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Other Causes of Pediatric Deformity

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Congenital scoliosis

The definition of congenital scoliosis is a lateral curvature of the spine attributable to congenital vertebral anomalies. Congenital anomalies of the spine result from abnormal vertebral development during weeks 4 to 6 of gestation. These anomalies result in asymmetric growth of the spine with development of deformity. Although the anomalies are present at birth, the deformity usually manifests later in life. The overall incidence of these anomalies is unknown. It has been estimated to be approximately 0.5 per 1000 in the thoracic spine. Isolated anomalies are likely sporadic with little genetic tendency. It is possible that multiple vertebral anomalies carry a 5% to 10% risk for future siblings [1]. Congenital scoliosis produces a spectrum of deformity ranging from mild to severe progressive curvature with dire cosmetic and cardiopulmonary consequences.

Classification and natural history

Congenital scoliosis can result from failure of segmentation, formation, or a mixed form (Fig. 1) [2]. Approximately 80% of anomalies may be classified as failures of segmentation or formation, with 20% being a mixed form. A block vertebra results from bilateral segmentation defects, with fusion of the disc spaces between the involved vertebra. When this occurs in the neck, it is a component of the Klippel-Feil syndrome [3]. A block vertebra in other parts of the spine is difficult to diagnose because it produces little deformity. Unilateral bars occur on the concave side of a curve; when they are associated with a contralateral hemivertebra, they produce the highest propensity for scoliosis progression. A unilateral unsegmented bar is a bony bar fusing the disc spaces and facets on one side of the spine. The unsegmented bar does not contain growth plates, and therefore does not grow. Segmentation defects involve bony bars between adjacent segments. Rib fusions are often observed on the side of the unilateral bar.

Failure of formation produces a wedge vertebra or a hemivertebra. A wedge vertebra represents partial failure of vertebral body formation on one side. The asymmetric vertebral body maintains two pedicles. In contrast, a hemivertebra represents complete failure of formation of half a vertebra. There are three main types of hemivertebrae: fully segmented (65%), partially segmented (22%), and nonsegmented/incarcerated (12%) [4]. A fully segmented hemivertebra possesses a normal disc above and below the anomaly. A partially segmented hemivertebra is fused to the neighboring vertebra on one side with an open disc space on the opposite side. An incarcerated hemivertebra lies in a niche in a neighboring vertebra. Its appearance is more ovoid than that of other hemivertebrae. The pedicles remain well aligned with minimal scoliosis.

Determining which curves are likely to progress rapidly is a difficult proposition. In general terms, 25% of curves are nonprogressive, 25% progress slowly, and 50% display rapid progression [5]. The determinants of curve progression depend on the type of anomaly, its location, and the age of the patient. Normal longitudinal

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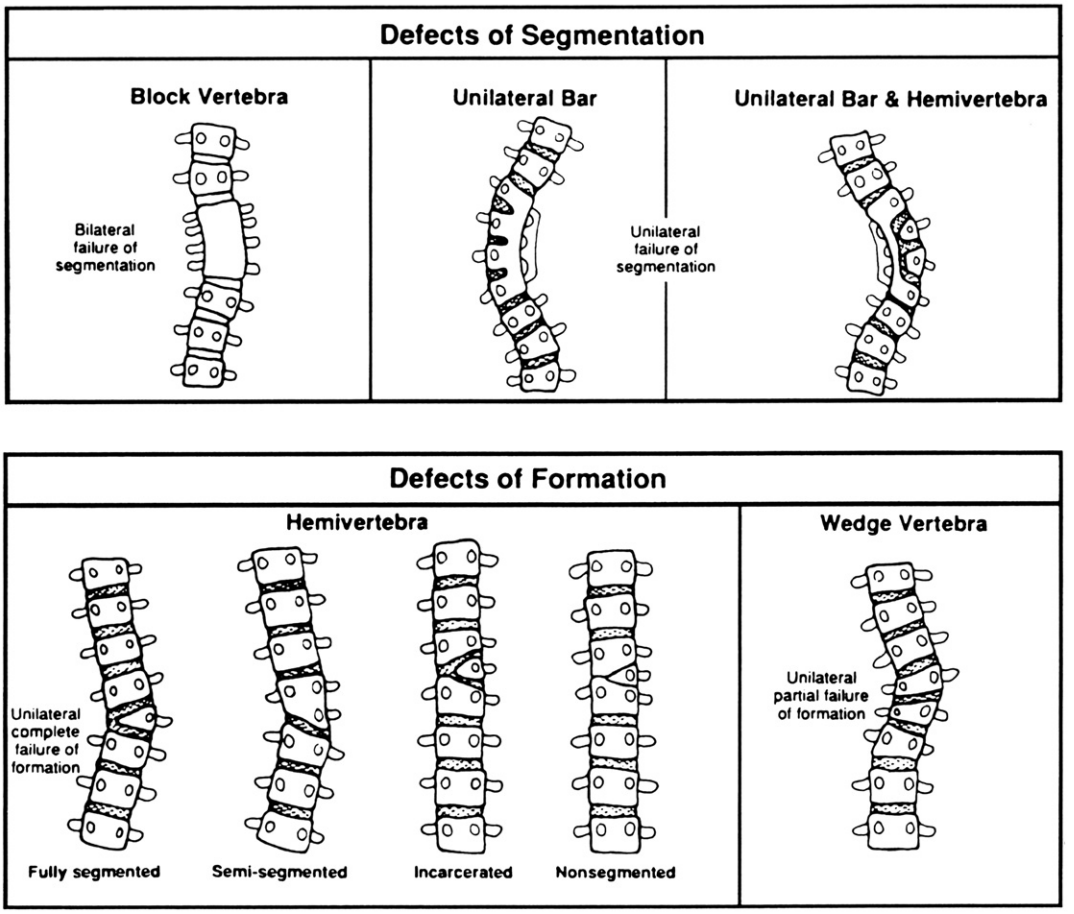

Fig. 1. Different patterns of congenital scoliosis. Defects of segmentation produce block vertebrae or unilateral bars. Defects of formation result in hemivertebrae or wedge vertebrae. (*From* McMaster MJ. Congenital scoliosis. In: Weinstein SL, editor. The pediatric spine: principles and practices. New York: Raven Press. p. 229; with permission.)

growth of the spine is a summation of the growth occurring at the superior and inferior end plates of the vertebral bodies. Usually, the spine grows symmetrically, except in the case of a congenital anomaly, in which there is asymmetry of the number of growth plates. Curve progression occurs from unbalanced growth of the spine. Thus, the quality of the disc spaces surrounding the anomalous segment predicts the potential for asymmetric growth. Good healthy discs portend curve progression. McMaster and Ohtsuka [2] reported on the rate of worsening in patients with various types of congenital spine anomalies. For those anomalies producing curve worsening, progression occurs during rapid periods of growth and continues until skeletal maturity. Curves in the thoracolumbar spine seem to progress more rapidly than those in the upper thoracic spine.

The most severe anomaly is the unilateral bar with a contralateral hemivertebra, which progresses between 5° and 10° per year. The block vertebra, with no healthy intervening discs, seems to be the most benign. An isolated hemivertebra may progress 2° to 5° per year, and a sole unilateral bar may progress approximately 5° per year. Location is a strong factor to consider when predicting the actual deformity, however. For example, a fully segmented hemivertebra at the lumbosacral junction may cause marked deformity because it causes the spine to take off obliquely from the sacrum. A long secondary thoracolumbar curve may develop that is initially mobile but later becomes fixed. To avoid this result, the lumbosacral hemivertebra should be treated early. A hemimetameric shift occurs when a hemivertebra on one side is balanced by another

on the opposite side of the spine, separated by one normal vertebra. Intuitively, it would seem that the number of growth plates should balance in this circumstance. These still progress in up to 30% of patients, however [6].

Associated anomalies

The spine develops between 4 and 6 weeks of gestation along with the genitourinary, musculoskeletal, and cardiovascular systems. Most patients with congenital scoliosis also manifest abnormalities in other organ systems. The prognosis for these patients is excellent if the anomalies are detected and treated. These anomalies may be isolated or in association with the VACTRL syndrome (vertebral anomalies, anorectal atresia, cardiac anomalies, tracheoesophageal fistula, and renal and limb anomalies) [7]. The musculoskeletal system may demonstrate congenital anomalies in the cervical spine (Klippel-Feil syndrome) and upper extremity, such as Sprengel's deformity or radial deficiency [8].

Genitourinary abnormalities are observed in 20% to 40% of these children [9,10]. These are usually anatomic abnormalities with normal renal function. A renal ultrasound scan or evaluation of the kidneys on spine MRI is recommended for all these patients, however. Cardiac abnormalities occurred in 26% of patients in one series of congenital scoliosis [7]. Ventricular septal defects tend to be the most common finding. Before any surgical intervention, an echocardiogram with an evaluation by a cardiologist is recommended.

Neural axis abnormalities occur in up to 40% of patients with congenital scoliosis [7,11]. A wide variety of abnormalities is observed, including diastematomyelia, intradural lipoma, syringomyelia, Chiari malformation, and tethered cord. Bradford and colleagues [11] reported a 38% incidence of neural axis abnormalities in 42 patients: 16 harbored a tethered cord, 4 had a diastematomyelia, 4 had a syringomyelia, 3 had a diplomyelia, and 1 had a sacral teratoma (some patients harbored more than one abnormality). Clinical indicators include skin stigmata, such as a hairy patch or skin dimple. Foot and leg abnormalities, such as cavus feet, vertical talus, and calf atrophy, also show a high association with intradural anomalies. These neural axis abnormalities occur with all types and locations of congenital spine anomalies. The highest association occurs with a unilateral bar and contralateral hemivertebra in the lower thoracic area, however, of which up to 52% harbor an intraspinal anomaly [12]. Neurosurgical intervention is required for any anomaly that tethers the cord before attempting surgical correction of the deformity [13]. An MRI examination of the entire spine from the occiput to the sacrum is recommended before any surgical intervention.

Patient evaluation

A thorough history and physical examination are essential because of the high incidence of associated anomalies. Height and weight are recorded at every visit. Note is made of head tilt, shoulder elevation, pelvic obliquity, overall trunk balance, and any cutaneous stigmata that may indicate an underlying pathologic condition. A thorough neurologic assessment, including reflex testing, is mandatory.

Anteroposterior and lateral plain films are obtained to follow curve progression. These are done supine (before child is able to stand). Once a child is able to stand, radiographs are done supine and standing. Thus, the supine radiograph serves as a comparison to the previous radiographs, and the standing radiograph functions as a new baseline. The Cobb angle is measured at every visit. In congenital scoliosis, the intraobserver error for the Cobb angle is 10°, because the landmarks may be difficult to discern [14]. It is advised to use the same landmarks on every radiograph. Lateral bend films are used to assess flexibility and rigidity of the curvature. CT with three-dimensional reconstructions is an invaluable tool to study the anatomy of the anomaly and is obtained before surgical intervention. In patients who are candidates for the vertical expandable prosthetic titanium rib (VEPTR), this provides an assessment of rib integrity for attachment sites. MRI is also performed before surgical correction to rule out any intraspinal anomaly requiring neurosurgical intervention.

Management

Nonsurgical

After diagnosis, children are initially followed every 3 months to assess curve progression. If the curve remains stable and the anomaly is one that is not likely to rapidly progress, the patient may be followed every 6 months with plain anteroposterior and lateral radiographs. During the adolescent growth spurt, however, plain radiographs are taken again every 3 to 4 months.

Bracing of congenital scoliosis is rarely indicated [15]. These curves are usually inflexible and unresponsive to bracing [16]. Bracing can be used to control compensatory curves that may develop in response to the congenital curve, however. The compensatory curves occur in areas of the spine that contain normal vertebrae and possess increased flexibility. In addition, bracing may be used after surgery until solid fusion occurs.

Surgical treatment

Treatment of congenital scoliosis is primarily surgical [17,18]. Surgery is indicated for curve progression or when an anomaly is deemed likely to progress. Thus, prophylactic surgery is indicated for young patients with curves likely to show severe progression. The prognosis for congenital scoliosis depends on the type of vertebral anomaly, its location, and the age of the patient. The location of the anomaly is paramount, because the cervicothoracic and lumbosacral junctions produce a larger cosmetic deformity than other areas of the spine. Surgical goals include attaining spinal balance while retaining maximal flexibility and growth potential. Numerous techniques to achieve these goals have been attempted, with good success for appropriately selected patients. These include in situ fusion [15], anterior/posterior hemiepiphysiodesis [19,20], and hemivertebra excision [21–23]. More recent fusionless techniques focus on stabilizing curves with growing rods [24] or VEPTRs [25,26] until skeletal maturity to allow maximal spine growth.

In situ fusion. The aim of performing an in situ fusion is to stabilize the curve. It can be done anteriorly, posteriorly, or combined. The compensatory curves may still progress after an in situ fusion of the deformity is performed. The fusion should extend one level above and below the deformity [27]. Ideally, this procedure is performed before the development of a major deformity, because no correction is attained from the procedure. Isolated posterior fusion may lead to the development of the crankshaft phenomenon [28]. This develops when continued anterior growth in the presence of a posterior tether produces worsening of the original curvature. The addition of an anterior release with bone grafting prevents this phenomenon from occurring.

Convex epiphysiodesis and posterior arthrodesis. Convex epiphysiodesis may provide stabilization or even correction of moderate-sized curves [29]. Convex epiphysiodesis relies on adequate growth potential on the concave side to correct the spinal curvature. Thus, this procedure is contraindicated in patients with a unilateral bar that has no growth potential on the concavity. The convex lateral halves of the discs are removed anteriorly. The patient then undergoes a posterior arthrodesis on the convexity with no exposure of the spine on the concavity. The patient is immobilized in a cast in the position of maximum correction. Winter and colleagues [20] reported that 38% of their patients were corrected an average of 10° and that 54% stabilized. The ideal patient is younger than 5 years of age, with a short curve less than 40° and scoliosis involving five segments or less. It is not recommended for patients with a kyphotic component to the deformity. The correction obtained from this procedure is gradual and unpredictable.

Hemivertebra excision. The best indication for excision is an isolated fully segmented hemivertebra at the lumbosacral junction. These cause marked deformity secondary to an oblique takeoff from the sacrum. When operating at the level of the thecal sac, one does not have to contend with the spinal cord. Still, neurologic risks are high, and the parents must understand all the alternatives. The surgical procedure may be performed through a posterior-only or combined anterior/posterior approach. The posterior-only approach involves removing the hemivertebra body and discs from an approach through the pedicle. The excised area is then compressed down, and correction is maintained with instrumentation followed by a cast for several months. Ruf and Harms [22] report on 28 patients treated with posterior-only excision of the hemivertebra with excellent results. Although, hemivertebra excision is safest inferior to the thoracolumbar junction, several authors have reported excellent results with excision in the thoracic [23,30,31] and even cervical spine [30].

Fusionless techniques. The goals of surgical treatment for scoliosis in the growing child include correction of the deformity with maintenance of a flexible spine. This allows the child to attain maximal trunk height. Surgery without fusion permits this to occur through placement of arthrodesis for correction, followed with interval expansions to allow attainment of maximal height.

Growing rods. Growing rods were first described by Harrington [32] in 1962. Harrington

used a subperiosteal approach and placed a single distraction rod connected to hooks at the distal ends. This technique was improved by Moe and colleagues [33]. The subperiosteal exposure was limited to the areas of hook placement, and these sites were not fused. Patients wore a Milwaukee brace after surgery and underwent expansion when greater than 10° of loss of correction occurred. Reported complications, including hook dislodgement and rod breakage, occurred in 50% of patients. In 1997, Klemme and colleagues [34] reported on their 20-year experience using the Moe technique. Sixty-seven young patients with a wide variety of diagnoses, including congenital scoliosis, underwent an average of 6.1 procedures. The average curve reduction was 30%, and the curve was stabilized in 44 of the 67 patients. Complication rates are high and include wound infection, hook and screw dislodgement, and rod breakage.

Akbarnia and colleagues [35] developed the dual-rod technique to improve on hardware-related complications. In this technique, subperiosteal dissection is limited to the upper and lower anchor sites. Hooks and screws are used in a claw-like configuration and may be fused with bone graft. The rod is placed subcutaneously and joined on each side with tandem connectors. Patients are braced until final fusion occurs. Lengthenings are done at approximately 4- to 6-month intervals. Thompson and colleagues [24] compared the dual-rod technique with the single-rod technique in a retrospective comparison and found that the dual-rod systems produced better initial correction and allowed more growth. The overall complication rate seemed similar, however (48%).

Vertical expandable prosthetic titanium rib. The spine, lung, and chest grow interdependently. Campbell and Hell-Vocke [26] developed the VEPTR to address the spine and chest wall deformity without the need for a fusion. The original indication for this device was for the treatment of thoracic insufficiency syndrome, which is defined as the inability of the thorax to support normal respiration and growth. This is diagnosed by a history of respiratory difficulties, on physical examination by lack of motion as demonstrated by the thumb excursion test, and radiographically by a restricted hemithorax. The major categories of this syndrome include flail chest, rib fusion and scoliosis, and hypoplastic thorax syndrome as seen in Jeune's and Jarcho-Levin syndromes.

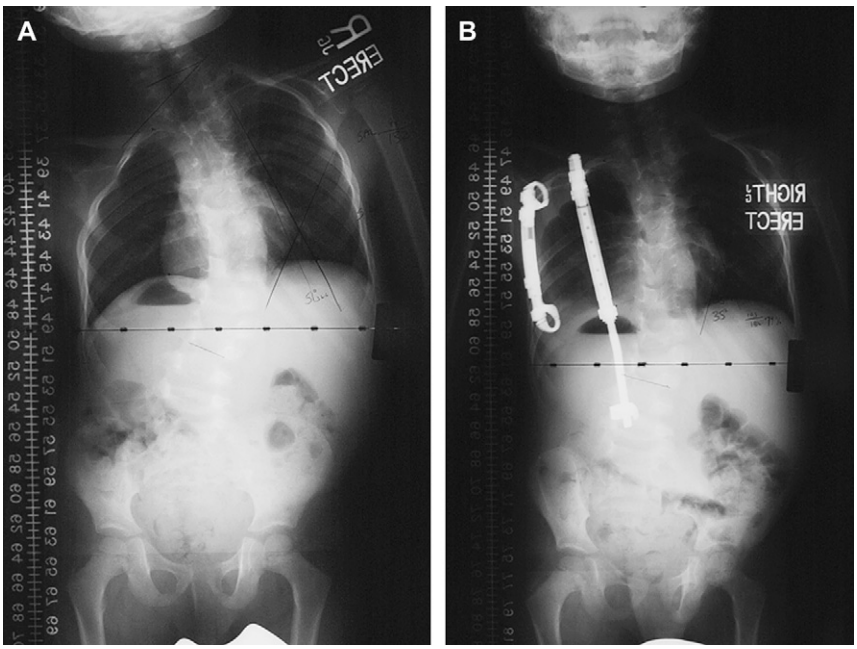

Fig. 2. (*A*) Anteroposterior spine radiograph of a 3-year-old child with progressive congenital scoliosis. (*B*) Patient underwent placement of a rib-to-rib and rib-to-spine VEPTR. His curvature has been maintained, and he undergoes expansion every 4 to 6 months.

The VEPTR device does not involve spinal arthrodesis but rather an opening wedge thoracostomy and implantation. The device can be attached rib to rib, rib to spine, or rib to pelvis, depending on the individual patient. After initial implantation and correction, the patient undergoes serial lengthenings every 4 to 6 months (Fig. 2). Once the original device has been maximized (after approximately six to eight lengthenings), the device is exchanged out. In patients with congenital scoliosis and fused ribs, the longitudinal growth of the thoracic spine after use of the VEPTR was 7.1 mm/y compared with a normal longitudinal growth of 6 mm/y. The concave and convex sides of the spine showed growth, as did the unilateral unsegmented bar [26].

Neuromuscular scoliosis

Pediatric scoliosis is seen in several conditions. The major categories include idiopathic, congenital, and neuromuscular. The Scoliosis Research Society has subdivided neuromuscular conditions into neuropathic and myopathic categories. The neuropathic etiologies include cerebral palsy, spinocerebellar degeneration, syringomyelia, spinal cord tumor, spinal cord injury, myelomeningocele, and spinal muscular atrophy. Myopathic conditions include Duchenne's and Becker's muscular dystrophy, arthrogryposis, and congenital myopathies. A large proportion of children with neuromuscular conditions develop scoliosis. This ranges from the 20% to 70% seen in cerebral palsy [36] to 100% in skeletally immature children with cervical or thoracic level spinal cord injury [37]. The etiology of the scoliosis is not completely understood but likely involves asymmetric muscle strength and tone. Numerous factors likely contribute to this phenomenon [38]. For example, after spinal cord injury, cord tethering or a syrinx may develop. Dorsal rhizotomy in patients with cerebral palsy may increase the risk of future deformity [39]. The natural history of the underlying disease helps to determine the timing and surgical procedure undertaken for treatment.

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