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Complications of Growth-Sparing Surgery in Early Onset Scoliosis

Behrooz A. Akbarnia, MD,*† and John B. Emans, MD‡

Study Design. Review of available literature, authors' opinion.

Objective. To describe complications associated with growth-sparing surgical treatment of early onset scoliosis (EOS).

Summary of Background Data. EOS has many potential etiologies and is often associated with thoracic insufficiency syndrome. The growth of the spine, thorax, and lungs are interrelated, and severe EOS typically involves disturbance of the normal development of all 3. Severe EOS may be treated during growth with surgical techniques, intended to preserve growth while controlling deformity, the most common of which are spinal "growing rods" (GR) or "vertical expandable prosthetic titanium rib" (VEPTR). Although presently popular, there is minimal long-term data on the outcome of growth-sparing surgical techniques on EOS.

Methods. Review.

Results. Potential adverse outcomes of GR or VEPTR treatment of EOS include failure to prevent progressive deformity or thoracic insufficiency syndrome, an unacceptably short or stiff spine or deformed thorax, increased family burden of care, and potentially negative psychological consequences from repeated surgical interventions. Neither technique reliably controls all deformity over the entirety of growth period. Infections are common to both GR and VEPTR. Rod breakage and spontaneous premature spinal fusion beneath rods are troublesome complications in GR, whereas drift of rib attachments and chest wall scarring are anticipated complications in VEPTR treatment. Indications for GR and VEPTR overlap, but thoracogenic scoliosis and severe upper thoracic kyphosis are best treated by VEPTR and GR, respectively.

Conclusion. Surgeons planning treatment of EOS should anticipate the many complications common to growth-sparing surgery, share their knowledge with families, and use complications as one factor in the complex decision as to when and whether to initiate the repetitive surgeries associated with GR or VEPTR in the treatment of severe EOS.

Key words: surgical complications, early onset scoliosis, early onset spinal deformity, thoracic insufficiency syndrome, VEPTR (vertical expandable prosthetic titanium rib), growing rods, spinal instrumentation without fusion. **Spine 2010;35:2193–2204**

From the *Department of Orthopaedic Surgery, University of California San Diego, LA Jolla, CA; †San Diego Center for Spinal Disorders, LA Jolla, CA; and ‡Children's Hospital, Harvard Medical School, Boston, MA.

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Address correspondence and reprint requests to Behrooz A. Akbarnia, MD, San Diego Center for Spinal Disorders,4130 La Jolla Village Drive—Suite 300, LA Jolla, CA 92037; E-mail: akbarnia@ucsd.edu

Early onset scoliosis (EOS) has been defined as a significant spinal deformity beginning before the age of 5.[1] Unlike adolescent spinal deformity, untreated progressive spinal deformity in early life can cause significant health problems for young children and adults, particularly later significant pulmonary compromise. Any treatment of EOS must focus not just on the spine, but also on the development of the chest to improve the child's long-term quality of life. The surgical options for EOS are complex. Complications are common, seemingly inevitable part of the surgical treatment of EOS. Children with EOS might also have associated medical problems and are at risk for even more complications. Most importantly, pulmonary growth and development can be affected when spine and chest deformities start very early in life.

The complexity and potential complications associated with surgical treatment of EOS begins with the varied etiologic diagnoses for EOS. EOS may be associated with congenital vertebral anomalies, bone dysplasias, connective tissue disorders, neuromuscular disorders, or idiopathic spinal deformities. The list of known etiologies is long, each with its own set of unique potential problems. In addition, EOS can be associated with lordosis, kyphosis, or any combination. All these variations in etiology and deformity share the same potential problems associated with untreated EOS. Lung development and pulmonary function are at risk.[2–4] Campbell *et al* has termed the potential pulmonary compromise associated with severe EOS "thoracic insufficiency syndrome" (TIS) defined as "the inability of the thorax to support normal lung growth and respiration."[5] Spine length is also at risk. In addition to spinal deformity which detracts from spine height, EOS has previously been treated with early fusion, to the detriment of final spine height.[2–4] Spine mobility is also at risk in EOS and less well documented.[3] On the basis of these adverse outcomes for EOS, objectives in the treatment of EOS should include the maximization of pulmonary function, spine length, and residual mobility, while minimizing hospitalizations, family burden of care, and complications.

Surgeons and families who are facing these decisions should balance the risk *versus* long-term benefits before initiating any treatment. Because of significant complications of untreated spine and chest wall deformity in this age group, treatments, even with higher complication rates are often chosen to improve the natural history. Some of the difficulty in treatment is because of the very nature of the growing spine and chest wall, and the need

for periodic revision while other difficulties represent true complications. Understanding the nature of the difficulties associated with the treatment of EOS as well as possible complications will allow early detection and timely management of complications.

The goal of treatment in children with EOS is to control the deformity, allow spinal and chest wall growth, lung development, and improve pulmonary function, therefore, provide a better quality of life. Treatment options such as casting or bracing are commonly used methods for early intervention but when the deformity is progressive and severe, surgical treatment is often indicated. Definitive spinal fusion with or without instrumentation has been used as standard approach for a long time, but such fusion, if performed before the growth is completed, results in a short trunk with possible lung underdevelopment and subsequent pulmonary complications.[2–4] There is also a possibility of increasing spinal deformity through adding on of deformity in unfused segments or within the fusion by the "crankshaft" phenomenon.[6,7]

Procedures that allow or encourage spine and chest growth rather than inhibit growth by early arthrodesis have gained popularity in the treatment of EOS. Such procedures are variously referred to as "fusion-less" or "growth-sparing" or "growth-friendly." Skaggs divided these procedures into distraction-based, growth-guided, and tension-based procedures.[8] The most common spine-based distraction techniques are "growing rods" (GR),[9] and rib-based distraction techniques include the use of "vertical expandable prosthetic titanium rib" (VEPTR)[10] with or without expansion thoracoplasty. These techniques follow the concept of periodic distraction to allow the spine and thorax to grow. Like all other procedures for EOS, GRs and VEPTR have been shown to have a high risk of complication. The high rates of complication may be the result of both multiple surgeries and the presence of critical health issues in these patients. In this article, we will discuss common complications and necessary actions to decrease the risk of complications for both GRs and VEPTR procedures. Growth-guided procedures are not discussed since the Luque technique is now rarely used for growth because of tendency for autofusion. Newer growth-guided procedures like Shilla[11] might provide better outcome when longer follow-ups are available. No long-term data are available for tension-based techniques in very young children.

■ General Complications in Growth-Sparing Techniques

Frequent surgeries increase the risk of general complications caused by surgery, such as undesired event due to anesthesia, postoperative period, and hospitalization. Other complications include failure to successfully control progressive spine deformity, ending growth with an unacceptably short or stiff spine, an unacceptably deformed, stiff or small chest, or failing to avert TIS as described by Campbell. Catching up and keeping pace with growth demands repetitive lengthening procedures. Current attitude has evolved into periodic lengthening to "drive" the growing spine.[9,12] Lengthening schedules vary depending on the extent of the spine instrumented and growth rate but may be as frequent as every 4 months in very small children, every 6 months in most children, and every 9 months when only a short section of spine is involved. Children exposed to multiple trauma are also more likely to experience severe symptoms of posttraumatic stress disorder and depression than those who experience a single event.[13] Additionally from a financial point of view more time, surgeries, and complications leads to enormous direct and indirect cost for the patient's family. To achieve satisfactory results with these techniques the treatment period is often long and may take many years to complete.

Complications in GRs

GR treatment necessitates regular rod lengthening; therefore, children receiving initial GR implants at a younger age are likely to undergo more procedures than children initially treated at an older age. Bess *et al*[14] reported a 13% decrease in the likelihood of suffering a complication for each year increase in age at initial surgery. Additionally, it is possible that younger children are less healthy and have more severe scoliosis than older children and accordingly are at greater risk for complications. Klemme *et al*[15] reported multiple complications in a group of 67 children, including 1 death but felt fusion without instrumentation appropriate for severe EOS. In 2002, Mineiro and Weinstein[16] questioned the worthiness of GR techniques. In their report on 16 patients, complication rate was notably high. Implant-related complications were the most common complications in their series with rod fracture as the most common implant-related complication. They also reported skin breakage, wound complication, and malalignment in their patients. Most recently, Bess *et al*[14] reported complications in 140 patients from Growing Spine Study Group (GSSG). Overall complication rate per procedure was 19%. Of patients, 58% had minimum of one complication (mean of 1.2 complications per patient). Fifty-eight percent of complications were managed at the time of planned surgery. Analysis of total complications demonstrated a linear decrease in survivorship (complication free rate) for each surgery performed, indicating increasing complication rates with increase number of surgeries. At 7 procedures, there was a 49% chance of having a complication. At 11 procedures, the complication risk increased to 80%. They concluded high complication rate in this group of patients is a function of sustained treatment duration and the number of procedures required during the treatment period.

Fusion Complications. In standard GR technique, limited fusion is carried out within the 2 upper and lower foundations.[17] This is usually performed at 2 or 3 adjacent levels at each end of the construct. Like other fusion techniques, although very low, there is always a possibility of nonunion within the foundation levels. Founda-

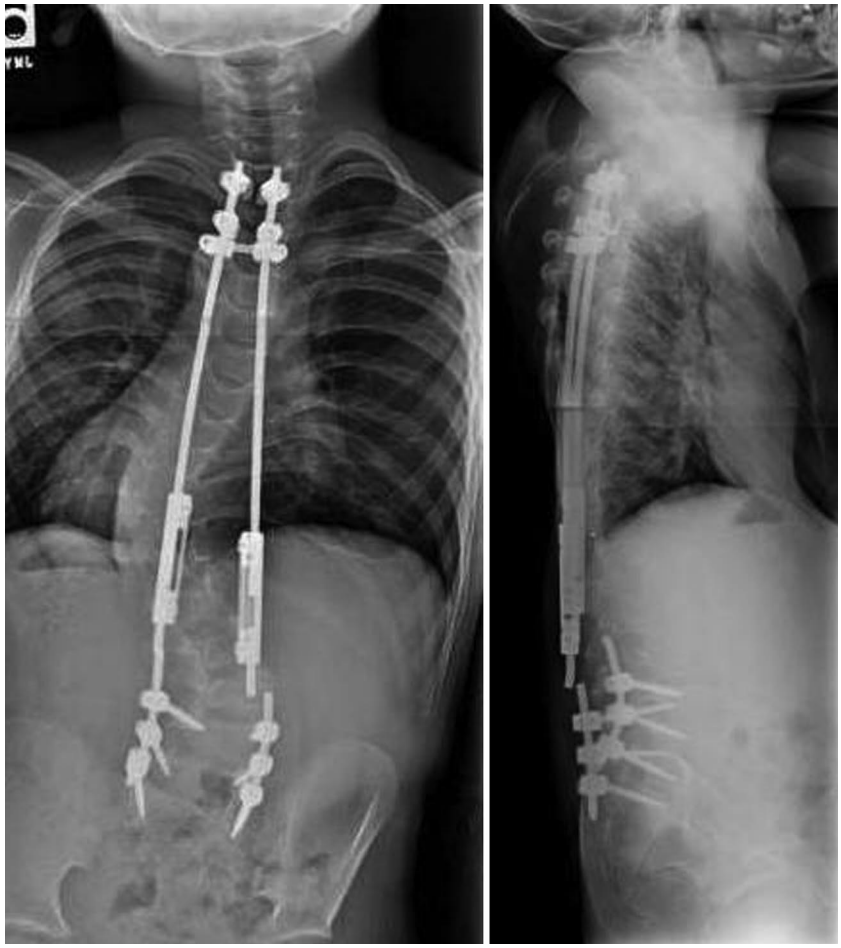


Figure 1. A 6-year-old boy with Beal's syndrome treated with dual GR for progressive scoliosis. Patient returned complaining of low back pain. Posterior-Anterior (PA) and lateral radiographs confirmed rod fracture. Revision surgery was done and the broken rods were replaced (Courtesy of Burt Yaszay, MD, San Diego).

tions are usually designed to carry weight and distraction forces. Motion within foundation anchors can increase the failure rate, which includes wide variety of complications from screw/hook loosening, implant prominence, to rod fracture.

There is also risk of unwanted fusion of adjacent levels if subperiosteal exposure extended beyond foundation levels.

Skin-Related Complications. Multiple surgeries, mostly through the same incision site, leave the skin tissue susceptible to infection and other skin problems. Impaired nutritional status can also increase the risk of skin-related complications and should be attended to before surgery. Tissue handling is extremely important during these multiple surgeries for providing adequate coverage and to reduce skin complications. Skin retraction should be minimized, and skin flaps should have been developed for full thickness skin coverage. At closure, skin should be under minimal tension and flaps are occasionally necessary for coverage. Implant prominence may be unavoidable in very thin and small child. Postoperative padding may be helpful to minimize pressure and possible skin dehiscence. Skin has to be carefully watched for any redness and signs of skin breakdown, and if seen, aggressive treatment should immediately be initiated. Submuscular application of the rod is also shown to decrease wound complications.[14]

Superficial and Deep Wound Infection. This complication may affect the treatment outcome significantly by increasing the number of unplanned surgeries and difficulties of controlling infection. Implants are an essential part of the treatment and implant removal in deep wound infections should be the last resort. The rods can usually stay if the infection is diagnosed early and treated with debridement and intravenous antibiotics. Occasionally with the dual-rod technique, 1 rod can be removed if prominent, with plans for reinsertion later. There is no data available at this time for the length of antibiotic therapy needed in a child with EOS following postoperative deep wound infection. With superficial wound infections, more aggressive surgical intervention and skin closure can reduce the chance of becoming a deep wound infection.

Implant-Related Complications. Implant-related complications are the most common complications in GR surgeries. These include rod fracture, anchor failure, or prominent implant, which can cause skin breakdown and even infection. (Figures 1, 2, 3) Among the implant-related complications, rod fractures are the most common problem.

Yang *et al*[18] reported the GSSG experience of 86 rod fractures in 46 patients. The overall rate of rod fracture was 15%; however, the risk was increased in patients with single rods, history of previous fracture, small diameter rods, stainless steel rods, proximity to tandem

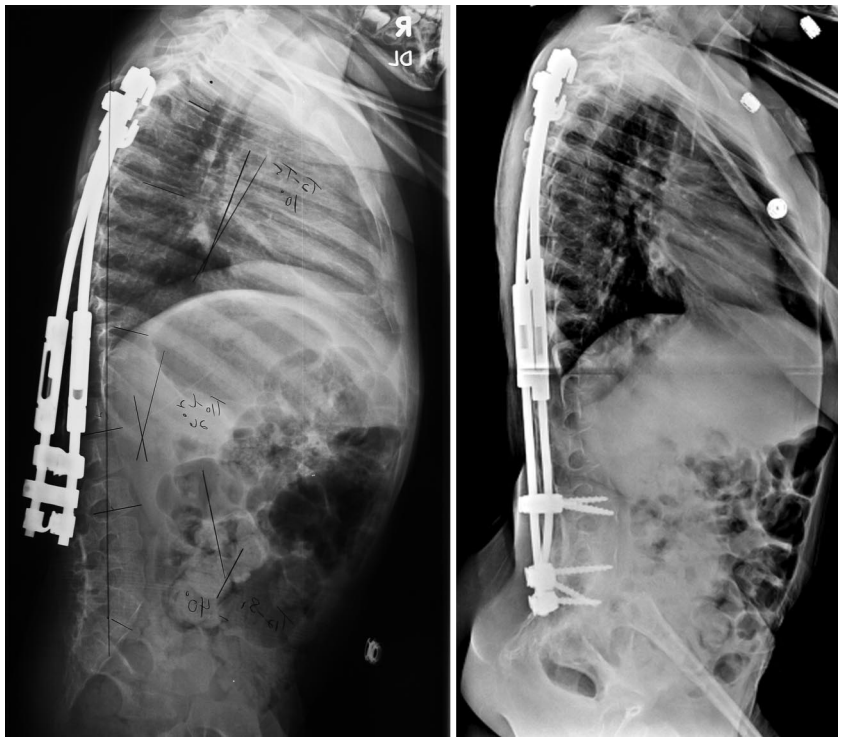


Figure 2. Hook dislodgement in lumbar area in a patient with arthrogryposis treated with dual GRs. Hooks were replaced with pedicle screws and instrumentation had to be extended.

connectors, smaller tandem connectors, and in ambulatory patients. The rate of rod fracture did not correlate with anchor type or degree of the curve. It is advised that replacing the rod may be a preferred strategy over connecting the broken rods.

Asymptomatic implant failure may be revised at the time of planned lengthening surgery. If only one rod in a dual-rod construct is broken, both rods should be changed to prevent early fracture of the second rod.

There are changes of the anchor sites that are expected because of normal spinal growth and are not considered true complications. These include hook and screw migrations, requiring revision which can be usually performed at the time of planned surgeries.

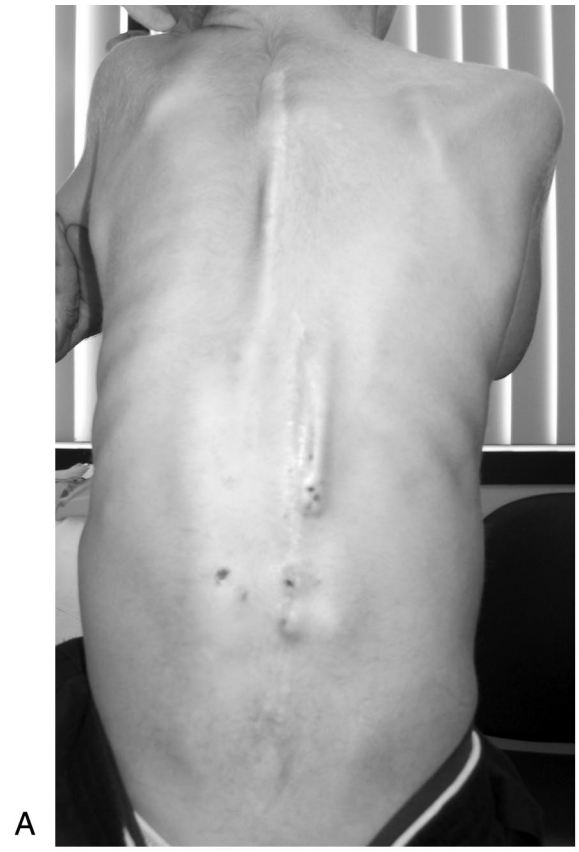
Alignment Complications. It is important to obtain and maintain acceptable coronal and sagittal balance at initial surgery. Different studies have shown improvement of coronal and sagittal plane deformity after initial surgery in both single- and dual-GR techniques.[19] To avoid proximal junctional kyphosis, the rods should be contoured into kyphosis at the top of the construct and the interspinal ligaments should be kept intact as much as possible. The upper foundation is usually extended to T2 and occasionally even higher to reduce the risk of proximal junctional kyphosis. This is especially true in children with nonidiopathic scoliosis. If there is thoracic hyperkyphosis, the rods should be contoured into kyphosis since excessive correction may lead to implant failure after surgery. The tandem connectors should be placed at the thoracolumbar junction and not at the lordotic or kyphotic segments of the spine unless it can be contoured. Short instrumentation, especially in patients with nonidiopathic scoliosis, should be avoided to prevent adding on to the curve as the child grows.

Another possible complication is curve decompensation. If the levels are selected carefully and initial instrumentation done accordingly, curve decompensation is unlikely. In the original report on dual GRs, Akbarnia *et al*[17] reported only 2 cases of curve decompensation following final fusion, both treated by extension of the instrumentation and fusion.

Neurologic Complications. Neurologic complications are uncommon in GR surgeries without associated procedures. Neurologic deficit may occur with excessive distraction or with significant deformity correction. The incidence of intraoperative neurologic injury is 0.1% in index surgeries, revision, and lengthenings.[20] Intraoperative neuromonitoring is a reliable way to monitor changes during surgery and is recommended for primary insertion and exchanges, but controversial for lengthening.[20]

Careful lengthening to avoid overdistraction at initial surgery and at lengthening procedures will reduce the risk of complications. In revision and exchange surgeries using dual rods, it is helpful to maintain a baseline length by keeping one side of the construct intact. Two rare cases of delayed cardiac event reported[21,22] and both recovered after immediate shortening of the rods. Therefore, the child should continue to be closely monitored during the immediate postoperative period for development of any neurologic deficit.

It is necessary to follow proper surgical technique to reduce the rate of complications and to achieve the best long-term results. This is especially important during the initial surgery to pay special attention to the details of



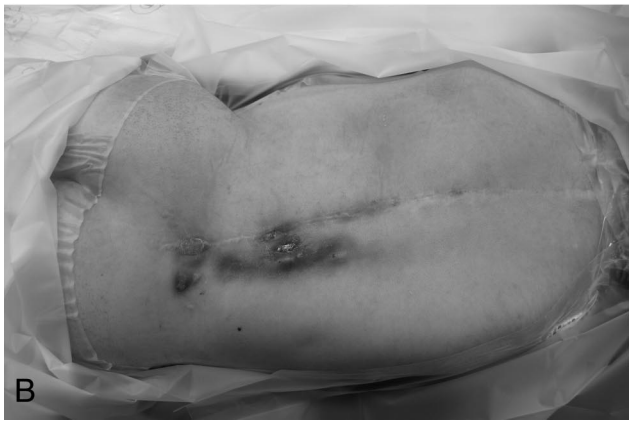


Figure 3. Prominent implants in an 8-year-old patient treated with dual GRs. Prominent implants can cause skin breakage and wound infection if not treated properly.

selection of the levels, exposure, anchor insertion, and rod contouring, and placement to reduce the complication rate in these complex surgical procedures.

Complications in VEPTR and Rib-Based Distraction Devices

VEPTR and GRs share many common problems inherent to repetitive surgery and the growing, deformed spine (Table 1). However, some complications are unique or more common in VEPTR than in GR constructs.[23–26] Knowledge of VEPTR-related complications will help the surgeon choose the most satisfactory growth-sparing surgical technique for a particular EOS and proactively avoid complications as much as feasible.

Anchor Point Problems. The cephalad attachments of VEPTR devices are circumferential rib "cradles," whereas the caudal attachments may be ribs, spine, or pelvis.[10] All anchor points are subject to failure, either acute or chronic, depending on bone quality, stresses applied, and duration of attachment. Migration through a rib by the VEPTR cradle is a common problem. Technical details and patient characteristics both contribute to this complication. Appropriate goals and selection of implants and implant locations can diminish but will likely not eliminate these complications.

Acute, short-term problems are principally rib fracture and rib cradle displacement associated with fracture and typically occurs in association with initial implant, exchange, or lengthening. Rarely dislocation of the entire rib at the costovertebral articulation occurs if excessive distraction force is applied. Simple fractures will usually heal but with some partial displacement if the fractured rib is 1 of 2 encircled. If only a single rib is encircled, or both encircled ribs are fractured, fracture is usually accompanied by significant displacement requiring reinsertion. In the author's experience, rib fracture is commonly associated with initial VEPTR insertion in patients with skeletal dysplasias, osteopenia, or poor nutrition and might be lessened by optimizing bone health. Preoperative assessment of bone density and treatment of osteopenia or osteoporosis will minimize this complication. Rib fracture occurs more commonly with the cephalad attachment of single rib-to-spine "hybrid" devices and can be minimized by load sharing with a second construct. The rib attachment optimally attaches perpendicular to the inferior surface of the rib. Choosing the wrong angle "rib cradle" device makes rib cut-out more likely. Kyphotic deformities corrected by cantilever forces applied through the upper rib attachment may fail acutely if too much deformity is corrected. Excessive distraction force can also fracture the ribs and is probably best avoided by performing an expansion thoracostomy[23] or intercostal muscle lysis so that correction can be achieved with less force. Acute laminar hook displacement with hybrid devices is rare.

Chronic, long-term "drift" or "migration" of rib attachments seem to be nearly universal in VEPTR rib attachments,[10,23–28] but is most common in single devices and in rib-to-spine devices. Campbell *et al*[29] reported drift of rib attachments in 7 of 27 patients. Often the drift is not functionally significant, as the rib cradle gradually drifts cephalad and dorsal and it pulls with it a solid bone attachment and remains functionally connected to its original rib. Some drifting attachments lose functional connection and become both loose and prominent. Reinsertion can usually be done at the time of elective lengthening as the original rib attachment area has usually reformed with new bone. Laminar hook drift in rib to spine devices is also common but is often not significant in that new bone forms around hook as it drifts with the hook remaining functionally stable. Campbell *et al*[29] noted significant laminar hook drift in 4 of 37 patients,

Table 1. Comparison of Indications, Treatment, and Complications in GR and VEPTR in EOS

	Growing Rods	VEPTR
Best indication	Normally segmented spine, flexible chest deformity	Thoracogenic scoliosis or fused ribs
Relative contraindication?	Primary chest wall deformity	Poor soft tissue coverage
Multiple operations needed?	Yes	Yes
Upper thoracic kyphosis?	Possible control	Poor control
Spine growth?	+	+
Chest deformity correction?	When flexible	Direct, invasive
Ease of final fusion	Difficult, scarred	Easier, unscarred
Final fusion needed?	Yes	Yes
Failures—common	Rods break	Rib attachments drift
Complication—severe	Spontaneous posterior spine fusion	Chest wall stiffness

GR indicates growing rods; VEPTR, vertical expandable prosthetic titanium rib; EOS, early onset scoliosis.

with 2 eventually disengaging completely and requiring revision. Revision is straightforward. Iliac S-hook drift is also common over time, particularly in unilateral devices. The drift of the iliac attachments is generally distal, not posterior or lateral in direction. Often this is not significant, but occasionally the hook may subside sufficiently to threaten integrity of the acetabulum. Revision is straightforward but might require significant exposure and should be undertaken before the device becomes so inferior as to approach the hip joint.

Brachial Plexus Problems. Among growth-sparing instrumentation, brachial plexus problems are unique to VEPTR and have been described in several series.[24,30,31] Their occurrence reflects the use of ribs on the chest wall as anchor points, the common use of expansion thoracostomy, and the use of VEPTR in congenital deformity with major anomalies of the chest wall and shoulder.[30] Two principal etiologies for VEPTR-related brachial plexus injury have been identified. The brachial plexus can be injured by direct trauma or impingement from an implant placed too cephalad and laterally in the uppermost thorax. Campbell *et al* has described the boundaries for safe upper rib cradle placement, suggesting devices should remain medial to the scalene muscles and never cephalad to the second rib.[10] The more common cause for injury, however, is compression of the plexus between upper chest wall and the clavicle or upper humerus at the time of the initial distraction and expansion procedure. Brachial plexus compression in this manner has been experimentally created by Nassr *et al*.[30]

Cephalad displacement of the upper thorax occurs with distraction of the VEPTR device and may be enhanced by adjacent expansion thoracostomy, trapping the brachial plexus between upper thorax and clavicle or shoulder. If the scapula and attached muscular flap are drawn caudally to cover a newly expanded hemithorax, this compression effect is more likely. The brachial plexus palsy may be delayed, as the compression gradually takes effect and postoperative swelling occurs. Understanding and awareness of this problem are the first steps to avoidance. Upper extremity motor and sensory monitoring during the initial procedure may provide early warning[31] and is strongly recommended for initial implantation, revision, or exchange, but controversial for lengthening. In patients in whom extensive displacement of the thorax is anticipated, preliminary clavicular osteotomy, performed in much the same way and for the same reason as for correction of Sprengel deformity, can be performed. The need for excessive caudal displacement of the scapula can be mitigated by creation of a more mobile chest wall flap or even by preliminary implantation of a tissue expander. When VEPTR is used as a pure distraction device without thoracostomy, such as in the minimal incisional technique described by Smith and coworkers,[27,28] brachial plexus injury is less likely, as there is no expansion thoracostomy, and the VEPTR device is placed only very medially.

Chest Wall Problems. Chest wall scarring and rib fusions have been identified after VEPTR but minimally reported.[24] Their broad clinical significance for VEPTR patients is as yet unclear. For the patient with a congenitally stiff, small chest such as that seen with congenital rib fusions, spondylocostal, or spondylothoracic dysplasia, continued expansion of the chest by VEPTR is the primary goal and areas of chest wall stiffness associated with the VEPTR device are not problematic unless they inhibit continued chest growth or VEPTR expansion. Many patients with severe myopathies, thoracogenic scoliosis, or burns also probably fit into this category as the affected chest wall shows little movement. However, if the original deformity was associated with a deformed but mobile chest, such as infantile idiopathic scoliosis, inadvertent stiffening of the chest wall in association with VEPTR may be detrimental to long-term chest wall mechanics. Scarring is seen around VEPTR devices, just as it is around GR constructs, particularly at the site of repeated lengthening. The leather-like scar beneath GR or VEPTR devices is readily apparent to the surgeon at the time of device exchange. Spontaneous, dense refusion of previously separated congenitally fused ribs can occur and is often clinically troublesome and signaled by inability to further lengthen the device. The incidence of these refusions is unknown, but the author has had to reosteotomize recurrent rib fusions in 4 of approximately 45 patients and reported on 2 such recurrent fusions in 31 patients,[24] with prior expansion thoracosto-

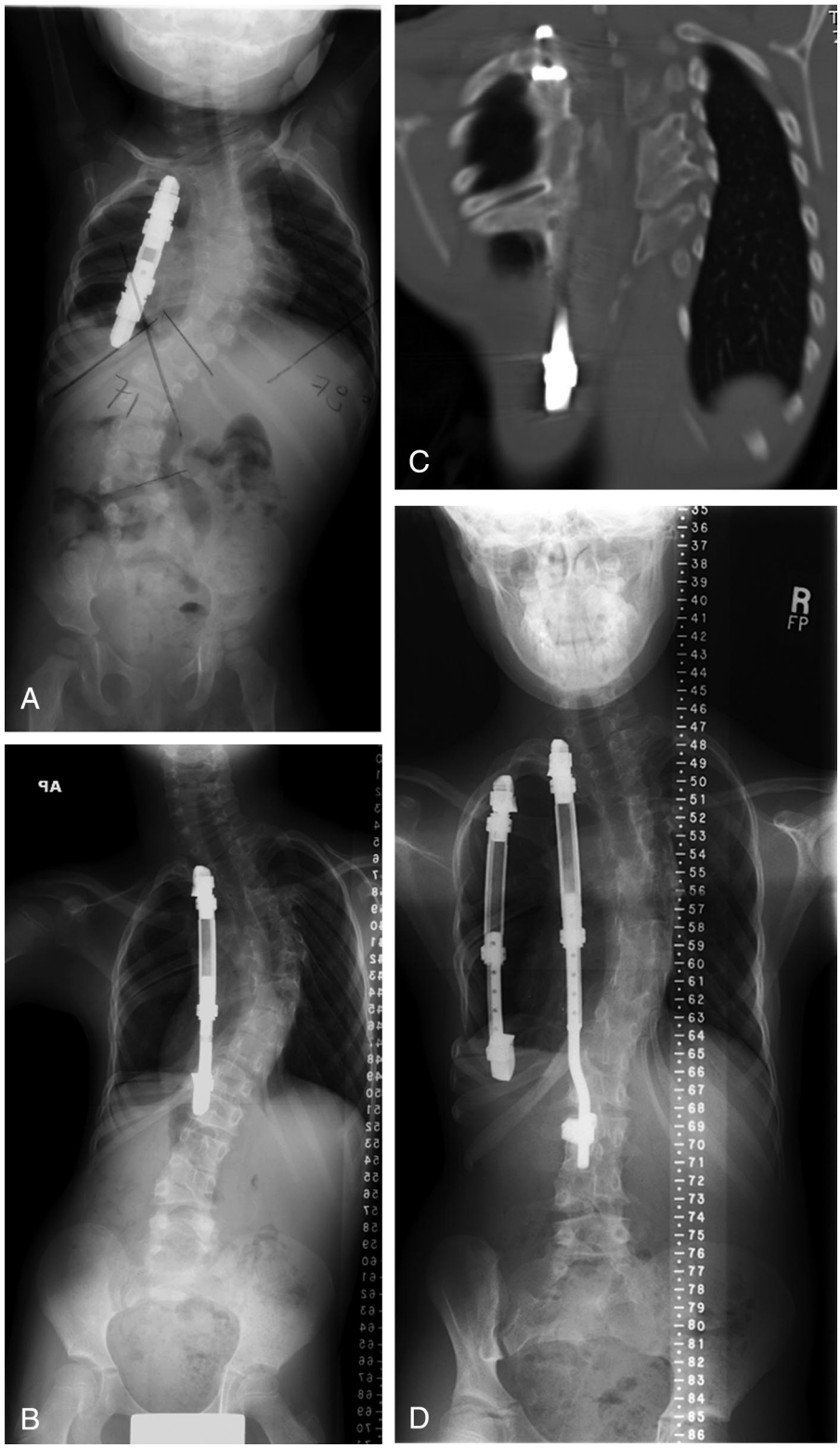


Figure 4. Recurrent rib fusion. This patient had been treated for congenital rib fusions and congenital scoliosis with expansion thoracostomy and insertion of VEPTR devices at age 13 months (**A**). At age 8 (**B**), periodic device lengthening became difficult and CT (**C**) revealed extensive recurrent rib fusions beneath the VEPTR device. Revision with resection of rib fusions and repeat expansion thoracostomy regained control of chest and spine deformity and has continued to withstand repeated lengthening though 4 years (**D**) follow-up. When lengthening of VEPTR devices becomes difficult, CT may reveal underlying rib fusions which may be amenable to repeat expansion thoracostomy.

mies done for congenital rib fusions. The fusions are typically medial in location, and their release with adjacent scar permitted the resumption of device lengthening (Figure 4). If VEPTR lengthening has become increasingly difficult, a search for rib fusion by computed tomography (CT) is appropriate and may reveal rib fusions. Also, spontaneous bridging calcification or bone between normal ribs beneath the VEPTR device has been reported anecdotally, much as one might see spontaneous laminar fusion beneath GR constructs. Bridging bone between ribs can also be seen beneath GR constructs which simply rest directly on the ribs. However, the author is not aware of any of the instances of localized "bridging bone" between normal

ribs underneath the device being of significance or inhibiting lengthening.

Shoulder Problems. Shoulder stiffness as well as spontaneous fusion of the scapula to the VEPTR device and ribs may occasionally occur. The location of upper VEPTR rib cradles beneath the scapula may contribute to shoulder stiffness. The initial surgical incision for expansion thoracostomy and insertion of the VEPTR device as well as repetitive incisions for lengthening probably increase the likelihood of scarring of the scapula or its controlling musculature Attention to surgical technique, early encouragement of range of motion exercises, and placement of lengthening incisions and the distraction lock away from the scapula may make this problem less likely. If scapulothoracic motion is limited, the surgeon should consider adhesions to the underlying thorax as an etiology. A CT might reveal actual bridging bone between scapula and ribs at the location of the VEPTR cradle. At the time of device exchange or as a separate procedure, it is possible to mobilize the scapula from the underlying thorax, freeing scar and adhesions, and excising bony bridges. Postoperative physical therapy in this circumstance is critical. In the author's limited experience, most scapulas will remain mobile after such a procedure.

Wound Problems and Infection. Wound integrity is critical both to the initial VEPTR procedure as well as multiple subsequent lengthening procedures. Wound dehiscence or superficial wound infection may lead to deep infection involving the implant that may end in a difficult problem most commonly managed at best with removal of the implant as a common outcome. Campbell and others[24,25,29,32–34] have recounted their experience with infection in VEPTR patients. Campbell *et al*[29] reported an overall incidence of infection of 1.9% per surgical procedure. Attention to preoperative nutrition and soft tissue handling is suggested as the principal deterrents to perioperative infection. Predisposing factors to wound problems are many, are often coincident with the diagnosis of EOS and a constricted, deformed chest wall, and can only partly be mitigated. Poor health of local soft tissue may be a problem in myelodysplasia, some neuromuscular conditions, or if there have been multiple prior operations in the vicinity. Low weight for age, congenitally absent musculature, and prominent devices all predispose to wound difficulties. Insensate skin or uncooperative patients may also predispose to breakdown by failing to protect the soft tissue overlying the device from trauma. Several steps can be taken proactively to minimize the risk of wound problems and secondarily, infection. Nutrition must be optimal before surgery. Achieving adequate nutrition may require gastrostomy in some children. Flaps should not only be planned to allow access to VEPTR insertion sites and thoracostomy(ies) but also to minimize the surgical scar overlying the device, particularly the most prominent portions of the device. The author prefers to create flaps in which the muscle layer is longer than the overlying skin, making dehiscence less likely. Excessive tension on the wound must be avoided. Prominent devices need to be protected from pressure in the postoperative period, and a donut-like padding is incorporated into the postoperative dressing.

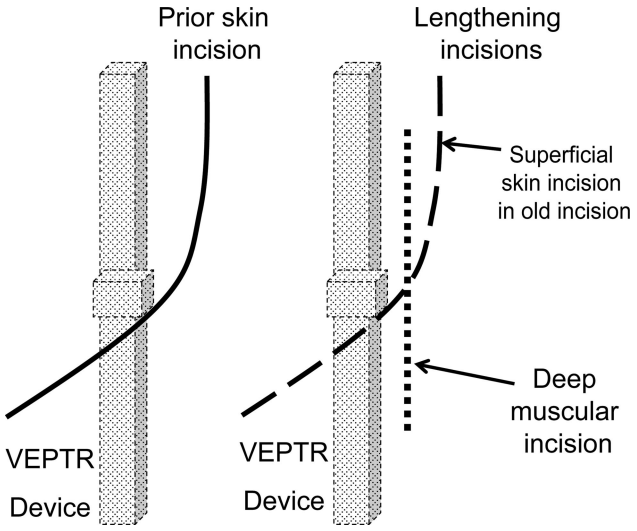


Figure 5. Separate superficial and deep incisions for device lengthening. Repetitive lengthening procedures create local scar and provide recurring opportunities for wound problems including infection or dehiscence. Careful treatment of soft tissues may make these complications less likely. One approach to minimize wound problems with lengthenings is to use separate superficial skin and deep wound incisions, so that if either layer is compromised, the device is still covered.

Each device lengthening represents another chance for a wound problem or infection. For lengthening, we try to avoid full thickness incisions, making separate superficial skin and deep muscular incisions (Figure 5), so that should there be partial wound dehiscence, the device is less likely be exposed. We try to handle the soft tissues carefully and avoid excessive trauma, emphasizing restoration of muscle coverage and obliteration of wound dead space with each closure.

Kyphosis and Sagittal Plane Problems. VEPTR, like most GR systems is distraction-based, relying on repetitive distraction forces to allow growth and maintain spinal alignment between cephalad and caudal anchors. For several reasons, such a system tends to alter the desired normal sagittal alignment. Lumbar lordosis may be inadvertently diminished, particularly if the caudal anchor point is in the lower lumbar spine or pelvis. Although VEPTR devices extending this low ("hybrid" devices with lumbar extensions or pelvic S-hooks) can and should be contoured into lordosis, distraction over time tends to push the lumbar spine into less lordosis. By contrast, the dorsally convex curved shape of the expandable portion of the VEPTR devices encourages the maintenance of thoracic kyphosis. This built-in prokyphotic feature can be effective to a fault over time with repeated distractions. As the curved expandable section of the device elongates, it occupies a larger arc on the

circle defined by the device's radius of curvature, and more kyphosis than desired may be seen. Planned device exchange at the limits of device expansion usually solves this in rib-to-spine devices by reestablishing the original length of expandable section and thereby the original arc and the desired thoracic kyphosis.

Excessive preoperative or postoperative upper thoracic hyperkyphosis is troublesome for both VEPTR and GR. Upper thoracic hyperkyphosis typically occurs in paralytic deformities such as myopathies or arthrogryposis but can also be present in infantile idiopathic and congenital deformities. Distraction-based systems often fail to initially correct or subsequently control upper thoracic kyphosis, yet may successfully treat low thoracic or thoracolumbar kyphosis. Repetitive distraction below or at the apex of an existing upper thoracic kyphosis may significantly worsen the deformity above. Although VEPTR devices can correct some mild upper thoracic kyphosis by cantilever correction at the time of insertion, they are unable to extend above the second rib as a point of purchase and are probably ineffective at controlling severe upper thoracic kyphosis. Although over time the ribs may be kept from deforming further into kyphosis by the VEPTR, the spine may independently deform and continue to collapse into an unacceptable progressive upper thoracic kyphosis, particularly in the setting of poor or absent upper thoracic or cervical musculature. In this author's opinion, severe upper thoracic kyphosis cannot be treated successfully by VEPTR and may be better managed by GR variations, where instrumentation can continue up into the cervical spine, after the severe kyphosis.

■ Discussion

Dual Versus *Single GRs*

Several studies have compared single- and dual-GRs. In the study by Bess *et al,*[14] patients with a single GR were 1.2 times more likely to experience complications compared with those with dual rods. Dual GRs also reduced implant-related complications and unplanned surgeries. Mechanical stress is reduced when 2 rods were used compared with a single rod. This is especially important in fusionless techniques, because the construct incurs continued loading and micromotion that makes the implants susceptible to fatigue and mechanical failure. Dual rods likely dissipate the amount of stress seen by the construct compared to single rods. Additionally, if 1 rod fails, the intact rod is likely to maintain stability and delay construct revision until the next subsequent planned lengthening procedure. Unplanned surgery has reduced the greatest among patients with submuscular dual GRs and has increased the greatest among patients with subcutaneous single rod.

Subcutaneous Versus *Submuscular Placement*

The rationale behind subcutaneous, rather than submuscular (subfascial) placement of GRs, was to reduce the risk for unwanted spine fusion, by minimizing subperiosteal exposure of the spine, thereby reducing the incidence of spontaneous fusion. However, Bess *et al*[14] reported more total complications and more wound complications after subcutaneous GR placement compared with submuscular placement. Patients with subcutaneous rods were 1.8 times more likely to experience complications compared to those with submuscular rod placement.

Submuscular placement of the GRs reduced wound complications because the implants had superior soft tissue coverage compared with subcutaneous GRs. The mechanical benefits provided by a dual-rod construct were negated by soft tissue problems when they were placed subcutaneously. Patients who had subcutaneous dual rods demonstrated more wound complications, more prominent implants, and more implant-related unplanned procedures compared with submuscular dual GRs, and demonstrated the greatest wound complication than all other treatment groups (11 of 37 patients or 31%). Unplanned surgeries were reduced the greatest among patients with submuscular dual GRs (planned to unplanned surgery ratio = 20:1). It appears that children treated with submuscular dual rods benefited from stable constructs that had adequate soft tissue coverage. Conversely, patients treated with single subcutaneous GRs demonstrated the worst planned to unplanned surgery ratio (7.4:1). These patients were at greatest risk for a complication, either due to tenuous construct stability or poor soft tissue coverage.

In an attempt to reduce the number of surgical procedures during the treatment period and therefore reducing the complications, in posterior nonfusion distraction-based techniques, the technology for external, remotely controlled GRs has been reported and is being investigated.[35,36] Growth-guided treatment methods and the remote lengthening techniques theoretically allow for even less invasive treatment than that was available using traditional GR procedure. These techniques are promising; however, there are no long-term outcomes available on their use.

GR Versus *VEPTR*

How should the surgeon decide between VEPTR and dual submuscular GR, which represent the currently most popular means of attempting to control EOS while allowing growth of the spine and chest. Each technique has its "ideal indications," and decision-making for GR and VEPTR is intertwined as the indications for each overlap in some areas (Table 1). Perhaps the best indication for GR is a progressive deformity in a normally segmented deformity such as infantile idiopathic scoliosis not controlled by bracing or casting. In this example, bone quality and soft tissue coverage should be optimal for GR, and chest deformity will likely improve with control of the spinal deformity. By contrast, the "ideal" indication for VEPTR (and expansion thoracostomy) is thoracogenic scoliosis or multiple fused ribs in association with congenital scoliosis. In this example, rib and

spine bone quality will be satisfactory and unless there are major congenital muscle deficiencies or prior operative scars, there will be sufficient soft tissue coverage for the VEPTR implant and expansion thoracostomy. Complications will be minimal in each of these "ideal" applications. However, there are many other diverse etiologies for EOS such as congenital scoliosis without rib fusion, cerebral palsy, myopathies, arthrogryposis, osteogenesis imperfecta, bone dysplasias, Ehlers-Danlos, neurofibromatosis, and myelodysplasia. Each individual patient may present at a different stage of growth, with a differing severity and direction of deformity. How is the surgeon to choose the appropriate use of VEPTR or GR in the individual patient? Choice of GRs *versus* VEPTR should be based on the origin of the deformity, preference, and experience of the surgeon, and associated problems such as soft tissue coverage and upper thoracic kyphosis. Both dual GRs and VEPTR are associated with a high rate of complications, but complications will perhaps be the least if the correct application is chosen. VEPTR with expansion thoracostomy is clearly preferred if the chest deformity is "primary" such as in thoracogenic scoliosis or multiple fused ribs. VEPTR with inferior pelvic attachments also might offer a unique advantage by not being spine-based at both ends, making it possible to control some myelodysplasia deformity without instrumenting the spine at all. Previous GR or VEPTR associated infections may also be a relative indication for the use of alternative technique. Spinal stenosis, absent posterior spinal elements, or other spine abnormalities might make use of a rib-based technique safer or easier than spine-based GRs. Contraindications[10] to VEPTR include the absence of adequate ribs to support distraction, inadequate soft tissue coverage, and an easier or better way to accomplish control of EOS. The authors believe excessive upper thoracic kyphosis is a relatively contraindication to VEPTR and is better handled with GR. Unlike VEPTR, GR may be continued into the uppermost thoracic or lower cervical spine for control of upper thoracic kyphotic deformity. EOS managed with VEPTR may have some advantage over that managed with GR at the end of treatment at the time of definitive fusion. Instead of encountering the distorted, partially fused and scarred posterior spinal elements typical after GR, following VEPTR the spine has not been in contact with spinal rods and is unscarred and unfused, making final definitive instrumentation and fusion easier.

Expansion Thoracostomy or Just VEPTR?

Campbell *et al's* original description of VEPTR[10] assumed an associated expansion thoracostomy as part of the surgical procedure. Clearly, a thoracostomy is needed in instances of thoracogenic scoliosis and fused ribs or where extensive correction of chest shape is desired. Some of the described complications of VEPTR, however, relate to the extensive approach associated with expansion thoracostomy and implant insertion. Smith and others have popularized a minimal incision technique with the use of VEPTR,[27,28] much as one would use GR, except with upper anchor points on ribs rather than the spine for neuromuscular or other collapsing scoliosis or myelodysplasia. Although experience is limited, this technique appears easier for flexible deformity than the traditional combination of VEPTR with expansion thoracostomy and offers an option for the treatment of myelodysplasia, neuromuscular, and other troublesome curves during growth, while making the final fusion easier than if GRs had been used.

■ Conclusion

The goals of EOS treatment can be broadly stated and will help the surgeon in decision-making. EOS treatment should seek to achieve by the end of growth: maximum spine length, maximum pulmonary function, as much residual spine motion as feasible, yet minimize operations, hospitalizations, overall complications, and family burden.

Balancing Risk and Benefit of Growth Sparing Treatment

Complications are a prominent feature of the treatment of EOS by growth-sparing techniques. The surgeon and family may be faced with a difficult choice between growth-sparing surgical intervention and continued nonoperative management of the deformity. In each case, a definitive fusion will likely be needed near maturity. Consideration of the options must include a discussion of complications as they relate to GR or VEPTR. Families need to understand that there will be unexpected events such as rod breakage or loss of anchor points or need for revision or exchange, and these complications might cause the premature cessation of treatment of EOS. Treating surgeons must find a balance for the number of procedures during the treatment period. On one hand, the surgeon must perform lengthenings in a timely basis to allow spinal growth and thoracic development; on the other hand, an excessive number of lengthening procedures may lead to an increased number of complications. This is especially applicable as growth velocity decreases and duration between lengthening can be extended. In a report by Sankar *et al,*[37] length achieved progressively diminished with subsequent lengthenings. Previous recommendations by the authors have advocated lengthening the GR construct every 6 months to facilitate growth and prevent spontaneous spinal fusion; however, future research is needed to determine the optimal interval for individual construct lengthening.

When to Start?

Longer experience with growth sparing treatment of EOS suggests that many patients will suffer treatment-halting complications. Infection, adverse patient reaction, extensive rib refusion in VEPTR, or spontaneous posterior fusion in GR patients might force lengthening to stop well before the planned final fusion near maturity. Campbell and Hell-Vocke have shown greater spine growth and pulmonary function in patients whose treat-

ment began earlier,[23] yet the report of Sankar *et al*[37] suggests less and less length is achieved with subsequent lengthenings. Spontaneous posterior fusions beneath GRs or VEPTR devices are presumed more common when procedures start earlier in life. The surgeon is thus faced with a dilemma: early operation may yield the best chance for lung growth and curve control, but also more operations, greater risk of complications, and a risk that if lengthening is halted because of infection or spontaneous fusion, cessation of growth-sparing treatment will occur at an early age, while large amounts of growth still remain. Although often there may be no choice as to when to intervene, if a deformity is worsening slowly it may be preferable to wait before starting the cycle of initial growth-sparing surgery followed by repeated lengthening. Two factors other than curve magnitude may help in decision-making: thoracic kyphosis and chest wall deformity. Because growth sparing treatments are problematic for the treatment of kyphosis, particularly upper thoracic kyphosis, treatment should not be delayed if kyphosis is worsening or severe. As no growth-sparing treatment is entirely successful in reversing chest wall deformity, the severity and evolution of chest wall deformity should be considered in the decision as to when to initiate treatment. Treatment should start before the chest wall deformity becomes so severe that reasonable thoracic shape and function can no longer be anticipated at the end of treatment.

Minimizing Complications

In this article, we have tried to set forth the common complications associated with the management of EOS by growth-sparing procedures. Although not always achievable, complications should be minimized by selecting the optimal surgical technique for the individual disease and deformity. Preoperative patient nutrition is critical for soft tissue health. Careful soft tissue handling techniques minimize complications in both VEPTR and GR surgery. Submuscular rod placement and careful creation of anchor bases will minimize complications related to the implant. In all instances, doing the initial surgery correctly is advantageous, as revisions are less successful. Multiple lengthenings will be needed, and lengthening operations should be done with the same care and respect for tissues as the initial procedure.

Early recognition of complications, particularly infection, may mitigate their effect. Experience with both GR and VEPTR suggests that early aggressive treatment of implant-related infection can often allow retention of the implants and subsequent continued lengthening.[14,24,25]

■ Key Points

- Growth-sparing surgical techniques such as GR and VEPTR are commonly used to manage spine and chest deformity and lessen the severity of TIS in EOS.
- Surgical decision-making includes both when to intervene in the evolution of EOS, as well as the choice of growth-sparing surgical technique.
- Complications are common when GR or VEPTR are used in EOS, and should be anticipated in planning and discussion with families.
- Some complications for growth-sparing surgery such as spontaneous spinal fusion beneath GRs, rib fusions under VEPTR, and severe infection may result in premature cessation of treatment well before the end of growth.
- Complications are likely lessened by maximizing patient health and nutrition, as well as attention to surgical planning and choice of correct surgical technique.

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