Early-onset scoliosis is defined as scoliosis diagnosed at the age of five years or less (Fig. 1). These patients may have congenital spine malformations and thoracic malformations, such as fused or absent ribs, but they may also have normal vertebral formation. Early-onset scoliosis is uncommon. The incidence of idiopathic scoliosis has been reported to be 1.5% and infantile scoliosis represents only 4% of this population. However, early-onset scoliosis is an important health issue. These children develop spinal deformities at young ages and if left untreated, are at risk for rapid deformity progression, cosmetic disfigurement and pulmonary insufficiency.

Pehrsson et al reported that in patients with untreated scoliosis, the mortality rate increased in infantile-onset scoliosis followed by juvenile-onset scoliosis mainly due to respiratory failure (40%). The average age at death was 54 years but some patients died as young as 16 years. Postmortem findings of these children would show very small lungs with markedly decreased numbers of alveoli.

**Bracing**

Early-onset scoliosis commonly leads to severe deformities at an early age. Most are double major curves, and some have substantial thoracolumbar kyphosis. There is no strict guideline for treatment but most clinicians support observation in children with mild deformities and bracing in children who have progressive noncongenital curves. Unfortunately, bracing is known to fail frequently in treatment of scoliosis in young children, especially the congenital or neuromuscular types. This is especially true for those patients with a rib-vertebral angle > 20°, indicative of a progressive curve. Spinal bracing is now commonly used for minor curves in the range of 20° to 30°. Bracing requires prolonged use, especially in patients with early-onset scoliosis, and effective bracing is dependent on patients’ compliance and proper fitting. Patients with proximal thoracic curves require the use of a Milwaukee Brace, which is usually not well tolerated by patients.

**Fusion Surgery**

For early-onset scoliosis, early definitive fusion is advocated in the past for children with severe curves, deteriorating congenital spinal deformities, or progression despite bracing. However, there is a growing consensus that early spinal fusion results in negative pulmonary consequences and most surgeons now use fusion as the last resort in the young patient.

The technique of fusion surgery usually requires an anterior approach to be effective due to the crankshaft phenomenon or worsening rotational deformity in young patients with solid posterior fusions resulting from persistent anterior growth of the vertebral bodies as recognised by Dubousset. Goldberg et al, Karol et al and Vitale et al who all looked into long-term results from fusion in patients with early-onset scoliosis and found that 37.2% to 39.3% underwent reoperation due to progression of deformity. Even some patients treated with anterior and posterior fusion to theoretically eliminate crankshaft progression required subsequent revision surgery. Achieving circumferential fusion of the spine in young children may still fail due to continued growth.

Besides failure of the fusion to control the deformity, fusion can have drastic consequences on overall growth of the child. In DiMeglio’s studies on spinal growth, the most rapid period of spinal growth occurred in the first five years of life, when the spine increased 50% of its length. It was also found that between the ages of five and ten years, the spine continued to grow, albeit at a slower rate. In summary, the height of the thoracic spine averaged 11 cm in the newborn, 18 cm at age five years and 22 cm at age ten years. Translating this knowledge to treatment of early-onset scoliosis, spinal fusion in a five-year-old child can result in a 12.5 cm loss of spinal growth.

In addition, fused spines in growing children may lead to arrested pulmonary development. This leads to thoracic insufficiency syndrome or inability of the thorax to support normal respiration or lung growth. Campbell et al showed that immobile or nonfunctional chest wall caused by small thoracic height and multiple congenital rib malformations interfered with normal respiration. Poor pulmonary function was also seen in patients who had undergone early spinal fusion for scoliosis, inhibiting normal thoracic growth. Normal development of the lungs in the growing child had been well established. The greatest increase in the number of alveoli in normal children occurred in the first two years of life. Although lung volumes continued to increase into adolescence, alveolar growth was complete by eight years of age. For this reason, surgery at this age could create the greatest disturbance in pulmonary development. Pulmonary volumes tend to decline with normal aging and the decline was expected to begin in the mid-30s. Patients who have poor pulmonary reserve as a baseline would expect more significant loss of volume with early definitive fusions. Goldberg et al showed that fusion performed in patients who were younger than ten years of age had a mean forced vital capacity (FVC) of 41% of the normal at maturity as compared to 68% of normal FVC in patients fused older than ten years of age.

**Fusionless Surgery**

Various methods without the need of fusion are being designed to control growth, to delay definitive fusion surgery, and to
increase the thoracic volume. Applying principles of managing limb length discrepancies and gradual limb deformity corrections to the spine, epiphysiodesis on the convex side of the deformity with or without instrumentation is a technique to provide gradual progressive correction and to arrest the deterioration of the curves. Hemiepiphysiodesis, hemivertebal resection, and short segment fusions can be used in selected patients to allow for gradual correction of these curves. Staples used are usually biocompatible shape-memory metal alloy staples. However, these techniques are still controversial with mixed results at short-term follow-up. One study by Marks et al found that anterior and posterior growth arrest alone was not effective for preventing progression of deformity in infantile scoliosis. In contrast, Betz et al showed that stapling the anterior vertebral spinal growth plates could control curve progression. In their study, six out of ten patients with a mean curvature of 35° were controlled and stabilised during a one year follow-up period.

**Growing Rods**

To address the limitations of spinal bracing or fusion for early-onset scoliosis, growing rods (distractible spinal implants) were developed. Techniques of instrumentation include Harrington, Cotrel-Dubousset, or Luque rods. More recently, Akbarnia et al developed a technique using Isola dual rod instrumentation. These growing rods are inserted across a segment of spinal deformity where no fusion is performed. Cranial and caudal anchoring foundations are made using hooks or pedicle screws. Each foundation is connected to a rod, and the rods are connected by cross-links. Distraction of the growing rods is performed usually every six months in which the surgical incision site must be reopened for the distraction procedure. Once maximum spinal growth or skeletal maturity is reached, definitive final fusion is performed. By this approach, growing rods could control progression of spinal deformity as well as gradually correct the deformity.

In addition to gradual correction of spinal deformity and allowing continuous spinal growth, thoracic insufficiency syndrome could also be treated by the Vertical Expandable Prosthetic Titanium Rib (VEPTR device) (Fig. 2). After an opening-wedge thoracostomy, the acute correction can be stabilised by this device. The VEPTR is extended from the cephalad rib to the caudal rib, the lumbar spine, or to the posterior iliac crest. These devices are also distracted at scheduled intervals of six months. At a mean follow-up of 5.7 years, the vital capacity was found in one study to have significantly increased with mean coronal deformity correction of 74° to 49° at the latest follow-up. Technically, single growing rod instrumentation could be placed on the concave side of the spine to expand the smaller hemithorax. Some studies report better radiographic correction and fewer complications with bilateral constructs.

Despite the advantages of growing rod surgery, the need for repeated surgeries under general anesthetics is a major drawback due to increased anaesthetic and wound complications. Other complications include proximal migration of devices with rib fractures, and skin problems secondary to recurrent surgery and hardware. The complication rates ranged from 8% to 50%. Instrumentation failure included...
rod fracture or anchor failure and the overall rate was reported to be 15%.\textsuperscript{49} Junctional kyphosis and curve decompensation were also noted but rarely occurred.\textsuperscript{39} In addition, growing rod surgery is associated with various socioeconomic concerns including longer periods of hospitalisation and increased time away from school for the child and time away from work for the parents.\textsuperscript{50,52} Repeated operations and hospitalisation may also affect the child’s psychological well-being and appearance with a cosmetically poor surgical scar.\textsuperscript{50-52}

Recently, there were reports of decreased spinal length gain from repeated lengthening of the growing rods.\textsuperscript{53,54} Sankar et al\textsuperscript{44} proposed a “law of diminishing returns” whereby the average T1-S1 gain from a given surgical lengthening decreased significantly with repeated lengthening. Forcing distraction in such cases would result in implant failure. This phenomenon was likely due to progressive stiffness or autofusion of the spine caused by sudden distractions.\textsuperscript{54}

**Magnetically Controlled Growing Rods**
With the limitations associated with the growing rod, natural development calls for the construction of an implant that allows less invasive distractions. A remotely distractable, magnetically-controlled growing rod system has been developed to facilitate out-patient rod distractions, eliminate frequent surgeries under general anesthesia and reduce wound complications, psychological and socioeconomic problems, and frequent hospitalisation in young children (Fig. 3). This procedure can also mimic normal physiological growth and provide continuous neurological monitoring in a conscious patient during distractions. In addition, frequent distractions may avoid the law of diminishing returns suggested by Sankar et al.\textsuperscript{54} The magnetically-controlled growing rod has already been validated in animal studies.\textsuperscript{55,56}

The first report of the safety and efficacy of this technology was reported by Cheung et al.\textsuperscript{57} Their study noted that the corrective power of the device was similar to the conventional growing rod at two year follow-up. It was found that consistent gains in spinal growth were obtained with each distraction. The monthly increase also matched the predicted monthly spinal growth in five- to ten-year-olds\textsuperscript{57} and other traditional growing rods.\textsuperscript{39} Furthermore, there were no major rod or wound complications and there were good clinical outcome scores. Wick et al\textsuperscript{58} also showed promising results with a similar magnetic growing rod (Phenix rod) in two case reports. As such, the preliminary findings of the magnetically-controlled growing rods in young children with early-onset scoliosis have shown to be safe and effective; however, longer and larger studies are warranted to further verify the current findings, assess complications on long-term follow-up, and refine patient selection.

**Conclusion**
Early-onset scoliosis is a difficult challenge. These patients are young with significant remaining growth potential. Thus, patients will likely develop progressive deformities, cosmetic disfigurement and cardiopulmonary consequences. Conventional management tools, such as spinal bracing and fusion, have not been shown to be effective. In addition, fusion may also cause significant inhibition of normal spinal and thoracic growth. Growing rods have been the mainstay treatment of early-onset scoliosis with the ability to allow periodical distractions to accommodate growth. However, these growing rods require repeated surgeries for distraction and are associated with exponential increase in complications, namely anesthetic and wound complications. With the development of the magnetically-controlled growing rod system, remote and non-invasive distractions can be performed, reducing the need for frequent follow-up surgeries and the complications associated with such treatment, quicker return to activities, decrease psychosocial consequences for child and parent, in the long-term are more cost-effective. However, as with any new technology, additional studies are needed to further reduce complications and improve outcomes in children undergoing treatment for early-onset scoliosis.
References

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