Early Definitive Spinal Fusion in Young Children

What We Have Learned

Lori A. Karol MD

Published online: 19 October 2010
© The Association of Bone and Joint Surgeons® 2010

Abstract

Background Early-onset scoliosis, when left untreated, leads to severe deformity. Until the last decade, treatment of progressive curves in young children often consisted of definitive spinal fusion. The recognition of thoracic insufficiency syndrome associated with definitive early fusion has led to the development of new surgical techniques developed to preserve spinal and thoracic growth in young patients with progressive scoliosis.

Questions/purposes We asked: (1) Does early definitive fusion arrest progression of spinal deformity? To what extent does early definitive spinal fusion influence (2) pulmonary function and (3) thoracic growth?

Methods A Medline search of the published literature on early-onset scoliosis, congenital scoliosis, and infantile scoliosis between 2008 and 2010 was performed on spinal fusion for early-onset scoliosis, focusing on studies reporting pulmonary function at followup.

Results Spinal deformity is apparently not well controlled by early fusion since revision surgery has been required in 24% to 39% of patients who underwent presumed definitive fusion in early childhood. Restrictive pulmonary disease, defined as forced vital capacity less than 50% of

normal, occurs in 43% to 64% of patients who undergo early fusion surgery with those children who have extensive thoracic fusions and whose fusions involve the proximal thoracic spine at highest risk. Thoracic growth after early surgery is an average of 50% of that seen in children with scoliosis who do not have early surgery. Diminished thoracic spinal height correlates with decreased forced vital capacity.

Conclusions The literature does not support routine definitive fusion of thoracic spinal deformity at an early age in children with scoliosis.

Level of Evidence Level IV, therapeutic study. See Guidelines for Authors for a complete description of levels of evidence.

Introduction

There are few areas in pediatric orthopaedic research that have garnered more interest in the last 5 years than the management of early-onset scoliosis. Over the last 2 years, 30 English-language manuscripts have been published on the treatment of children younger than 10 years with scoliosis (Table 1) [1, 3, 4, 8, 11, 18, 21–23, 25, 26, 30–32, 36–42, 44–48, 51, 54–56]. It is important in this emerging innovative field to remember why this topic has become so important and why earlier forms of treatment led to the surge of innovative techniques.

Early-onset scoliosis occurs in a group of diagnoses unified only by age at presentation. Children with early-onset scoliosis may have congenital spine malformations that produce spinal deformity, often associated with thoracic malformations such as fused or absent ribs. Patients may also develop early-onset scoliosis despite normal vertebral formation. Those with normal vertebral development

---

Each author certifies that he or she has no commercial associations (eg, consultancies, stock ownership, equity interest, patent/licensing arrangements, etc) that might pose a conflict of interest in connection with the submitted article.

Electronic supplementary material The online version of this article (doi:10.1007/s11999-010-1622-z) contains supplementary material, which is available to authorized users.

L. A. Karol (✉)
Texas Scottish Rite Hospital for Children, 2222 Welborn,
Dallas, TX 75219, USA
e-mail: lori.karol@tsrh.org

Table 1. Studies of pulmonary function in children with early spinal fusion surgery

Study	Etiology	Number	Average age at surgery (years)	Average age at testing (years)	Anterior surgery?	Average number of thoracic levels fused	Number greater than one surgery	Average percent forced vital capacity
Bowen et al. [4]	Congenital	13	2.9	9.7	0/13	7.3	0	67%
Goldberg et al. [19]	Infantile onset	11	4.1	20.53	9/11	N/A	1	40.8%
Karol et al. [23]	Mixed (most congenital)	28	3.3	14.6	27/28	7.04	11	57.8%
Vitale et al. [51]	Congenital	21	4.7	12.6	15/21	5.09	5	74.4% (64.2% if thoracic fusion)

N/A = not available.

can be divided into patients whose scoliosis is the result of either a syndromic or neuromuscular condition and those with true infantile idiopathic scoliosis. Early-onset scoliosis is rare. Although the incidence of idiopathic scoliosis is approximately 1.5% of all teens, babies with infantile scoliosis represent only 4% of all patients with idiopathic scoliosis [27]. As a result of the lack of radiographic screening, the incidence of congenital spine malformations is unknown.

Although rare, children with progressive curves at young ages are at risk for severe pulmonary consequences attributable to both the spinal deformity and the surgical management of this deformity [5]. Pehrsson et al. [34, 35] studied the mortality rate in 115 women with untreated scoliosis. Mortality was most increased in the 29 patients with infantile-onset followed by 32 juvenile-onset patients compared with adolescent patients. The cause of death was listed as respiratory failure in 40% of the patients who had died. The average age at death was 54 years but included patients as young as 16 years. Nineteen of the 21 patients (90%) who died of respiratory failure had diagnosis of their scoliosis before age 9 years [34, 35]. A 1955 study by Scott and Morgan [43] reported four deaths in 28 children with infantile scoliosis. Although one was within the postoperative period, the remaining three were from cardiopulmonary failure resulting from the severe untreated spinal and thoracic deformity. These patients ranged from 17 to 19 years of age at the time of death [43]. Davies and Reid described postmortem findings in four patients who died of cardiopulmonary failure whose age at the time of death ranged from 8 to 18 years. These patients had scoliosis resulting from polio, muscular dystrophy, hemivertebra, and either infantile or juvenile idiopathic scoliosis. All four had very small lungs with markedly decreased numbers of alveoli [12]. Based on the prevailing belief that neglect and progression led to pulmonary failure, surgeons embraced the concept that a spine that was “short and straight,” as a result of early fusion, was preferable to one that was “longer but crooked” because growth had been allowed and progressive deformity had resulted [13, 53].

The treatment of early-onset scoliosis therefore typically involved observation in children with mild deformities, bracing in children who had progressive noncongenital curves, and early definitive fusion in children who initially presented with curves of severe magnitude, had worsening congenital spinal deformities, or who demonstrated substantial progression despite bracing. The importance of anterior fusion was not recognized until Dubousset et al. described the crankshaft phenomena, defined as worsening of rotational deformity in young patients with solid posterior fusions resulting from persistent anterior growth of the vertebral bodies [16]. Thus, early combined anterior/posterior fusion became accepted as the definitive treatment for preadolescent children with severe spine deformity, although its ability to arrest progression and avoid complications in the early-onset patient population remains unknown.

We therefore asked: (1) Does early definitive fusion in patients with early-onset scoliosis arrest progression of spinal deformity? To what extent does early definitive spinal fusion influence (2) pulmonary function and (3) thoracic growth?

Search Strategies and Criteria

A Medline search of the recent English language literature from 2008 through 2010 was performed using the key word scoliosis. This search yielded 1512 articles. The search was repeated by adding early onset, pulmonary function, congenital, infantile, titanium, cast, and spirometry as additional key words. These searches yielded 370 articles. I reviewed the titles of all these articles and then excluded case reports, collections of abstracts, lectures, those articles published in anesthesia journals, nursing journals, state medical journals, and foreign journals not readily accessible in North America. I also excluded articles on specific rare syndromes, neuromuscular scoliosis, adolescent scoliosis, surgical technique descriptions, and genetic reports. (Appendix 1; supplemental materials are available with the

online version of CORR.) I then included the articles on pulmonary outcome in patients with early-onset scoliosis from the historical literature.

Does Early Definitive Fusion Arrest Progression of Spinal Deformity?

If early definitive fusion is to be recommended, it is important for modern spine surgeons to know how effective this approach is in eliminating further progression of deformity. To do so, the reoperation rates of papers reporting the results of early fusion must be studied. In 2002, Goldberg et al. published long-term results from in situ fusion in 43 patients with congenital scoliosis. Sixteen of 43 patients (37.2%) who were at least 15 years of age at the time of final followup underwent reoperation as a result of “adding-on” and/or progression of their deformity after in situ fusion. Even four of 10 patients who were more recently treated with anterior and posterior fusion to theoretically eliminate crankshaft progression required subsequent revision surgery [20]. We reported 11 of 28 patients (39.3%) who underwent early spinal fusion before age 8 years were revised by an average 11.2 years followup. Again, 26 of the 28 patients had undergone combined anterior and posterior fusion yet still required revision [23]. Vitale et al. reported the results of “definitive” fusion in 21 patients with congenital scoliosis at an average of 7-year followup ranging from 3- to 13-year followup. At the time of publication, five of 21 patients had undergone more than one spinal procedure to achieve fusion. The age at the time of followup averaged only 12.6 years ranging from 7 to 19 years [51]. Because this study includes young patients who have yet to complete their spinal growth (or even achieve peak growth velocity), additional patients may experience deformity progression and require revision surgery in the future. Despite attempts to achieve circumferential fusion of the spine in young children, curve progression may still occur. Whether this is the result of pseudarthroses because of difficulty in achieving fusion in very young children, adding on of the deformity at the cephalad or caudal ends of the initial fusion, or evolution of the deformity as a result of continued growth of the neurocentral synchondroses [56] is unresolved to date.

To What Extent Does Early Definitive Spinal Fusion Influence Pulmonary Function?

In 2003, Campbell et al. published their seminal paper describing thoracic insufficiency syndrome [10]. Thoracic insufficiency is defined as the inability of the thorax to

support normal respiration or lung growth. Campbell et al. found there were groups of children who had unsustainable pulmonary function as a result of severe spinal and thoracic malformations resulting in immobile or nonfunctional chest wall mechanics, which interfere with normal respiration such as that seen in patients with Jeune’s syndrome or in children with abnormally small thoracic height resulting from multiple congenital rib malformations. However, they also described cases of poor pulmonary function in patients who had undergone early spinal fusion for scoliosis, which resulted in inhibition of thoracic growth [10].

Dimeglio and Bonnel have published their studies on spinal growth [15]. The most rapid period of spinal growth occurs in the first 5 years of life, when the spine increases in length 50%. Between the ages of 5 and 10 years, the spine continues to grow, albeit at a slower rate. At age 10 in girls and 12 in boys, the rate of growth again increases with spinal growth typically stopping at age 13 in girls and age 17 in boys. The height of the thoracic spine averages 11 cm in the newborn, 18 cm at age 5 years, 22 cm at 10 years, and 26.5 to 28 cm in the female and male adult, respectively [14]. Thus, using this information, one can estimate that complete thoracic fusion at age 5 should result in a 10-cm loss of thoracic height in an otherwise normal adult male. This results in diminished space available for the lung with pulmonary implications.

Similarly, the normal development of the lungs in the growing child has been studied [6, 12, 49]. The greatest increase in the number of alveoli in normal children occurs in the first 2 years of life [49]. Although lung volumes continue to increase into midadolescence, alveolar multiplication typically is complete by age 8 years. For this reason, surgery during the first 8 years of life is proposed to create the greatest disturbance in pulmonary development.

Patients with early-onset scoliosis, however, are frequently not otherwise normal. Patients with congenital scoliosis often have fused ribs, which will limit the ability of the already small chest to change its volume as a result of decreased mobility of the chest wall [33, 50]. Children with neuromuscular scoliosis resulting from the congenital muscular dystrophies, myelomeningocele, and such conditions as spinal muscular atrophy are weak, which again influences the ability to breathe deeply and forcibly exhale. The disturbance of thoracic growth in these children prone to respiratory compromise is most worrisome.

Several studies have been published documenting the pulmonary status of patients after early definitive fusion for scoliosis (Table 1). Goldberg et al. reported on 23 patients who had undergone spinal fusion for treatment of infantile scoliosis [19]. They performed pulmonary function testing and found mean forced vital capacity (FVC) averaged 41% of normal (range, 12%–67%) at maturity in 11 patients fused before age 10, whereas those who were fused at an older age

averaged 68% of normal FVC (range, 48%–88%). They concluded that early spinal fusion in patients with infantile scoliosis resulted in a worrisome diminution of pulmonary function as a result of growth inhibition [19].

At an average of 11 years (range, 6.4–20.5 years) followup in patients treated with spinal fusion before the age of 8 years, we found 71% of the patients had congenital scoliosis, but also included were patients with neurofibromatosis, infantile, and syndromic scoliosis [23]. Age at surgery averaged 3.3 years ranging from 4 months to 8.4 years. The FVC of our patients averaged 57.8% of age-matched normal values, ranging from a low of 27% to normal (99%). Forty-three percent of the 28 patients had restrictive lung disease defined as a FVC measuring less than 50% of age-matched normal values. Additionally, 44% of patients had a reduced maximum inspiratory pressure, which we believed indicated diaphragmatic weakness, chest wall weakness, or air trapping. Diminished pulmonary function correlated with the percentage of the thoracic spine that had been fused, with decreased T1-T12 height at followup, and with the inclusion of T1 or T2 in the fusion. Sixteen of 27 patients had thoracic height of less than 18 cm at followup, which is less than the average height of a normal 5 year old. A threshold of 18 cm of thoracic height, which represents the normal height of a 10 year old, was identified when substantial restrictive lung disease did not occur. Three patients were already symptomatic with two requiring the use of bilevel positive airway pressure. We concluded extensive thoracic fusion in young children should be avoided.

Bowen et al. recently published a study comparing pulmonary function in a group of 13 patients with congenital scoliosis who had posterior fusions before age 5 years with a group of patients with congenital scoliosis who had not undergone spinal surgery [4]. They found no difference in percent normalized FVC between the two groups with the surgical group averaging 67% and the nonoperated group 69% normal FVC. Differences between this study and the study of Karol et al. [23] include length of time after surgery to pulmonary function testing (PFT), which averaged 6.7 years in the Los Angeles study and 11.2 years in the study of Karol et al., and more importantly, minimum length of postoperative followup, which was 2 years in the study of Bowen et al. and 6.4 years in the TSRH study, yet important information is presented in this study, including the correlation between a higher thoracic apex of deformity and worsening PFTs and the presence of three of 13 patients with %FVC less than 50% (including one with approximately 23% FVC) at followup after posterior spinal fusion [4, 23]. Although this study concluded patients who had undergone earlier fusion were not worse from a respiratory standpoint than those who did not, one could also conclude patients with early posterior

fusion were not “protected” from pulmonary restrictive disease by their treatment of progressive spinal deformity with definitive fusion.

Finally, Vitale et al. published their results after definitive fusion in 21 patients with congenital scoliosis operated at an average age of 4.9 years and followed on average 7 years postoperatively [51]. They divided their patient population into two groups. The first group underwent fusion for scoliosis with a thoracic apex, whereas the second group had curves with apices distal to the thoracolumbar junction. They found the thoracic fusion group had more extensive fusions, lower pulmonary functions, greater Cobb angles at followup, and lower scores on quality-of-life measures. The percent predicted FVC for patients with thoracic fusions averaged 64.2% and 88.5% for patients with lumbar fusions. More worrisome was the correlation with low pulmonary function volumes and diminished quality of life. Not only is the lifespan of children with early fusion for scoliosis diminished, but the quality of their lives as evidenced by their ability to participate in activities with their peers and families is affected as well.

Equally ominous is the realization that the restrictive pulmonary disease that has been documented in the previous studies is inevitably going to worsen during midadulthood in these patients. The average age at the time of study was 10.5 years in the study by Bowen et al. [4], 12.6 years in the study of Vitale et al. [51], and in the TSRH study was only 14.6 years. Because many of the patients in these studies are not yet skeletally mature, their percent predicted pulmonary volumes will likely worsen because their spines do not grow and the space available for the lung does not elongate. Furthermore, the normal aging process causes pulmonary volumes to decline in individuals who have not had spinal surgery. This expected deterioration in FVC begins when normal adults reach their mid-30s [7]. Kory et al. found vital capacity decreases by 700 mL in the normal adult male by age 60 years [24]. If patients who had early definitive fusions have diminished pulmonary reserve as their baseline, the expected loss of volume during adulthood carries profound health implications.

To What Extent Does Early Definitive Spinal Fusion Influence Thoracic Growth?

Fewer studies have reported the influence of early definitive spinal fusion in patients with early-onset scoliosis on growth of the thoracic spine. Bowen et al. found patients who had fusions before the age of 5 years for treatment of their congenital scoliosis had thoracic growth, measured as T1 to T12 height, of 0.48 cm per year at 6.7 years followup, whereas those with congenital scoliosis who did not have fusion grew an average of 0.97 cm [4]. Karol et al.

found that 57% of patient who were fused before age 8 years had a thoracic height that was less than the average height of a 5-year-old child. These patients were 7.3 to 17.8 years of age at the time of followup. The authors observed a relationship between a diminished thoracic spinal T1 to T12 height and decreased FVC [23]. From these studies, it appears that thoracic growth is negatively influenced by early spinal fusion.

Discussion

Although early-onset scoliosis is a rare condition, the treatment of children with these conditions may be a large portion of a pediatric orthopaedic surgeon's practice. In light of the growing spine and the developing pulmonary system, it is of vital importance to understand the implication of treatment on not only the spinal deformity, but also on the ultimate respiratory function of the patient at maturity. We therefore asked: (1) Does early definitive fusion arrest progression of spinal deformity? To what extent does early definitive spinal fusion influence (2) pulmonary function and (3) thoracic growth?

There are limitations in the available literature on early-onset scoliosis that merit discussion. First, although the existing studies report diminished pulmonary function compared with normal values, none of the studies presently available report longitudinal data on pulmonary function. Preoperative data are not available as a result of the fact that these children were operated on at an early age before pulmonary function data could be collected. Furthermore, the studies by Goldberg et al. [19], Karol et al. [23], Bowen et al. [4], and Vitale et al. [51] all report a single pulmonary function measurement rather than repeated testing over time. We can predict that pulmonary function will continue to decline as these children age, but their chests do not grow accordingly as a result of their fusions. There are no Level I studies in the literature that prospectively compare outcomes in treated (or fused) patients with patients who are observed. The natural history of severe early-onset scoliosis is one of progression of deformity. For ethical reasons, withholding treatment from very young children with serious spinal deformities cannot be proposed. It is known that early-onset scoliosis, either from infantile scoliosis or congenital malformations, can result in diminished pulmonary function. We may never know how much of a role treatment of these severe curves in young children exacerbates the pulmonary compromise when compared with the diminished pulmonary function inherent in the condition of early-onset scoliosis if untreated. The closest report in the literature is that by Bowen et al., which compares a group of patients that underwent early fusion with another group that was observed [4]. This study was

not prospective and did not match patients for curve severity. The authors state those children in the surgical group presented with scoliosis at an earlier age and had more severe deformities than the nonsurgical group. The question of how much early surgery disturbs pulmonary function remains unanswered.

My review of the literature establishes that definitive spinal fusion in early childhood may not successfully prevent future spinal deformity as the child grows and matures. Although early surgery may result in success in limited congenital deformities such as single hemivertebra [38], the long-term outcome for most congenital deformities seen in the young child cannot be predicted at present. In fact, both Goldberg et al. [19] and Karol et al. [23] suggest early fusion, even with both anterior and posterior procedures, cannot be considered definitive in a significant number of cases, as evidenced by the continued progression noted in 45% of the patients of Goldberg et al. fused before age 10 and by the 39% revision rate after anterior/posterior fusion series reported by Karol et al. [23]. Causes for failure of anterior/posterior fusion to control deformity in the early-onset patient are unclear but probably include continued growth of vertebral elements in regions not arrested by conventional anterior interbody or posterior decortications and facet excision techniques. Other suspected causes would include pseudarthrosis and lack of internal fixation to assist in correction maintenance. Unfortunately, clinical studies to evaluate these causes are unavailable.

Second, early fusion has a negative effect on pulmonary function. The inhibition of thoracic growth results in a smaller chest with decreased vital capacity and restricted pulmonary disease in many children with early fusion. Series of patients who had early spinal fusions show that FVC ranges from 41% of normal in the study of Goldberg et al. [19] to 67% of normal in the study of Bowen et al. [4]. The key difference in these two studies is the length of followup. The former patients [19] were tested at skeletal maturity, whereas followup averaged only 6.7 years in the latter and most of the children were still growing at the time of this report [4]. It is also important to recognize that the presence of congenital spinal malformations, rib anomalies, and scoliosis even without fusion may lead to diminished pulmonary function [12, 34, 35, 37, 43]. It is impossible at present to separate the influence of the surgery from the influence of the deformity on the development of the pulmonary system in young children with early-onset scoliosis. The literature does demonstrate that traditional early spinal fusion of the thoracic spine does not allow for continued pulmonary development in young children [4, 10, 19, 20, 23, 51]. Similar studies of pulmonary function in large numbers of children with lumbar deformities have not been published, because the published studies include only rare children with lower

thoracic and lumbar scoliosis. Similarly, pulmonary function has not been measured in children who undergo resection of congenital malformations such as hemivertebra rather than early fusion.

Third, definitive spinal fusion in youngsters with early-onset scoliosis has been shown to inhibit thoracic growth, as evidenced by a decreased T1 to T12 height. Both studies by Karol et al. [23] and Bowen et al. [4] documented decreased growth of the thorax in patients with early spinal fusion compared with the normal values published by Dimeglio and Bonnel [14]. Fifty-seven percent of the patients in the Karol et al. [23] study have a thoracic height smaller than that of a normal 5-year-old child (less than 18 cm) despite the fact they were 7 to 18 years at the time they were measured. A total of 62.5% of these patients with short thoracic height already had less than 50% of normal FVC, indicating they have restrictive lung disease. The literature supports the fact that early thoracic fusion creates inhibition of thoracic growth, which in turn is associated with the likelihood of diminished FVC.

The current literature does not support routine definitive fusion of spinal deformity at an early age in young children with scoliosis. Although isolated reports of patients followed well into adulthood who are leading productive and pain-free lives exist [52], there are now several reports of increased mortality, diminished pulmonary function, and a decreased ability to carry out activities of everyday life. Reoperation after what was intended to be a definitive circumferential fusion in young children is not rare. Although early definitive fusion does still have a role in isolated vertebral malformations, particularly in the lumbar spine, the effect of fusion before age 5 on the growth of the thoracic cage is life-altering for many children. Nonoperative measures to control or correct spinal deformity such as casting should be pursued in young babies with idiopathic scoliosis with the goal of preventing deformities that will require surgical management [28, 41]. New surgical techniques that allow expansion of the thorax and growth of the spine through thoracoplasty with implantation of expandable rib devices or dual growing spinal rods without fusion offer the potential for continued pulmonary development in young children with severe deformities [1, 2, 9, 17]. Early reports of maintenance and even improvement in pulmonary function have recently been published [29, 45, 46]. Continued research is needed to establish more effective treatments for the youngest children with severe spinal deformity.

References

1. 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.
2. 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–S57.
3. Betz RR, Ranade A, Samdani AF, Chafetz R, D'Andrea LP, Gaughan JP, Asghar J, Grewal H, Mulcahey MJ. Vertebral body stapling: a fusionless treatment option for a growing child with moderate idiopathic scoliosis. *Spine*. 2010;35:169–176.
4. Bowen RE, Scaduto AA, Banuelos S. Does early thoracic fusion exacerbate preexisting restrictive lung disease in congenital scoliosis patients? *J Pediatr Orthop*. 2008;28:506–511.
5. Branthwaite MA. Cardiorespiratory consequences of unfused idiopathic scoliosis. *Br J Dis Chest*. 1986;80:360.
6. Burri PH. Structural aspects of prenatal and postnatal development and growth of the lung. In: McDonald JA, ed. *Lung Growth and Development*. New York: Dekker; 1997.
7. Burrows B, Cline MG, Knudson RJ, Taussig LM, Lebowitz MD. A descriptive analysis of the growth and decline of the FVC and FEV1. *Chest*. 1983;83:717–724.
8. Campbell RM. Spine deformities in rare congenital syndromes: clinical issues. *Spine*. 2009;34:1815–1827.
9. Campbell RM, Hell-Vocke AK. Growth of the thoracic spine in congenital scoliosis after expansion thoracoplasty. *J Bone Joint Surg Am*. 2003;85:409–420.
10. Campbell RM, Smith MD, Mayes TC, Mangos JA, Willey-Courand DB, Kose N, Pinero RF, Adler ME, Duong HL, Surber JL. The characteristics of thoracic insufficiency syndrome associated with fused ribs and congenital scoliosis. *J Bone Joint Surg Am*. 2003;85:399–408.
11. Caubet JF, Emans JB, Smith JT, Vanbosse H, Ramirez N, Flynn J, Vitale M, Smith M, St Hilaire T, Klinge S. Increased hemoglobin levels in patients with early onset scoliosis: prevalence and effect of a treatment with VEPTR. *Spine*. 2009;34:2534–2536.
12. Davies G, Reid L. Effect of scoliosis on growth of alveoli and pulmonary arteries and on the right ventricle. *Arch Dis Child*. 1971;46:623–632.
13. Day GA, Upadhyay SS, Ho EK, Leong JC, Ip M. Pulmonary functions in congenital scoliosis. *Spine*. 1994;19:1027–1031.
14. Dimeglio A. Growth of the spine before age 5 years. *J Pediatr Orthop B*. 1993;1:102–107.
15. Dimeglio A, Bonnel F. *Spine Growth* [in French]. Paris, France: Springer; 1990.
16. Dubousset J, Herring JA, Shufflebarger H. The crankshaft phenomenon. *J Pediatr Orthop*. 1989;9:541–550.
17. 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 (Phila Pa 1976)*. 2005;30:S58–S68.
18. Garrido E, Tome-Bermejo F, Tucker SK, Noordeen HN, Morley TR. Short anterior instrumented fusion and posterior convex non-instrumented fusion of hemivertebra for congenital scoliosis in very young children. *Eur Spine J*. 2008;17:1507–1514.
19. Goldberg CJ, Gillic 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.
20. Goldberg CJ, Moore DP, Fogarty EE, Dowling FE. Long-term results from in situ fusion for congenital vertebral deformity. *Spine*. 2002;27:619–628.
21. Hasler CC, 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.
22. Hedequist DJ. Instrumentation and fusion for congenital spine deformities. *Spine*. 2009;34:1783–1790.

23. 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.
24. Kory RC, Callahan R, Boren HG, Syner JC. The Veteran's Administration-Army cooperative study of pulmonary function. I. Clinical spirometry in normal men. *Am J Med.* 1961;30:243–258.
25. Mahar AT, Bagheri R, Oka R, Kostial P, Akbarnia BA. Biomechanical comparison of different anchors (foundations) for the pediatric dual growing rod technique. *Spine J.* 2008;8:933–939.
26. McCarthy RE, Sucato D, Turner JL, Zhang H, Henson MA, McCarthy K. Shilla growing rods in a caprine animal model: a pilot study. *Clin Orthop Relat Res.* 2010;468:705–710.
27. McMaster MJ. Infantile idiopathic scoliosis: can it be prevented? *J Bone Joint Surg Br.* 1983;65:612.
28. Mehta MH. Growth as a corrective force in the early treatment of progressive infantile scoliosis. *J Bone Joint Surg Br.* 2005;87:1237–1247.
29. Motoyama EK, Deeney VF, Fine GF, Yang CI, Mutich RL, Walczak SA, Moreland MS. Effects of lung function of multiple expansion thoracoplasty in children with thoracic insufficiency syndrome: a longitudinal study. *Spine.* 2006;31:284–298.
30. Motoyama EK, Yang CI, Deeney VF. Thoracic malformation with early-onset scoliosis: effect of serial VEPTR expansion thoracoplasty on lung growth and function in children. *Paediatr Respir Rev.* 2009;10:12–17.
31. Newton PO, Farnsworth CL, Faro FD, Mahar AT, Odell TR, Mohamad F, Breisch E, Fricka K, Upasani VV, Amiel D. Spinal growth modulation with an anterolateral flexible tether in an immature bovine model: disc health and motion preservation. *Spine.* 2008;33:724–733.
32. Noordeen MH, Garrido E, Tucker SK, Elsebaie HB. The surgical treatment of congenital kyphosis. *Spine.* 2009;34:1808–1814.
33. Owange-Iraka JW, Harrison A, Warner JO. Lung function in congenital and idiopathic scoliosis. *Eur J Pediatr.* 1984;142:198–200.
34. 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.
35. Pehrsson K, Nachemson A, Olofson J, Ström K, Larsson S. Respiratory failure in scoliosis and other thoracic deformities. A survey of patients with home oxygen or ventilator therapy in Sweden. *Spine.* 1992;17:714–718.
36. 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 B.* 2009;18:197–203.
37. Redding G, Song K, Inscore S, Effmann E, Campbell R. Lung function asymmetry in children with congenital and infantile scoliosis. *Spine J.* 2008;8:639–644.
38. Ruf M, Jensen R, Letko L, Harms J. Hemivertebra resection and osteotomies in congenital spine deformity. *Spine.* 2009;34:1791–1799.
39. Sabourin M, Jolivet E, Miladi L, Wicart P, Rampal V, Skalli W. Three-dimensional stereoradiographic modeling of rib cage before and after spinal growing rod procedures in early-onset scoliosis. *Clin Biomech (Bristol, Avon).* 2010;25:284–291.
40. Samdani AF, Ranade A, Dolch HJ, Williams R, St Hilaire T, Cahill P, Betz RR. Bilateral use of the vertical expandable prosthetic titanium rib attached to the pelvis: a novel treatment for scoliosis in the growing spine. *J Neurosurg Spine.* 2009;10:287–292.
41. 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.
42. Sankar W, Skaggs DL, Emans JB, Marks DS, Dormans JP, Thompson GH, Shah SA, Sponseller PD, Akbarnia BA. Neurologic risk in growing rod spine surgery in early onset scoliosis: is neuromonitoring necessary for all cases? *Spine.* 2009;34:1952–1955.
43. Scott JC, Morgan TH. The natural history and prognosis of infantile idiopathic scoliosis. *J Bone Joint Surg Br.* 1955;37:400–413.
44. Skaggs DL, Choi PD, Rice C, Emans J, Song KM, Smith JT, Campbell RM Jr. Efficacy of intraoperative neurologic monitoring in surgery involving a vertical expandable prosthetic titanium rib for early-onset spinal deformity. *J Bone Joint Surg Am.* 2009;91:1657–1663.
45. 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.
46. Smith JT, Jerman J, Stringham J, Smith MS, Golligorsky S. Does expansion thoracoplasty improve the volume of the convex lung in a windswept thorax? *J Pediatr Orthop.* 2009;29:944–947.
47. Sponseller PD, Thompson GH, Akbarnia BA, Glait SA, Asher MA, Emans JB, Dietz HC 3rd. Growing rods for infantile scoliosis in Marfan syndrome. *Spine.* 2009;34:1711–1715.
48. Sponseller PD, Yang JS, Thompson GH, McCarthy RE, Emans JB, Skaggs DL, Asher MA, Yazici M, Poe-Kochert C, Kostial P, Akbarnia BA. Pelvic fixation of growing rods: comparison of constructs. *Spine (Phila Pa 1976).* 2009;34:1706–1710.
49. Thurlbeck WM. Postnatal human lung growth. *Thorax.* 1982;37:564–571.
50. Tsirikos AI, McMaster MJ. Congenital anomalies of the ribs and chest wall associated with congenital deformities of the spine. *J Bone Joint Surg Am.* 2005;87:2523–2536.
51. Vitale MG, Matsumoto H, Bye MR, Gomez JA, Booker WA, Hyman JE, Royce DP Jr. 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.
52. Winter RB, Lonstein JE. Congenital thoracic scoliosis with unilateral unsegmented bar and concave fused ribs; rib osteotomy and posterior fusion at 1 year old, anterior and posterior fusion at 5 years old with a 36-year follow-up. *Spine.* 2007;32:E841–E844.
53. Winter RB, Moe JH. The results of spinal arthrodesis for congenital spinal deformity in patients younger than 5 years old. *J Bone Joint Surg Am.* 1982;64:419–432.
54. Yang JS, McElroy MJ, Akbarnia BA, Salari P, Oliveira D, Thompson GH, Emans JB, Yazici M, Skaggs DL, Shah SA, Kostial PN, Sponseller PD. Growing rods for spinal deformity: characterizing consensus and variation in current use. *J Pediatr Orthop.* 2010;30:264–270.
55. 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.
56. Zhang H, Sucato DJ. Unilateral pedicle screw epiphysiodesis of the neurocentral synchondrosis. Production of idiopathic-like scoliosis in an immature animal model. *J Bone Joint Surg Am.* 2008;90:2460–2469.