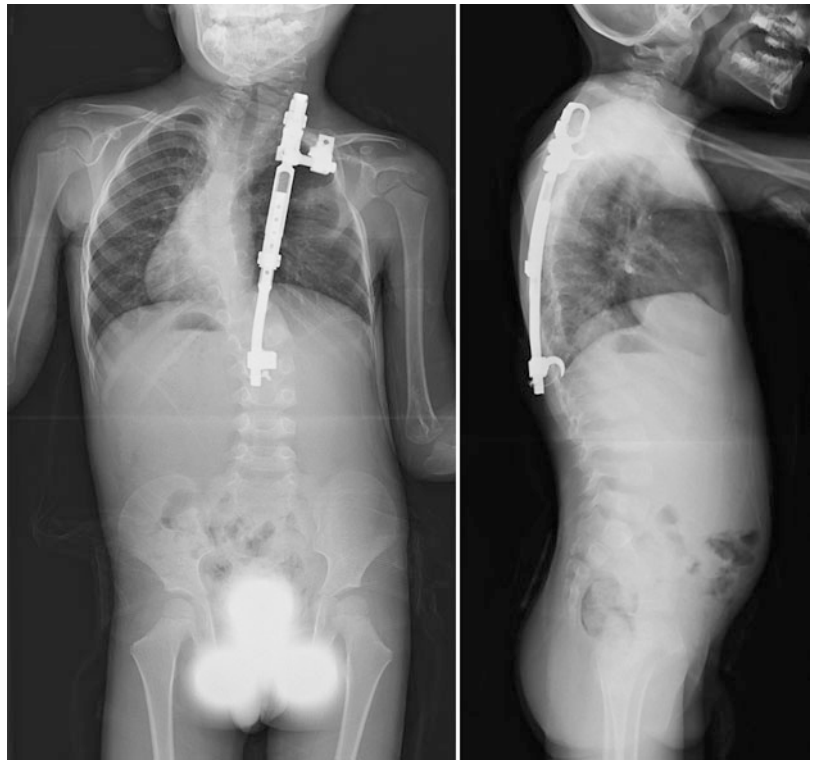

Fig. 2 PA and lateral x-ray demonstrating the use of VEPTR rib to spine construct for correction of congenital thoracic curve

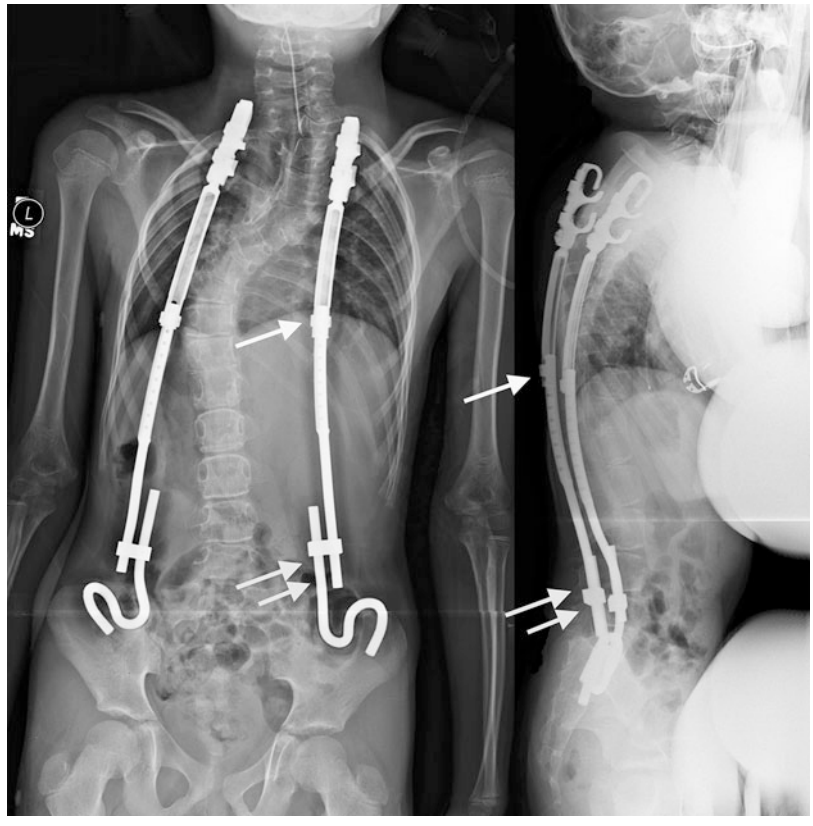

Fig. 3 PA and lateral x-ray demonstrating bilateral hybrid VEPTR growing rod construct for a congenital curve. Note that the VEPTR portion of the construct is maximally expanded (*single arrow*), and further lengthening would be accomplished by distracting at the domino connection distally (*double arrow*)

plexus injury (2.6% of cases), bone fractures of the rib or lamina, infections, or hook migration may be seen. In addition, ossification of the anchor or implant may approach 67% of cases, which may stiffen the curve and impair thoracic compliance (Zivkovic et al. 2014).

Various other growth guidance systems exist and have been used in other scoliotic etiologies including neuromuscular, syndromic, idiopathic, and congenital scoliosis (Andras et al. 2015). These are not as well understood, and although they might carry similar complication rates, they do not typically show as much Cobb angle improvement and result in less growth over time (Andras et al. 2015).

Conclusion

The optimal treatment strategy for congenital scoliosis is still not absolute. Latitude can be taken by treating physicians on the optimal time to treat and whether to consider long-segment or short-segment fusions or growing constructs. There has been a trend towards early identification and intervention of these congenital defects to minimize development of large compensatory curves that increase treatment morbidity. Management of these patients requires the understanding of the natural history of the specific congenital spine deformity as well as anticipation of curve evolution. There is now better appreciation that the younger the patient is at the time of initial presentation, thoracolumbar deformities and unilateral segmentation failure portend a worse curvature progression. Patients with congenital scoliosis may have involvement of multiple organ systems, and the neurologic, cardiac, and genitourinary systems should be evaluated. Preoperative MRIs are also obligatory because of a high rate of concurrent neurologic anomalies. In addition, pulmonary function should be assessed prior to any surgical intervention.

Once the diagnosis of congenital scoliosis has been made, a wide array of treatment options is available. Considerations include the variability of the disorder as well as a lack of cohesive evidence in management and dissimilarities in surgical technique. Observation may be warranted when the curve is minimal and there is not a high propensity for progression due to a mild underlying disorder. Convex hemiepiphysiodesis and instrumented fusion may be appropriate as a prophylactic intervention for progressive short-segment deformities. Osteotomies and vertebrectomies may correct severe deformities but carry a higher risk of neurological injury. Growing systems may address long-segment deformities when there is concern for axial skeletal growth. Growing rods or an expansion thoracoplasty with vertical expandable prosthetic titanium rib (VEPTR) are the treatments of choice when thoracic insufficiency syndrome is of concern. The goal of surgery is to minimize pulmonary compromise or any additional neurological deficit while maintaining adequate spinal alignment. Ultimately, any surgical intervention attempts to achieve a balanced spine while maximizing the underlying spine growth.

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