

An insight into early onset of scoliosis: new update information – A review

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Abstract. – Early-onset scoliosis is an onerous challenge to physicians. These patients are young with significant remaining growth potential. Thus, patients are likely to develop progressive deformities, cosmetic disfigurement and cardiopulmonary consequences warrant early intervention in many cases. The purpose of this review is to provide the readers with brief description of the disease, therapeutic modalities available and their indications and use. Publications and abstracts related to EOS in the last decade were carried out and synthesized into a review “an insight into early onset of scoliosis.” A comprehensive understanding of the scoliosis, its impact on the thoracic development may guide in treatment, which is often required at a young age in these children to prevent irreversible pulmonary insufficiency. Current treatment techniques are based on multiple factors may include non-surgical strategies, such as Derotational body cast or brace in younger patients with curve <50 degrees. Surgical treatment of spinal deformity should be considered when nonoperative measures are failed to arrest curve progression. Growing rods have been the mainstay treatment of early-onset scoliosis which require repeated surgeries for distraction and are associated with exponential increase in complications. The vertical expandable prosthetic titanium rib may be beneficial for those patients with congenital scoliosis and fused ribs and thoracic insufficiency syndrome. Shilla technique is an alternative to growing rods that avoids the morbidity of repeated lengthening. Growth modulation using staples or tethers shows promise for milder curvatures, but further follow-up is needed to define their use. Although new technologies have improved the treatment of children with EOS but it continues to be challenging with high complication rates.

Key Words:

Scoliosis, Bracing, Casting, Traction, Fusion less surgery, Vertical expandable prosthetic titanium rib.

Introduction

Structural scoliosis during infancy was initially described by Harrenstein¹ in 1936 implicating

rickets as the most likely cause in a majority of young children. Subsequently in 1954, James² described three types of idiopathic scoliosis according to the age of onset: 1) infantile denotes curves that develop at the age of three years or less, 2) juvenile between four and nine years of age and 3) adolescent between ten years of age until the end of growth. These three time periods correspond to distinct periods of growth during childhood and adolescence. The infantile and adolescent periods are marked by an increased growth velocity, whereas most of the juvenile period, in contrast, correlates with a deceleration of spinal growth, which is steady growth period and curve progresses slowly, thus making juvenile-onset scoliosis relatively rare form of the disorder^{3,4}. An alternative classification of scoliosis was coined by Dickson categorizing scoliosis into “early onset scoliosis” (EOS) and “late onset scoliosis”. EOS was defined as scoliosis present in children younger than 5 years of age including idiopathic, neuromuscular, congenital or syndromic scoliosis whereas the term late onset scoliosis was assigned to scoliosis at the age of six years and beyond⁵.

The age of onset of scoliosis remains a key factor in the management of this disorder as major thoracic deformity before five years of age is associated with twice the mortality rate compared to the general population because of the compromised cardiopulmonary reserve emanating from substantial thoracic deformity⁶. In addition, the traditional methods of definitive fusion for halting curve progression limit the growth of spine and lungs and may further impair respiratory function which contribute to thoracic insufficiency and increased mortality⁷⁻⁹. It was primarily due to the complications associated with early definitive fusion in young patients that innovative techniques were developed for controlling spinal deformity without impeding thoracic growth. This review addresses the etiology, pathophysiology, diagnosis and treatment options of growth sparing techniques of EOS.

Etiological and Epidemiological Considerations

Infantile idiopathic scoliosis (IIS), one of the many causes of scoliosis among patients of three years of age or less, comprises less than 1% of all cases of idiopathic scoliosis in the United States¹⁰. Although rare but it is more common in boys than girls (ratio, 3:2). The majority of the curves (75% to 90%) tends to be left-sided rather than right-sided and may be associated with congenital anomalies such as hip dysplasia, congenital heart disease, and mental retardation^{10,11}. Hooper¹² reported 6.4% prevalence of developmental dislocation of the hip among 156 patients with infantile scoliosis, which represents ten times the prevalence of developmental dislocation of the hip found in the general population. Ceballos et al¹³, on the other hand, reported an even greater (25%) prevalence of hip dysplasia. Compared to USA, the incidence of IIS before the age of 2 years appears to be higher in England¹⁴. Similarly the prevalence of infantile idiopathic scoliosis as high as 12.8% reported from Europe appears to be much higher than 0.25% prevalence reported from North America^{1,11,12}. Recently a declining trend in the prevalence of IIS has been reported from Edinburgh which is comparable to prevalence of IIS in Boston¹⁵. This has been attributed to either removal of adverse or introduction of unknown beneficial environmental factor in Edinburgh as both Boston and Edinburgh share common genetic pools. These regional variations indicate that environmental factors which remain to be identified may be involved in the development of IIS.

Early onset scoliosis appears to have a multifactorial etiology. Initially intrauterine molding theory was hypothesized by Browne¹⁶ which was thought to be the main cause of not only IIS but was also believed to be responsible for accompanying deformities such as plagiocephaly, decreased hip abduction or rib molding¹⁷. This hypothesis was later fortified by Mehta¹⁸ and Lloyd-Roberts and Pilcher¹⁹. These beliefs were subsequently set aside because of the absence of scoliosis and other associated abnormalities at birth and developed later. Following this Mau²⁰ proposed the postnatal external pressure molding theory as a cause of IIS in Europe because of the traditional oblique supine placement of the infant at night as opposed to North America where the infant was usually placed in prone position while asleep. This trend of placing the infant in prone

position later shifted to supine position in accordance with the recommendations of American Academy of Pediatrics to avoid the risk of sudden infant death syndrome²¹. In order to testify Mau's hypothesis, a current estimate of the prevalence of EOS in the USA will provide very useful information. Moreover Wynne-Davies also supports postnatal pressure theory based on the observation that out of 134 babies 97 developed scoliosis and plagiocephaly during the first six months of post natal period²².

A number of population based studies have implicated hereditary factors as a higher prevalence of IIS among relatives of patients with scoliosis has been reported compared to the general population²³. Similarly the likelihood of having scoliosis among siblings and the parents of patients with late onset scoliosis is thirty time higher than the controls²². Despite these observations supporting the involvement of hereditary factors as etiological agents a definite pattern of inheritance remains obscure. There is, however, some evidence that in idiopathic scoliosis, the transmission pattern may be dominant or there is either an autosomal or multiple-gene inheritance pattern¹¹. Similarly Robin and Cohen²⁴ while investigating the pattern of inheritance of adolescent idiopathic scoliosis over five generations within a family found that the pattern of inheritance is either autosomal or multiple-gene inheritance. In addition, Wynne-Davies²² reported prevalence of scoliosis as 2.6% among the first degree relatives of early onset patient which was higher than that in general population (0.39%); however, the association was not strong. Large scale population based studies have revealed that 11% of first-degree relatives of patients with scoliosis, 2.4% of second-degree and 1.4% of the third degree relatives tend to have scoliosis^{10,11,25}. Studies on identical (monozygous) twins exhibit a 73% and fraternal (dizygous) twins show 36% concordance rate²⁶⁻³⁰. Genome-wide scanning and linkage analyses of the families with scoliosis have shown potential linkage to regions on the autosomes^{31,32} where candidate regions on chromosomes 6,9,16 and 17 have been shown to have strong evidence for linkage³³. Genetic locus on 19p13 chromosome has been implicated in the etiology of scoliosis^{34,35}. In addition DNA markers associated with progression to severe curve in idiopathic scoliosis have also been identified³⁶.

There is no strong scientific evidence implicating any particular biomechanical factor in the eti-

ology of EOS or adolescent scoliosis. A possible role of growth hormones or growth-modulating chemical factors (e.g., melatonin and clamodulin) cannot be ruled out³⁷. The reported abnormalities of connective tissue, platelets, skeletal muscle, spinal column and rib cage are all thought to be secondary to the deformity itself^{38,39}.

Pathophysiology of EOS

Spinal deformities are known to adversely affect thorax development by changing its shape and reducing its normal mobility. Intact rib-vertebral-sternal complex fits the thorax three-dimensionally into an elastic cube shape cavity. In the presence of scoliosis this cavity becomes a flat, rigid and adopts an elliptical shape preventing the expansion of lungs⁴⁰. These deformations adversely affect the patients though yet to be discovered mutual interactions and influences among various skeletal and organic components^{41,42}. There is, however, sufficient evidence suggesting a relationship between the growth of spine and progression of spinal deformity resulting in alterations in the size and shape of the thoracic cage^{43,44}. Respiratory failure is the main cause of high mortality rate among patients with untreated infantile-onset scoliosis and juvenile-onset scoliosis⁴⁵ resulting not only from the extrinsic disturbance of the chest wall function but also from the intrinsic factor in the form of associated alveolar hypoplasia⁴⁶.

This extrinsic disturbance of respiratory function due to congenital and acquired chest wall, spine and other syndromic deformities, has an adverse effect on the function and growth of the lungs. If these deformations are left untreated at young age, risk of suffering from rapid deformity progression, deformed chest, compression on pulmonary parenchyma, pulmonary hypertension and cor-pulmonale is significantly higher⁴⁷. The condition in which thorax is unable to support normal respiration or lung growth is referred as *thoracic insufficiency syndrome*⁴⁸. In the recent years the focus has been shifted from spine alone to spine, chest wall and lung because of the fact that treatment directed towards well aligned spine and a thoracic cavity support adequate pulmonary development and function⁴⁰. Spinal growth at the rate of 2.2 cm/y achieves maximum length by the age of 5 years from T1 to S1 that slows down during the next 5 years to 1 cm/y followed by another peak growth after the age of 10 years with the growth velocity of 1.8 cm/y^{3,49}. At-

tainment of T1-T12 length of at least 18 cm at maturity is considered to support an adequate pulmonary function⁹. The circumference of thorax which is only 7% of adult size at birth, increases to 30% by age 5 years and 50% by 10 years⁴⁹. The greatest increase in the number of alveoli in normal children occurs in the first two years of life⁵⁰ and subsequent expansion of lung volume up to age 5 years occurs by a rapid increase in peripheral airway conductance that accompanies airway enlargement^{51,52}.

Most of the researchers^{3,49,53-55} agree that lung growth is essentially complete by 8 years of age with a "golden period" of maximum growth occurring before the age of 5 years thus deformities developing after this age have little effect on lung growth^{6,56}. This golden period of alveolar hyperplasia is accompanied by a rapid growth of the bony thorax as well⁴⁹. It is, therefore, important to preserve both thoracic growth and lung volume during this critical period of life. Patients with early onset deformities have been shown to have fewer alveoli than expected due to failure of alveolar multiplication and are more likely to be victims of respiratory demise. Post mortem studies also have suggested that the mechanical compression of the alveoli due to chest wall deformity, is not a factor in reduction in the number of alveoli and this is probably due to premature cessation of alveolar proliferation^{55,57}.

Clinical and Radiographic Assessment

Clinical Evaluation

Patient evaluation should begin with history focusing on prenatal and perinatal complications and hospitalizations for intensive care needs and other pulmonary issues. Parents should be interviewed about the discovery of the curvature and the rate of progression. Frequently occurring EOS associated co-morbidities such as neuromuscular, dysplastic, or syndromic conditions should also be characterized. An assessment of the general health of the patient may help in early understanding of the overall effect of the disease process. Physical examination is mandatory for analysis of the spinal deformity and exclusion of associated conditions. Initial inspection should include the skin, the entire spine, the head, the pelvis, and the extremities. Examination of skin may reveal cutaneous abnormalities such as café au lait spots or axillary freckles as seen in neurofibromatosis, midline patches of hair as seen in

spinal dysraphism, or bruising as seen with trauma. The spinal examination should include inspection and palpation of the spine. Examination of children with EOS may be difficult because of age imposed limitations and different techniques must be applied. In young children, the Adams forward bend test is not possible, but physicians should assess the flexibility of the curvature by holding the infant prone on their knees with convex side downward. If the child is walking, then one may suspend the child by holding under the arms and assess the correction of curve with lateral pressure.

In addition, physical examination must also include notation of chest or flank asymmetry, chest excursion, and abdominal reflexes. Limitation in chest excursion may indicate syndromic scoliosis and thoracic insufficiency syndrome⁴⁸. Abdominal reflex abnormalities should initiate a thorough neurologic evaluation to rule out neurological causes of scoliosis (e.g., syringomyelia or Chiari 1 malformation)⁵⁸. It is also important to thoroughly examine the head. Plagiocephaly, bat-ear deformity and congenital muscular torticollis are common conditions affecting IIS. Although these conditions frequently occur without scoliosis, it is important to be aware of the association.

Examination of the pelvis should be done to rule out developmental hip dysplasia, which is associated with infantile idiopathic scoliosis¹⁶. The lower-extremity examination must exclude limb-length inequality as the cause of scoliosis. When scoliosis is secondary to a limb-length inequality, the lumbar prominence is found on the side of the longer limb. The diagnosis of functional scoliosis due to limb-length inequality is confirmed by having the patient perform a sitting forward bend or by placing a lift under the short limb to equalize the limb lengths.

Radiographic Evaluation

Anteroposterior and lateral radiographs of the entire spine at initial visit to evaluate both the Cobb angle and rib-vertebral angle difference (RVAD) should be performed to rule out congenital vertebral abnormalities. In children who are too young to stand, the radiographs should be made with the child placed supine. The cervical spine should be evaluated for fusions and instability. The lumbosacral junction, hip joints and pelvis should be carefully examined to rule out congenital anomalies or developmental hip dys-

plasia. Measurement of the Cobb angle is performed to determine the magnitude of the spinal deformity⁵⁹. The (RVAD) is a method available to predict curve progression or resolution in children with infantile idiopathic scoliosis (Figure 1).

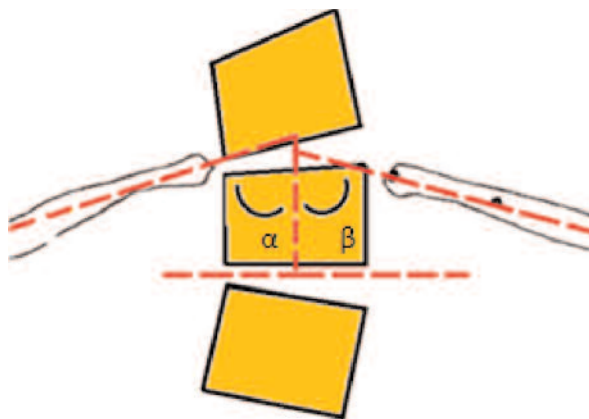
Mehta⁶⁰ in 1972, described the method that now bears her name. This measurement is performed by drawing a line perpendicular to the endplate of the most translated apical vertebrae and then a line down the concave and convex rib at this same level. The angle created on the convexity is then subtracted from that on the concavity to create the RVAD. The RVA difference (RVAD) is the difference of the angle created on the convexity is, then, subtracted from that on the concavity and is useful in distinguishing resolving versus progressive curves.

Mehta⁶⁰ found that a RVAD less than 20° indicates a curve that is likely to resolve (85% to 90% of patients), while an RVAD of greater than 20° is frequently associated with a curve that will progress.

Mehta⁶⁰ also introduced new radiographic criteria based on the relationship of the apical ribs with the apical vertebra known as the “phase of the rib head” useful for prognostication of infantile scoliosis (Figure 2). A “phase-1” relationship indicates that the rib head is distinct from the upper corner of the apical vertebra on a coronal radiograph and the curves that have a phase-1 relationship, the RVAD may be calculated and used to determine the likelihood of progression. In a phase-2 relationship, the head of the rib on the convex side of the apical vertebra overlaps the upper vertebral corner and the RVAD is not measured because the curve is certain to progress.

In the initial description of the RVAD and phase of the rib head, Mehta⁶⁰ studied 138 cases of IIS, 83% resolution in phase-1 relationships with an RVAD of $\leq 20^\circ$. Conversely, 84% of the group that progressed had an RVAD of $\geq 21^\circ$. Ferreira and James⁶¹ confirmed these findings in their own patient population and reported results, curves in patients with phase-1 ribs and of less than 20° RVAD resolved in 99% of patients, whereas 98% of patients with an RVAD of greater than 20° or a phase-2 relationship progressed.

Computed tomography (CT) provides unique information that can be invaluable in evaluation of congenital curves or curves with complex anatomy of the spine as well as the thoracic cage and lung volumes⁶². Three dimensional reconstructions have been utilized to understand the deformity in all planes as well as the pedicle



α = Rib Vertebral Angle (RVA)
 $\beta - \alpha$ = Rib Vertebral Angle Difference (RVAD)

Figure 1. Rib Vertebral Angle Difference (RVAD). The rib vertebral angle (RVA) measures the angle formed by a line perpendicular to the endplate of the most apically translated vertebrae and a line drawn down the middle of the concave and convex ribs. The RVA difference is the difference between these angles.

morphology for choice of the implants. A whole-spine MRI scan should be obtained on all patients with EOS as approximately 20% of these children will have an underlying intraspinal anomaly^{63,64}. Future applications of MRI include functional MRI and dynamic MRI, both of which may provide real-time understanding of lung function and the impact of thoracic deformity on the lung⁶⁵.

Treatment

Nonoperative Delaying Tactics

Among patients at risk for thoracic insufficiency delay in surgical interventions until the age of 10 years, once sufficient growth of spine has occurred, is usually the foremost non-operative approach if possible⁶⁶. Management principles are based on age of the child, the magnitude of the curvature and RVAD measured on plain radiographs. Most patients are at low risk of progression if the Cobb angle is under 25° with an RVAD of less than 20°. Clinicians should observe these patients with serial radiographs every four to six months¹³. If the progression of more than 10° was observed at any stage during the follow-up, then Active treatment should be initiated. If there was minor or no progression then

the curve may undergo resolution. Once curve is resolving, one to two-year follow-up may be sufficient to watch for any possible recurrence during the adolescent growth spurt.

Various nonoperative treatment approaches to early onset scoliosis include bracing, derotation cast and traction.

Bracing

In the absence of coexisting neuromuscular or other medical conditions adversely affecting respiratory function by circumferential chest or abdominal compression bracing is considered as a traditional and a time-honored method for controlling non-congenital deformities in EOS. The success rate in preventing progression of spinal curve is variable as most curves may exhibit a transient radiological improvement after application of well-constructed orthoses, even amongst very young patients that may progress with future growth. A brace functions predominantly to stabilize rather than reverse the deformity and thus is less likely to result in permanent correction of spinal curvature than serial casting in patients with infantile idiopathic scoliosis. Bracing is more commonly used in older patients with larger curves that would not be expected to correct with casting. It is also indicated in patients with progressive scoliosis and who are unable to tolerate casting, and also among the patients where surgical intervention is contraindicated for medical or other reasons. In children with curvatures of over 35° bracing can be considered successful if progression is prevented for several years, thereby, delaying the need for fusion until

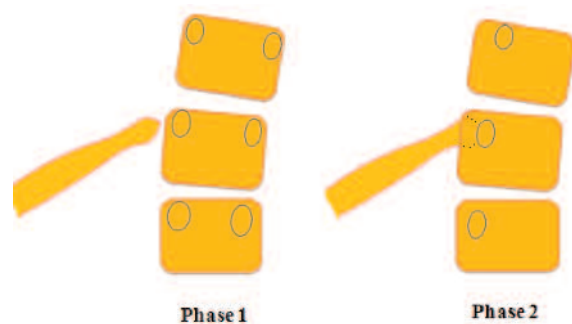


Figure 2. Phases of curve patterns. Phase 1: no overlap of rib head and vertebral body. Phase 2: note the overlap of the rib on the vertebra.

the child is older. If fusion can be delayed until 10 years of age, the need for anterior fusion may be avoided⁶⁷. The brace is worn for at least 23 h per day and for at least 2 years until there is no evidence of curve progression. If there is progression of more than 5° in these patients then active surgical treatment should be initiated. The Milwaukee brace is preferred to an underarm thoraco-lumbo-sacral orthosis (TLSO) in early-onset patients because of its ability to apply corrective forces directly at the apex of the curve, at the pelvis, and at the neck while minimizing the constrictive aspects of a TLSO.

Casting

Before the advent of effective spinal instrumentation cast treatment used to be the standard treatment for all patients with scoliosis and was associated with several complications including superior mesenteric artery syndrome, pressure sores and adverse effects on cranial nerves and brachial plexus if the neck portion was applied with too much head or shoulder distraction^{68,69}.

These problems were addressed by the replacement of the Risser cast, which was based on a 3-point bending principle with the de-rotational technique introduced in the 1960s in Europe and recently popularized by Mehta⁷⁰.

Serial de-rotation cast treatment has an effective growth-preserving strategy in the early management of EOS with benefits of improved patient compliance, reduced risk of spontaneous fusion, and avoidance of complications associated with serial lengthening procedures^{70,71}. Common indications for serial casting include, progression of 10-20 degrees, progression past 20 degrees with low magnitude (<50 degrees) coronal deformity and anticipated progression (RVAD of >20 degrees, rib phase 2) in whom no surgical intervention have been attempted. A series of casts applied under anesthesia can provide significant curve correction and may improve the flexibility of the spine as well. When the child resumes brace wear after several months in a cast or casts, the deformity is of lesser grade (<10 to 20 degrees), the brace is more likely to be comfortable in controlling the curve. In addition, because of the cast being equivalent to a full-time brace many parents prefer using it. Casting eliminates the problem of poor compliance and the difficulty of donning braces in uncooperative young children.

Currently, the evidence supporting de-rotational casting in non-idiopathic infantile scoliosis

is limited. Elongation Derotation Flexion (EDF) casting has shown to be curative in patients aged less than 17 months with relatively small curvatures⁷¹. Curative casting has been reported in some centers for older children with larger curves as a part of delay tactic devised to put off surgery as long as possible rather than a curative treatment. This is particularly important given the “law of diminishing returns” noted by Sankar et al⁷², whereby a steady decrease in spinal length is obtained with each subsequent lengthening using growing rods. Sanders et al⁷¹, have recently reported good results with casting as a delay tactic in a large series of children with EOS. Fletcher et al⁷³, concluded that serial casting is a viable alternative to surgical growth sparing techniques in moderate-to-severe early-onset scoliosis and may help delay eventual surgical intervention.

Traction

Despite the fact that traction is rarely used today, it does remain an important option for the treatment of children with spinal disorders⁷⁴. Halo-gravity traction is an invaluable method to restore coronal and sagittal balance in severely decompensated curves and may decrease the neurologic risks associated with the surgical correction of these severe deformities, be it by casting, subcutaneous rods, VEPTR, or definitive instrumentation and fusion. Patients who have stiff curves or severe spinal deformities causing secondary cor-pulmonale may benefit from a period of pre-operative traction to allow aggressive pulmonary toilet. It may help in reducing postoperative complications by Improvements in vital capacity, arterial blood gases, and right heart failure^{47,75,76}. Patients with progressive deformities with weakness, skin or chest wall defects, or mental retardation are unsuitable candidates for casting. In addition large, stiff curves may not benefit from serial casting, and the cast may be poorly tolerated. In these instances, halo-gravity traction is an invaluable method to achieve correction of the deformity and improvement in respiratory mechanics⁷⁶.

Operative Treatment

The surgical management of EOS aims is to check progression of the curve while allowing maximum growth of the spine, lungs, and thoracic cage. It is usually recommended for progressive curves of $\geq 45^\circ$ in an immature child

with infantile idiopathic scoliosis⁷⁷. The choice of surgery depends on the age of the patient, type, severity and progression of curve. Several procedures have been described and practiced at various centers around the world. Historically, the treatment for early-onset scoliosis has been posterior spinal fusion to halt curve progression. Isolated posterior spinal fusion in young children was found to be unsuccessful for long-term correction of deformity. This was mainly due to crankshaft phenomenon resulting from continued anterior growth of the spine following successful posterior fusion alone in skeletally immature patients^{43,78}. Sanders et al⁷⁹, reported that patients who have open triradiate cartilages and are at Risser grade-0 spinal maturity have a high likelihood of curve progression due to the crankshaft phenomenon following isolated posterior fusion. In order to prevent the crankshaft phenomenon in patients with an open triradiate cartilage, an anterior arthrodesis must be included with the posterior fusion. A preferable successful outcome was believed to be a straight and a shortened spine rather than a deformed spine of near normal length. However, anterior and posterior fusion results in considerable loss of both spinal growth and sitting height. Because of the two stages of maximal spinal growth velocity occurring at zero to five years of age and at ten to fifteen years of age, early fusion for EOS may result in substantial loss of expected height. In addition to the detrimental effects on spinal growth, premature fusion may also have potentially deleterious effects on developing thoracic cage and lungs. In the backdrop, the limitations of early fusion, current research has focused on development of fusionless methods, with limited anchoring to the spine that could periodically be lengthened with additional surgical intervention.

Fusion Less Surgery

A number of spinal and chest wall fusion less techniques have been developed and refined to control growth, to delay definitive fusion surgery, and to increase the thoracic volume. Most widely used techniques are dual growing rods and the vertical expandable prosthetic titanium rib device, guided-growth systems, and vertebral staples, which represent an effort to avoid the potentially severe respiratory effects associated with early spinal fusion, that limits spinal growth⁸⁰.

Growing Rods

The concept of a rod that is lengthened at regular intervals after initial implantation was introduced over 25 years ago⁸¹⁻⁸³. Current growing rod techniques combine the principles of fusion less surgery to control curve magnitude while achieving spinal growth using modern implant systems. More recently, Akbarnia and McCarthy⁸⁴ developed fusion less 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. 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 is reopened for the distraction procedure. Once maximum spinal growth or skeletal maturity is achieved, definitive final fusion is performed. This approach of using growing rods is not only useful in controlling the progression of spinal deformity but can also gradually correct the deformity⁸⁵⁻⁸⁷. Akbarnia and McCarthy⁸⁴ reported the results of a multicenter study of 23 patients with 2 years follow-up where curve magnitude improved from 82 degrees preoperatively to 38 degrees after the first surgery and 36 degrees at final follow-up. Spinal growth averaged 1.21 cm per year with similar improvements in the space available for the lung. Patients were lengthened a mean of 6.6 times and initial surgery was at 5 years 5 months of age. Eleven of twenty-three patients suffered a complication during treatment. In addition, Thompson et al⁸⁸ found that using dual rods resulted in fewer complications and provided better correction due to the increased rigidity in the system when compared to single rods.

Complications are frequent and are related to the prolonged treatment required by distraction-based techniques and repeated anaesthesia. Despite the advantages of growing rod surgery, the need for repeated surgeries under general anaesthesia is a major drawback due to increased anaesthetic and wound complications^{89,90}. Other complications include proximal migration of devices with rib fractures, and skin problems secondary to recurrent surgery and hardware^{87,91}. The complication rates range from 8% to 50%^{82,87,91}. Instrumentation failure included anchor failure or rod fracture and the overall rate

was reported to be 15%⁹². Junctional kyphosis and curve decompensation have also been rarely reported⁸⁴.

Despite the apparent successes with this technique, significant obstacles still limit its overall effectiveness in managing scoliosis in the very small or young child. Perhaps the most concerning evidence was reported by Sankar et al⁷², who demonstrated a “law of diminishing return” that crescendos after seven lengthening. This phenomenon was likely due to progressive stiffness or autofusion of the spine caused by sudden distractions⁷². Similarly Nordeen et al⁹³ found that the force required to distract the spine doubles by the fifth lengthening procedure with less than 8 mm of spinal growth achieved with each lengthening after this point. Distraction forces were 40% higher in patients with apical fusions in addition to growing rods.

Vertical Expandable Prosthetic Titanium Rib (VEPTR)

Vertical expandable prosthetic titanium rib (VEPTR), made by Synthes Spine of West Chester (PA, USA) is a titanium alloy longitudinal rib distraction device. VEPTR implantation is a technique that is aimed at correcting the chest wall deformities along with scoliosis, which may be either, congenital, syndromic or early-onset idiopathic types. Thoracic spine and chest wall deformity are usually correlated; therefore, elongation of the chest wall will increase the space available for the lung and improve respiratory mechanics in patients with thoracic insufficiency syndrome (TIS)⁹⁴⁻⁹⁶. This vertically placed device uses distraction to indirectly elongate the spine and chest, stabilizing the progression of the spinal deformity while preserving spinal growth. The device may be attached from rib to rib, rib to spine, or rib to pelvis⁹⁴. By limiting the exposure of spine to maintain normal anatomy, it is also viable option for patients with IIS with larger, stiffer curves.

The use of VEPTR is perhaps best supported in conditions such as scoliosis with multiple rib fusions, absent ribs, or severe thoracic hypoplasia, which were all previously associated with a high risk of early mortality and otherwise not amenable to more traditional bracing, casting, or even growing rod techniques.

Campbell et al⁹⁶ reported on 27 patients treated with open wedge thoracoplasty and found a mean correction of the scoliosis from 74 to 49 degrees, and thoracic spine height increased by a

mean of 7.1 mm/y. Common complications included 1.9% having infections, 15% having skin sloughing requiring rotational flaps, two patients having brachial plexopathy and “asymptomatic” proximal migration of the device through ribs in 7 patients. Emans et al⁹⁷ also follow pulmonary development using CT scan in 31 patient who had fused ribs and scoliosis that had caused TIS. Lung volumes, pulmonary function and Spinal growth all were improved using VEPTR expansion thoracoplasty.

Hasler et al⁹⁸ studied 23 children with noncongenital scoliosis and found a significant increase in space available for lungs (SAL) and mean correction of Cobb from 68 degrees preoperatively to 54 degrees at the final follow-up. Smith et al⁹⁹ used VEPTR like the growth rods of Akbarnia and colleagues by fastening the proximal anchors to the ribs rather than to the spine. This procedure was adapted with an assumption that the technique would result in less autofusion and scarring allowing maximal growth, without disturbing the native spine.

The use of VEPTR for idiopathic scoliosis (Figure 3), particularly with associated kyphosis remains controversial. Proponents of its use highlight the positive impact on pulmonary growth as measured by space available for the lung and a lower complication rate than in sicker patient populations⁹⁸⁻¹⁰⁰. On the contrary, the critics have noted that early implantation in healthy children may lead to decreased chest compliance as the thoracic cage stiffens. Others⁷³ have noted the high level of complications seen in some series and continue to emphasize the need for delayed surgery in an attempt to minimize the number of procedures.

The use of VEPTR in myelomeningocele both for distal kyphosis (gibbus) deformity and neuromuscular scoliosis has been recommended¹⁰¹. The use of VEPTR in kyphoscoliosis has also been fraught with complications as proximal junctional kyphosis often develops and proves extremely difficult to treat. Use of hybrid systems with proximal up-going rib hooks or proximal fixation on the second rib may prove to be more suitable in this situation^{100,102}. Future research is required to define the use of chest wall procedures in this patient population.

Alternative Techniques and Future Directions

Physicians continue to seek surgical options that do not require regular lengthening proce-

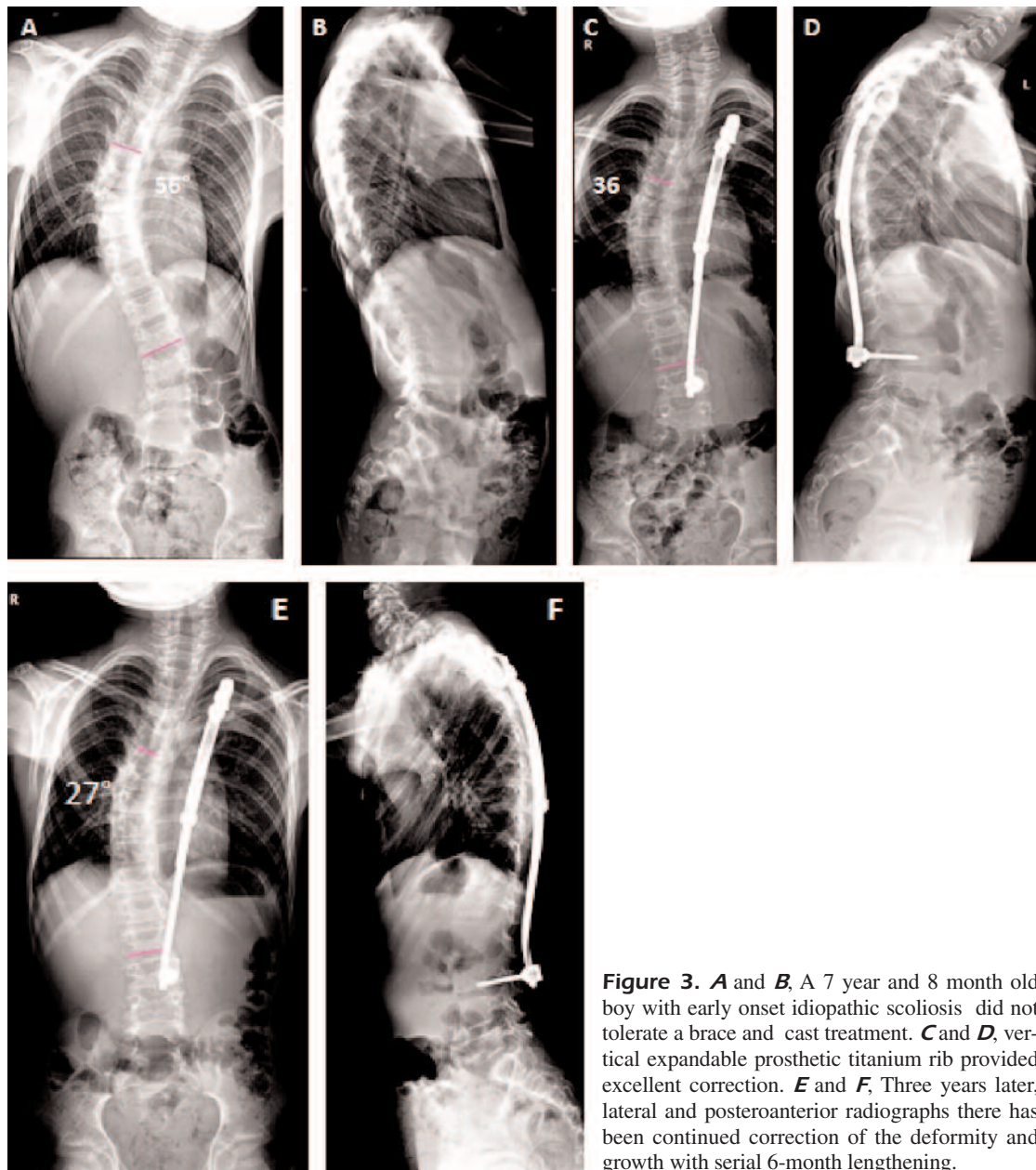


Figure 3. *A* and *B*, A 7 year and 8 month old boy with early onset idiopathic scoliosis did not tolerate a brace and cast treatment. *C* and *D*, vertical expandable prosthetic titanium rib provided excellent correction. *E* and *F*, Three years later, lateral and posteroanterior radiographs there has been continued correction of the deformity and growth with serial 6-month lengthening.

dures and to construct an implant that allows less invasive distractions. Reported techniques for human use are externally controlled expandable devices and guided growth techniques include hemiepiphysiodesis, and spinal tethers and Shilla.

Magnetically Controlled Growing Rods

A remotely distractible, magnetically-controlled growing rod system [Phenix and MAGEC

(Magnetic Expansion Control System) Ellipse Technologies, Inc., Irvine, CA, USA] has been developed to facilitate out-patient rod distractions, eliminate repeated surgeries required with the usual distraction methods, under general anesthesia, and frequent hospitalization in young children (Figure 4). This procedure theoretically diminishes the complications of wound infection. It may be especially helpful in children with comorbidities and those with psychological and so-

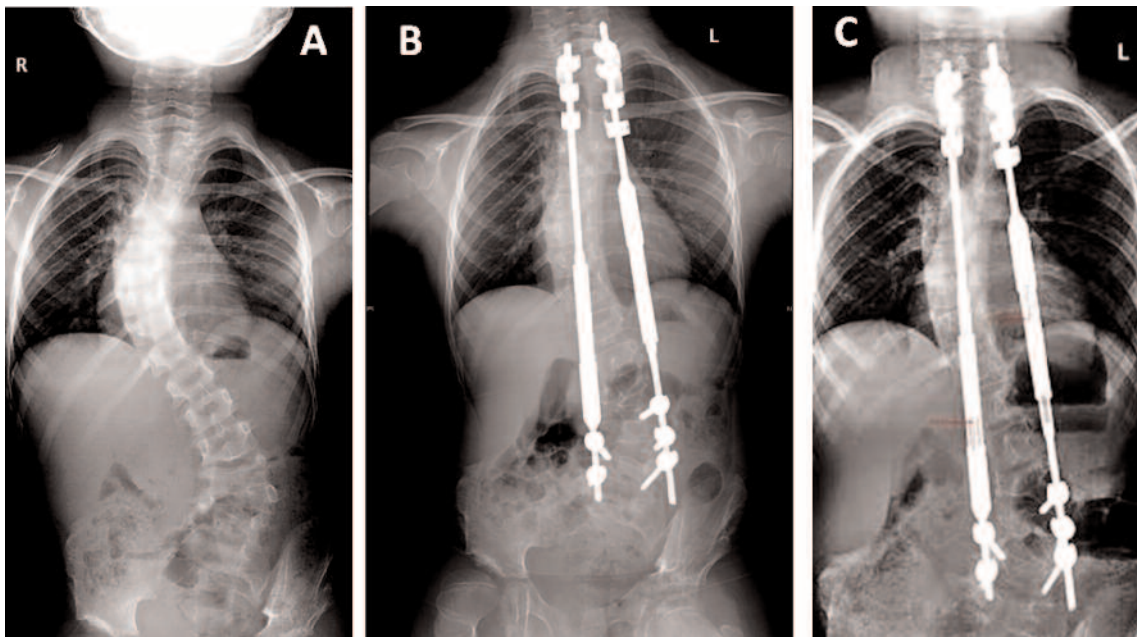


Figure 4. A 4 years old girl with early-onset scoliosis who underwent curve correction management with bilateral magnetic remote-controlled growing rods. Posteroanterior **A**, view of preoperative spine; immediately postoperative spine **B**, and spine at 2 years follow-up **C**.

cioeconomic problems. In addition, frequent distractions may avoid the law of diminishing returns suggested by Sankar et al⁷². The magnetically-controlled growing rod has already been validated in animal studies by Akbarnia et al, using MAGEC system^{103,104}. The Phenix rod system, was developed by Drs Lotfi Miladi, Arnaud Soubeiran, and Jean Dubousset that uses magnets to produce gradual lengthening without repeated surgeries allowing rib or spine fixation. In a preliminary study¹⁰⁵, there was correction of the scoliosis from 63 to 33 degrees and a growth rate of 2 mm/mo.

The first report of the safety and efficacy of this technology was published by Cheung et al¹⁰⁶. 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-olds³ and other traditional growing rods⁸⁴. Furthermore, there was no major rod or wound complications with favorable clinical outcome scores. Wick et al¹⁰⁷ also showed promising results with a similar magnetic growing rod (Phenix rod) in two case reports. Large scale follow up studies are needed to determine

the efficacy and safety of this technology and for verification of current findings, assessment of complications on long-term follow-up and refinement of patient selection.

Convex hemiepiphysiodesis

Convex hemiepiphysiodesis, commonly used for multilevel congenital deformities, is a safe but somewhat unpredictable method of “guiding” spinal growth. The concept of hemiepiphysiodesis is based on the Hueter-Volkman Law, involving inhibition of growth by compression of the convexity of the deformity to arrest growth on the convexity of the deformity, allowing the concave side to grow. More than forty years ago, Roaf¹⁰⁸ proposed that the deformity seen in early onset scoliosis was due to asymmetric growth of the convex (faster-growing) and concave (inhibited) sides of the curve.

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, hemivertebral

resection, and short segment fusions can be used in selected patients to allow for gradual correction of these curves.

Staples

Biocompatible shape-memory metal alloy staples have been used in the clinical practice. However, these techniques are still controversial with mixed results at short-term follow-up. One study by Marks et al¹⁰⁹, reported on convex epiphysiodesis with or without Harrington instrumentation and found no significant improvement in the measured deformity. They recommended instrumentation at the time of convex epiphysiodesis as the best option to control but not reverse the progression of the curve. In contrast, Betz et al¹¹⁰ reported that staples placed thoracoscopically for convex epiphysiodesis could control curve progression. In their study, six out of ten patients with a mean curvature of 35° were controlled and stabilized during one year follow-up period.

Spinal tethers

This comprises a group of devices that can limit growth on the convex side of progressive curves, both those in the frontal plane, involving scoliosis, and those in the sagittal plane, involving kyphosis. Efficacy of growth modulation by using a flexible spinal tether has been shown in animal models¹¹⁰⁻¹¹³.

Luque trolley and Shilla technique

The precursor to the Shilla system was the Luque trolley system^{81,82,109,114,115} which was described initially in 1977 by, Luque and Car-

doso. It did not appear to provide reliable spinal growth or deformity correction¹¹⁵. In the Shilla procedure, multiplanar correction is obtained by pedicle fixation at the apex of the deformity. This usually involves an apical fusion with non-locking polyaxial screws proximally and distally to guide a rod that is purposefully left long to minimize the need for subsequent surgery as spinal growth occurs, the rod slides through the non-locking screws. The effectiveness of this concept has been proven in a caprine model¹¹⁶. The Shilla technique has no long-term follow-up series to date. So far, the results of 10 patients with more than 2 year follow-up from a cohort of 36 patients have been reported¹¹⁷. Curve magnitude was reduced from a mean of 70.5 degrees preoperatively to 27 degrees postoperatively and 34 degrees at 2 years with increment in truncal height to a mean of 12% and the SAL increased to an average of 13%. Recently, with modern Luqué trolley technique (Figure 5) Jean Ouellet¹¹⁸ combined stable base (end vertebrae) fixation with apical control and gliding anchors, permitting self-lengthening of the construct by guided growth and avoiding repetitive and scheduled surgeries. In this small series of five patients, only one was an outright failure, suggesting the initial experience was favorable enough to justify this preliminary report and continue evaluating this procedure.

Fusion Surgery

For early-onset scoliosis, early definitive fusion was advocated in the past for children with severe

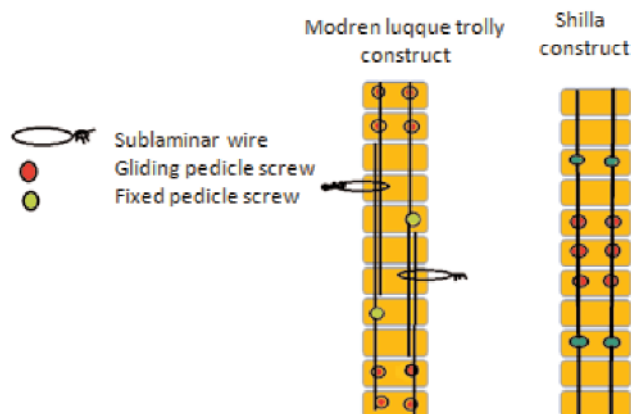


Figure 5. Schematics Illustrations show the two different self-lengthening growing constructs.

curves, deteriorating congenital spinal deformities, or progression despite bracing^{119,120}. This traditional posterior spinal arthrodesis with or without instrumentation in this age group should be supplemented by anterior arthrodesis to prevent crankshaft phenomenon⁷⁸. Circumferential arthrodesis is likely to halt curve progression and 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 young patients⁸⁸. Children in whom spinal fusion may be indicated is often based on the type and location of the anomaly and the age of the patient. A limited apex arthrodesis and/or resection may be possible if the deformity is limited to a few segments and still allow growth of other spinal segments. Definitive Spinal fusion becomes appropriate when the patient has achieved sufficient spinal length and thoracic volume to stop growth of the spine and thus achieve permanent correction. Timing of the procedure is controversial, but in general, patients who are at least 10 years of age and have completed the greatest part of their thoracic growth, are considered suitable candidates for definitive fusion to finish their scoliosis treatment⁶⁶.

Conclusions

Understandings of current growth sparing surgical techniques have improved the morbidity and mortality of children with progressive EOS. Treatment continues to be challenging with high complication rates because of the repetitive nature of the lengthening surgeries, additional studies are needed to further reduce complications and improve outcomes in children undergoing treatment for EOS.

References

- 1) HARRENSTEIN RJ. Die Skoliose bei Säuglingen und ihre Behandlung. *Zeitschr Orthop Chir* 1929; 52: 1-40.
- 2) JAMES JI. Idiopathic scoliosis; the prognosis, diagnosis, and operative indications related to curve patterns and the age at onset. *J Bone Joint Surg Br* 1954; 36-B: 36-49.
- 3) DIMEGLIO A. Growth of the Spine Before Age 5 Years. *J Pediatr Orthop B* 1992; 1: 102-107.
- 4) DICKSON R. Early-onset idiopathic scoliosis. In: Al-Arrayad S (ed). *The Pediatric spine: principles and practice*. Raven Press: New York, 1994, pp. 421-429.
- 5) DICKSON RA. Conservative treatment for idiopathic scoliosis. *J Bone Joint Surg Br* 1985; 80: 176-181.
- 6) BRANTHWAITE MA. Cardiorespiratory consequences of unfused idiopathic scoliosis. *Br J Dis Chest* 1986; 80: 360-369.
- 7) GOLDBERG CJ, GILIC I, CONNAUGHTON O, MOORE DP, FOGARTY EE, CANNY GJ, DOWLING FE. Respiratory function and cosmesis at maturity in infantile-onset scoliosis. *Spine* 2003; 28: 2397-2406.
- 8) VITALE MG, MATSUMOTO H, BYE MR, GOMEZ JA, BOOKER WA, HYMAN JE, ROYE DP. A retrospective cohort study of pulmonary function, radiographic measures, and quality of life in children with congenital scoliosis: an evaluation of patient outcomes after early spinal fusion. *Spine* 2008; 33: 1242-1249.
- 9) KAROL LA, JOHNSTON C, MLADENOV K, SCHOCHET P, WALTERS P, BROWNE RH. Pulmonary function following early thoracic fusion in non-neuromuscular scoliosis. *J Bone Joint Surg Am* 2008; 90: 1272-1281.
- 10) RISEBOROUGH EJ, WYNNE-DAVIES R. A genetic survey of idiopathic scoliosis in Boston, Massachusetts. *J Bone Joint Surg Am* 1973; 55: 974-982.
- 11) WYNNE-DAVIES R. Familial (idiopathic) scoliosis. A family survey. *J Bone Joint Surg Br* 1968; 50: 24-30.
- 12) HOOPER G. Congenital dislocation of the hip in infantile idiopathic scoliosis. *J Bone Joint Surg Br* 1980; 62-B: 447-449.
- 13) CEBALLOS T, FERRER-TORRELLES M, CASTILLO F, FERNANDEZ-PAREDES E. Prognosis in infantile idiopathic scoliosis. *J Bone Jt Surg* 1980; 62: 863-875.
- 14) SCOTT JC, MORGAN TH. The natural history and prognosis of infantile idiopathic scoliosis. *J Bone Joint Surg Br* 1955; 37-B: 400-413.
- 15) McMASTER MJ. Infantile idiopathic scoliosis: can it be prevented? *J Bone Joint Surg Br* 1983; 65: 612-617.
- 16) BROWNE D. Congenital postural scoliosis. *Proc R Soc Med* 1956; 49: 395-398.
- 17) BROWNE D. Congenital deformities of mechanical origin. *Arch Dis Child* 1955; 30: 37-41.
- 18) MEHTA M. Infantile idiopathic scoliosis. In: Dickson RA, Bradford DS (eds). *Management of spinal deformities*. Butterworth: London, 1984, pp. 101-120.
- 19) LLOYD-ROBERTS GC, PILCHER MF. Structural idiopathic scoliosis in infancy: a study of the natural history of 100 patients. *J Bone Joint Surg Br* 1965; 47: 520-523.
- 20) MAU H. Does infantile scoliosis require treatment. *J Bone Joint Surg Br* 1968; 50: 881.
- 21) AMERICAN ACADEMY OF PEDIATRICS AAP TASK FORCE ON INFANT POSITIONING AND SIDS: Positioning and SIDS. *Pediatrics* 1992; 89: 1120-1126.
- 22) WYNNE-DAVIES R. Infantile idiopathic scoliosis. Causative factors, particularly in the first six months of life. *J Bone Joint Surg Br* 1975; 57: 138-141.

- 23) LOWE TG, EDGAR M, MARGULIES JY, MILLER NH, RASO VJ, REINKER KA, RIVARD CH. Etiology of idiopathic scoliosis: current trends in research. *J Bone Joint Surg Am* 2000; 82-A: 1157-1168.
- 24) ROBIN GC, COHEN T. Familial scoliosis. A clinical report. *J Bone Joint Surg Br* 1975; 57: 146-148.
- 25) COWELL HR, HALL JN, MAC EWEN GD. Genetic aspects of idiopathic scoliosis. A Nicholas Andry Award essay, 1970. *Clin Orthop* 1972; 86: 121-131.
- 26) ESTEVE R. Idiopathic scoliosis in identical twins. *J Bone Joint Surg Br* 1958; 40-B: 97-99.
- 27) FISHER R, DE GEORGE F. A twin study of idiopathic scoliosis. *Clin Orthop* 1965; 55: 117-126.
- 28) KESLING KL, REINKER KA. Scoliosis in twins. A meta-analysis of the literature and report of six cases. *Spine* 1997; 22: 2009-2014; discussion 2015.
- 29) MCKINLEY LM, LEATHERMAN KD. Idiopathic and congenital scoliosis in twins. *Spine* 1978; 3: 227-229.
- 30) MURDOCH G. Scoliosis in twins. *J Bone Joint Surg Br* 1959; 41-B: 736-737.
- 31) WISE CA, BARNES R, GILLUM J, HERRING JA, BOWCOCK AM, LOVETT M. Localization of susceptibility to familial idiopathic scoliosis. *Spine* 2000; 25: 2372-2380.
- 32) SALEHI LB, MANGINO M, DE SERIO S, DE CICCO D, CAPON F, SEMPRINI S, PIZZUTI A, NOVELLI G, DALLAPICCOLA B. Assignment of a locus for autosomal dominant idiopathic scoliosis (IS) to human chromosome 17p11. *Hum Genet* 2002; 111: 401-404.
- 33) MILLER NH, JUSTICE CM, MAROSY B, DOHENY KF, PUGH E, ZHANG J, DIETZ HC, WILSON AF. Identification of candidate regions for familial idiopathic scoliosis. *Spine* 2005; 30: 1181-1187.
- 34) CHAN V, FONG GC, LUK KD, YIP B, LEE MK, WONG MS, LU DD, CHAN TK. A genetic locus for adolescent idiopathic scoliosis linked to chromosome 19p13.3. *Am J Hum Genet* 2002; 71: 401-406.
- 35) ALDEN KJ, MAROSY B, NZEGWU N, JUSTICE CM, WILSON AF, MILLER NH. Idiopathic scoliosis: identification of candidate regions on chromosome 19p13. *Spine* 2006; 31: 1815-1819.
- 36) OGILVIE J. Adolescent idiopathic scoliosis and genetic testing. *Curr Opin Pediatr* 2010; 22: 67-70.
- 37) ACAROGLU E, AKEL I, ALANAY A, YAZICI M, MARCUCIO R. Comparison of the melatonin and calmodulin in paravertebral muscle and platelets of patients with or without adolescent idiopathic scoliosis. *Spine* 2009; 34: E659-663.
- 38) MOREAU A, WANG DS, FORGET S, AZEDDINE B, ANGELONI D, FRASCHINI F, LABELLE H, POITRAS B, RIVARD CH, GRIMARD G. Melatonin signaling dysfunction in adolescent idiopathic scoliosis. *Spine* 2004; 29: 1772-1781.
- 39) LOWE T, LAWELLIN D, SMITH D, PRICE C, HAHER T, MEROLA A, O'BRIEN M. Platelet calmodulin levels in adolescent idiopathic scoliosis: do the levels correlate with curve progression and severity? *Spine* 2002; 27: 768-775.
- 40) CANAVESE F, DIMEGLIO A, GRANIER M, STEBEL M, PITTONI E, CAVALLI F. Arthrodesis of the first six dorsal vertebrae in prepubertal New Zealand white rabbits and thoracic growth to skeletal maturity: the role of the "Rib-Vertebral-Sternal complex". *Minerva Orthop Traumatol* 2007; 58: 369-378.
- 41) CANAVESE F, DIMEGLIO A, VOLPATTI D, STEBEL M, DAURES JP, CANAVESE B, CAVALLI F. Dorsal arthrodesis of thoracic spine and effects on thorax growth in prepubertal New Zealand white rabbits. *Spine* 2007; 32: E443-450.
- 42) MEHTA HP, SNYDER BD, CALLENDER NN, BELLARDINE CL, JACKSON AC. The reciprocal relationship between thoracic and spinal deformity and its effect on pulmonary function in a rabbit model: a pilot study. *Spine* 2006; 31: 2654-2664.
- 43) HEFTI FL, MCMASTER MJ. The effect of the adolescent growth spurt on early posterior spinal fusion in infantile and juvenile idiopathic scoliosis. *J Bone Joint Surg Br* 1983; 65: 247-254.
- 44) SONG KM, LITTLE DG. Peak height velocity as a maturity indicator for males with idiopathic scoliosis. *J Pediatr Orthop* 2000; 20: 286-288.
- 45) PEHRSSON K, LARSSON S, ODEN A, NACHEMSON A. Long-term follow-up of patients with untreated scoliosis. A study of mortality, causes of death, and symptoms. *Spine* 1992; 17: 1091-1096.
- 46) BEREND N, MARLIN GE. Arrest of alveolar multiplication in kyphoscoliosis. *Pathology (Phila)* 1979; 11: 485-491.
- 47) SWANK SM, WINTER RB, MOE JH. Scoliosis and cor pulmonale. *Spine* 1982; 7: 343-354.
- 48) CAMPBELL RM, SMITH MD, MAYES TC, MANGOS JA, WILLEY-COURAND DB, KOSE N, PINERO RF, ALDER ME, DUONG HL, SURBER JL. The characteristics of thoracic insufficiency syndrome associated with fused ribs and congenital scoliosis. *J Bone Joint Surg Am* 2003; 85-A: 399-408.
- 49) DIMEGLIO A. *Le rachis en croissance: scoliose, taille assise, et puberté*. Springer-Verlag: Paris; New York, 1990.
- 50) ZELTNER TB, CADUFF JH, GEHR P, PFENNINGER J, BURRI PH. The postnatal development and growth of the human lung. I. Morphometry. *Respir Physiol* 1987; 67: 247-267.
- 51) THURLBECK WM. Postnatal human lung growth. *Thorax* 1982; 37: 564-571.
- 52) HOGG JC, WILLIAMS J, RICHARDSON JB, MACKLEM PT, THURLBECK WM. Age as a factor in the distribution of lower-airway conductance and in the pathologic anatomy of obstructive lung disease. *N Engl J Med* 1970; 282: 1283-1287.
- 53) DIMEGLIO A. GROWTH IN PEDIATRIC ORTHOPAEDICS. IN: MORRISY T, WEINSTEIN S (EDS). *Lovell & Winter's Pediatric Orthopaedics*. Lippincott William & Wilkins: Philadelphia, 2005, pp. 35-66.
- 54) DIMEGLIO A. Growth in pediatric orthopaedics. *J Pediatr Orthop* 2001; 21: 549-555.
- 55) DEGROODT EG, VAN PELT W, BORSBOOM GJ, QUANJER PH, VAN ZOMEREN BC. Growth of lung and thorax dimensions during the pubertal growth spurt. *Eur Respir J* 1988; 1: 102-108.

- 56) PEHRSSON K, BAKE B, LARSSON S, NACHEMSON A. Lung function in adult idiopathic scoliosis: a 20 year follow up. *Thorax* 1991; 46: 474-478.
- 57) GOLLOGLY S, SMITH JT, WHITE SK, FIRTH S, WHITE K. The volume of lung parenchyma as a function of age: a review of 1050 normal CT scans of the chest with three-dimensional volumetric reconstruction of the pulmonary system. *Spine* 2004; 29: 2061-2066.
- 58) ZADEH HG, SAKKA SA, POWELL MP, MEHTA MH. Absent superficial abdominal reflexes in children with scoliosis. An early indicator of syringomyelia. *J Bone Joint Surg Br* 1995; 77: 762-767.
- 59) COBB J. Outline of study of scoliosis. *Am Acad Orthop Surg Instr Course Lect* 1945; 5: 261-275.
- 60) MEHTA MH. The rib-vertebra angle in the early diagnosis between resolving and progressive infantile scoliosis. *J Bone Joint Surg Br* 1972; 54: 230-243.
- 61) FERREIRA JH, DE JANEIRO R, JAMES JI. Progressive and resolving infantile idiopathic scoliosis. The differential diagnosis. *J Bone Joint Surg Br* 1972; 54: 648-655.
- 62) GOLLOGLY S, SMITH JT, CAMPBELL RM. Determining lung volume with three-dimensional reconstructions of CT scan data: A pilot study to evaluate the effects of expansion thoracoplasty on children with severe spinal deformities. *J Pediatr Orthop* 2004; 24: 323-328.
- 63) GUPTA P, LENKE LG, BRIDWELL KH. Incidence of neural axis abnormalities in infantile and juvenile patients with spinal deformity. Is a magnetic resonance image screening necessary? *Spine* 1998; 23: 206-210.
- 64) PAHYS JM, SAMDANI AF, BETZ RR. Intraspinous anomalies in infantile idiopathic scoliosis: prevalence and role of magnetic resonance imaging. *Spine* 2009; 34: E434-438.
- 65) SMITH J, DUBOSSET J. Imaging of the Growing Spine. In: Akbarnia B, Yazici M, Thompson G (eds). *The Growing Spine: Management of Spinal Disorders in Young Children*. Springer: Germany, 2010, pp. 87-96.
- 66) MEHTA M, MOREL G. The non-operative treatment of infantile idiopathic scoliosis. In: Zorab P, Siezler D (eds). *Scoliosis*. Academic Press: UK, 1979, pp. 71-84.
- 67) JOHNSTON CE, HAKALA MW, ROSENBERGER R. Paralytic spinal deformity: orthotic treatment in spinal discontinuity syndromes. *J Pediatr Orthop* 1982; 2: 233-241.
- 68) SCHWARTZ DR, WIRKA HW. The cast syndrome. A case report and discussion of the literature. *J Bone Joint Surg Am* 1964; 46: 1549-1552.
- 69) EVARTS CM. The cast syndrome. Report of a case after spinal fusion for scoliosis. *Clin Orthop* 1971; 75: 164-166.
- 70) MEHTA MH. Growth as a corrective force in the early treatment of progressive infantile scoliosis. *J Bone Joint Surg Br* 2005; 87: 1237-1247.
- 71) SANDERS JO, D'ASTOUS J, FITZGERALD M, KHOURY JG, KISHAN S, STURM PF. Derotational casting for progressive infantile scoliosis. *J Pediatr Orthop* 2009; 29: 581-587.
- 72) SANKAR WN, SKAGGS DL, YAZICI M, JOHNSTON CE, SHAH SA, JAVIDAN P, KADAKIA RV, DAY TF, AKBARNIA BA. Lengthening of dual growing rods and the law of diminishing returns. *Spine* 2011; 36: 806-809.
- 73) FLETCHER ND, McCLUNG A, RATHJEN KE, DENNING JR, BROWNE R, JOHNSTON CE. Serial casting as a delay tactic in the treatment of moderate-to-severe early-onset scoliosis. *J Pediatr Orthop* 2012; 32: 664-671.
- 74) CRAWFORD HT. In: WEINSTEIN S (ED). *Pediatric spine surgery*. Lippincott, Williams & Wilkins: Philadelphia, 2001.
- 75) ARLET V, PAPIN P, MARCHESI D. Halo femoral traction and sliding rods in the treatment of a neurologically compromised congenital scoliosis: technique. *Eur Spine J* 1999; 8: 329-331.
- 76) SINK EL, KAROL LA, SANDERS J, BIRCH JG, JOHNSTON CE, HERRING JA. Efficacy of perioperative halo-gravity traction in the treatment of severe scoliosis in children. *J Pediatr Orthop* 2001; 21: 519-524.
- 77) LENKE L, DOBBS M. IDIOPATHIC SCOLIOSIS. In: FRYMOYER J, WIESEL S (EDS). *The adult and pediatric spine*. Lippincott Williams and Wilkins: Philadelphia, 2004, pp. 337-360.
- 78) DUBOUSSET J, HERRING JA, SHUFFLEBARGER H. The crankshaft phenomenon. *J Pediatr Orthop* 1989; 9: 541-550.
- 79) SANDERS JO, HERRING JA, BROWNE RH. Posterior arthrodesis and instrumentation in the immature (Risser-grade-0) spine in idiopathic scoliosis. *J Bone Joint Surg Am* 1995; 77: 39-45.
- 80) SKAGGS D, AKBARNIA B, YAZICI M, THOMPSON G. Hybrid distraction-based growing rods. In: *The Growing Spine: Management of Spinal Disorders in Young Children*. Springer: New York, 2010, pp. 601-612.
- 81) LUQUE ER. Paralytic scoliosis in growing children. *Clin Orthop* 1982; (163): 202-209.
- 82) MOE JH, KHARRAT K, WINTER RB, CUMMINE JL. Harrington instrumentation without fusion plus external orthotic support for the treatment of difficult curvature problems in young children. *Clin Orthop* 1984; (185): 35-45.
- 83) HARRINGTON PR. Treatment of scoliosis. Correction and internal fixation by spine instrumentation. *J Bone Joint Surg Am* 1962; 44-A: 591-610.
- 84) AKBARNIA BA, MARKS DS, BOACHIE-ADJEI O, THOMPSON AG, ASHER MA. Dual growing rod technique for the treatment of progressive early-onset scoliosis: a multicenter study. *Spine* 2005; 30: S46-57.
- 85) SPONSELLER PD, THOMPSON GH, AKBARNIA BA, GLAIT SA, ASHER MA, EMANS JB, DIETZ HC. Growing rods for infantile scoliosis in Marfan syndrome. *Spine* 2009; 34: 1711-1715.

- 86) AKBARNIA BA, BREAKWELL LM, MARKS DS, MCCARTHY RE, THOMPSON AG, CANALE SK, KOSTIAL PN, TAMBE A, ASHER MA, GROWING SPINE STUDY GROUP. Dual growing rod technique followed for three to eleven years until final fusion: the effect of frequency of lengthening. *Spine* 2008; 33: 984-990.
- 87) THOMPSON GH, AKBARNIA BA, CAMPBELL RM. Growing rod techniques in early-onset scoliosis. *J Pediatr Orthop* 2007; 27: 354-361.
- 88) THOMPSON GH, AKBARNIA BA, KOSTIAL P, POE-KOCHERT C, ARMSTRONG DG, ROH J, LOWE R, ASHER MA, MARKS DS. Comparison of single and dual growing rod techniques followed through definitive surgery: a preliminary study. *Spine* 2005; 30: 2039-2044.
- 89) AKBARNIA BA, EMANS JB. Complications of growth-sparing surgery in early onset scoliosis. *Spine* 2010; 35: 2193-2204.
- 90) BESS S, AKBARNIA BA, THOMPSON GH, SPONSELLER PD, SHAH SA, EL SEBAIE H, BOACHIE-ADJEI O, KARLIN LI, CANALE S, POE-KOCHERT C, SKAGGS DL. Complications of growing-rod treatment for early-onset scoliosis: analysis of one hundred and forty patients. *J Bone Joint Surg Am* 2010; 92: 2533-2543.
- 91) HELL AK, CAMPBELL RM, HEFTI F. The vertical expandable prosthetic titanium rib implant for the treatment of thoracic insufficiency syndrome associated with congenital and neuromuscular scoliosis in young children. *J Pediatr Orthop Part B* 2005; 14: 287-293.
- 92) YANG JS, SPONSELLER PD, THOMPSON GH, AKBARNIA BA, EMANS JB, YAZICI M, SKAGGS DL, SHAH SA, SALARI P, POE-KOCHERT C, GROWING SPINE STUDY GROUP. Growing rod fractures: risk factors and opportunities for prevention. *Spine* 2011; 36: 1639-1644.
- 93) NOORDEEN HM, SHAH SA, ELSEBAIE HB, GARRIDO E, FAROOQ N, AL-MUKHTAR M, AL MUKHTAR M. In vivo distraction force and length measurements of growing rods: which factors influence the ability to lengthen? *Spine* 2011; 36: 2299-2303.
- 94) CAMPBELL RM, SMITH MD, HELL-VOCKE AK. Expansion thoracoplasty: the surgical technique of opening-wedge thoracostomy. *Surgical technique. J Bone Joint Surg Am* 2004; 86-A Suppl 1: 51-64.
- 95) YAZICI M, EMANS J. Fusionless instrumentation systems for congenital scoliosis: expandable spinal rods and vertical expandable prosthetic titanium rib in the management of congenital spine deformities in the growing child. *Spine* 2009; 34: 1800-1807.
- 96) CAMPBELL RM, SMITH MD, MAYES TC, MANGOS JA, WILLEY-COURAND DB, KOSE N, PINERO RF, ALDER ME, DUONG HL, SURBER JL. The effect of opening wedge thoracostomy on thoracic insufficiency syndrome associated with fused ribs and congenital scoliosis. *J Bone Joint Surg Am* 2004; 86-A: 1659-1674.
- 97) EMANS JB, CAUBET JF, ORDONEZ CL, LEE EY, CIARLO M. The treatment of spine and chest wall deformities with fused ribs by expansion thoracostomy and insertion of vertical expandable prosthetic titanium rib: growth of thoracic spine and improvement of lung volumes. *Spine* 2005; 30: S58-68.
- 98) HASLER C-C, MEHRKENS A, HEFTI F. Efficacy and safety of VEPTR instrumentation for progressive spine deformities in young children without rib fusions. *Eur Spine J* 2010; 19: 400-408.
- 99) SMITH JR, SAMDANI AF, PAHYS J, RANADE A, ASGHAR J, CAHILL P, BETZ RR. The role of bracing, casting, and vertical expandable prosthetic titanium rib for the treatment of infantile idiopathic scoliosis: a single-institution experience with 31 consecutive patients. *Clinical article. J Neurosurg Spine* 2009; 11: 3-8.
- 100) RAMIREZ N, FLYNN JM, SERRANO JA, CARLO S, CORNIER AS. The Vertical Expandable Prosthetic Titanium Rib in the treatment of spinal deformity due to progressive early onset scoliosis. *J Pediatr Orthop Part B* 2009; 18: 197-203.
- 101) SMITH JT, NOVAIS E. Treatment of Gibbus deformity associated with myelomeningocele in the young child with use of the vertical expandable prosthetic titanium rib (VEPTR): a case report. *J Bone Joint Surg Am* 2010; 92: 2211-2215.
- 102) SANKAR WN, ACEVEDO DC, SKAGGS DL. Comparison of complications among growing spinal implants. *Spine* 2010; 35: 2091-2096.
- 103) AKBARNIA BA, MUNDIS GM, SALARI P, YASZAY B, PAWELEK JB. Innovation in growing rod technique: a study of safety and efficacy of a magnetically controlled growing rod in a porcine model. *Spine* 2012; 37: 1109-1114.
- 104) AKBARNIA B, MUNDIS GM, SALARI P. A technical report on the ellipse technologies device: a remotely expandable device for non-invasive lengthening of growing rod. *J Child Orthop* 2009; 3: 530-531.
- 105) MILADI L. Magnetic powered extensible rod for thorax or spine. Kyoto, Japan, 2010.
- 106) CHEUNG KM, CHEUNG JP, SAMARTZIS D, MAK KC, WONG YW, CHEUNG WY, AKBARNIA BA, LUK KD. Magnetically controlled growing rods for severe spinal curvature in young children: a prospective case series. *Lancet* 2012; 379: 1967-1974.
- 107) WICK JM, KONZE J. A magnetic approach to treating progressive early-onset scoliosis. *AORN J* 2012; 96: 163-173.
- 108) ROAF R. The treatment of progressive scoliosis by unilateral growth-arrest. *J Bone Joint Surg Br* 1963; 45: 637-651.
- 109) MARKS DS, IQBAL MJ, THOMPSON AG, PIGGOTT H. Convex spinal epiphysiodesis in the management of progressive infantile idiopathic scoliosis. *Spine* 1996; 21: 1884-1888.
- 110) BETZ RR, KIM J, D'ANDREA LP, MULCAHEY MJ, BALSARA RK, CLEMENTS DH. An innovative technique of vertebral body stapling for the treatment of patients with adolescent idiopathic scoliosis: a feasibility, safety, and utility study. *Spine* 2003; 28: S255-265.
- 111) NEWTON PO, UPASANI VV, FARNSWORTH CL, OKA R, CHAMBERS RC, DWEK J, KIM JR, PERRY A, MAHAR AT. Spinal growth modulation with use of a tether in an immature porcine model. *J Bone Joint Surg Am* 2008; 90: 2695-2706.

- 112) PATEL A, SCHWAB F, LAFAGE R, LAFAGE V, FARCY JP. Does removing the spinal tether in a porcine scoliosis model result in persistent deformity? A pilot study. *Clin Orthop* 2011; 469: 1368-1374.
- 113) UPASANI VV, FARNSWORTH CL, CHAMBERS RC, BASTROM TP, WILLIAMS GM, SAH RL, MASUDA K, NEWTON PO. Intervertebral disc health preservation after six months of spinal growth modulation. *J Bone Joint Surg Am* 2011; 93: 1408-1416.
- 114) RINSKY LA, GAMBLE JG, BLECK EE. Segmental instrumentation without fusion in children with progressive scoliosis. *J Pediatr Orthop* 1985; 5: 687-690.
- 115) PRATT RK, WEBB JK, BURWELL RG, CUMMINGS SL. Luque trolley and convex epiphysiodesis in the management of infantile and juvenile idiopathic scoliosis. *Spine* 1999; 24: 1538-1547.
- 116) MCCARTHY RE, SUCATO D, TURNER JL, ZHANG H, HENSON MAW, MCCARTHY K. Shilla growing rods in a caprine animal model: a pilot study. *Clin Orthop* 2010; 468: 705-710.
- 117) MCCARTHY R, LUHMANN S, LENKE L. Greater than two year followup Shilla growth enhancing system for the treatment of scoliosis in children. Montreal, Canada, 2008.
- 118) OUELLET J. Surgical technique: modern Luqué trolley, a self-growing rod technique. *Clin Orthop* 2011; 469: 1356-1367.
- 119) DAY GA, UPADHYAY SS, HO EK, LEONG JC, IP M. Pulmonary functions in congenital scoliosis. *Spine* 1994; 19: 1027-1031.
- 120) WINTER RB, MOE JH. The results of spinal arthrodesis for congenital spinal deformity in patients younger than five years old. *J Bone Joint Surg Am* 1982; 64: 419-432.