In few areas does orthopedic care have a more profound impact than on early-onset scoliosis, a condition that can diminish a child’s life expectancy and quality of life if not treated promptly and appropriately.

**Classification**

In the United States, scoliosis diagnosed before the age of 4 years is categorized as infantile scoliosis. Idiopathic scoliosis diagnosed between ages 4 and 10 years is classified as juvenile idiopathic scoliosis. In the United Kingdom, scoliosis diagnosed before the age of 5 years is termed early-onset scoliosis. Although such semantics may be important for research purposes, the clinician must simply bear in mind the gravity of treating scoliosis with an onset at a young age. Because scoliosis progression is related to growth and peak growth velocity, scoliosis in infants and preschool-aged children has the potential for rapid and severe progression.

Early-onset scoliosis may be further subdivided based on a variety of parameters. As in other forms of scoliosis, the etiology is an important factor in determining treatment and prognosis. Mehta reported less success in the treatment of infantile scoliosis in patients with underlying syndromes and neuromuscular conditions.

Infantile scoliosis is also classified as resolving or progressive, and this distinction is critical. Curves in approximately 80% of infants with scoliosis will resolve without intervention. This resolution usually begins as the children develop trunk musculature around the time they begin cruising. Mehta reported radiographic findings that are associated with progressive infantile scoliosis. These parameters include documented progression of the Cobb angle on successive radiographs, a difference in the rib–vertebral angle at the apex of the deformity of more than 20°, rib phase 2, and a double major curve pattern.

Congenital anomalies of the bony elements are managed similarly to congenital scoliosis diagnosed later in childhood. Risk of progression is increased by the presence of concave bars, concave rib fusions, and fully segmented hemivertebrae. The main differences in the management of congenital scoliosis are that younger patients have more growth potential (and thus more potential for deformity progression) and that the pattern of congenital anomalies is difficult to discern (bones are less ossified, and thus the ability to discern segmentation is imprecise). The children must be evaluated for renal anomalies (missing kidney or horseshoe kidney), cardiac defects, and neural axis problems (tethered cord, Chiari malformation, and syrinx). Often, the renal tree is visualized on a spinal magnetic resonance image, obviating the need for an additional study.

**Natural History**

The natural history of early-onset scoliosis is bleak. Pehrsson et al reported a significantly increased mortality rate in patients with infantile and juvenile idiopathic scoliosis, likely resulting from respiratory failure. Several studies have shown that humans cease growing new alveoli after age 7 years. Thus, deformity leading to decreased chest volume before this age has a permanent, negative effect on pulmonary function regardless of restoration of alignment later in life. Early-onset scoliosis is a serious condition that requires early intervention.
TREATMENT

Historically, the treatment for early-onset scoliosis has been early fusion. The typical treatment involved an uninstrumented fusion immobilized with a body cast followed by exploration of the fusion mass 6 months later. Repeat bone grafting and casting would be performed if a solid fusion was not achieved. This treatment was less than ideal in that it required repeat surgery in young patients, prolonged immobilization in a cast, caused shortening of the trunk, and had the potential for crankshaft phenomenon. Nonetheless, no other options were available. However, as patients reached maturity, an increase in mortality was noted. Decreased growth of the spine, particularly the thoracic spine, led to decreased pulmonary function in adulthood.

In response to the limitations of early fusion, Akbarnia and McCarthy developed fusionless instrumentation strategies to address spinal deformity in children, which used growing rods with limited anchoring to the spine that could periodically be lengthened with additional surgery. Akbarnia et al subsequently reported the evolution of the surgical techniques for growing rods. They learned that bilateral rods were better than 1, larger diameter rods failed less frequently than smaller diameter rods, and subcutaneous rods led to less fusion of the spine but more infections. They also found that best results were obtained when lengthening procedures were performed more frequently than biannually.

The Vertical Expandable Prosthetic Titanium Rib (VEPTR) (DePuy Synthes Spine, Inc, West Chester, Pennsylvania) is often used in patients with early-onset scoliosis (Figure 1). To date, the device has only been granted Humanitarian Device Exemption status by the US Food and Drug Administration; thus, it may only be used for thoracic insufficiency syndrome in pediatric patients. Thoracic insufficiency syndrome is a congenital condition where severe deformities of the chest, spine, and ribs prevent normal breathing and lung growth and development. The device may be attached from rib to rib, rib to spine, or rib to pelvis.

Several studies have demonstrated increased thorax and lung volume in patients treated with the VEPTR. The VEPTR is lengthened every 6 months to allow for spinal growth. In patients with congenital scoliosis and fused ribs, Campbell et al demonstrated that the longitudinal growth of the thoracic spine after VEPTR instrumentation was 7.1 mm per year, compared with a normal growth of 6 mm per year. A common problem that is uniquely seen with the VEPTR is a slow erosion of the anchors through their bony attachment sites, including the ribs, laminae, and ilia. Patients are usually asymptomatic, although some have reported painless prominent instrumentation. The erosion is often found incidentally on routine radiographs.

Growing rods and VEPTRs, although an improvement over early fusion, are fraught with complications. Infections and instrumentation failures and dislodgement are frequent. Over half of patients will experience at least 1 complication. Furthermore, evidence has emerged that a finite period of time exists in which growing instrumentation can facilitate growth. Cahill et al reported that nearly all patients treated with growing rods had autofusion of the spine at the time of definitive fusion. Autofusion increases the complexity of the definitive fusion at maturity and decreases the ability to obtain additional lengthening. Sankar et al reported that diminishing length was gained with subsequent lengthenings. Similar limitations of the VEPTR device are also emerging, although the fact that it is anchored away from the spine may allow for more micromotion through rib facet joints, thus avoiding disturbance of the perispinal musculature that could lead to differentiation of pluripotent stem cells into osteoblasts.

The finite period of effectiveness of growing instrumentation has led to an expanded role for serial body cast treatment of early-onset scoliosis. Mehta reported a series of 139 patients with progressive infantile idiopathic scoliosis treated with serial casting. Approximately two-thirds of the patients had complete resolution of their curves. One of the most important factors associated with this level of success was age at initiation of treatment: The average age was 18 months for those with treatment success. However, those whose curve progression did not completely resolve had significant reduction in curve magnitude and were able to reach adolescence before surgery was required. Thus, although Mehta may consider patients in that group as having failed treatment, serial body casting helped avoid early surgery. Therefore, serial body casting may play an important role in delaying or even eliminating the need for growth sparing surgery with the VEPTR or growing rod implants.

Technologies are emerging that are likely to significantly alter the treatment of scoliosis in children. The limitations of the VEPTR and growing rods, such as high infection rates, implant loosening, and early fusion, may be eliminated or decreased by using implants that do not require repeat surgery to lengthen. The MAGEC MAGnetic Expansion Control (Ellipse Technologies, Inc, Irvine, California), used successfully in Europe, lengthens externally via battery power. This allows for more frequent lengthenings, avoiding

Figure 1: The Vertical Expandable Prosthetic Titanium Rib device in a rib-to-spine orientation. (Image used with permission from DePuy Synthes Spine, Inc, West Chester, Pennsylvania.)
the prolonged periods of immobilization that are necessary with serial casting.

Two other procedures that do not require repeat lengthening are vertebral body stapling and vertebral body tethering. Vertebral body stapling involves insertion of shape memory alloy brackets across the disk space on the convexity of the curvature. The staples crimp further on reaching body temperature, imparting continuous corrective force and slowing growth on the convexity. Experience at our institution demonstrated good success at minimizing progression in the vast majority of patients with thoracic curves less than 35° and lumbar curves less than 40° (Figure 2). For thoracic curves between 35° and 45°, we have had excellent early results with vertebral tethering using a polyethylene-terephthalate braided cord tensioned through anterior vertebral screws. At our institution, vertebral body stapling is performed in patients as young as 7 years and vertebral tethering in patients as young as 9 years.

Early-onset scoliosis is a condition with the potential for severe adverse consequences. However, science in this area is young as 9 years.

Figure 2: Preoperative (A), 4-day (first erect) postoperative (B), 1-year follow-up (C), and 2.5-year follow-up (D) posteroanterior radiographs of a patient treated with vertebral body stapling.

REFERENCES


