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Pediatric scoliosis

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Abstract Pediatric scoliosis is a relatively uncommon condition typically first noticed due to altered stature or by routine spine screenings by a school nurse or pediatrician. The formal diagnosis is made with spine radiographs, with coronal curvature measurement of 10° or greater. Treatment may consist of serial observation, bracing until skeletal maturity, or surgery for correction and fusion/stabilization of severe or progressive deformity. Overall success for non-operative management of scoliosis is affected by the etiology for the deformity, close follow up and monitoring for evolution of the deformity, and patient compliance with their treatment regimen. The most common surgical technique is a posterior approach spine fusion with implanted instrumentation, and patients are typically back to their activities of daily living by 6 months postoperatively. Continued intermittent monitoring of the scoliosis throughout adulthood is recommended, to detect late deformity progression, development of arthritis symptoms, or other associated issues.

Keywords Adolescent idiopathic scoliosis · Neuromuscular scoliosis · Congenital scoliosis · Observation · Bracing · Instrumented posterior fusion

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Introduction

Since early human history, descriptions of patients with spine deformity and methods for their treatment have been recorded by medical professionals. Scoliosis as we use the term today is defined as a lateral spinal curvature of 10° or greater, assessed using the Cobb method [1]. This definition comes from the coronal curvature being the most easily appreciated aspect of scoliosis observed on radiographs, but it must be understood that scoliosis is actually a 3-dimensional deformity, and the typical scoliotic spine is also both axially rotated and has altered sagittal contour through the length of the curve [2, 3]. Scoliosis nomenclature has been historically used to indicate the age of onset, and the etiology for the deformity. Infantile, juvenile, and adolescent descriptors correlate to scoliosis diagnosed at ages 0–3, 4–9, and 10–17 years, respectively. Similarly, neuromuscular, congenital, and idiopathic are descriptors reflecting the attributed etiology responsible for the spinal deformity. Neuromuscular diseases (eg. cerebral palsy or muscular dystrophy) and congenital failure of formation or segmentation in the spine can predispose affected children to develop scoliosis, but for the vast majority of pediatric patients the etiology for the scoliosis is unknown. In those patients where a causative association can not be attributed to the spine deformity, the term "idiopathic" is used, although there is mounting evidence for a genetic basis for idiopathic scoliosis despite the moniker. Overall, scoliosis is an uncommon diagnosis in children, yet population studies have shown that 2–3% of children meet criteria for the diagnosis and should be assessed and followed clinically [4, 5]. Clinical concern for unrecognized scoliosis progression includes pulmonary or cardiac embarrassment, neurological injury, or difficulty maintaining skin

integrity over bony prominences or delivering care to severely involved syndromic and neuromuscular scoliosis patients.

For the majority of patients with idiopathic scoliosis, it is uncommon to need more treatment than intermittent clinical and radiographic observation [6]. However, for patients with non-idiopathic scoliosis, there is a significantly higher chance that more aggressive treatments will be needed. Physical therapy to strengthen muscles and to reinforce posture is a typical first treatment offering for all patinents that can tolerate such a regimen, but bracing of the trunk to minimize curve progression is typically the first definitive treatment for non-operative management scoliosis in idiopathic and selected non-idiopathic patients. Clinical research for a variety of bracing strategies and clinical success rates is somewhat conflicting, and more standardized and better controlled large clinical trials are currently being conducted in association with the Scoliosis Research Society and the NIH. As bracing is the only non-operative treatment modality that is generally accepted as effective in managing pediatric spinal deformity, when this modality fails, surgical intervention is frequently required as the definitive intervention. In this review we will discuss common examples of pediatric scoliosis: adolescent idiopathic, neuromuscular, and congenital.

Adolescent idiopathic scoliosis

Affecting otherwise normal and healthy children, adolescent idiopathic scoliosis (AIS) does not have an obvious clinical etiology that drives the spine deformity. Children may have intermittent back pains, tend to be tall and slim, but are otherwise as active and athletic as their peers. Girls are affected slightly more commonly than boys for small curves (1.4:1) but the gender disparity widens dramatically for curve magnitudes in excess of 30° (10:1), producing an average of 7–8 fold more girls affected than boys overall [7]. The diagnosis of AIS is one of exclusion, where other etiologies must be considered and "ruled out". Multiple clues during the clinical evaluation may help the clinician recognize that the scoliosis has an underlying etiology, including a detailed birth and developmental history indicating developmental delays consistent with neuromuscular etiologies, history of being "double jointed" possibly consistent with Marfan's disease or a collagen vascular disease, physical exam notable for midline pigmentation or hairy patch consistent with dysraphism and congenital scoliosis, or the constellation of skin nodules, café ole marks, and axillary freckles consistent with neurofibromatosis. Patients with previously undiagnosed Marfan's disease or neurofibromatosis can be misclassified as idiopathic scoliosis, and could lead to mis-management or possible catastrophic surgical outcomes from aortic or neurological complications, respectively. Similarly, either the clinical exam or spine radiographs demonstrating a left sided thoracic curve should raise suspicion for a neurological etiology for the scoliosis such as Charcot Marie Tooth, a tethered cord or spinal cord syrinx, and appropriate further testing including electrodiagnostic studies (CMT) or MRI of the full spinal axis should be obtained. If a non-idiopathic etiology for the scoliosis is suspected, it is the responsibility of the clinician to perform appropriate testing to confirm or refute the diagnosis, possibly including X-Rays and MRI of the entire spine as described, but also renal ultrasound and echocardiogram to assess for other congenital malformations, as may be found in patients with asymmetric facies or limb development anomalies.

That there is no apparent etiology for the spine deformity of AIS led investigators to search for a more subtle basis to explain the disease. It was well established that scoliosis tended to concentrate in family kindreds, and this led to the observation for the importance of obtaining a thorough family history when evaluating patients with AIS. Harrington reported that in women with scoliotic curves greater than 15°, the incidence of scoliosis in their daughters was 27%, markedly higher than by chance alone [8]. Similarly, studies of identical twins have shown concordance rates as high as 73%!, further suggesting the importance of genetics in the disease [9]. Numerous investigators have reported associations of genetic loci that link to scoliosis in specific family kindreds, but no one gene has been identified as a "master molecule" to explain AIS, leaving experts to the conclusion that the process driving deformity progression is multifactorial on a genetic basis, and may also be influenced by environment. Nonetheless, a new genetic test (ScoliScore ®™) has come to market with the aim of identifying AIS patients with mild to moderate curves that may progress to severe curves requiring surgery. The test involves the patient providing a saliva sample that is then tested against 53 genetic markers correlated with scoliosis, and an algorithm involving current curve magnitude, age, and positivity for the number of different gene markers generates a ScoliScore Number. The Number can be compared to a standard curve generated during extensive pre-market research to determine the patient's expected risk for progression to a severe curve magnitude. Ward et al. studied this score in three cohorts with known AIS treatment outcomes, and the method produced very high positive and negative predictive values for screening AIS patients [10]. As regards clinical use, it is not clear yet if this genetic test will allow practicioners to release AIS patients with very low scores from serial follow up, or to require AIS patients with very high scores to submit to aggressive and invasive treatments earlier than currently clinically utilized.

Scoliosis curves have the highest risk for progression during adolescent growth spurts. It is therefore important to identify when patients are in their peak growth velocity, and to tailor treatment strategies to mitigate curve progression during this period. Once skeletal maturity is achieved, the risk for progression decreases dramatically [11]. With the concepts of growth rate and growth potential in mind, it is important for the treating physician to estimate the amount of growth remaining and how "skeletally mature" the patient has become with each office visit. This can be done clinically with serial measurements of overall standing and seated trunk height measurements with each office visit, Tanner staging, and recording the timing of menarche for female patients. Radiographic methods include obtaining a left hand radiographs for comparison with a Greulich and Pyle atlas, open or closed status of the triradiate cartilage, or with use of Risser staging of the iliac apophysis [12–14]. Of these three, the most commonly used clinically the Risser sign described in 1958 [15]. Dr Risser described the progressive anterior to posterior opacification and closure of the iliac apophysis as the patient approaches skeletal maturity. The iliac apophyses are apparent on the screening full spine x-rays that are already routinely obtained for monitoring the scoliosis, making use of the Risser sign logical and safe.

Bracing

Treating scoliosis with bracing has been attempted throughout the centuries. Modern bracing with removable orthoses came about as an alternative to cast immobilization of the spine. Despite the materials used for the braces, the concept for the treatment remains the same: apply forces through the ribs or soft tissues of the trunk to enact a reduction of the scoliosis so that as the patient grows the spinal deformity progression is prevented or minimized. The goal of brace treatment is to prevent or minimize progression of the deformity during longitudinal spine growth, but despite the improvement of curve magnitude in the brace, it is not anticipated to reduce scoliosis magnitude once the brace treatment is discontinued. As bracing is an attempt at deformity prevention by modulation of growth, it should not be offered to patients who are skeletally mature. There is controversy as to when to initiate bracing of scoliosis, with most authors indicating the 20–26° curves as providing both indication of requirement for intervention and a small enough curve to be controlled in a brace. As curves advance into the 40–50° range, the spinal deformity is more difficult to effectively manage using bracing, and these patients may have lost the opportunity for a bracing trial. In general, braces can be classified based on the area of the spine that they contact. A TLSO (thoraco-lumbar-sacral orthoses) spares the cervical spine while a CTLSO (cervical-thoracic-lumbar-sacral orthosis) includes the cervical spine. The Milwaukee brace developed and popularized in the 1940s by Drs Walter Blount and Albert Schmidt is an example of a CTLSO, it was worn around the clock and was thought to be effective, but is rarely utilized today. Currently, the most commonly prescribed braces are thermoplastic rigid TLSOs that are either "off the shelf" and can be modified with foam pads to optimize correction of the scoliosis in the brace (eg the Boston® brace), or the TLSO can be custom molded to the patient as the patient is held in longitudinal traction to correct their scoliosis. In either option, the goal is to obtain nearly 50% correction of the scoliosis while the in the brace, and regimens for use are typically full time. An alternate to full time brace wear is the nighttime bending rigid TLSO brace, where the goal is for scoliosis overcorrection in the thermoplastic orthosis, and with resultant brace wear in the evening and during sleep where the dramatic trunk bend and curve correction would be tolerated by the patient. Nighttime bending braces are best utilized in scoliosis affecting the thoracolumbar and lumbar spine, as it is technically challenging to obtain the dramatic corrections required higher in the thoracic segment. A more recent option is the tension-based spinal orthosis (eg SpineCor®, a TLSO composed of soft materials capable of producing corrective forces and decreased magnitude of the patient's scoliosis while in the appliance). Clinical research organized and funded through the National Institute of Health and the Scoliosis Research Society is being conducted to prove the efficacy of bracing in general, and for the relative success for particular orthoses.

In order for a scoliosis brace to be effective, it must be worn. Compliance has been an important problem with every brace design. The CTLSO braces such as the Milwaukee brace have especially poor compliance, due to immobilization of the cervical spine, and inability for patients to "hide" the orthosis under clothing. It must be understood that in addition to the mild physical discomfort from the brace itself, there are also psychological stressors during the adolescent years including wanting to "fit in" that deters brace wear in children. Clinical research utilizing temperature or load sensors in spinal orthoses have been used to measure brace regimen compliance, with measured compliance rates reported as 33–82% of full time wear as assessed measuring forces in the brace [16] and 47–78% as assessed by thermal sensor [17, 18]. Surprisingly, TBSO braces that would be more easily worn under clothing and possibly more comfortable secondary to their soft materials, showed compliance rates of 54% in a recent study of twelve patients with thermosensor-modified braces [19], not apparently different from rigid TLSOs. As regards how much brace wear is required to alter the natural history of curve progression, Katz et al. in a prospective study using heat-sensor modified rigid TLSOs showed that there is a

dose-response relationship for the amount of time the brace was worn during the day and control of scoliosis progression, that 82% of patients wearing their brace for 12 h or more showed no curve progression, and that patients prescribed 16 or 23 h of brace wear per day actually wore their braces about the same amount of time [20••]. Their findings led to the suggestion that patients be advised to wear their rigid TLSO braces for more than 12 h per day.

Surgery

In patients whose spinal deformities have not been controlled with non-surgical management, and where the scoliosis is either relentlessly progressing or is over 50°, surgical intervention may be required. These criteria reflect the desire to stop and correct ongoing spinal deformity in the present and future, respectively. Ongoing deformity progression in the setting of a patient being appropriately managed in a brace should prompt the clinician to consider brace modifications, alteration in the brace wear regimen, change to a different brace, or consideration of surgery if the brace implementation alterations have been attempted unsuccessfully. The clinical literature reflects different curve magnitudes for which to intervene surgically for the AIS patient, but the standard at our institution is 50°, a curve measurement shown to predispose skeletally mature patients to long-term deformity progression [21]. Despite expected slow progression of curves at ~1° annually as an adult [22], over the course of the adult lifespan, a 50 degree scoliosis in a teenager could progress to an 80–100° deformity, predisposing the patient to cardiac and or pulmonary compromise [23, 24]. The primary goals of surgical intervention are to achieve a balanced, solidly fused, and pain-free spine.

A complete preoperative evaluation is required for each patient with AIS, including detailed history and physical, detailed neurologic examination, and routine laboratory work. For patients with focal neurological findings or abnormal curve patterns (left thoracic, absence of hypho-kyphosis, severe rotation) a full spine MRI is suggested to evaluate for potential pathology within the spinal canal (tethered cord, lipoma, diastematomyelia) or central nervous system (syrinx, Chiari malformation, diplomyelia). Other radiographic studies required for surgical planning are full spine standing PA and lateral views, and flexibility studies (traction, push-prone, or side-bending films). For areas where pedicle or other bony anatomy is difficult to interpret on plain radiographs, a CT scan through the area may provide valuable information to allow hooks or wires to be available on the operating room in addition to pedicle screw implants. Classifications for AIS curve patterns have undergone multiple generations of improvement, and the most widely used classification currently in use is that devised by Lenke et al. [25]. The Lenke classification has been shown to have good inter and intra-observer reliability, and has given surgeons a comprehensive and reliable system for communication as well as pre-surgical planning [26, 27, 28•]. The Lenke classification has also provided a means to logically choose distal fusion levels and perform thoracic-only lumbar-sparing fusions in appropriately selected patients with flexible curves, an intervention expected to reduce postoperative long-term morbidity secondary to adjacent segment degeneration below fusions and due to decreased lumbar mobility.

Surgical treatment of scoliosis has evolved tremendously over time. Initially, scoliosis patients requiring surgery had non-instrumented spine fusion and postoperative immobilization provided by body casts and long-term bedrest. Harrington instrumentation evolved in the 1960s allowing surgeons to achieve two points of fixation and greatly improved deformity correction, however body casts and bedrest was still the standard [29]. Segmental instrumentation, allowing multiple points of fixation began with hooks and wires [30], but evolved into the pedicle screw constructs used today, and which provide more power for deformity correction and shorter fusion lengths [31, 32]. With modern segmental instrumentation, no requirement for postoperative bracing or bedrest is routinely necessary, and patients are typically back to activities by 6–12 months postoperatively. Surgical management of AIS has improved dramatically in the past several decades, but should be understood to be the “last resort” for management of spinal deformity that has not responded to other more conservative treatment options.

Neuromuscular scoliosis

Neuromuscular scoliosis is spinal deformity that results from improper functioning of the nervous system or the muscles. The improper functioning of the muscles that serve to support the spine, or the nerves that control those muscles, can lead to spinal deformity. Neuromuscular scoliosis, in contrast to AIS, has a different “motor” that drives the spinal deformity, and these neuromuscular scoliosis patients can have rapid and relentless progression of their spinal deformity throughout life. Classical neuromuscular curve patterns are of the “long C-shape” and may begin at the cervicothoracic junction and continue to the pelvis, where pelvic obliquity and sitting difficulties are common problems. Cerebral palsy, muscular dystrophy and neurofibromatosis are examples of neuromuscular diseases that affect the spine and have high rates of associated scoliosis. Bracing and wheelchair modifications for non-ambulators may allow patients with initially small magni-

tude curves to be managed nonsurgically, but most patients will eventually require surgical intervention if their spinal deformity remains progressive.

Cerebral palsy

Cerebral palsy (CP) is a nonprogressive neurologic disorder that affects movement. It is thought to be a result of incomplete brain development, or a sequela of brain damage, frequently associated with birth trauma. The severity with which the patient manifests their CP symptoms correlates with their risk for scoliosis. Monoplegic ambulators (children with one weak limb capable of walking independently) have much lower risk for progressive scoliosis than quadriplegic non-ambulators. In the young patient with CP newly diagnosed with scoliosis, bracing may be considered in an effort to minimize scoliosis progression during their longitudinal trunk growth, to allow a more proportionate body to limb length ratio when definitive fusion is required. For the more severely involved CP patients, pulmonary function may decrease as the scoliosis worsens due to reflux or poor clearance of secretions at baseline, and their worsening ability to participate with coughing and chest physical therapy. Retained secretions may in turn lead to pneumonia and worsening of other underlying respiratory issues (asthma, bronchiectasis, chemical pneumonitis). For patients with repeated bouts of reflux or pneumonia, fundoplication and placement of an abdominal wall gastrostomy for feeding may be a means to prevent pulmonary embarrassment while maintaining caloric needs for adequate nutrition. Nutrition is a key consideration when managing patients with CP, and affects the patients' abilities to mount immune responses to fight infection as well as heal skin areas affected by pressure/stasis from improper positioning or by surgical trauma [33].

Management of patients with scoliosis associated with CP should be global, with the patient evaluated for all of their orthopaedic issues and managed accordingly. This may alternately require optimization form a pediatric medicine vantage for pulmonary and nutrition issues, with use of orthotics or wheelchair modifications to allow optimal sitting posture, injections or releases of extremity contractures, or directed consideration for their spinal deformity. Physical examination of the patient will help to assess general flexibility of the scoliosis, and advisability for a bracing trial. Ambulators with flexible curves will typically tolerate rigid TLSO bracing of small magnitude deformities (20–30°) until closure of the triradiate cartilages, a radiographic parameter that can help to predict risk for crankshaft deformity postoperatively. Ambulatory patients with rigid curves, and non-ambulatory patients may not tolerate bracing as well, although nonambulators may be candidates for braces to be fabricated that incorporate into their wheelchairs that may make this intervention more tolerable. Sitting balance and skin pressure leading to decubiti are important considerations in nonoperatively treating patients with CP in braces, as these patients may be less able to reposition, particularly in the nonambulatory population. Although bracing is a temporizing measure, patients and caregivers achieve high satisfaction with TLSOs [34].

Surgery is indicated when curves progress to greater than 45–50°, when functioning such as assisting with transfers is impeded due to the deformity, when the scoliosis is interfering with skin-fold hygene or other care delivery, or when the patient has skin breakdown as a result of the deformity (ischial or other decubiti). Once the decision has been made to pursue surgical management, a thorough nutrition and pulmonary evaluation are required, and a global orthopaedic assessment of the patient should be performed. A complete discussion regarding each of the medical, nutritional, and orthopaedic issues facing the patient should be conducted with them and their family, and a multidisciplinary team should be assembled to develop and enact a plan from necessary preoperative evaluations and procedures, to the surgical stay, and then on to the postoperative convalescence that may require subacute skilled nursing. Parents and caregivers should be warned that patients who have hip contractures and that require spine fusion to the pelvis may have what appears to be worsening of the hip pathology postoperatively, due to the imposed rigidity of the lumbar spine and pelvis and inability to accommodate the hip problems. Classic fusion levels are T2 to L5 for ambulators, and T2 to pelvis for non-ambulators, with the thought that the residual mobility at L5/S1 may help ambulators to more easily continue to maintain this ability postoperatively. Sparing the lumbosacral is controversial, with at least some authors advocating for T2 to pelvis due to concerns for progressive deformity below the fused levels [35], but others stopping at L5 reporting good results [36]. Perioperative concerns include routinely high blood losses, challenging nutrition and pulmonary status, and increased risk for infection. Pseudarthrosis is notoriously high in patients with CP, with rates ranging from 7–20% [37, 38]. With the advent of segmental spine instrumentation, pseudarthrosis has been decreased substantially [39, 40].

Duchenne's muscular dystrophy

The incidence of scoliosis ranges from 75–90%in patients with Duchenne's muscular dystrophy [33]. There is typically rapid progression after diagnosis, and children

also tend to have competing medical issues such as deteriorating pulmonary function due to their gradually worsening muscular physiology. Efforts to brace patients with Duchenne's muscular dystrophy are only effective for smaller curves, and may further compromise respiratory function, requiring most patients and their families to consider spinal surgery, stabilization, and fusion for deformity magnitudes considered small when compared to AIS or CP [41].

Typical management of spinal deformity in Duchenne's patients includes very close monitoring with serial clinical and radiographic exams, and definitive fusions T2-pelvis when curves progress to the 25–30° range, prior to the anticipated precipitous decline of pulmonary function. Spinal surgery improves quality of life overall, and sitting comfort for non-ambulatory patients [42]. Spinal stabilization however, does not provide a protective role for pulmonary function, as this is a factor controlled by the status of the muscles, not the chest wall architecture [43, 44]. A thorough medical evaluation is needed preoperatively since muscular dystrophy is a systemic disease, involving multiple organ systems, including the heart. Intraoperatively high bleeding rates may be encountered and post-operative complication rates are also higher [45]. Overall patients with Duchenne's muscular dystrophy have shortened life expectancy with 12% achieving 25 years of life [46].

Neurofibromatosis

Neurofibromatosis (NF) is a multisystem disease involving nerve tissue, and may present with nerve tumors (optic, acoustic, or peripheral). The most common form of NF is type-1, also known as von Recklinghausen's disease, and is an autosomal dominant inherited genetic disorder where the Neurofibromin tumor suppressor gene (NF-1) is dysregulated. Clinical manifestations of NF type-1 include the presence of café-au-lait spots, Lisch nodules, gliomas, and cutaneous or deep neurofibromas. Spine deformities occur in NF-1 in up to 50% of patients, are described as either dystrophic or nondystrophic, are associated with dural ectasia, and classically have high rates of pseudarthrosis [47]. Nondystrophic scoliotic curves are most common, and the deformity is managed similarly to that for idiopathic scoliosis. Treatment should follow standard bracing guidelines with surgical intervention for curves over 50°. There is a small risk of nondystrophic scoliosis modulating into a dystrophic curve, so close radiographic observation is necessary [48]. Dystrophic curves tend to be highly rotated with sharp angles involving a short segment, penciling of ribs, scalloping of the vertebral bodies, and widening of the interpedicular distance. There is no role for brace treatment in the dystrophic curve pattern, as their relentlessly progressive nature dictates early surgical intervention with fusion. Strict care must be taken in the zone containing the dystrophic changes, to prevent iatrogenic neurological injury.

Classically, patients with scoliosis associated with NF underwent anterior/posterior surgery to obtain thick and stable fusion masses in an attempt to prevent pseudarthrosis in the short to intermediate follow up, and to minimize dural ectasia from eroding the fusion and producing instability in late follow up. Multiple procedures may be required in this patient population, with additional procedures to re-graft and thicken existing fusions, or to repair pseudarthroses or fusions that deteriorated. Neurological risks, including paraplegia or paralysis, are higher in patients with neurofibromatosis following deformity correction and fusion, particularly in patients with dystrophic curve patterns. Intra-spinal pathology must be ruled out preoperatively with MRI and CT scans to investigate the soft tissue and bone architecture, with greatest concern at the location of dystrophic changes. Pseudarthrosis rates are higher in patients with both dystrophic and nondystrophic curves, with classical surgical treatment regimens involving a return to the operating room for fusion exploration and augmentation 6 months following the index procedure. Modern techniques involving pedicle screw instrumentation and use of rhBMP-2 may improve outcomes [49].

Congenital scoliosis

Congenital scoliosis encompasses a spectrum of structural disorders that are present at birth, and which cause progressive deformity as the child grows. This is in contrast to infantile idiopathic scoliosis where structural abnormalities are absent. The disorders comprising congenital scoliosis can be grouped into defects of segmentation, formation, or mixed, and should be considered in the context for how they alter vertebral geometry and potential for growth. Congenital malformations that produce higher liklihood for asymmetric growth tend to have the highest probability for inducing clinically significant scoliosis curves. Perhaps the simplest congenital deformity is the block vertebra, where 2 or more vertebrae fail to segment symmetrically, and therefore provide minimal risk for progression of deformity. Unilateral bars are the result of failure of segmentation unilaterally, with near normal growth potential preserved contralaterally, and a tether incapable of growth on the affected side involving 2 or more vertebrae. Estimates for scoliosis curve progression for unilateral bars can be as high as 9° per year, which would produce unacceptable deformity even after just a few years of normal growth [50, 51]. Similar descriptions for relentless curve progression in the setting of hemivertebrae

(unilateral failure of formation), and unsegmented bar with contralateral hemivertebra (mixed defect), will frequently prompt early surgical intervention when these congenital defects are noticed to prevent progression. Congenital scoliosis is frequently associated with other systemic abnormalities, including cardiac, genitourinary, and the gastrointestinal system. When the diagnosis of congenital spinal deformity is made, a search for other congenital defects in other locations of the spine, as well as the viscera is mandatory.

Treatment goals for congenital scoliosis are the same as for other scoliosis diagnoses: to maintain spine balance in the coronal and sagittal plane, to stop or minimize spine deformity, and to have a pain free spine. Bracing may be attempted for very young patients, or to improve posture and temporize until surgery can be arranged, but typically plays little role in the management of patients with congenital scoliosis. Surgical intervention is tailored both to the age of the patient as well as to the nature of the defect. Prophylactic intervention while the curve is small is appropriate if prognosis is poor, as would be expected for a low lumbar hemivertebra, or a hemivertebra with contralateral bar. The range of surgical interventions include anterior hemiepiphysiodesis to ablate the "normal" growth opposite an unsegmented bar, excision of hemivertebra to prevent asymmetric growth, spinal posterior fusions to control aberrant growth over longer areas of congenital deformity, and complex osteotomies to allow correction of the spine in addition to instrumented fusion. Hemiepiphysiodesis is suitable to patients younger than 5 with a fully segmented hemiverteba that corrects to less than 40° [52]. Through convex growth arrest of the hemivertebra, growth on the concave side allows gradual improvement of spinal alignment. However, concave growth and deformity correction can be unpredictable, and close follow up is mandatory. Hemivertebra excision removes the driving force that creates the deformity, allows maximal immediate correction, and has become much more commonly used than epiphysiodesis. The risks of hemivertebra excision surgery from an all-posterior approach can decrease the risks associated with the classically used anterior/posterior technique [52].

Conclusions

Pediatric scoliosis encompasses a broad range of conditions with or without a clearly defined etiology for the spine deformity. Treatment involves careful initial and serial clinical and radiographic assessments, and frequently involves bracing of the scoliosis curves to prevent progression. Surgery for relentlessly progressive or for severe curves typically involves posterior approach surgery with instrumentation and fusion to correct and stabilize the spine. Using modern nonoperative and operative treatments, management of pediatric scoliosis has become much better tolerated and less anxiety provoking for patients.

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- • Of importance
- •• Of major importance

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