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Management of Severe Spinal Deformity

Scoliosis and Kyphosis

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Study Design. Review of the literature and author's experience with the treatment of severe spinal deformity.

Objective. To define the anatomic and physiologic challenges in treating severe spinal deformity and to describe the preoperative, intraoperative, and postoperative strategies to achieve the optimal safe result.

Summary of Background Data. Severe pediatric spinal deformity is a relatively uncommon condition that often arises following treatment of early onset scoliosis. Patients most often present with severe clinical and radiographic deformity with poor pulmonary function. In contrast to the more common adolescent idiopathic scoliosis which is a primary spinal deformity, patients with severe spine deformity have the added chest wall deformity which may need to be addressed at the time of treatment. Previous literature has identified the challenges in the treatment of these patients and the higher risk for complications.

Methods. A literature review and review of the author's personal experience in the treatment of these patients was performed. An assessment of the preoperative, intraoperative, and postoperative factors leading to an optimal result was analyzed and reported.

Results. The early evaluation should include a multidisciplinary approach from the orthopaedic surgeon, pulmonologist, anesthesiologist, and perhaps the neurologist to provide a baseline assessment. Advanced imaging of the spine with computed tomography is useful especially when previous surgery has been performed and/or when plain radiography is limited. Magnetic resonance imaging of the spinal cord and brain stem is important to ensure that no neural axis abnormalities are present and can determine if spinal cord compression is present. Severe spinal deformity should be distinguished from the more common adolescent idiopathic scoliosis deformity in that both the spine and the chest wall are affected. Preoperative halo-gravity traction is an invaluable tool to improve the flexibility of the spine and chest, to improve pulmonary function, and to stress the spinal cord while the patient is awake and provides feedback as to the neurologic assessment. Surgical treatment should be divided into 3 phases. First, anchor placement which should be predominantly pedicle screws placed in a segmental fashion and also use of reduction screws when performing vertebral column resections. Second, steps should be performed to increase the flexibility of the spine and chest with incremental releases from simple posterior soft tissue releases to posterior facet resections, to vertebral column resections for the most severe deformity. The third phase is the correction of the spine and chest wall deformity. Many strategies can be used to correct these deformities and relies on good anchor point fixation and good releases of the spine and chest wall. Provisional rod fixation is critical when performing resection of the spine to allow for safe correction of the deformity. Improvements in the clinical and radiographic appearance, pulmonary function, and self image are often dramatic.

Conclusion. The treatment of severe spinal deformity is challenging and requires careful assessment of the patient by the orthopaedic surgeon, anesthesiologist, pulmonologist, and neurologist especially when neurologic deficits are present. Proper planning and execution of the correct surgical procedure for the surgeon provides an outstanding life-changing result in these patients.

Key words: severe spine deformity, vertebral column resection, prone thoracoscopic release, halo-gravity traction. **Spine 2010;35:2186–2192**

Pediatric spinal deformity can be classified into scoliosis, kyphosis, or the combination kyphoscoliosis. These spinal conditions can be further classified into mild, moderate, or severe with respect to both the radiographic and the clinical deformity. This article focuses on those patients who have the most severe deformities which require extensive surgical management. The severe spinal deformity should be distinguished from more mild-or-moderate spinal deformity requiring surgical treatment because the preoperative assessment, evaluation, and preoperative management for these curves can be substantially different from the more common moderate adolescent idiopathic scoliosis (AIS) curves. Preoperative assessment by the orthopaedic surgeon, as well as the pulmonologist, anesthesiologist, and even neurologist is worthwhile to obtain full perspective for each of these patients and their specific medical challenges. Advanced imaging is always necessary for these patients to fully understand their spinal anatomy, to rule out neural axis abnormalities, and/or cord impingement. The treatment for these curves is often a combination of nonoperative and operative management. Presurgical treatment methods especially include the use of halo-gravity traction (HGT) which is well tolerated by the patient and the family. The surgical strategy for treatment generally falls into 3 stages: anchor point placement, spinal mobilization through a variety of techniques, and finally, correction strategies.

The management of severe curves can potentially carry significant risk, especially with respect to neurologic deficits, significant blood loss, and pulmonary issues. However, the benefit of this surgical treatment is

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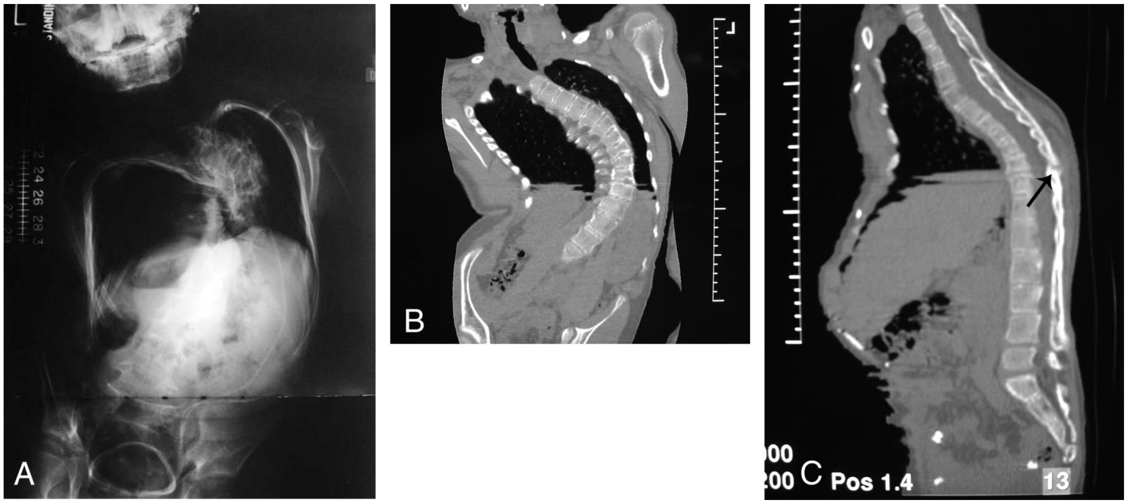

Figure 1. Utility of the CT scan. **A**, PA radiograph an 11 year-old boy who had multiple previous surgical treatments for spinal deformity. His radiographs are very difficult to visualize to understand his anatomy. **B**, The multiplanar reconstruction CT image in the coronal plane with greater visualization of the anatomy demonstrating multiple sites of nonunion from the previously-attempted anterior fusion **C**, The lateral multiplanar reconstruction CT image demonstrating a posterior pseudoarthrosis (arrow).

potentially far reaching from a cosmetic, pulmonary, and general health viewpoint which may prolong many of these patient's lives.

Comparison Between the Common AIS Curve and "Severe Spinal Deformity"

AIS is the most common type of scoliosis we see with a prevalence of 1/1000 patients. This is more common in females than males, especially with larger curves. Generally, AIS curves present with relatively mild-to-moderate curves. The common surgical preoperative curve magnitude threshold to perform surgery is usually 50° to 55°. The surgical strategy for these curves includes anchor point placement followed by correction strategies with distraction, translation, cantilever, rotation, and direct vertebral rotation maneuvers. Generally, deformity correction of 60% to 70% with modern segmental spinal instrumentation can be achieved safely. The reported neurologic deficits with this type of surgery is <1%.

In contrast, severe spinal deformity is relatively rare and a distinction from AIS should be made. First, severe deformity is not only in the spine but also in the chest wall. Long standing severe spinal deformity results in chest wall stiffness with decreased compliance of chest wall mechanics resulting in poor pulmonary function (Figure 1) (Supplemental Digital Content 1, Figures, online only, available at: http://links.lww.com/BRS/A485). The surgical treatment for severe deformity is a very large operation which carries significant high neurologic risk and is, in part, dependent on the type of surgical procedure performed. In addition to the anchor point placement and corrective forces applied to the spine, severe deformity requires an intervening step to "loosen" up the spine and chest, using a variety of techniques. These include anterior release, rib osteotomy, rib head resection, pedicle subtraction osteotomy, and finally the most advanced and aggressive surgical release—a vertebral column resection (VCR). Anecdotally, the benefit of these procedures is exceptionally high with a recognizable improvement by the patient in their clinical appearance.

Comparison Between Kyphosis and Scoliosis

Similar to scoliosis, kyphosis deformity comes in a wide range of severity from the moderate Scheuermann kyphosis to the more severe congenital kyphosis. Distinguishing these types of deformities is important for the preoperative evaluation which may not include advanced imaging in the patient with Scheuermann kyphosis, however, is mandatory for the patient with congenital kyphosis to include computed tomography (CT) and magnetic resonance imaging (MRI). The intraoperative treatment of these curves is also far different since most patients with Scheuermann kyphosis can be readily treated with a posterior approach that includes Ponte-type wide releases,[1] while the patient with severe kyphosis due to a butterfly vertebra, for example, may require an anterior-posterior approach or an all-posterior approach with a pedicle subtraction osteotomy or VCR. The risk for neurologic deficits is also significantly higher in the patient with a congenital kyphosis. A comparison between patient with a primary kyphotic deformity and one with a primary scoliotic deformity reveals some significant differences. Kyphosis of the spine always adds significant neurologic risk since the spinal cord can be draped over the apex of the deformity and there is some disturbance of spinal cord perfusion from the anterior vertebral artery.[2,3] However, the correction strategies to treat primary kyphosis are fairly straightforward because it is usually single plane sagittal deformity.

Pertinent Issues for Severe Deformity

The risks for treatment of severe spinal deformity are similar, but more common than in AIS. These include neurologic deficit, pulmonary issues, and blood loss intraoperatively. The classic articles on neurologic deficits following AIS surgery are from the Scoliosis Research Society database morbidity and mortality statistics. MacEwen *et al* in 1975 reported an incidence of neurologic deficit following AIS surgery of 0.72%, with 50% of those patients having complete paraplegia.[4] More recently, Coe *et al* used similar methodology in reviewing the morbidity and mortality statistics from the Scoliosis Research Society and reported a 0.54% overall incidence following surgical treatment for AIS.[5]

In contrast to AIS surgery, the incidence of neurologic deficit following VCR for severe spine deformity is much higher. Suk *et al* reported his experience with 16 patients with severe scoliosis who underwent a posterior vertebral column resection with a significant improvement in the radiographic deformity from 109° to 45°. However, he reported 1 patient (6.3%) who had a permanent complete paralysis following this surgery. Bradford and Tribusreported their experience in 24 patients who had rigid coronal decompensation who underwent both an anterior and posterior VCR with 4 of 24 (16.6%) having transient neurologic complications which improved with time so that there were no permanent deficits. The most recent publication by Lenke *et al* reported their experience of 35 consecutive patients, reporting 8.5% of patients who lost intraoperative neuromonitoring data during correction. However, no patient (0%) had permanent spinal cord-related complications with have 2 patients (5.7%) who had transient nerve root palsies.

Preoperative Evaluation for Severe Spine Deformity Surgery

The preoperative assessment for these patients with severe deformity requires a multidisciplinary approach. The patients who have severe deformity often have low body mass index secondary to significant energy required for breathing and activities of daily living. The assessment by a developmental pediatrician, as well as a dietician is important since patients often require nutritional supplements and may require feeding tubes to increase their nutritional status. The prealbumin, albumin, and protein levels should be normalized and the total lymphocyte count should be greater than 1500 before surgery to ensure healing.

Assessment by a pulmonologist is critical for these patients to fully understand their pulmonary function. They often will need a formal sleep study to assess for sleep apnea, as well as evaluation for needing bilevel positive airway pressure. The bilevel positive airway pressure fitting is often used both before and after surgery in these patients.

It is critical to have the involvement of the anesthesiologist before surgery to evaluate the patient's airway, since occasionally the severe deformity of the spine influences the trajectory and path of the trachea, and may make intubation and maintenance of the patient's airway difficult. In this setting, a tracheostomy may be necessary to ensure a safe airway, and to maintain ventilation intraoperatively, as well as after surgery.

Preoperative Imaging for Severe Spine Deformity

The imaging of patients who have severe deformity includes a standard posterior-anterior (PA) and lateral standing radiographs. Additional plain radiography should include traction films to assess the flexibility of the deformity. An anterior posterior and lateral radiographs of the cervical spine, especially for those patients who have syndromic conditions, or those who are going to be placed into preoperative traction is important. Flexion–extension lateral views to identify any instability should also be obtained in the syndromic patient.

Advanced imaging is mandatory for all patients with severe deformity. For those patients who have previous surgical treatment, or if a severe deformity is present and cannot be fully visualized with plain radiography, a CT should be obtained of the entire area of spine involvement of the deformity. The CT scan allows one to understand the pedicle anatomy including the width, depth, and trajectory of the pedicles. It allows one to identify pseudarthroses when previous fusion has been performed. Multiplanar reconstruction of the axial images is important, especially when trying to understand the very complex anatomy in these patients and will allow one to visualize those levels of planned osteotomies or resections (Supplemental Digital Content 1, Figures, online only, available at: http://links.lww.com/BRS/A485). Finally, the CT scan can be used to make a model of the spine for those patients who have very severe deformity (Supplemental Digital Content 1, Figures, online only, available at: http://links.lww.com/BRS/A485). The model is useful to evaluate the patients before surgery, to fully understand the anatomy before any surgical treatment, and can be used intraoperatively to visualize the spine including the pedicles.

MRI is essential for all patients who have large curves, especially for those patients who are undergoing large operative correction, to assess the neural axis. It is important to understand whether syrinx or Chiari malformations are present, as well as any stenosis or cord impingement secondary to severe deformity (Supplemental Digital Content 1, Figures, online only, available at: http://links.lww.com/BRS/A485). If spinal stenosis is identified then a CT myelogram is important to understand the specific levels of compression and whether dye will pass to understand whether formal decompression of those levels is warranted. It should be remembered that in kyphoscoliosis the level of cord compression is always on the concave pedicle and may require a resection of that pedicle with the remaining body.

The future of imaging, especially when a VCR is being planned, may be in understanding the blood flow to the spinal cord over the planned level of resection. Selective angiography has not been used to my knowledge in this setting; however, it will most likely have some role in the future. In addition, MRI, contrast-enhanced gradient echo, and 3-dimensional pulse sequence may have a role in these patients in the future.

Preoperative treatment of spinal deformity should be considered for those patients with severe deformity of both the spine and chest with the primary mode of treatment being traction. Traction has a long history in the treatment of spinal deformity. It initially started with the use of halo femoral traction, then going to halo pelvic ring traction, and more recently HGT. HGT can be applied using pulleys and weights. However, the safest and greatest use of HGT uses a spring-loaded device (Supplemental Digital Content 1, Figures, online only, available at: http://links.lww.com/BRS/A485). This has a built in safety neurologic feature so that when impending neurologic problems occur, the patient simply lifts up and decreases the amount of traction across the spine.

The benefits of traction have been previously documented and play a large role in the treatment of many patients who have very severe deformity.[6–8] In addition to increasing the flexibility of the spine, it also increases the flexibility of the chest and anterior structures. It has been documented to improve pulmonary function which is important in these patients who have critically low pulmonary function. In addition, it stresses the spinal cord in a patient who is awake and can verbalize any impending neurologic deficits. Some of the subjective benefits include the fact that one sees these patients each and every day and allows one to think about the patient's spinal deformity and it allows one to see the family each day and to speak to them and to continue the ongoing relationship before a very large surgery which may carry significant risk. In addition, the traction may change the surgical plan. It is important to realize that most severe spine deformities will demonstrate significant improvement radiographically and clinically following traction (Supplemental Digital Content 1, Figures, online only, available at: http://links.lww.com/BRS/A485). The indications for traction include those patients who have severe pulmonary dysfunction, severe spine deformity, especially when previous surgery has been performed, and severe chest wall deformities and constriction. It is generally contraindicated with congenital kyphosis,[6] with previous spinal cord tumor resections and residual neurologic deficits, and when cord tethering is present. Intraoperative temporary traction is another modality to use the distractive forces to safely gain correction.[9]

Surgical Planning

Since the incidence of neurologic deficit can be high in these patients, several methods can be used to limit these complications. The first is to decide on the appropriate procedure on the basis of the severity of the deformity, the goals of the patient, and family with respect to deformity correction and their tolerance for risk, and just as importantly, the surgeon's experience performing these procedures. Certainly, the goal should be to choose the procedure with the lowest incidence risk for complications while achieving adequate deformity correction. The author's contraindications for a VCR are as follows: suboptimal or no baseline spinal cord monitoring, when less risky surgeries can achieve the same goals, and for those patients whose spine cannot be instrumented with segmental pedicle screw fixation.

It is important to have other specialty services for the patients including a neurologist to perform a thorough neurologic examination, anesthesiologists to evaluate them from a medical standpoint and to assess their airway, and a pulmonologist especially when previous pulmonary issues have been present.

Preoperative planning includes identifying the type of implants that will be available at the time of surgery. The surgical treatment of these deformities requires a variety of anchor types to include pedicle screws, hooks, and wires as well as having a variety of rods with various stiffness and strength characteristics. The author prefers titanium implants in the event a postoperative MRI is necessary with greater ability to visualize the spinal cord, and the rod stiffness and strength characteristics allow one to correct the spine safely without undue stress on the bone-screw interface.

Intraoperative neuromonitoring is critical for the surgical treatment of severe deformity and includes multimodal monitoring using somatosensory evoked potentials (SSEPs) and motor evoked potentials. Although transcranial motor evoked potentials (TcMEP) is an accurate and physiologic method to monitor the motor tracts of the spinal cord, neurogenic motor evoked potentials (NMEP) continues to be very useful in spinal deformity surgery at some institutions.[10,11] For severe deformity, the author uses somatosensory evoked potential, TcMEP, as well as NMEP. The dual modality MEP strategy has been used by the author especially during VCR cases relying on the various levels of sensitivity of the 2 methods. Since the TcMEP are more sensitive than the NMEP, the amplitude of the TcMEP generally begins to decline first with things such as retraction of the spinal cord during the resection, or during the initial closure of an osteotomy. This generally occurs without a decline in the NMEP data and allows one to then move to the contralateral side and continue the resection or to begin to look for tension across the spinal cord or translation of the spine with cord compression, *etc.* during the reduction of the spine. Most commonly, the NMEP data will remain stable during the initial drop in the TcMEP and provides some confidence to the surgeon that clinical function of the cord is intact. The author has found that TcMEP data commonly diminishes during a VCR, and is often associated with tension or compression on the spinal cord which, when addressed, results in a normal neurologic examination at the completion of the surgery especially when the NMEP data are at the baseline.

Surgical Treatment

Surgery for correction of severe deformity occur in 3 stages. First is anchor placement, second spine mobilization, and finally, correction strategy. All 3 phases are critical to a successful surgical treatment.

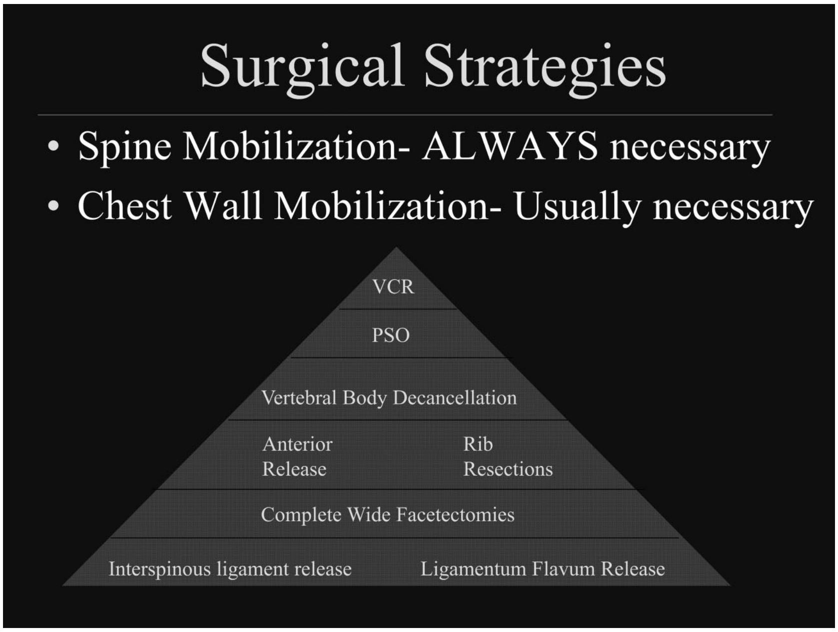

Figure 2. Pyramid of increasing degrees of "spine and chest mobilization" strategies, which can be used when performing operative correction of severe spinal deformity.

Anchor placement requires segmental fixation for these deformities. The preferred fixation method is pedicle screws since they provide 3 column fixation and are versatile in obtaining and maintaining correction of the spine and providing a stable fixation point. The availability of long tulip head screws (reduction screws) is critical, especially when treating severe deformity and when performing VCRs. Typically, these are used distal to the apex of the deformity and correction strategies allow the rod to be reduced to these reduction screws sequentially. Alternative fixation methods include hooks and wires when pedicle screws are impossible to place secondary to small pedicles, when pedicle fixation is tenuous, or when loss of fixation occurs with the pedicle screws.

Mobilization of the spine is always necessary in severe deformity. In addition, it is critical to evaluate whether these long-standing spinal deformities with previous surgery require chest wall mobilization. If this is necessary then often the more aggressive VCR is needed. The authors use a pyramid of spine mobilization strategies to sequentially consider greater mobilization of the spine (Figure 2). The standard baseline release should always be performed for these deformities and includes soft tissue release of the interspinous ligament release and ligamentum flavum as well as bony resection with wide facetectomies at all levels at the apex of the deformity.

The indications for performing an anterior release and rib resection continue to deminish with greater use of pedicle screws in both the thoracic and lumbar spines.[12,13] However, the author starts to consider anterior release *via* a prone thoracoscopic approach in the thoracic spine and an open release in the thoracolumbar–lumbar spine for those curves which are >85° in which the flexibility index is <40% to 50%.[14,15] An additional indication is for a skeletally immature patient with open triradiate cartilage and before peak height velocity. In doing an anterior release, a complete disc excision and rib head resection is appropriate for these patients who have severe deformity. Pulmonary function is not detrimentally affected with anterior release, when performed in the prone position with double lung ventilation.[15] The combination of severe deformity and skeletal immaturity is an acceptable indication to perform an anterior release and fusion in combination with a posterior fusion and instrumentation (Figure 3). The next level for release is vertebral body decancellation. This allows one to gain correction on the convexity of the curve especially when there is wedged vertebra and decancellation of the bone allows one to close down these levels.

The most aggressive and often used surgical release strategy is a VCR which has been described for severe deformity. This can be performed using a combined anterior and posterior approach[16,17] or more recently *via* an all-posterior approach.[18,19]

The VCR technique requires stable fixation proximal and distal to the levels of resection. The level of resection is determined by the deformity and is performed at its apex. After clean muscle stripping of the spine, anchor placement is performed. Rib resections are necessary over the levels of resection. Concave rib resections are performed over those levels of resection as well as proximal and distal to the apex to allow for visualization of the resected vertebral levels and serves to improve the flexibility of the spine and chest. On the convexity of the deformity, less rib resection is necessary since visualization of the spine is easier and a greater space is available between ribs. The author performs a posterior laminectomy at this point to fully expose the spinal cord and to identify the nerve roots of the vertebral levels to be resected. The nerve roots of the resected levels are then clamped and tied off with silk suture trying to stay medial

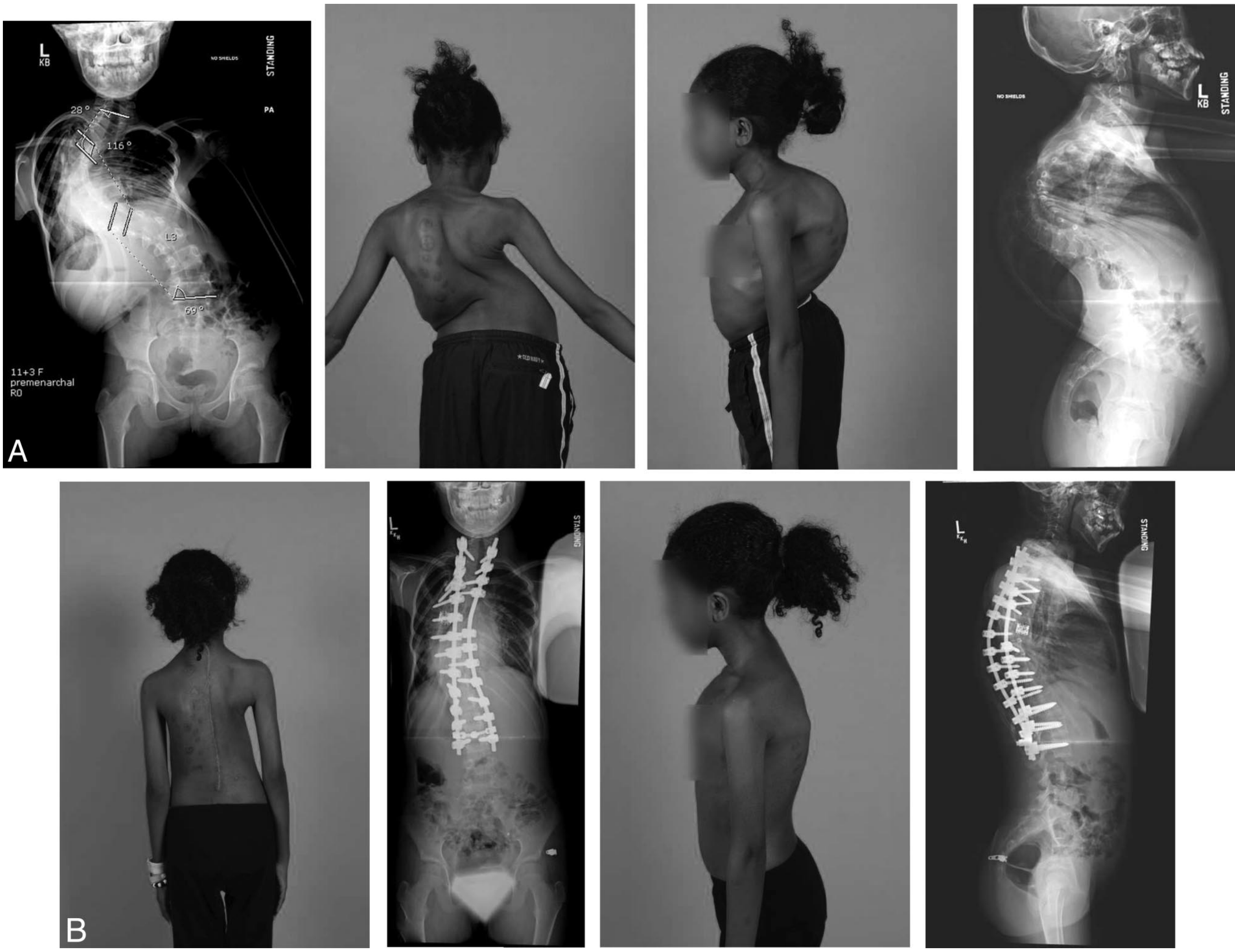

Figure 3. Posterior vertebral column resection. **A,** PA and lateral radiograph of a 14 year-old girl who has thoracogenic scoliosis. Her clinical pictures demonstrate her severe deformity. **B,** Postoperative radiographs following a posterior vertebral column resection over two levels and posterior instrumentation and fusion from T1–L3. The clinical appearance demonstrates marked improvement in her trunk height, coronal balance and overall appearance of her spine and chest.

to the dorsal root ganglion. The author has not found ligation of these nerve roots to effect the chest wall sensation or development of neuromas.

The order of resection is important to allow one to have stable provisional fixation at all times. The concave resection is first performed and is the most difficult because the pedicles on this side are very small and are most commonly all cortical bone. The spinal cord is always draped over the concave pedicles and care is taken to ensure the spinal cord is protected. Following completion of the concave resection, a provision rod is then placed on the concave side and the convex side. The remaining resection can then be performed with both the concave and the convex rod in place since rotational deformity of the spine will allow one to work under the convex rod to remove the convex vertebral body.

The correction strategies following resection of the bone should always include maintenance of fixation to the spine to prevent translation, distraction, or other stresses to the spinal cord. The authors prefer to seat the rod proximally, and provisionally engage the set screws to the tip of the tulip head reduction screws. With the rod still provisionally fixed in the top portion of the tulip head screws, manual compression of the 2 spine segments across the apex is performed. This allows one to maintain control of both segments of the spine while being free to close down the gap created by the resection. Anterior structural support in the form of a cage is placed to prevent translation and excessive shortening of the anterior column. Gentle pressure on the distal aspect of the rods allows them to disengage from the set screw on the reduction screws and providing in-line stability since the rod is still in the channel of the screw and allows the surgeon to manually compress the spine. Reduction of the rod to the remaining anchor points is then performed so that further compression can be performed using the manual compressors spanning across the pedicle screws.

The surgical results of treating severe spinal deformity include outstanding radiographic results and clinical results as reported by many authors. The overall patient satisfaction with these aggressive large surgeries cannot be overestimated since many of these patients have very severe deformity of the spine and chest and surgical treatment provides them with a truly life-changing event with significant improvement in their pulmonary function, their ability to perform activities of daily living, their sense of self-image, and cosmesis. The significant neurologic risk associated with these surgeries requires meticulous preoperative planning, intraoperative surgical performance with good spinal cord monitoring, and maintenance of postoperative perfusion to the spinal cord to achieve success.

■ Key Points

- Severe spine deformity is less common than adolescent idiopathic and involves spine and the chest wall deformity which both need to be addressed at surgery.
- There is a higher risk for neurologic deficits after surgery for severe spine deformity and preoperative, intraoperative, and postoperative strategies should be used to limit this complication.
- The surgical treatment of severe spinal deformity requires one to increase the flexibility of the spine and chest through a graduated level of release which is dependent on the severity of deformity.
- VCR is the most powerful technique to gain correction for severe deformity and requires careful preoperative planning to fully understand the anatomy, meticulous surgical technique, and requires multimodal intraoperative neuromonitoring.
- The successful outcome after surgical treatment for severe deformity is a life changing event with improvement in thoracic height, pulmonary function, and a profound affect on cosmesis and self image.

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