Congenital scoliosis develops as the result of anomalous vertebral development and growth. The spine, therefore, grows more rapidly on one side, resulting in a progressive deformity with spinal imbalance. The vertebral anomalies are the result of the failure of formation, segmentation, or both of these during the embryonic stage of development. Unilateral failure of formation usually results in an unsegmented or partially segmented unilateral hemivertebra. This situation can be responsible for progressive congenital scoliosis resulting in spinal imbalance. When the hemivertebra is situated near the lumbosacral junction, a particularly difficult and severe trunk shift with spinal imbalance and pelvic obliquity can result.

Typically, progression of the deformity requires surgical treatment. A number of options are available for the treatment of congenital scoliosis, including fusion in situ, posterior fusion with instrumentation, hemiepiphyseodesis, and hemivertebra excision. Lumbosacral hemivertebra producing lateral spinal decompensation has been well documented as an indication for hemivertebra excision. Early intervention appears to yield the best results. Fixation can present a major problem in infants, in small and young patients because they often lack adequate pedicle size and bone stock to safely and effectively accommodate the commonly used fixation devices such as pedicle screws.

We present a technique of fixation to the ilium after excision of a lumbosacral hemivertebra that has been successful in reducing the trunk decompensation and progression of the curve. The technique uses cables, which can compress down the residual space from the excision and can be used regardless of the size of the lumbar vertebra and sacrum. It is particularly suitable for infants and very young children when the lumbar and sacral pedicles are too small and the bone inadequate to easily accommodate the pedicular fixation. Furthermore, it offers efficient biplanar (coronal and sagittal plane) correction with restoration and improvement of lumbar lordosis.

Surgical Technique

Intraoperative neurologic monitoring is standard for all patients. The hemivertebra excision is addressed first from an anterior retroperitoneal approach, followed by the completion of the excision posteriorly with a lumbar exposure of the spine. The posterior iliac crest is exposed on the side of the planned instrumentation. The paraspinal muscles are reflected from their insertion into the sacrum and the adjacent ilium to allow an unobstructed course from the lumbar vertebra to the iliac crest. The fascia is divided and the pelvic apophysis is split either sharply or with electrocautery. The pelvic brim and the outer table of the ilium are exposed subperiosteally with a Cobb elevator with care to avoid penetrating into the gluteal musculature or into the sciatic notch. The posterior column of the ischiium is palpated and identifies the course of the screw. An awl or curette is used to develop the channel as a path for placement of the screw. A ball probe is now used to sound and determine the length of the screw (the diameter depending on the thickness of the


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posterior ilium). We advise using the widest possible screw diameter. A small area of the crest at the point of entry allows countersinking of the screw. The screw is advanced and seated, followed if necessary by placement of the top nut to close the head of the screw. Laminotomies prepare the laminae of the adjacent normal vertebrae cranial to the hemivertebra on the side of excision. Two cables are passed under the lamina of the adjacent and caudal normal vertebra and then inserted through the closed head of the screw. The cables are then tensioned to close down the space created by the excision of the hemivertebra using not more than 30 ft-lbs of force. Manual bending of the spine helps to easily close this gap. A bilateral posterior lateral arthrodesis completes the procedure. Local bone and medial iliac crest is used as bone graft and may be augmented if necessary by cancellous allografts.

The paraspinous muscles and the fascia are closed over the construct with a no. 0 absorbable suture. The remainder of the wound is closed in a standard fashion. The postoperative care includes a 1½ spica cast or brace for 3 months, followed in time by removal of the implant when the fusion is secure. We advise an average waiting period of 12 months before implant removal, although the decision is best taken on a case-to-case basis.

Clinical Examples
Details of patient demographics, hemivertebra levels, curve characteristics, pre- and postoperative parameters, and findings at latest follow up are presented in Table 1.

Table 1. Patient Characteristics, Pre- and Post-Treatment Parameters and Comparison With Two Other Reported Series (Using Casting Technique Alone)

<table>
<thead>
<tr>
<th>Series</th>
<th>Case</th>
<th>Sex</th>
<th>Age at Surgery</th>
<th>Level of Hemivertebra</th>
<th>Magnitude of Curve</th>
<th>Coronal Balance (mm)</th>
<th>Lumbar Lordosis*</th>
<th>Hardware Removal</th>
<th>Latest Follow-Up</th>
</tr>
</thead>
<tbody>
<tr>
<td>Current study</td>
<td>1</td>
<td>F</td>
<td>10 yrs 6 months</td>
<td>S1</td>
<td>33°</td>
<td>7°</td>
<td>16</td>
<td>2 ½ years</td>
<td>5 ½ years: 24° (unchanged for 3 years)</td>
</tr>
<tr>
<td></td>
<td>2</td>
<td>F</td>
<td>2 yrs</td>
<td>L5</td>
<td>30°</td>
<td>0°</td>
<td>25</td>
<td>9</td>
<td>1 ½ years: 0°</td>
</tr>
<tr>
<td></td>
<td>3</td>
<td>F</td>
<td>4 yrs</td>
<td>L5</td>
<td>40°</td>
<td>8°</td>
<td>11</td>
<td>33</td>
<td>3 years: 8°</td>
</tr>
<tr>
<td>Bradford et al61</td>
<td>1</td>
<td>M</td>
<td>10 yrs 1 month</td>
<td>L5</td>
<td>38°</td>
<td>15°</td>
<td>—</td>
<td>—</td>
<td>7 years: 14°</td>
</tr>
<tr>
<td></td>
<td>2</td>
<td>M</td>
<td>4 yrs 1 month</td>
<td>L4</td>
<td>56°</td>
<td>22°</td>
<td>—</td>
<td>—</td>
<td>2 years 3 months: 20°</td>
</tr>
<tr>
<td>Klemme et al68</td>
<td>1</td>
<td>—</td>
<td>1 yr 3 months</td>
<td>L3</td>
<td>35°</td>
<td>10°</td>
<td>—</td>
<td>—</td>
<td>5 years 10 months: 5°</td>
</tr>
</tbody>
</table>

* Lumbar lordosis was measured between the superior endplate of L1 and the superior endplate of S1.

Figure 1. A and B, Hemivertebra excision with internal fixation postsurgery showing well-balanced spine (A) with a curve of 7° (reduced from 33° to 7°). Lateral radiograph (B) demonstrating the fixation and restoration of the lumbar lordosis.
Patient No. 1  A 10½-year-old girl with a sacral hemivertebra at S1, partially fused to the sacrum, developed progressive scoliosis. There was a documented curve progression from 15° to 26° between the ages of 8 and 9 and then progressed to 33° when she presented to us. There was a coronal imbalance of 35 mm (distance measured between the plum line passing through the first thoracic vertebra and the central sacral line). Through a combined anterior and posterior approach, she underwent excision of the hemivertebra and closure of the resulting defect as previously described with a 6.5-mm cancellous screw and a single cable. The patient was not treated with additional immobilization using an orthosis/cast. Her postoperative curve measured 7° after surgery and her spinal balance was improved (Figure 1). The coronal imbalance improved to 10 mm and was considered a clinically acceptable result. She had failure of the cable (in that the cable itself broke) and underwent hardware removal 2 ½ years after surgery. At latest follow up 5½ years after the primary procedure, she appears fused. With prolonged orthotic wear, she went on to fusion although with a 17° loss of correction. Her curve measures at 24° and is unchanged over the last 3 years.

Patient No. 2  A 2-year-old girl had a right L5 fully segmented hemivertebra with trunk shift and coronal imbalance of 25 mm. Her curve measured 30° from L4–L5. There was a documented progression from 16° to 30° as recorded from her previous radiographs at the age of 11 months. After excision of the hemivertebra, the spine was instrumented with a 6.5-mm cancellous screw into the ilium and 2 cables under the lamina of L4. She was placed into a spica cast. At her follow up 1 year after surgery, her curve measured 0°. The coronal imbalance was corrected to less than 5 mm from the central sacral line. Her implants have subsequently been removed and there has been no loss of correction. She continues to maintain correction and has solid fusion at 3-year follow up after surgery.

Patient No. 3  A 4-year-old girl had a left semisegmented hemivertebra at L5. There had been a documented pro-
gression of her curve from 22° to 40° over a period of 18 months before surgery. Before surgery, the curve measured 40° with a coronal imbalance of 25 mm (Figure 2). After the excision, 2 cables were passed beneath the lamina of L4 and secured to a 6.5-mm iliac screw, which closed the defect and corrected the imbalance. The patient was immobilized into a 1½ spica cast. Six weeks after surgery, the curve measured 8° and the coronal imbalance was corrected to 8 mm (Figure 3). Hardware was removed 1 year after surgery. At 3 years follow up, she continues to do well and has a sound fusion with a radiographic curve of 8° (Figure 4). The clinical progress has been outlined in Figure 5.

**Biomechanical Rationale**

Conceptually, screw placement into the ilium demonstrates the biomechanical advantage of this construct (Figure 6). Our biomechanical model illustrates the the-

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**Figure 4.** A and B, Three years postsurgery, the radiograph continues to demonstrate a well-balanced spine with sound fusion.

**Figure 5.** A and B, Clinical photographs of the patient at 6 weeks (A) and 3 years postsurgery (B), demonstrating the well-balanced curve and symmetry.
oretically enhanced corrective forces (Figure 7). When analyzed by vector forces, the construct exerts almost twice the bending force in the coronal plane as compared with a sacral pedicular construct. Hence, it should help improve the lateral curve correction. Furthermore, this construct works at an obtuse angle with respect to the interface point at the cable–lamina junction in the sagittal plane, allowing for improved lumbosacral lordosis (Figure 8). Sagittal plane correction of the curve in all 3 cases leading to restoration and improvement in the lumbar lordosis substantiates this rationale in our series of patients (Table 1). Finally, the bone stock found in the ilium is more abundant to that of the sacral pedicles, which are small in this age group.

Discussion

The natural history of congenital scoliosis has been well documented.1–4 The extent of progression clearly relates to the location of the curve as well as the type of vertebral anomaly. A study by Winter et al. revealed curve progression of $\geq 31^\circ$ in 26 of their 57 patients untreated for $\geq 2$ growth years or more, with thoracic and thoracolumbar curves showing more severe deformity.5 In 1975, Nasca et al. concluded that the major determinants of progression were the location of hemivertebrae and the
presence or absence of a unilateral bar. McMaster and Ohtsuka found that a unilateral unsegmented bar with a contralateral hemivertebra at the same level caused the most severe deformity. McMaster and David studied 104 patients with congenital scoliosis resulting from hemivertebrae and divided them into 3 groups: fully segmented, semisegmented, and nonsegmented. A single, segmented, nonincarcerated hemivertebra was the most common deformity (65%) and demonstrated the greatest potential for growth.

A hemivertebra at the lumbosacral level presents a significant decompression problem in addition to the potential for progression. Standard methods of treatment require fusion of many segments to attain proper curve control. Early treatment is preferred to prevent progression of the spinal imbalance and worsening pelvic obliquity.

The goals of early treatment are avoidance of progression of the deformity and achievement of spinal balance in both coronal and sagittal planes, with the least inhibition of growth. Ideally, early excision of the hemivertebra with arthrodesis of 1 or 2 segments at one time provides for correction and spinal balance without excessive multisegmental fusion.

The lumbosacral hemivertebra is well documented as an indication for hemivertebra excision. Royle first reported removal of hemivertebrae in 1928. Early attempts at correction were discouraging, and patients commonly experienced serious complications. In 1979, Leatherman described a 2-staged procedure and reported an average correction of 47% without neurologic complications. That approach allowed direct visualization of the hemivertebra and complete vertebrectomy. Leatherman and Dickson warned that the 1-stage procedure was dangerous, but modern literature supports the relative safety of the 1-stage approach when performed by a skilled spinal surgeon.

Neurologic complications are a source of great concern in this procedure. Recently, Slabaugh et al. reported on 8 patients undergoing lumbosacral hemivertebrae removal with 3 cases experiencing transient quadriiceps weakness. King et al. observed 3 cases of neurologic problems out of 7 cases with 1 L5 root paresis. On the other hand, in a recent reported series, Deviren et al. noted no neurologic damage in their 10 cases. Lazar et al. reported no long-term neurologic injuries in their series of 11 cases. Careful intraoperative monitoring of the nerve roots at risk may help prevent such complications. The patients in our series demonstrated no neurologic complications or other serious early or midterm complications.

One of the major challenges encountered in the young and small patient with hemivertebra excision is attaining stable fixation. It is also necessary to try to close the space created by removal of the hemivertebra. Fixation is especially difficult with the relatively small and porous bones of infants and young children. As a result, non-standard constructs such as vertebral body screws passed through a plate or coupled with a twisted wire may be used anteriorly. At more proximal levels of the spine, wiring or a small-sized compression system can help attain closure of space. These techniques are less well suited to lumbosacral fixation. The technique described here overcomes most of these problems.

If the child is older and larger, a more standard construct such as a pedicle screw fixation may represent the best option. In the younger child, the pedicles are small, and the potential for penetration through the pedicle is relatively high. The bone stock (particularly sacral) is less plentiful, and the sacral skin may not really provide adequate coverage for sacrum after fixation. Failure of instrumentation may occur when applied to bones that are small or osteopenic. The posterior column of ischium has reasonably good bone stock and offers an optimal site for fixation. Regardless of the particular device used, application of an orthosis/cast for 2 to 3 months appears to facilitate adequate fusion in these patients. Our first case attests to the wisdom of this.

There have been reports in the literature on treatment of hemivertebrae with surgical excision and cast immobilization. Both series included patients having hemivertebrae at varied anatomic levels. Because our paper is focused on the specific problem of lumbosacral hemivertebrae, we chose to compare and contrast these specific cases with those of Bradford et al. and Klemme et al. (Table 1). Klemme et al. did attempt segmental stabilization in their cases with sublaminar suture tapes and used intraoperative compression clamps. We believe that our technique presents a better attempt at stabilization of the segment in this age group. Bradford et al. agree that instrumentation is desirable if and when possible and when the bone stock is adequate. They primarily offered casting for stabilization as instrumentation was not feasible in their cases. We have some reservations about application of physical force without electronic monitoring (Bradford et al.) and again the use of early postoperative wedging casts without neurophysiologic monitoring for correction (Bradford et al.). We used acute correction with complete sensory and motor-evoked potential monitoring in all our cases. Also, restoration with improvement of lumbar lordosis is possible with this construct and has not been specifically addressed in any previous studies with the casting technique.

We have been unable to identify other reports of iliac fixation for lumbosacral hemivertebra excision. The placement of fixation into the pelvis is comfortable for surgeons who are familiar with Galveston spinopelvic fixation. The orientation of the screws may also confer a biomechanical advantage. In conclusion, we believe that hemivertebra excision with iliac fixation represents a safe and effective treatment for congenital scoliosis resulting from lumbosacral hemivertebra. It is ideally suited to the young and small child when the local anatomy prevents the use of more traditional methods for fixation. Because the fixation is not completely rigid, there is the potential
for fatigue failure of the braided cables as observed in patient no. 1. We therefore advise 2 cables with supplemental cast/orthosis protection. Finally, to free the sacroiliac joint, we suggest removal of the implants when fusion is secure.

Key Points

- Failure of formation with hemivertebra is a common occurrence in congenital scoliosis.
- Progressive scoliosis with spinal imbalance and decompensation may be best treated by vertebral excision, correction of lumbosacral deformity, and surgical stabilization.
- Standard constructs for surgical stabilization and fixation may not be optimal in infants and young children as a result of smaller pedicles, poor bone stock, and soft bones.
- A new technique of fixation after hemivertebra excision is described that appears to overcome most of these problems with good prospective results.

References