Fusionless procedures for the management of early-onset spine deformities in 2011: what do we know?

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Abstract While attempts to understand them better and treat them more effectively, early-onset deformities have gained great pace in the past few years. Large patient series with long follow-ups that would provide high levels of evidence are still almost non-existent. That there is no safe treatment algorithm defined and agreed upon for this patient population continues to pose a challenge for pediatric spine surgeons. In this review, authors who are well known for their research and experience in the treatment of early-onset scoliosis (EOS) have come together in order to answer those questions which are most frequently asked by other surgeons. The most basic eight questions in this field

have been answered succinctly by these authors and a current overview is provided.

Keywords Growing spine · Pediatric orthopedics · Fusionless · Early-onset scoliosis · Treatment

Introduction

Early-onset spine deformity has become a highly controversial field of spinal surgery in the last few years. With the advances in implant technology, small children have entered the scope of spinal instrumentation and severe deformities have become easily correctable. However, in small children, correcting the curve is not always synonymous with treating the disease. Again, the documentation of long-term results of early spinal fusion in the recent years and the possibility of life-threatening side effects have turned the focus of research to methods that can control deformity without the need for fusion. The first results are promising. However, the lack of long-term results regarding new treatment methods, the inability to determine standard indications and contraindications for them, and the lack of large patient series due to the relative rarity of these conditions cause some caution in the evaluation of these results. In this paper, researchers who have taken on important roles in studies regarding the treatment of early-onset deformities in the last decade have come together to find answers for the basic questions in the minds of surgeons treating small children with spinal deformities. We hope that the frequently asked questions (FAQ) format, quite common in the Internet world but rarely used in scientific journals, will be an important tool in conveying expert opinion to clinicians and researchers and increase interest in this subject, where evidence-based

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knowledge is so scarce. This paper should be accepted as a first step to write a ‘white paper’ in this field.

Can we manage spine deformities at young ages like we do in older children? Behrooz A. Akbarnia

There has been significant advancement over the past decade in understanding the natural history and treatment options for early-onset scoliosis (EOS) in young children. We now understand that, if untreated, progressive EOS may lead to significant pulmonary complications, including thoracic insufficiency syndrome (TIS) [1]. However, the fusion of multiple segments of a young child’s spine, especially in the thoracic region, may lead to similar unsatisfactory, and even catastrophic, outcomes by preventing normal growth of the spine and thorax [2]. Recent developments in growth-friendly techniques have equipped physicians and their patients with revolutionary treatment options for progressive EOS.

Due to the shortage of evidence-based clinical research in EOS, clinical experience and knowledge-based information are primarily relied upon by the treating physician to formulate a treatment plan. Consequently, there is significant variation between surgeons in deciding on an appropriate non-operative and operative treatment method [3–5]. The selection of the optimal treatment is even more difficult due to the distinctly different etiologies of EOS. For example, should a 20-month-old patient with infantile idiopathic scoliosis be observed, treated with a cast, or undergo surgery? While there is disagreement even among very experienced surgeons about how and if such a patient requires treatment, all would agree that the ultimate goal is to improve the natural history of the patient’s spinal deformity and quality of life.

In recent years, there has been a growing interest for expanding non-operative and operative alternatives for EOS, and many new techniques have emerged [6–9]. Skaggs [10] introduced a classification of growth-friendly procedures based on the mechanism by which they modulate the spinal and chest wall growth. The classification included distraction-based, compression-based, and growth-guided techniques. Each of these techniques has advantages and disadvantages which will be discussed in this issue. Sankar et al. [11] also studied patients from the Growing Spine Study Group (GSSG) who had growing rods placed and underwent multiple subsequent lengthenings, and they found that there was a law of diminishing returns as the number of lengthenings increased. It appears that, after seven lengthenings, the gain in spinal length was minimal, calling into question if the increasing risks of additional surgeries justify the diminishing benefits. Bess et al. [12] reviewed complications in a large cohort of

patients from the GSSG database and found that, after multiple lengthenings, the rate of complication was significantly higher. Bess et al. concluded there was a clear correlation with multiple surgical procedures and a relatively higher rate of complications. From these recent studies, it appears that there may be an indication for prolonging observation or selecting non-operative treatment in an attempt to reduce the risk of complications associated with repeated surgeries. However, one must also consider the critical developmental changes that the spine and lungs undergo from birth to 5 and 8 years of age, respectively. Ultimately, a balance of allowing normal spinal and lung growth, preventing progression of the spinal deformity, and minimizing complications must be attained. It is possible that this delicate balance may only be achieved when the treatment can be individualized for each patient based on diagnosis, age, and severity of the deformity. Unfortunately, the EOS patient population is small and heterogeneous, making it difficult to study the results of different treatment outcomes in a large and meaningful number of patients. The lack of evidence-based research studies distinguishing favorable versus unfavorable outcomes is further complicated by the scarcity of outcome assessment tools for this complex group of patients. An attempt to develop and validate quality of life measurements is currently underway, which may provide new information on patient outcomes and facilitate future outcomes-based research [13].

We are seeing a notable improvement in the treatment of EOS with a rapid pace in innovation. Future research efforts, both basic and clinical, must match this pace in order to offer real-time information and to objectively assess the results of surgeons’ clinical judgment. The assembly of multicenter study groups is an attempt to overcome the challenges of studying a relatively rare pediatric disorder by collecting a large volume of data in a relatively short period of time. These data are often stored in a central database, where longitudinal patient data from initial visit to final follow-up can be queried and analyzed. Additional enhancements to our research tools will offer new methods to gather information, with the hope that the quality of life of children with EOS will be much improved in the future.

Does the deformed spine grow like the normal one? Alain Dimeglio

Growth of the spine and thorax in the child is evaluated by many parameters, including standing and sitting height, arm span, weight, thoracic perimeter, T1–S1 spinal segment length, and respiratory function. A thorough analysis of these parameters will allow the surgeon to plan the best

treatment at the right moment. Normal values for these parameters have been published previously [1, 14–16].

Only a perfect knowledge of normal growth parameters allows a better understanding of the pathologic changes induced on a growing spine by an early-onset spinal deformity. These deformities have negative effects on standing and sitting height, thoracic cage shape, volume and circumference, and lung development. All growths are synchronized, but each one has its own rhythm. As the spinal deformity progresses, by a ‘domino effect’, not only is spinal growth affected, but the size and shape of the thoracic cage are modified as well. This distortion of the thorax will interfere with lung development. Over time, the scoliotic disorder changes its nature: from a mainly orthopedic issue, it becomes a severe pediatric, systematic disorder with TIS [1, 14], cor pulmonale [15], and hypotrophy. In the most severe cases, these alterations can be lethal.

The growing spine is a mosaic of growth plates and it is characterized by changes in rhythm. During growth, complex phenomena follow each other with significant speed. This succession of events, this setting up of elements is programmed according to a hierarchy. Growth is harmony and synchronization. The slightest error, the slightest slip or modification, can lead to a malformation or to a deformity. Abnormal growth alters this virtuous circle. Spinal growth is the product of more than 130 growth plates working at different speeds, but is strict synchronism. Symmetric and harmonious growth is typical for normal spines. However, in severe scoliosis, the growth plate disorganization leads to asymmetrical growth. Complex spinal deformities alter growth cartilages of the spine and—according to the Hueter–Volkmann Law, which states that “compression forces inhibit growth and tensile forces stimulate growth”—vertebral bodies become distorted and can perpetuate the disorder. Therefore, all scoliotic deformities become, over time, growth plate disorders [16–20]. While cessation or modulation of growth in vertebral growth plates under experimental conditions has been shown to cause deformity, this modulation of growth has also become a modality of treatment safely used in the clinical setting.

Does a deformed spine grow normally? Several studies have been published in the last few years reporting that near-normal growth has been attained with the growing rod or vertical expandable prosthetic titanium rib (VEPTR) treatment of EOS of a variety of etiologies [2, 18, 21–23]. These studies have bolstered the hope that effective control of deformity will help in the restoration of normal growth. However, it should not be forgotten that children with congenital scoliosis often have deficient or supernumerous growth plates in their spines compared with their normal peers, resulting in abnormal patterns of growth. Also, syndromic patients with their numerous comorbidities

often have deficiencies of general health and nutrition, adversely affecting the spine just as much as the rest of their bodies. Putting together these points into consideration for an answer to the titular question, for the time being, this answer is most undoubtedly no.

Spinal and thoracic growth both obey strict rules and can be controlled only by following their requirements. Only the critical analysis of all growth parameters over time allow unmasking and understanding the magnitude of the deficits induced by an early-onset spinal deformity. Four different scenarios can be identified:

1. The clinical picture gets worse. Abnormal growth leads to a deficit that sustains the deformity (‘snowball effect’). Hypotrophy due to weight loss, weakens—among others—the respiratory muscle, making breathing more difficult.
2. The clinical picture is stable.
3. The clinical picture gets slightly better with improvement of the different clinical parameters, such as weight, vital capacity, and sitting height.
4. The clinical picture returns to normal. In this ‘ideal’ scenario, all clinical parameters catch up the deficit induced by the deformity. Unfortunately, this is not likely to happen, as most of the children with severe spinal deformities will end up at skeletal maturity with a short trunk, a significant loss of vital capacity, and disproportionate body habitus.

Therefore, surgical strategies should consider the whole life span of the patient and should provide answers to two basic questions: (1) For what functional benefit? (2) For what morbidity?

How can we understand the child with a crooked spine? Michael G. Vitale

The child with a crooked spine presents a multitude of complex inter-related health issues. In contrast to children with adolescent idiopathic scoliosis, children with EOS represent a heterogeneous population. Children may have infantile idiopathic scoliosis with no other associated comorbidities or may have a primary thoracic insufficiency with severe pulmonary problems, as is present, for example, in Jarcho–Levin syndrome [24, 25]. In order to understand the child with a crooked spine, we need a basis for differentiating the many different groups inherent in this population. Unfortunately, no such useful and comprehensive classification of EOS exists, although efforts by the authors of this article are currently underway to develop such a classification.

One way of understanding the patient with a crooked spine is to assess whether the child has thoracic

insufficiency or not. We are all familiar with a growing preponderance of data showing a relationship between lung function, scoliosis, and fusion, yet, perturbations in pulmonary function are also quite variable in children with EOS [26, 27]. At the broadest cut, perhaps we should understand whether the child with a crooked spine is currently, or soon to be, at risk of suffering pulmonary problems. Such information would logically guide treatment. As defined by Dr. Robert Campbell, TIS is the inability of the thorax to support normal lung function [1, 28].

On another level, the child with a crooked spine may or may not have significant comorbidities. We need to develop a way to consider and categorize such comorbidities. The child with neuromuscular scoliosis at a young age presents a different set of challenges, problems, and, perhaps, different treatment opportunities than the child with idiopathic scoliosis. We need to consider differences between progressive neuromuscular disease, such as spinal muscular atrophy or various other muscular dystrophies and static encephalopathies, such as cerebral palsy. All of these patients can develop scoliosis at an early age, but the manifestations of scoliosis and complications of treatment vary [29]. Another group of patients with EOS are children with comorbid cardiac conditions, including thoracogenic scoliosis. Finally, there is a large group of heterogeneous syndromes, ranging from Marfan's disease to osteogenesis imperfecta, which present with EOS, and, again, this informs our understanding of the disease and our choice of treatment [30, 31].

We must consider the age of the child. Clearly, the 14-month-old child, the 4-year-old child, and the 8-year-old child represent different problems and different opportunities for care. Growing evidence suggests stalling intervention in the younger children and the use of casts [6]. For an intermediate age group, growth rods using rib-based foundations such as the VEPTR may be more appropriate. Traditional growth rod systems which utilize spinal fixation may be preferable on somewhat older children, and opportunities for growth modulation using staples and other devices will likely be increasingly prevalent in the juvenile age child [32, 33].

In contrast to adolescent idiopathic scoliosis, where the curve pattern is the largest source of variability among patients, children with EOS have a plethora of other differences. Nevertheless, characteristics of the curve pattern need to be understood, appreciated, and treatment appropriately, customized for specific curve patterns. Does the child have pelvic obliquity or subluxation of the hip? Is this a long, sweeping curve or a double or triple major curve pattern? Is this a curve which would lend itself to an apical fusion and Shilla-like strategy, or do we need to span from proximally to distally, with concern about progression over time?

It is critical to appreciate the entire available bone stock in the young child with scoliosis. Bone stock varies depending on age, diagnosis, ambulatory status, and other variables. In considering bone stock, we must look not only at the lamina, pedicles, and transverse processes of the spine, but importantly at the ribs, which present an opportunity to avoid the spine and, thus, avoid fusion in a young child. There is still significant uncertainty about the ideal means of pelvic fixation in growing constructs, with multiple options available, including S hooks and screws. None of these are ideal in the youngest patients [34].

We know so little about the relationship between thoracic structure and scoliosis in a young child. On a very superficial level, we have been informed by Dr. Mehta's observation about the prognostic effect of the rib/vertebral angle difference, yet we know very little about how the ribs relate to the apex of spinal deformity in young children, the effect of rotation of the thorax and/or spine relative to each other, and the validity and reliability of such measurements as measured by traditional X-rays [35].

Finally, we must understand something about the child's psychosocial framework and that of their parents before we enter into what tends to be a long and involved contract of care of these children [36–38]. Quality of life scores have been shown to be very low in patients with EOS and thoracic insufficiency, and we need to maintain an appreciation for the broad ways in which this disease state affects the lives of patients and families. We have developed an EOS questionnaire which reflects many aspects of health that are important to this patient group—lung function, sleeping, play behavior, activities of living, physical and psychological function, etc. We have also shown adverse psychosocial effects and issues of anxiety in a subset of children who undergo repetitive surgery for EOS. In some situations, repetitive surgery may not be worth the potential gains, and we may consider fusion to avert psychosocial or even physical morbidity in some children.

In summary, the child presenting with EOS is not a single child, but a population of children. This complex heterogeneous patient population presents a significant diagnostic challenge. For this reason, there is an urgent need for a classification system which integrates a host of variables that will inform us about the optimal treatment strategies of this population. In fact, the authors of this article are working towards the development of just such a classification system.

How do the lungs and thorax interact with the spine during postnatal growth? Gregory J. Redding

It is intuitive that growth of the normal lung and thoracic cage parallel one another after birth. The volumes of both

structures increase in a non-linear fashion over the first two decades of life, with rapid growth occurring before 3 years of age and again during the pubertal growth spurt [19]. The volumes of both structures are proportional to height when spine disease is absent, and norms for lung function in children, including lung volumes, are based primarily on standing height [39]. Lung growth is a complex topic, as different pulmonary structures and regions grow at different rates. At birth, the newborn has the same number of conducting airways as an adult. Tracheal caliber increases two- to threefold between birth and adulthood [40]. In contrast, the peripheral regions of the lung that contain alveoli and pulmonary capillaries, known collectively as the acinar regions, undergo substantial postnatal growth and development. From infancy to adulthood, the alveolar number increases by up to sixfold and the alveolar-capillary surface area increases more than tenfold as a result of increased alveolar number, complexity and septation, and capillary development [40, 41].

In patients with prenatal lung hypoplasia, such as in children with diaphragmatic hernias at birth, the potential for postnatal lung growth is limited, with reduced alveolar number and alveolar-capillary surface area, despite a catch-up to normal lung volumes [42]. Lung carbon monoxide diffusion capacity, a clinical measure of alveolar-capillary surface area, remains abnormally low in these patients, despite the normalization of vital capacity values [42]. This suggests that lung hypoplasia at birth will limit the alveolar and capillary number, while alveolar volume increases with postnatal thoracic cage and spine growth. Likewise, certain thoracic cage disorders, such as Jeune's syndrome, develop prenatally and have small thoracic cage dimensions and lung volumes at birth. Lungs at autopsies in the past were described as 'hypoplastic', but morphometric analyses are lacking [43]. Recent advances in thoracic cage expansion have increased the survival of these patients, but lung function and diffusion capacities have not been described postoperatively over time in order to discover how much more lung growth can occur.

Other clinical conditions, which begin after birth, suggest that there is a bidirectional interaction between the postnatal growth of the lung and the thorax. Recent work in young rabbits undergoing unilateral rib tethering and, hence, mild scoliosis shortly after weaning demonstrates a complex relationship, with reduced concave lung volumes on the tethered side but compensatory increases in lung volume on the contra-lateral side [44]. However, histology in this model demonstrates that alveoli appear to be simplified in structure throughout and not localized to one side. This suggests that chest wall movement restriction and reduced hemithorax size may have a global effect on alveolar and capillary development throughout both lungs [44]. There is limited histological data in the literature on

this topic. The lungs of four patients with severe scoliosis who died at 8–15 years of age were described at autopsy three decades ago by Davies and Reid [45]. The authors described severe pulmonary hypoplasia and pulmonary vascular changes consistent with severe pulmonary hypertension. The alveoli were described as fewer in number, simpler in shape, but variable in volume compared to normal features for age, suggesting that postnatal alveolar development and proliferation were impaired by limited spine and thoracic growth.

More recently, Goldberg et al. [46] reported the lung volumes and lung diffusion capacities of 21 patients with infantile idiopathic scoliosis who were >15 years of age at the time of study and subdivided them into three subgroups. Group 1 required no intervention as they had mild non-progressive scoliosis. Group 2 required fusion after the age of 10 years for progressive spinal deformity. Group 3 required fusion prior to the age of 10 years for rapidly progressive deformity. The table in that paper illustrates that preoperative Cobb angles were worse in those undergoing spine fusion but no different between those undergoing late and early fusion. However, both vital capacity and lung diffusion capacity were worse in the group fused early compared to the other two groups. Assuming that diffusion capacity was corrected for lung volume, these results suggest that the timing of the spine and chest wall deformity and its rate of progression impact lung growth in the acinar region. The earlier the deformity and its progression, the worse the lung function, based on vital capacity, and the more profound the impact on postnatal lung growth, as depicted by the volume-corrected lung diffusion capacity. Early spine fusion per se, could also have contributed to these pulmonary abnormalities.

In addition, the respiratory tract can dictate the shape of the thoracic cage. Children with airway obstruction and gas trapping, such as those with asthma, develop an increased chest wall depth and barrel-shaped chest, which may or may not reverse over a period of months to years after treatment is begun. Similarly, fixed airway obstruction, such as subglottic stenosis, can produce a secondary pectus excavatum deformity, presumably as a result of long-term increased intrathoracic pressures during inspiration [47]. Up to 27% of children with congenital diaphragmatic hernia and unilateral lung hypoplasia develop scoliosis concave to the hypoplastic side by adulthood [42, 48]. Whether this is related to the thoracotomy performed to correct the diaphragm defect early in life or the degree of unilateral lung hypoplasia or both is unclear. Thoracotomy unrelated to lung hypoplasia in children with cardiac disease also increases the risk of scoliosis postoperatively [49]. The chest wall is most deformable early in childhood and the changes in thoracic shape and dimensions due to primary lung/airway disease usually occur early in life.

It remains unclear what potential exists for compensatory lung growth, as opposed to lung expansion, following the surgical correction of the spine and expansion of the thoracic cage. Reports by Karol et al. document that early spine fusion for progressive scoliosis limits further spine growth and leads to diminished thoracic height, and, hence, a long-term loss of vital capacity [2]. Growth-sparing techniques to minimize the progression of spine curvature have been developed recently, but is not clear that such techniques reverse all aspects of scoliosis, e.g., spine rotation, despite improvement in the Cobb angle. Serial lung function measurements before and after expansion thoracoplasty and the placement of VEPTRs demonstrate that lung volumes are preserved and increase in absolute terms almost commensurate with growth over a 4-year postoperative interval [50]. Earlier intervention with VEPTRs, before the age of 4 years, appears to preserve lung growth better based on one small series [51]. To date, lung diffusion capacities have not been serially measured using growth-sparing surgical devices to discover if postnatal acinar development is similarly preserved.

Numerous animal studies have demonstrated that the lung is capable of postnatal compensatory growth following lobar resection [52]. What remains unclear is the potential for compensatory growth and development in a lung that is progressively constrained over time as spine and thoracic cage deformities worsen. Maximal compensatory lung growth may well be limited to a specific time after birth and diminish after the period of alveolar multiplication is complete. Published estimates of this period of alveolar multiplication vary from 1 to 8 years of age [53–55]. The optimal timing of surgical interventions to expand the thoracic cage to both minimize progressive postnatal pulmonary hypoplasia and maximize compensatory lung growth still need to be determined, but are likely to be early rather than late in childhood.

The rod is growing. What about the spine? **Muharrem Yazici**

Although breathtaking developments have taken place in the field of spinal surgery in the last three decades, the main principles of treatment have remained the same: maximum safely attainable correction, the restoration of physiological spinal contours and trunk balance, and that this is done by fusing the fewest mobile segments possible. With current advances in surgical, anesthesiologic, and intensive care techniques and improvements in implant technology, these goals are attained with less difficulty. In adolescent and adult deformities, a near-normal appearance can be achieved when spinal mobility is sacrificed. In the future, research in adolescent and adult deformity surgery

will focus on the development of techniques which will stop deformity before it forms and control it without giving up spinal mobility.

With contemporary spinal implants sized down appropriately for pediatric use, the same success in deformity correction has been achieved in EOS as well. In EOS, however, correction of the deformity is only one facet of the problem. The protection of the child's potential for growth is as important, and perhaps even more so, than deformity correction itself. This problem, which remains to be solved, has made EOS the area of interest most open to progress and one which has shown a great deal of development in the last several years.

“A short but straight spine is better than a long and crooked spine” has, for the longest time, been the undisputed and many-times repeated motto of spinal surgery. This motto is unquestionably consistent in itself. If one of these options must be chosen, shortness should be accepted for a well-aligned spine. However, in recent years, it has been shown that the problems caused by a shortened spine or one not permitted to grow affect more than the spine itself, and that they cause negative effects on every aspect of the child's growth from the thorax to the cardiac system has been proven beyond doubt [2]. In order to avoid these two unfavorable outcomes, this evidence has led researchers to investigate the possibility of a third option. Is it possible to attain a long and well-aligned spine? This is the most pertinent question regarding EOS today.

Since the 1970s, spinal implants as internal braces have been in use for the treatment of early childhood deformities not controllable by conservative means [56]. While received with great enthusiasm, Moe's subcutaneous Harrington rod application lost its popularity in the 1980s. This is due more to the disappointment because of the preservation of growth potential not meeting expectations rather than the high rate of complications experienced with this technique. This disappointment led to the abandonment of the rigid, fused spine that did not grow quite as much as expected and experienced many complications with the classic subcutaneous Harrington instrumentation and the adoption of studies recommending the use of conservative treatment to its limits and then achieving full correction with early fusion [57, 58]. However, with the beginning of the 1990s, Akbarnia et al.'s new technique of double growing rod instrumentation with strong anchor foundations at either end and their modification of routine lengthening every 6 months without waiting for the deformity to increase has shown good results and led to the dissipation of the earlier disappointment [7]. The technique, known as the ‘subcutaneous rod’ until then, became the ‘growing rod’ at this point.

While the meaning of the word ‘growth’ includes ‘increase in size, number, value, or strength’, in biological sciences, it means the accomplishment of a previously

defined rhythm and that it does this on its own without outside intervention. The length of the rod in the growing rod technique does not grow on its own, nor does it do it with any kind of rhythm. Instead, it is lengthened with a surgical procedure, an outside intervention. For this reason, the term ‘growing rod’ has been in dispute, and, in the beginning, it was suggested that the technique should rather be called ‘lengthened rod technique’ for the sake of accuracy. Yet, the appeal of the concept that the word ‘growth’ communicates has led to the acceptance of the new term and its safe settlement in the spinal surgery literature. This optimistic claim became scientific truth with the publication of well-documented patient series treated with this technique and the verification of continued spinal growth during the duration of treatment.

In the studies regarding normal spinal growth by Dimeglio et al., it has been shown that the T1–S1 segment grows 1.2 cm per year between the ages of 5 and 10 [59]. In Akbarnia et al.’s series, where 23 patients treated with the growing rod technique were carefully evaluated for the change in vertebral height, it was shown that the T1–S1 segment in these patients exhibited a growth of 1.2 cm per year as well [7]. This paper is the first publication that proves that the growing rod technique allows normal development of the spine while providing effective correction. In a subsequent study by the same group of authors [60], a similar group of patients was assessed for the relationship between spinal growth and the frequency of lengthenings; it was found that, with increasing frequency of the distractive force applied to the spine by the rods (via routine lengthening), more growth could be achieved. While 1.8 cm/year growth was attained in patients lengthened every 6 months, less growth was shown to have occurred in patients who received less frequent lengthenings. This study has encouraged the belief that growth can not only be preserved with the growing rod technique, but it can be stimulated by more frequent lengthenings as well.

VEPTR is another fusionless instrumentation technique that, although indirectly, also applies distraction on the spinal column. While it does not focus on the spinal growth per se, it has been found in clinical studies that, after repeated attempts at the lengthening of this system, significant growth occurs not only in vertebral bodies but also in the anatomic regions that are designated as unsegmented bars on plain films [14]. While the idea of growth in a region lacking a growth plate such as the unsegmented bar was received with skepticism at first, the same observation has been repeated in congenital deformities treated with the growing rod as well [61]. This situation has been explained with distraction stimulating appositional growth or the structure thought to be an unsegmented bar on two-dimensional X-rays has, in actuality, remnants of growth plates that are induced to grow.

Continuation of the normal growth of the spine is expected in situations where spinal fusion has not occurred. Is it possible that distraction stimulates vertebral development? The Hueter–Volkman law is a principle described many years ago that has been repeatedly proven correct on long bone epiphyses by many experimental as well as clinical models [62]. Is the response of the growth that occurs at the vertebral apophysis, which does show certain histological and anatomic differences, similar to that of the appendicular skeleton?

Stokes et al. have applied Ilizarov-like devices to mouse tails and shown that, with the employment of specifically designed springs exerting distraction, vertebral growth is stimulated and, with compressive forces, it is impeded [63]. This study proves that the Hueter–Volkman law applies to the apophyses in the tails of mice at least. Yilmaz et al. [64] have devised a model in immature pigs that simulates the growing rod technique used in humans and researched the effects of distraction on the growth of the vertebral body. In this study, the speed of growth of vertebral segments under distraction was found to be significantly higher than that in the control segments. While the direct extrapolation of this observation to the human is hindered by its being carried out on an animal model and in non-scoliotic spines, this finding is essential in that it shows that vertebral growth can be stimulated with distraction in the lumbar spine. Lastly, Olgun et al. [65] have presented a study in which 20 patients treated essentially by the growing rod modification as described by Akbarnia et al. were assessed for the comparison of growth rates between vertebrae within instrumentation levels and those without. In this study, it has been shown that segments within distraction grow faster than those outside it. Measurements were performed on lower thoracic vertebrae which were designated as intermediate segments, while control segments were lumbar. The lumbar vertebrae in these patients have shown slower rates of growth compared to the lower thoracic vertebrae, although it is known that lumbar vertebrae grow faster than thoracic vertebrae in the normal spine [59]. Or, to put it in a better way, the growth rate of the lower thoracic vertebrae has surpassed that of the lumbar vertebrae.

However, distraction is not the only factor that affects the mobility, health, and growth of the spinal column; immobilization of the motion segments within instrumentation may also play a role. Kahanovitz et al. [66], in an animal study, applied fusionless instrumentation and reported histologically identifiable changes in the facet joints that have been immobilized by the rods but not included in the arthrodesis. Therefore, the growing rod technique is still being criticized in that it will not result in more physiological motion after the rods were removed, because the spanned (not included in the arthrodesis)

segments would undergo fibrosis, ankylosis, and even autofusion [57, 58, 67]. Soft tissues, as well as bones, are affected by these forces. Histological analysis in animal studies of intervertebral discs showed that compression leads to degeneration and distraction to regeneration [68–70]. A more recent study suggests that endplates are also effected by compression and distraction forces [71]. In this study, Hee et al. showed that compression led to a decrease of vascular channel volume in the endplates and distraction led to recovery, and that after the application of compression, discs showed ossification of the cartilaginous endplates. The 4-mm-diameter rods usually preferred for the pediatric age group are fairly flexible and do not cause absolute immobilization in the motion segments that are spanned by the instrumentation, especially if many levels are included, and allows some degree of motion. Again, by performing frequent lengthenings, long-term immobilization is avoided. For all of these reasons, it can be speculated that the pediatric growing rod will not be afflicted by the disadvantages of fusionless instrumentation in the adult. Long-term results of contemporary growing rod techniques have not yet been reported. Therefore, it remains to be seen whether this speculation will, in the future, become scientific fact or remain wishful thinking.

In conclusion, strong evidence exists regarding growing rod treatment preserving spinal growth and that the spine grows along with the rod (continues its normal growth). With the increase in frequency of distraction applied to the vertebral column (via routine lengthening), fusion/ankylosis rates have been shown to decrease, and with the introduction of self-lengthening or externally driven devices that will allow more frequent distraction into routine practice, it is hoped that this growth will take place with fewer problems. The observation that extra spinal growth can be achieved with the growing rod treatment should be confirmed with larger patient series followed for longer amounts of time.

Are fusionless procedures another type of ‘birthday party syndrome’? Social and psychological aspects of multiple interventions and hospital stays

Paul D. Sponseller

EOS is a challenging condition for patients and caregivers. It may seriously impair the eventual quality of life [26]. The indications for observation, casting, bracing, and various fusionless procedures are becoming established [26, 32]. Several types of distraction and growth-guiding techniques have been developed to alleviate the effects of early fusion, including the use of growing rods and VEPTR, and there will certainly be new growth-guiding options in the future [26]. However, the introduction of these procedures

has introduced the phenomenon of repetitive surgery to the pediatric spine world. Mercer Rang coined the term ‘birthday party syndrome’ for the phenomenon of children with cerebral palsy who required multiple, unplanned surgical procedures in successive years, causing them to often celebrate their birthdays in the hospital. The implication was that the surgeon did not appreciate the effect of one surgery necessitating a later one. This has led to the anticipation of such consequences and the performance of multilevel single-event surgery to minimize occurrence of the ‘syndrome’. However, in growing spine surgery, we acknowledge a priori that there will be multiple planned surgical procedures. In addition, there are even more unplanned procedures, due, in part, to the 15% rod fracture rate, as well as other complications.

We have tried to quantify the magnitude and the consequences of the ‘birthday party syndrome’ in growing rod surgery. We studied the age of patients when they start growing rods, age at until final fusion, and the mean lengthening interval. From 1994 to 2007, 265 patients underwent growing rod surgery at 16 international centers. The mean treatment time for active patients was 4.5 ± 1.9 years. Patients who had completed treatment and reached final fusion had an average treatment time of 5.1 ± 2.4 years. In the database, the mean age at initial surgery was 6.0 ± 2.5 years, with 94% of patients <10 years of age at growing rod insertion. The mean lengthening interval for the 265 patients in the database was 8.6 ± 5.1 months. Five of 16 centers had experienced familial resistance towards regular lengthenings. Their concerns included concerns of risk/reward after initial procedure, perception of psychological effects, parents not perceiving a clinical change in the child to necessitate lengthening, and realization of the burden of care. The scheduling of lengthening was the responsibility of the family in seven practices, of the surgeon in six, and of both in three. A trend approaching statistical significance which we noted was a decrease in age at growing rod insertion.

The database included 61 patients who finished the lengthening phase of treatment at a mean age of 12 ± 1.8 years. This implies that patients who have growing rods inserted have the potential of undergoing up to 12 procedures before final fusion. Our database showed that few patients reached this level.

Because growing rods are a complicated and long course of treatment, several authors emphasize the importance of having an understanding of and an agreement with the patient’s family [7, 26, 60, 72–74]. This is critical when it comes to adhering to the recommended lengthening intervals. However, the theoretical discussion of the program may not be matched by a willingness to carry through with repeated lengthenings in all families once they experience the process. Studies of repeated procedures of other types

(such as voiding cystourethrograms and even general anesthesia [73, 74]) have demonstrated this.

Akbarnia et al. and others have shown dual-rod treatment to be the most effective when the lengthening interval is 6 months or less, regardless of progression [57, 60, 75–78]. Our survey showed that most surgeons are in agreement with this practice and have a preferred lengthening interval of 6 months. However, only 23% of intervals fell within this time. The mean interval was 8.6 ± 5.1 months. The survey indicated that both scheduling factors and reluctance by families may be factors in causing the intervals to be longer than preferred. The same factors are likely to be operating in VEPTR populations. Until the development of procedure-free lengthening (the ultimate solution to the problem), further efforts will be needed in defining and carrying out lengthening at appropriate intervals. Education and support programs for families will need to be built into clinical protocols.

Does a normal shape of the thorax mean a normal function? Robert M. Campbell

The thorax is a complex, dynamic anatomic structure, which serves to protect the heart and lungs, powers respiration through diaphragmatic contraction and rib cage expansion, and provides support for the girdles of the shoulders, cervical spine, and the cranium. The anatomic definition of the thorax includes the ribs, the sternum, with the thoracic spine as its posterior boundary, and the diaphragm as its lower boundary.

The diaphragm is continuous with the muscle layer of the abdominal wall [79]. The arterial supply of each hemidiaphragm originates from the internal mammary, intercostal, and phrenic arteries, and collateral circulation is so abundant that only severe vascular compromise affects diaphragmatic contractility [80].

The gross shape of the adult thorax is complex, roughly elliptical in cross-section, and widest in the coronal plane at the level of the 8th/9th ribs in the mid-axillary line, narrowing proximally up to the 1st rib, and slightly tapering inward distally. The lateral/anterior inner surfaces of the ribs from the 1st to the 9th face relatively downward, while the ribs more distal face more medial. The anterior wall of the thorax, the sternum, is approximately 50% the height of the thoracic spine.

The first rib articulates solely on the body of T1, and the 11th and 12th ribs also only articulate on their respective vertebral bodies. Ribs 2–9 articulate with the spine between vertebral bodies, through a fibrous disk bridging across the intervertebral disk that supports the convex articular superior and inferior facets of fibrocartilage, with the corresponding rib head concave facets attached within a

synovial cavity. The joint is stabilized by a strong radiate ligament fanning out from the rib head to the disk, and the vertebral bodies above and below. A second synovial joint exists more laterally at the articulation of the tubercle of the rib and the transverse process of the vertebra, the joint being stabilized by three strong ligaments: two ligaments stabilizing the articulation directly to the same transverse process, while a third, the superior costo-transverse ligament, passes from the neck of the rib to the transverse process above it. The facet joints of the rib tubercles differ in orientation based on location. The upper seven bilateral rib–transverse process facet joints are oriented in a vertical plane, but, distally, the facet joints are primarily horizontal. The 11th and 12th ribs have no articulation with the transverse processes of the respective vertebra.

Growth of the thorax is complex, with changes in both function and geometry over the time. The gross thoracic volume is 6.7% of the adult size at birth, enlarging to 25% by the age of 5 years, further increasing to 50% by the age of 10 years, finally reaching full adult size by skeletal maturity [17]. The growth of the thorax and lungs have to parallel each other closely in order to ensure normal pulmonary development [1]. Lung growth by alveolar cell multiplication is maximum during the first 2 years of growth, continuing to a lesser degree until the age of 8 years, with later lung increase in size due to alveolar cell hypertrophy.

The cross-sectional geometry of the thorax changes considerably during growth as a function of rib growth and the relative inclination of the ribs from the spinal plane. In the fetal stage, the thorax is narrowed in the transverse plane, but, at birth, becomes more circular in cross-section, then gradually assumes the adult elliptical shape through poorly understood mechanisms related to individual rib orientation and growth. In infancy, there is transverse orientation of the ribs, with respiration almost totally diaphragmatic without any significant contribution of the rib cage to lung expansion during inspiration. By the age of 4 years, the ribs begin to angle downward, narrowing the anteroposterior (AP) diameter of the thorax, with the change completed by the age of 10 years [81]. This new orientation allows the ribs to contribute to active respiration with outward motion of the rib cage during inspiration. By this time, the thoracic cross-section begins to resemble an oval configuration. By skeletal maturity, the ribs further incline downward, with a cross-section assuming the adult configuration of an ellipse. This change in thoracic symmetry is measured by the ‘thoracic index’, the thoracic transverse diameter/AP diameter $\times 100$, which is less than 100 at birth, increasing to 103–135 by adulthood, with the index in males being slightly higher.

The movement of the thorax with postural changes is complex, with the thoracic spine capable of only slight

rotation, lateral and forward flexion, and extension. The osseus–chondral ribs/sternum complex have an intricate pattern of motion with respiratory rib cage expansion based on complex movement of the rib–vertebral body articulations and eccentric motion of the rib shaft anteriorly, with mechanisms differing based on the level of the thorax considered. Galen (129–200 AD), through animal experiments, first began to define the thoracic respiratory pump through postmortem dissections and nerve ablation experiments in live specimens [82]. He classified muscles as either expiratory or inspiratory, defined the innervations of the diaphragm, and analyzed both diaphragmatic and chest wall motion with respiration.

The dynamic shape of the thorax changes considerably with inspiration and expiration, reflecting complex rib cage expansion mechanics and diaphragmatic contraction. The thoracic respiratory pump depends on muscle force, gravity, and the structural properties of the chest wall to function. In adults, the rib cage expansion of the lungs is responsible for 20% of vital capacity, while the diaphragm provides the additional 80% of lung expansion.

The change in cross-sectional thoracic shape with respiration depends on the level of the thorax. Proximally, with inspiration, the first rib rotates upward, pivoting on the rib head on an axis running along the rib neck, with the rib tubercle rotating at the costotransverse joint, which extends obliquely posteriorly. The anterolateral portion of the rib shaft moves up and down in what is described as a ‘pump handle’ movement, with the anterior section of the rib shaft moving upward and slightly forward with almost no excursion laterally, with the sternum anteriorly moving upward. As one proceeds distally in the thorax, the axis of rotation of the costovertebral joint gradually shifts to a more anterior-posterior orientation, so that inspiration causes the ribs to not only rotate anteriorly, but also laterally, with an increase in both anterior, posterior, and transverse cross-section of the chest, typified by the term ‘bucket handle’ movement. The costal cartilages connecting the osseous ribs to the sternum move freely with inspiration, and supply elastic recoil to help the chest wall return to normal with expiration. The lower two floating ribs articulate only with the vertebral body and rotate posteriorly with deep inspiration.

The volume of the normal growing thorax is provided by growth in height of the thoracic spine, normal anterior and transverse outward growth of the rib cage, and normal orientation of the ribs appropriate for age. By skeletal maturity, the height of the thoracic spine is normally 26.5 cm for females and 28 cm for males [17]. A decrease in that height, due to either congenital malformation of the thoracic vertebra or iatrogenic shortening by early spine fusion, could possibly decrease the thoracic volume as well as the lung volume. Karol et al. [2] noted

that patients undergoing spine fusion early in life began to have significant risk of severe restrictive lung disease when their thoracic spinal height at skeletal maturity was 22 cm or less, probably reflecting decreased thoracic volume and TIS. A primary chest abnormality such as rib fusion could also negatively impact thoracic volume and shape.

The function of the thorax in respiration can be affected by deformity. In EOS, there is a distortion of the convex hemithorax from rotation of the spine, forming a ‘rib hump’, stiffening the chest with a negative effect on vital capacity, as well as loss of volume of the convex lung because of the windswept deformity of the chest.

Both the shape and the function of the thorax have practical applications for the treatment of children with spinal deformity. Two important goals of growth-sparing surgical techniques in EOS should be to prevent adverse change in the shape and function of the thorax in order to promote optimal pulmonary function that would remain stable throughout the patient’s lifetime. The ideal surgical technique for these goals currently does not exist, but awareness of the need to consider ideal thoracic shape and function in the treatment of spine and chest wall deformity is becoming more prevalent, and, some day, technology will exist to address all of these issues.

Is this an endless story? When do we stop lengthening? Jack M. Flynn

Over the last few decades, as surgeons around the world recognized the profound negative consequences of early spinal fusion in very young children, an increasing number of spinal instrumentation strategies have been developed to ‘grow the spine’ as the child grows. Naturally, these various strategies all reach an ‘endpoint’ as a child progresses through adolescence toward skeletal maturity. Spine-based growing rods and VEPTR have surged in popularity, and, thus, pediatric spine surgeons around the world have slowly accumulated an ever larger population of older children who had spinal instrumentation since they were very young. Families, weary from repeat surgery and the inevitable unplanned procedures and complications, begin to press the surgical team regarding the long-term plan. Will the instrumentation be removed? Will there be a final fusion? Will part of the instrumentation be retained while other portions are removed? What is the danger long-term of leaving in the instrumentation across the unfused spine? There are many unanswered questions that need to be explored, not only to answer the family questions, but also to make strategic decisions early in growing treatment that will make sense a decade or two hence when the child passes to the skeletal maturity.

In some cases, instrumentation placed early in childhood could logically be left in place. Perhaps the most straightforward example are VEPTR devices used to expand the chest wall of a child with Juene's syndrome or similar sorts of significant thoracic hypoplasia. As these children reach skeletal maturity and the chest wall ceases to grow, the VEPTR devices could be left in place as long as they are not causing skin breakdown or pain. It is difficult to envision a situation where these devices would be a risk to surrounding structures, or a risk to breaking over time. On the other hand, spine base growing instrumentation spanning many segments of the unfused spine would likely be at risk of implant failure over time. One would expect that the rods would be subjected to continuous micromotion across the unfused spine, and proximal and distal anchors would be at risk of pullout given the lack of adjacent bony fusion. This scenario, then, is quite the opposite of the VEPTR example above: empiric evidence would suggest that growing rods spanning the unfused, flexible spine should probably be removed prior to skeletal maturity. Most difficult cases involve those between these two extreme examples. There are many children who have VEPTR devices spanning long, stiff congenitally abnormal spines. Although there may be some micromotion across this instrumentation, the lack of significant motion and rib-based fixation likely put the patient at much less risk of long-term implant failure compared to patients with implants spanning the flexible, non-congenital spine.

Collaborative research efforts by several of the leaders in early-onset spine and chest deformity surgery have recently provided some important initial data regarding the final stage of growing spine treatment. A paper entitled "Is definitive spinal fusion, or VEPTR removal, needed after VEPTR expansions are over?" uncovering an analysis of 39 'VEPTR graduates' was presented by the Chest Wall and Spine Deformity Study Group (CWSDSG) at the Scoliosis Research Society (SRS) 43rd Annual Meeting in Salt Lake City, UT, September 2008. The authors reported on the 39 VEPTR graduates between the ages of 12 and 25 years (mean age 16.6 years). Eighteen had a spinal fusion, 11 will have only VEPTR treatment, and ten were undetermined. Of the patients, 68% with congenital scoliosis/fused ribs or progressive scoliosis had a fusion, while only 16% with hypoplastic or flail chest had been fused. The VEPTR devices were retained in 10/18 'fusion' and 9/11 'VEPTR-only' patients. Two patients had device failure (hook or sleeve breakage) waiting for their fusion. According to their surgeon, only 3/10 'undetermined' patients are likely to have a future spinal fusion; thus, most of the 'undetermined' group will probably become 'VEPTR-only' in the future. The authors concluded that VEPTR endpoint management varies by underlying diagnosis. VEPTR can be the definitive treatment for children

with hypoplastic or flail chest, but most children initially treated with VEPTR for congenital scoliosis, or progressive scoliosis without fused ribs, will have a definitive spinal fusion after the expansions are complete. Regardless, most VEPTR devices are not removed at the end of treatment.

At the SRS 45th Annual Meeting in Kyoto, Japan, September 2010, the GSSG presented their initial results of growing rod patients who have reached the end of treatment. The authors found that, of the 58 patients who reached final fusion, 53 (91%) had a final instrumented fusion, three were observed with growing rods in place, one had implant removal only, and one had a final instrumented fusion aborted for intra-operative neuromonitoring changes. Most patients had more levels fused than the number of levels spanned by their growing rods. The Cobb angle correction at final fusion varied considerably—in some cases, a fusion in situ was performed, in others, maximum correction was sought with osteotomies, compression, and distraction. At final fusion, 25% had minimal correction, 53% had moderate correction, and 15% had substantial correction; there was no correction information on 7%. The overall duration of growing rod instrumentation varied in the same way and required similar grouping into three categories: short (≤ 2 years), moderate (3–6 years), and substantial (≥ 7 years). At the time of final fusion, 22% had short instrumentation time, 57% had moderate instrumentation time, and 21% had substantial instrumentation time. Indications for final fusion varied, but common reasons were 'minimal spinal growth remaining' or 'broken rod'. There was a case of infection triggering the final fusion, and at least one case that documented the final fusion was performed because the family had grown tired of repetitive lengthening. Of the operative notes that specifically commented on spinal mobility at the time of growing rod removal for final fusion, 20% noted the spine to be mobile, 40% noted decreased flexibility with certain areas of autofusion, and 40% noted the spine to be completely stiff (or completely autofused). Smith–Peterson osteotomies were noted in 15% of operative notes; thoracoplasty was noted in seven cases. In most cases, the growing rod anchor sites were useful for the final fusion. Growing rod hooks or screws were often (but not always) exchanged for larger sizes. In several cases, proximal hooks were found to be attached to bony fusion but not to the transverse processes or lamina. In early cases, anterior fusion was often performed at the final fusion, reportedly to 'prevent crankshafting'. The final fusion is most often performed between 11 and 13 years of age, although there were several children older or younger, for a variety of reasons. The decision to move to final fusion was triggered by a problem (such as a broken rod or infection), or by the assessment that there was not much spinal growth left.

Conclusion

To summarize, we are finally obtaining some early data on the status and management of children who have reached the end of the expansion phase of their growing instrumentation. The data show that final treatment varies with underlying diagnosis, the condition of the spine and chest wall, and the instrumentation used. Further prospective studies will generate a better understanding of the status of the spine at the conclusion of growth treatments, and, perhaps, provide some treatment algorithms that can guide pediatric spine surgeons and the families under their care.

While there are many unresolved issues regarding the fusionless instrumentation methods used in the treatment of early-onset spinal deformity, it is obvious that they have improved the day-to-day life of these small children by negating the requirement for long-term external immobilization and allowing them to have an almost normal life with regular play, mostly uninterrupted school attendance, and uncomplicated daily hygiene. Again, it is obvious that this kind of treatment results in the effective control of deformity, while allowing the chest cage and spine to grow at a near-normal rate. Despite all of these advantages, it remains an exhausting, lengthy treatment for the family, the physician, and the child, with repeated surgeries taking their toll on the general health of the patient, as well as the growth and development of the spine and chest cage. Although the current methods have come a long way in the past, they remain far from perfect and still require more improvement.

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