

Corrective Surgery For Severe Kyphoscoliosis In Neurofibromatosis Type 1: Operative Challenges And Risks During Revision Surgery

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INTRODUCTION:

We present a case of a young male with neurofibromatosis type 1 with severe kyphoscoliosis who underwent vertebral column resection for correction of his deformity. Treatment of a dystrophic curve poses many challenges, both pre, post and intraoperatively and here we discuss the use of navigation in spine surgery to help in establishing pedicle screw entry points in cases with distorted anatomy to establish a solid fixation in an extreme situation such as this.

CASE REPORT:

Our patient is a 19 year old male diagnosed with neurofibromatosis type 1 since 1 year old. He had undergone multiple surgeries since the age of 3, which included convex epiphysiodesis and growing rod insertion, distraction and anterior hemiepiphysiodesis. Previous implants used were hooks and sublaminar wires, but all had to be removed at the age of 11 due to recurrent infection. Over the years his kyphosis progressed and he developed pseudoarthrosis at T12/L1. He had reduced effort tolerance when walking more than 30 mins and kyphosis of 100 degrees. There was no motor deficit of both lower limbs. He underwent posterior instrumentation and fusion of T4 to L4 with vertebral column resection of T12 and L1. Intraoperatively, the anatomy was distorted and poorly defined because all pedicle entry points were obscured by solid bone due to his multiple previous fusion surgeries. There was a solid fusion block over the thoracic spine. These made spinal column anchorage points difficult to establish. Facet screws had to be inserted for the lumbar spine instead of pedicle screws as most pedicle were small or absent. Neuromonitoring (Motor evoked potential) was normal through out the instrumentation

procedure and intra-op computed tomography (CT) showed accurate placement of implants.

Postoperatively, patient developed paraplegia of the lower limbs from L2 downwards with partial improvement of left L4 and L5 myotome to power 2 on day 7. He is currently still undergoing rehabilitation.

DISCUSSION:

The use of intraoperative navigation system, especially CT based navigation is a useful and sometimes lifesaving tool for accurate placement of pedicular screws, especially in severe deformities. They have been proven to increase the accuracy in the cervical and thoracic spine by about 90%.¹ Without the use of this navigational system, it would be impossible to identify any pedicular entry points, and we would have to resort to using hooks, which do not have the biomechanical strength to support a vertebral column osteotomy. We used the *Brainlab* CT Navigation system that provided a 3 dimensional view of the entire operative field. We had to resort to the use of facet screws for the lumbar spine to due to the extremely small or absent pedicle, possibly due to dural ectasia. The use of facet screw in the lumbar spine as a salvage screw is a useful alternative to traditional pedicle screw placement as they are both biomechanically equivalent.²

Severe kyphoscoliosis with the vertebral rotation, and the additional distortion of surface anatomy by the previous fusion surgery makes traditional free hand surgery impossible. The dysplastic nature of the spine, pedicle penciling, vertebral bodies excavation and osteopenia is what make this case most challenging to perform.

CONCLUSION:

Treating a dystrophic scoliotic curve is a high risk surgery and is very technically demanding. Accurate and safe pedicle screw placement can