
Osteogenesis Imperfecta and Scoliosis - EXPEDIUM™ 4.5 mm System

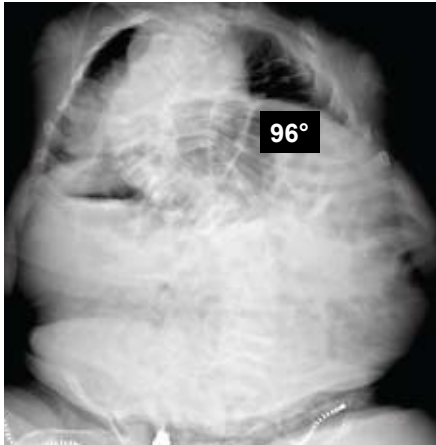
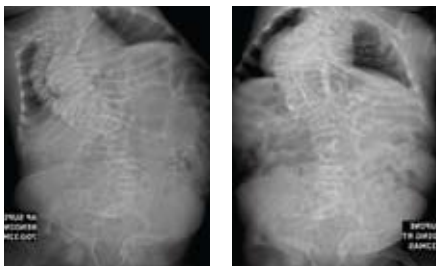


FIGURE 1. Pre-op radiography.



FIGURE 2. Lateral pre-op radiography.



FIGURES 3, 4. Bending Film.

History

The patient is male with severe osteogenesis imperfecta (OI, type III) and scoliosis; he is nonambulatory due to severe long bone deformities and is dependent for all aspects of his care. He has sustained over 80 fractures (including spinal compression fractures) with minimal trauma due to severe osteopenia. His scoliosis had progressed during the last 2 years to the point that he was having difficulty sitting for any length of time due to back pain and he was developing respiratory difficulties and labored breathing. PFTs showed moderately severe restrictive lung disease with FVC of 54%. He is intelligent, articulate, attends the 10th grade and lives at home with his parents.

Radiology

His preoperative x-rays demonstrated a thoracic curve of 96°, as shown in *Figure 1*, which had progressed from 80° just 7 months prior. The lateral film demonstrated a proximal thoracic kyphosis of 38° and a main thoracic kyphosis of 35°, as shown in *Figure 2*. Bending films and a traction film showed the thoracic curve was quite rigid, as shown in *Figures 3 and 4*. Significant chest wall and rib deformities were noted.

Surgical Procedure

Numerous surgical options were discussed with the patient and family: anterior release, halo gravity traction, posterior only surgery with instrumentation, vertebral column resection, staged procedures, etc. After contemplating his medical comorbidities, pulmonary function, low bone mineral density and the family's desire to avoid halo traction, the surgical plan was to perform a wide posterior release, osteotomies where necessary and posterior instrumentation. Due to the patient's small stature and low bone density, the implant choice was EXPEDIUM 4.5 mm Ti. He was given a 3 day course of intravenous pamidronate prior to surgery.

In the operating room, the patient was positioned prone on the OSI Table with special pads to accommodate his size. His upper extremities were difficult to position on the standard arm boards due to contractures and deformity. Total intravenous anesthetic technique was used to allow TcMEP, SSEP and EMG monitoring without the use of hypotension.



FIGURE 5. Post-op radiography.



FIGURE 6. Six month post-op radiography.



FIGURE 7. Six month post-op radiography.

After the spine was exposed from T2 to L3, wide posterior releases were performed at almost every segment, and these were extended to Ponte-type osteotomies from T6-T12. The concave rib heads were also excised along the apical levels to facilitate translation of the curve by improving flexibility. Segmental fixation from T2-L3 was achieved with the use of polyaxial pedicle screws in the thoracic and lumbar spine. The reduction-type screws were used in the concave apex to allow slow, controlled translation of the spine to the rod. Polyaxial screw heads would allow the most modularity and ease of rod engagement with the lowest stress at the bone-screw interface.

After the spinal anchors were placed in the freehand method and checked with fluoroscopy and EMG stimulation, the 4.5 mm titanium rods were cut and contoured to restore thoracic kyphosis and preserve lumbar lordosis for sitting. The rods were engaged with translation and cantilever correction modes. The reduction screws were helpful with translating the apex to the rod in a controlled manner, over time to take advantage of viscoelastic relaxation. A balanced correction was achieved, as shown in *Figure 5*; spinal cord monitoring remained at baseline. Cross links were applied to add torsional rigidity to the construct. Estimated blood loss was 1600 cc.

Results

He was extubated in the operating room, mobilized on postoperative day 1 and was discharged from the hospital on postoperative day 5 in good condition. His hospital course was complicated only by some left upper extremity paresthesias (numerous re-positioning attempts were made intraoperatively due to upper extremity SSEP changes) and a mild ileus.

At three weeks, his incision was well healed; he had discontinued all narcotics and was interested in returning to school. At 6 weeks, the left hand paresthesias had resolved. He has returned for a 6 month postoperative visit with no subjective complaints, no implant prominence, greatly improved sitting tolerance and improved respiratory mechanics. His SRS 22 outcomes scores show improvement in all domains and profound satisfaction with his treatment. His x-rays show continued maintenance of correction and no instrumentation failure, as shown in *Figures 6 and 7*.