

**Orthopedics Abstracts**

**Title: ADVANCES IN THE MANAGEMENT OF SPINAL DEFORMITIES - KYPHOSIS**

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**Background:** Prevalence of Spinal Deformity

Multiple studies document the so-called incidence of the various types of spine problems in children with myelomeningocele at different ages.<sup>2-7</sup> Whether these numbers represent an actual incidence or a prevalence is unknown. Cobb measurements also have been used to define developmental scoliosis. The prevalence ranges from 52% to 89% in this population. The prevalence of congenital scoliosis ranges from 7% to 20%. More than 80% of patients aged >10 years will have scoliosis. Using rigid criteria, Trivedi et al<sup>2</sup> recently defined developmental scoliosis in this patient population as a Cobb magnitude >20°. This figure was chosen because curves of less than this magnitude often were observed to improve or even to resolve. The authors concluded that when a scoliosis did not develop by age 15 years, the child would not develop a curve later in life.

**Results:** Associated Health Issues

The global health concerns in these children are numerous and may dramatically influence the care of the spinal deformity. Common issues include central nervous system involvement, such as mental retardation, hydrocephalus requiring shunting, and tethering problems of the brain and spinal cord. Insensate skin, latex allergy, renal anomalies, bacterial colonization of the urinary tract, bowel and bladder incontinence, and lower extremity malalignment are other factors that often require evaluation and treatment.

**Developmental Scoliosis**

Frequently seen in young children, developmental scoliosis primarily resulting from paralysis typically is a long, sweeping, C-shaped curve with or without pelvic obliquity.

Many factors correlate with the occurrence of developmental scoliosis. Clinical motor level is an important predictor.<sup>2,6</sup> Trivedi et al<sup>2</sup> found the prevalence of scoliosis to be 93%, 72%, 43%, and <1%, respectively, in patients with thoracic, upper lumbar, lower lumbar, and sacral motor levels. The level of the last intact laminar arch (LILA) is another important predictive factor in the development of scoliosis.<sup>4,7</sup> Trivedi et al<sup>2</sup> found the prevalence of scoliosis to be 89%, 44%, 12%, and 0%, respectively, in patients with thoracic, upper lumbar, lower lumbar, and sacral LILAs. The LILA is not always synonymous with the motor level.

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Overall, it appears that the three most important factors in predicting the development of scoliosis are the motor level, ambulatory status, and LILA.

Muller and Nordwall<sup>15</sup> reported on the use of the Boston brace in the management of scoliosis and found that when treatment was instituted early and before the curve reached 45°, the brace could arrest progression of the curve. These results, however, have not been reported by others. The brace we have recommended is custom-molded and does not interfere with pulmonary function, lower extremity bracing, self-catheterization, or sitting—a challenge in some patients. Obesity may be a relative contraindication. The brace aids in sitting balance and frees the hands for function. Patients who use a brace need to have their skin checked daily for areas of pressure and breakdown, although a custom-fitted brace usually avoids these problems.

#### Surgical Indications and Principles

Listing the absolute indications for surgical intervention is difficult because the long-term natural history of untreated spinal deformity in this population is relatively unknown. Most agree, however, that progressive scoliosis >50° that causes sitting imbalance is an important indication. McMaster<sup>16</sup> thought that loss of function as an indication was more important than the degree of curvature. Ideally, spinal reconstruction would be done after most adult sitting height is attained.

Medical comorbidities, such as shunt function, pulmonary function, skin condition, and urinary tract infection, require evaluation and treatment before surgery. When poor tissue coverage is a concern, consultation with a plastic surgeon is advised to consider the use of preoperative tissue expanders.

Combined anterior and posterior arthrodesis and instrumentation provide the best chance to achieve a durable fusion.<sup>16-20</sup> Anterior discectomy improves the preinstrumentation flexibility and correctability of the curve, and anterior interbody fusion increases the strength of the entire fusion mass. The addition of allograft bone may be necessary in patients who have had prior bone graft harvesting, in those with small ilia, and in those requiring long fusions. Bone graft substitutes and growth factors may play an important role in the future.

Anterior fusion and instrumentation alone is considered for selected curves. Sponseller et al<sup>21</sup> revisited this technique and had good results with anterior fusion alone only when the thoracolumbar curve was <75°; when there were no syrinx, no increased kyphosis, and no compensatory curve >40°; and when there was independent sitting balance. Parsch et al<sup>19</sup> recommended instrumented anterior and posterior fusions, especially in patients with thoracic level paralysis, to decrease the rate of implantation failure and to prevent postoperative loss of correction. In the series of Stella et al,<sup>22</sup> the best corrections were obtained in patients who had instrumented anterior and posterior fusions.

The fusion should extend from the upper thoracic vertebrae to the sacrum.

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Instrumentation without spinal fusion is not recommended in this patient population; neither is fusion without instrumentation, except for congenital anomalies requiring in situ fusions. Segmental posterior instrumentation provides a means of curve correction and all but eliminates postoperative immobilization. Posterior spinal fusion with instrumentation alone has unacceptably high rates of failure.<sup>20,23</sup> Instrumentation of the dysraphic spine is difficult, and the surgeon must use skill and experience in determining surgical strategies and in choosing the vertebral elements to the implant. When laminae are present, sublaminar wires or cables are passed in the standard caudocephalad fashion; when laminae are absent, drill holes may be made in the vertebral bodies for anchor sites.

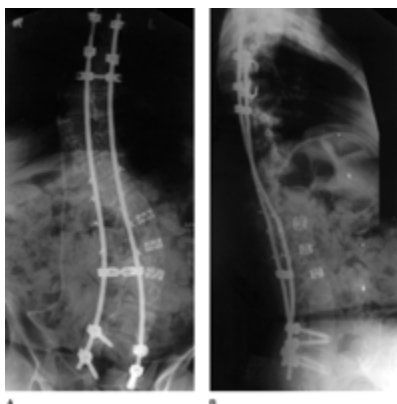
Pedicle screw fixation offers many solutions; in this population, however, the pedicles often are small, dysplastic, and maloriented. Rodgers et al<sup>24</sup> have shown that pedicle screw instrumentation allowed preservation and correction of lumbar lordosis and that the anterior approach possibly could be avoided. This technique also may allow the surgeon to end the fusion above the sacrum, which may be beneficial in select ambulatory patients. Multihook systems are effective in the thoracic spine in which the anatomy is more nearly normal. Consideration should be given to the use of titanium implants if MRI studies of the region will be needed later. Sacral and pelvic fixation generally is thought to be mandatory in patients with fixed pelvic obliquity. However, Wild et al<sup>25</sup> reported spontaneous correction of the pelvic obliquity following anterior and posterior spinal fusion.

### Complications

Spinal surgery in this challenging patient population is associated with higher rates of complication.<sup>18,23</sup> Wound infection may occur in up to half of these patients, as well as incisional necrosis (commonly seen when a triradiate incision is used). However, Ward et al<sup>18</sup> found no long-term disability from incisional skin necrosis in their patients. Infection rates have approached 43% and are highest when surgery is performed with concurrent urinary tract infection.<sup>18</sup>

Foley catheters should be removed as soon as the patient is medically stable. Intravenous antibiotics should be continued postoperatively until discharge. The rate of neurologic deficit is low.<sup>18,30</sup> Cerebrospinal fluid leaks may occur as a result of the surgical dissection or tethering of the spinal cord. Progression of the curve may occur above and below the fusion mass when selection of the fusion levels is inappropriately short. Pseudarthrosis occurs in up to 76% of patients and is related to the surgical approach, type and presence of instrumentation, or use of a posterior approach alone.<sup>20,22</sup> The pseudarthrosis rate is 0% to 50% with an isolated anterior arthrodesis, 26% to 76% with an isolated posterior arthrodesis, and 5% to 23% with a combined anterior and posterior arthrodesis (Figure 1). Pseudarthrosis secondary to implant failure has occurred in up to 65% of cases.<sup>16,18,30</sup> Fractures of the extremities secondary to disuse osteoporosis from immobilization are common. Shunt malfunction may occur following acute correction of large curves.<sup>23</sup>

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**Figure 1** Postoperative images of a patient with myelomeningocele scoliosis. **A**, Anteroposterior radiograph following anterior fusion with placement of interbody cages and posterior fusion/instrumentation to the sacropelvis. Note restoration of coronal balance. **B**, Lateral radiograph demonstrating restoration of sagittal balance.

In more than half of patients in some series, reduced walking ability in preoperative ambulators after surgery has been reported.<sup>30,31</sup> There may occasionally be an improvement in activities of daily living (eg, sitting balance), but hip flexion contractures may increase. A greater potential for ambulation exists when the scoliosis is  $<40^\circ$  and pelvic obliquity is  $<25^\circ$ . The evaluation of postoperative patient activity (and function) is multifactorial and can be affected by older age, obesity, neurologic level, central axis lesions, and motivation of the patient. Improved pulmonary function has been reported after anterior and posterior spine fusion procedures.<sup>32</sup> Patients may have problems with self-catheterization after spinal surgery; however, in these situations, modalities such as mirrors with central holes can be used by the patient. As an alternative, urologic bladder diversion procedures may be performed. Skin sores may develop when changes in sitting balance redistribute pressure on the skin.

#### Rigid Lumbar and Thoracolumbar Kyphosis

Banta and Hamada<sup>33</sup> found that 46 of 457 patients had developmental kyphosis, rigid congenital kyphosis, or kyphoscoliosis that progressed an average of  $8^\circ$ ,  $8.3^\circ$ , and  $6.8^\circ$  per year, respectively.<sup>34</sup> The prevalence of rigid kyphosis of the lumbar spine ranges from 8% to 15%, depending on the series.<sup>35-38</sup> The curve may be initially large at birth, and progression can range from  $4^\circ$  to  $12^\circ$  per year.<sup>39</sup> Mintz et al<sup>35</sup> reviewed 51 children who had a rigid kyphosis at birth; 40 of these patients had a thoracic level paralysis, and 9 of the remaining 11 patients had grade 3 motor strength in the quadriceps. Progression becomes more rapid after the first year of life, when the child begins to sit. The fixed compensatory thoracic lordosis, so commonly seen in older patients, is not present at birth and progresses by approximately  $2.5^\circ$  per year.<sup>36</sup>

Children with rigid lumbar kyphosis have a characteristic clinical appearance: they sit on the posterior aspect of the sacrum with a protuberant abdomen and kyphotic gibbus. Occasionally, an extension deformity of the cervical spine may develop to balance the trunk. The legs appear to be long because of the flexed position of the pelvis, and the lower ribs are splayed laterally. These children usually are more severely neurologically involved, have a higher prevalence of hydrocephalus, and have a poorer quality of life. Thoracolumbar

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kyphosis is characterized by a collapsing C-shaped curve with its apex found in the lower thoracic or lumbar region; it is supple early but can become rigid.

Atrophic or absent erector spinae muscles allow the quadratus lumborum muscle to become a flexor of the spine.<sup>37</sup> The erector spinae muscles, when present and functioning, act as flexors of the spine in their position anterior to the pedicles. Hypertrophic psoas muscles also may act as flexors of the spine, along with the crura of the diaphragm. The dysplastic laminae and pedicles are directed laterally, and the intervertebral articulations are absent or rudimentary. With the development of sitting, the increased moment arm and physiologic load lead to a progressive kyphotic deformity, which continues until the vertebral bodies become wedge-shaped anteriorly and the rib cage rests on the pelvis.

The rationale for surgical treatment of rigid lumbar kyphosis is based on many functional factors, but the absolute criteria remain ill-defined. The deformity is progressive in all cases and is recalcitrant to nonsurgical treatment. The abnormal sitting posture often forces the child to rely on the hands for support, thus diverting their use from functional activities. Repeated episodes of skin breakdown occurring over the apex of the kyphosis are difficult to prevent and create risk for the patient. These two clinical scenarios—abnormal sitting and skin breakdown—are perhaps the most compelling reasons for surgical intervention. Compression of the abdominal contents from the kyphotic deformity also has been suggested as a theoretic concern, yet no study has documented functional benefits related to this parameter following kyphectomy. Families often worry about shortened trunk height; however, performing multilevel corpectomies and osteotomies may exacerbate this problem. Respiratory compromise in untreated deformity also is a concern. However, most of these patients have low aerobic demand, and adaptive changes (eg, flared ribs, barrel-shaped chests) may partially compensate. Martin et al<sup>38</sup> showed that, with wheelchair modifications, these children can do well and may complain only of the cosmetic deformity.

The most appropriate timing and the optimal type of surgery are areas of controversy. Bracing can be used early to slow the progression of deformity, but a surgical intervention is nearly always required. As the child ages, the deformity becomes more rigid, and a compensatory fixed thoracic lordosis develops. Initial attempts at correction involve resection or osteotomy of the apical vertebrae, with little attention directed to the proximal thoracic lordosis. Proponents of neonatal kyphectomy at the time of closure of the myelomeningocele report that the procedure is safe and provides good initial correction.<sup>40</sup> Even though recurrence of the kyphosis was common, the new deformity was less rigid and easier to address.<sup>40</sup> More extensive fusion and instrumentation are required in the older child, and complication rates are higher.<sup>41</sup>

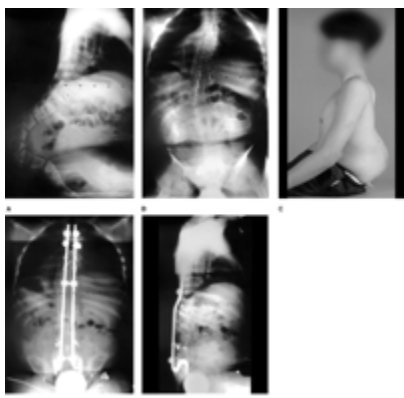
These children also require an extensive preoperative evaluation. An area of interest has been determining the course of the abdominal aorta<sup>42</sup> as well as the method of most effective evaluation (ie, aortography, MRI, ultrasound, computed tomography). All published studies noted here have shown that the abdominal aorta does not follow the path of the kyphosis and is at little risk during kyphectomy. Preoperative shunt function should be tested. When cordotomy is to be performed, the procedure should not be done at the same level of the dura. This will allow the cerebrospinal fluid to circulate and avoids an increase in intracranial

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pressure. Lalonde and Jarvis<sup>43</sup> showed that cordotomy allows for better correction, potentially decreasing spasticity and potentially positively affecting bladder function. However, undertaking a cordotomy increases surgical time and the degree of blood loss.

For thoracolumbar kyphosis, Lindseth and Stelzer<sup>39</sup> described removal of the cancellous bone from the vertebra above and the one below the apical vertebra, which can be performed at any age. The posterior elements are removed, and an eggshell-type procedure is performed without violating the end plates. The apical vertebra is then pushed forward, thus correcting the kyphosis. Fusion is done posteriorly only so that continued anterior growth may provide further correction of the kyphosis. Fixation is with tension-band wiring around the pedicles in younger children and with sublaminar wires, pedicle screws, and rods in older children.

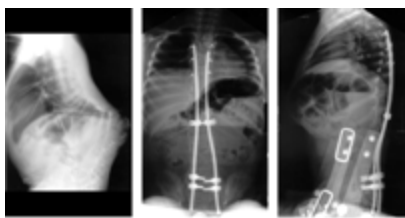
For rigid lumbar kyphosis, the procedures include kyphectomy as described by Lindseth and Stelzer.<sup>39</sup> This entails resection or osteotomy of the proximal portion of the apical vertebra of the gibbus and of the distal segments of the adjacent lordosis, with limited fusion and wire fixation.<sup>44</sup> In 39 patients (average follow-up, 11.1 years), Lintner and Lindseth<sup>44</sup> reported that 34 had a partial loss of correction, but only 2 patients settled into a position worse than their preoperative deformity. The three remaining patients maintained their correction. In the younger child, the kyphectomy is followed by a limited fusion to preserve growth of the adjacent vertebrae. In the older child, kyphectomy should be accompanied by more extensive fusion and instrumentation to the pelvis or sacrum (Figure 2). The optimal instrumentation and distal fixation technique have yet to be determined.<sup>45-48</sup>



**Figure 2** Rigid lumbar kyphosis in a 13-year-old boy. **A**, Preoperative lateral radiograph showing a 119° curve. **B**, Anteroposterior radiograph showing minimal deformity in the coronal plane. **C**, Clinical photograph of the deformity. Eighteen months postoperatively, there is excellent correction in the anteroposterior (**D**) and lateral (**E**) planes following resection of the first and second lumbar vertebrae and instrumentation with Dunn-McCarthy rods. (Case courtesy of B. Stephens Richards, MD, Dallas, TX.)

Sarwark<sup>49</sup> reported on the subtraction osteotomy of multiple vertebral bodies at the apex, which creates lordosing osteotomies at each level.<sup>50</sup> The vertebral body can be entered and subtracted via the pedicles with a curette, distal to proximal. A closing osteotomy is then done posteriorly to obtain correction. This procedure is done in children younger than age 5 years and is supplemented with full sagittal instrumentation from the midthoracic level to the sacrum. Excellent correction and restoration of the sagittal alignment can be obtained, but long-term results after completion of growth are needed to observe all related losses of correction (Figure 3). Reported advantages include less blood loss, decreased morbidity, no need for cordotomy, and continued growth because the end plates are not violated.

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**Figure 3** A, Preoperative lateral radiograph of an 8-year-old patient with rigid kyphosis. B, Anteroposterior radiograph taken 2 years postoperatively showing that there has been continued growth, as noted at the proximal instrumentation, following sagittal reconstruction using the subtraction technique and long instrumentation. C, Postoperative lateral radiograph.

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### Conclusion

Care of the child with myelomeningocele who has a spinal deformity, such as paralytic scoliosis or rigid lumbar kyphosis, is challenging because of the presence of medical comorbidities, such as central nervous system involvement, renal anomalies, and potential latex allergy. Evaluation and management of these children requires a team approach with physicians from multiple specialties. Preoperative discussions with the patient and family must address their perceived as well as the actual benefits of treatment.<sup>51</sup> Early treatment, which may include combined anterior and posterior arthrodesis and instrumentation, anterior discectomy, anterior interbody fusion, or the addition of allograft bone, is required to avoid the progression of curves to severe deformity that later may require extensive measures. Besides using skill and experience during surgical procedures of the spine, the surgeon must be familiar with and be prepared to use different kinds of implants. Although the surgical treatment of these patients remains difficult and is associated with higher complication rates, new implant designs, careful attention to detail, and preoperative planning can yield successful results with minimal associated problems and complications.

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