Progressive Neuromuscular Scoliosis Secondary to Spinal Cord Injury in a Young Patient Treated With Nonfusion Anterior Scoliosis Correction

Laury A. Cuddihy, MD,¹ M. Darryl Antonacci, MD,¹ Awais K. Hussain, MD,² Khushdeep S. Vig, BA, MBBChB,³ Mary Jane Mulcahey, PhD,⁴ and Randal R. Betz, MD¹

¹Institute for Spine and Scoliosis, Lawrenceville, New Jersey; Department of Orthopaedic Surgery, University of Illinois College of Medicine, Chicago, Illinois; ²Department of Orthopedic Surgery, Albany Medical Center, Albany, New York; ³Jefferson College of Rehabilitation Sciences (Philadelphia University & Thomas Jefferson University), Philadelphia, Pennsylvania

Ninety-eight percent of skeletally immature patients with spinal cord injury (SCI) suffer from progressive neuromuscular scoliosis (NMS). Operative treatment has typically been limited to posterior spinal fusion (PSF), but a newer technique as described may be less invasive and preserve more function. A PSF of the entire spine to the pelvis is standard of care. However, maintenance of spinal flexibility, motion, and potential growth is desirable. We present a case for proof-of-concept of utilizing a surgical motion-preserving technique to treat progressive NMS in an 11-year-old girl with T10 level (AIS B) paraplegia with a progressive 60° NMS of the lumbar spine. She had anterior scoliosis correction (ASC) from T11-L5 without fusion. Over 24 months, the curve growth-modulated to a residual of 12° with continued modulation to 7° at 3-year follow-up (skeletal maturity). **Key words:** anterior scoliosis correction, neuromuscular scoliosis, posterior spinal fusion, spinal cord injury

While children account for a relatively small number of new spinal cord injuries (SCIs) each year,¹⁻³ progressive neuromuscular scoliosis (NMS) is arguably one of the most prevalent and challenging secondary complications of pediatric-onset SCI.⁴ Canavese et al⁵ noted scoliosis in 100% of children with SCI. Parent et al⁶ did a systematic review and found scoliosis to be one of the major issues of pediatric SCI. Lancourt et al⁷ found that children from birth to age 10 with an SCI had a 100% incidence of scoliosis. Children 11-16 years old had a 19% incidence, and children older than 16 had a 12% incidence. All children injured prior to skeletal maturity will develop NMS,⁵⁻⁷ 67% of whom will require surgery to arrest the progression.⁸ Age at the time of injury is implicated as the strongest predictor of developing scoliosis, with children injured prior to 12 years of age being almost four times more likely to require surgical intervention.⁴

Secondary conditions of pediatric SCI that co-occur with progressive NMS include pelvic obliquity, pulmonary compromise, and functional decline.^{6,9,10} Pelvic obliquity results in asymmetric sitting and increases risk for pressure injury, poor trunk balance, compromised upper extremity reachable work-space, and gastrointestinal dysfunction.¹¹

Bracing with a thoracolumbosacral orthosis (TLSO) is a mainstay of treatment.¹²⁻¹⁶ Our previous work demonstrated that bracing of children with SCI before development of NMS may delay time to surgical correction by slowing curve progression, but it rarely prevents surgery.¹⁷

Surgical intervention is recommended for progressive NMS secondary to SCI, similar to children with myelodysplasia, when the curves progress to >40° and/or significant functional limitations are encountered related to the deformity.¹⁸ The current standard of care is posterior spinal fusion (PSF) performed on

Corresponding author: Randal R. Betz, MD, Institute for Spine and Scoliosis, 3100 Princeton Pike, Lawrenceville, NJ 08648; phone: 609-912-1500; email: randalrbetz@gmail.com

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children older than 10 years of age with curves $>40^\circ$; whereas for skeletally immature patients younger than 10, specialized instrumentation allowing for continued spine and chest wall growth (magnetically controlled growing rods and spine and rib-based posterior distraction, etc) is considered until the spine is finished growing, at which time a definitive fusion is performed.¹¹ Spinal fusion at an early age limits spinal motion and growth and has potentially detrimental effects on pulmonary function. In the presence of large curves, spinal fusions are also associated with a high prevalence of complications, regardless of age at time of surgery.¹⁹

Over the last 15 years, surgical techniques for fusionless correction of progressive scoliosis have been developed. Unlike a PSF, these newer fusionless techniques (utilizing anterior approaches) provide excellent scoliosis correction without fusion.^{20,21} Anterior scoliosis correction (ASC) with a cord-screw construct is a less invasive fusionless technique for scoliosis that has been shown to provide growth modulation as well as scoliosis correction in patients with AIS^{22,23}; however, it has never been used in cases of NMS. The following case report is a proof-of-concept of ASC for NMS secondary to SCI.

Case Presentation

A skeletally immature 11-year-old girl with a history of an automobile accident 3 years prior presented with neuromuscular scoliosis. Based on previous medical records, as a result of the motor vehicle accident at age 8, the child sustained a T10-T12 SCI causing paraplegia and traumatic brain injury (TBI). In addition to the paraplegia and mild TBI, she also had other systemic injuries, including a pneumothorax and multiple fractured ribs. Following the accident, she had a ventriculoperitoneal (VP) decompression shunt placed. At the time of initial evaluation, she was appropriately engaged in discussion about her medical history without evidence of symptomatic sequela of TBI, had a T10 neurological level of injury with lower extremity spasticity and neurogenic bladder and bowel, and was classified as sensory incomplete (B) according to the American Spinal Injury Association Impairment Scale (AIS).

During her postaccident course, she also developed NMS as expected, as she was 8 years old at the time of injury. When her curve reached 60° , a PSF from T2 to the pelvis was recommended. However, the family reported an interest in fusionless techniques due to concerns about the potential detrimental effects a spinal fusion may have on her current function, including independence in bladder and bowel management. In addition, they had concerns for her future such as recovering walking ability if and when clinical trial therapies advance to clinical application.

On general exam, the patient was noted to sit with a troublesome pelvic obliquity. A range of motion examination of her hips revealed that the right hip had normal range of motion with no contractures. The left hip, however, had restricted motion in both flexion and abduction secondary to heterotopic ossification.

Preoperative imaging (**Figure 1A-B**) showed a right thoracolumbar curve from T11-L5 of 60° with significant pelvic obliquity of 20° . Her pre-op thoracolumbar kyphosis from T7 to L4 measured $+65^\circ$. Radiographs of her hips demonstrated evidence of resected heterotopic ossification and a healed fracture of her left hip. Her bone age was determined to be Risser 0 and Sanders 4.

Because the patient was 11 years old, skeletally immature, and her family was interested in a treatment alternative to PSF, surgical treatment with ASC was presented as an option. ASC would eliminate the issues associated with shortened trunk as well as preserve motion for activities such as driving, feeding, balance, bowel and bladder care, pressure relief, and participation in sports. The risks and potential benefits were discussed at length with the patient and her family, who wished to proceed with ASC.

Operative Intervention and Postoperative Course

Surgery was performed by one of the senior authors (M.D.A.). The patient was placed in the left lateral decubitus position. Due to SCI, neither motor evoked potentials (MEPs) nor somatosensory evoked potentials (SSEPs) could be obtained in the lower extremities. Both MEPs and SSEPs were obtained in the upper extremities.

Figure 1. (A) Preoperative coronal radiograph with a Cobb angle of 60° and pelvic obliquity of 20°. Heterotopic ossification is present but not visible on this x-ray. (B) Demonstrates the sagittal deformity in the patient with a thoracolumbar kyphosis of +65°.

A mini-open thoracotomy approach was utilized to access right T11-12; after a partial take-down of the diaphragm, a retroperitoneal approach was utilized for right L1-L5. An FDA-approved posterior lumbar spine implant system (Dynesys®, Zimmer Biomet, Warsaw, IN) was used off-label for scoliosis. Vertebral body screws were inserted. The polyethylene terephthalate (PET) cord was then introduced, seated, and secured into the screw heads of T11-L5 after appropriate correction of the curve. Under fluoroscopic guidance, significant correction of the scoliosis curve was obtained (reduced from 60° to her planned residual curve of 25°) (**Figure 2A**). The planned residual curve was 25° to allow for 3 years of spine growth remaining.

Blood lost during the 3-hour surgery was 150 mL. Postoperatively, she was in the ICU for 1 day and had a chest tube in place for 3 days. There were no complications, and postoperative neurological examinations (preserved spasticity) were identical to those obtained at baseline.

At the time of discharge on postoperative day 7, she remained pain-free, with no fever and no signs of nausea, vomiting, diarrhea, constipation, chest pain, or shortness of breath. With her family's assistance, she was able to transfer from bed to wheelchair and was back on her preoperative bowel and bladder regimen. Her postoperative urine cultures were negative for growth; however, the decision was made to place her on Keflex for

3 months following surgery for UTI prophylaxis given her history of bacteriuria.

Subsequently her curve corrected through growth modulation from 60° preoperatively (**Figure 1A**) to 25° on initial post-op radiograph (**Figure 2A**) to 13° by 2 years post-op (**Figure 2B**) and to 7° by 3 years (**Figure 2C**) at which time she was skeletally mature (Risser 4+ and Sanders 8). Her pelvic obliquity corrected from 20° preoperatively (**Figure 1A**) to 5° on initial post-op radiograph (**Figure 2A**) to 0° by 2 years (**Figure 2B**) and 3 years (**Figure 2C**). Her sagittal plane shows a nice correction of the thoracolumbar kyphosis of +65° (**Figure 1B**) to +20° on initial post-op radiograph (**Figure 3A**) to a normal mild thoracolumbar kyphosis of +13° (**Figure 3B**), which is normal for patients with SCI.²⁴

The patient has had 3.5 years of follow-up to date. She is doing well and is able to perform all transfers to and from her wheelchair by herself. She also participates in physical therapy and walks with the “exoskeleton.”

Discussion

This case study describes the first application of ASC for neuromuscular scoliosis, and in doing so demonstrates proof-of-concept of ASC for NMS. Unlike idiopathic scoliosis, spinal fusion in children with SCI usually occurs at a younger age with the fused portion of the spine including more levels, often involving the sacrum and pelvis, because many of these children lack sitting balance or have pelvic obliquity. The posterior approach is most often used.²⁵

Further research has been directed toward growth and motion sparing strategies to treat deformity and seek curve stabilization to delay or prevent morbidity. Early procedures included the modified wedge osteotomy.^{20,21} Growth modulation techniques using anterior screw-cord devices have shown promising results with AIS in 1- and 2-year follow-up.^{22,23} Unlike spinal fusion, the use of the screw-cord device to achieve scoliosis correction allows for controlled growth modulation. Samdani et al²² at 2 years of follow-up showed a 70% correction in thoracic curves and 71% correction in lumbar curves, as well as

improvements in axial rotation. These growth modulation techniques offer a potential solution to progressive scoliosis in skeletally immature patients that allows for the controlled correction of scoliosis curves.

Radiographic curve measurements of the patient in this case report taken preoperatively and immediately postoperatively demonstrated modest curve correction from 60° to 25°. This was purposeful, due to the fact that gradual curve correction is expected with growth. She achieved a final curvature of 7° (88% correction) by 3 years and skeletal maturity. The maintenance of mobility and growth is potentially very important to preserving functional status and improving outcomes in patients with SCI.

The biggest advantage of ASC is the preservation of growth and, potentially, preserved spinal flexibility while still allowing for curve correction. It also allows for later fusion or implementation of other therapies, an advantage not afforded by PSF. For this particular patient, maintaining flexibility allows for easier transfers and general wheelchair mobility as well as participation in adapted sports. The flexibility afforded by ASC also allows for kyphosis in the sitting position and lordosis when standing, in marked contrast to the rigidity of the fused spine. The ability to compensate for loss of balance while seated or standing is compromised in PSF but is preserved with ASC. Spinal motion is also a general requirement for activities of daily living such as feeding, driving, bowel management, and pressure relief. Loss of truncal growth following PSF may compromise a healthy body image. Another important consideration is that lack of spinal motion superimposed on traumatic paralysis (as in this patient) may further magnify the functional and social complications associated with “locked-in” syndrome such as the inability to perform perineal hygiene, inability to wear leg braces, and the inability to correct sitting posture in the event of balance loss and spasticity.^{8,20,21}

Patients with neuromuscular scoliosis experience rapidly progressive musculoskeletal deformities, and we present a unique case of severe neuromuscular scoliosis treated with ASC from T11-L5. The excellent correction in the lumbar curve, the maintenance of sagittal balance, and

Figure 2. Postoperative radiographs demonstrating growth modulation of scoliosis secondary to SCI (coronal views). Gradual coronal correction, beginning from (A) the immediate post-op image measuring 25°, (B) 2 years 13°, and (C) 3 years 7°. Pelvic obliquity corrected to 5° on immediate post-op image (A) to 0° (B and C).

Figure 3. Postoperative lateral views demonstrating growth modulation and maintenance of her thoracolumbar kyphosis. Postoperative radiographs at (A) immediate post-op measuring +20° and (B) 3 years measuring +13°, which is ideal for a patient who is a primary sitter. Because she occasionally stands and does walking therapy, she has flexible instrumentation to allow lumbar lordosis.

the minimal complications seen in this case are promising. The advantages of this technique include the potential for preservation of motion and flexibility.

Conflicts of Interest

Dr. Betz and Dr. Antonacci have a royalty contract with a different company with a similar but different device than that used in this case report. The other authors declare no conflicts of interest.

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