

Proximal spinal dislocation with complete spinal cord injury after posterior instrumented fusion for scoliosis in Beals Syndrome

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Background

There's only one case reported in the literature about spontaneous proximal spinal dislocation after scoliosis surgery

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Sudden proximal spinal dislocation with complete spinal cord injury 1 week after spinal fusion in a child with Prader-Willi syndrome: a case report.

de Baat P et al

Aim of the study is to describe a complete spinal cord injury in a girl with scoliosis in Beals syndrome 10 days after posterior T4-L4 instrumented fusion.

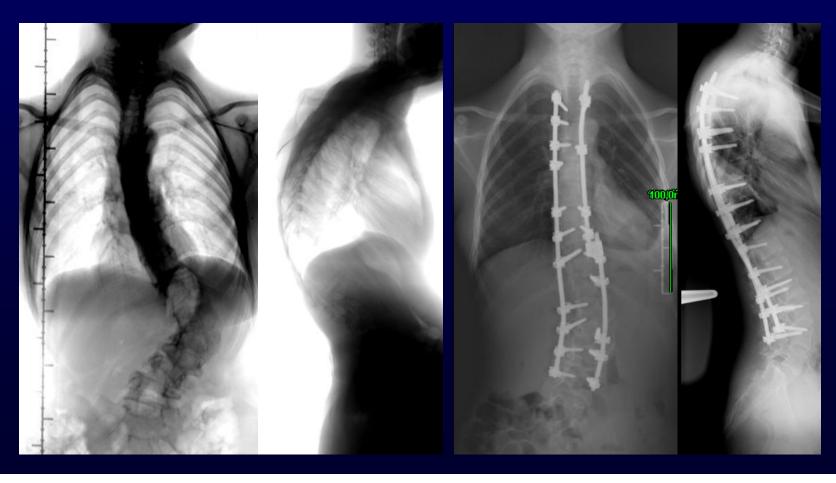
Materials and Methods

- ➤ Beals syndrome is an autosomal dominantly inherited connective tissue disorder characterized by:
- multiple flexion contractures
- arachnodactyly
- severe kyphoscoliosis
- abnormal pinnae
- muscular hypoplasia

similar to Marfan syndrome, but without significant aortic root dilatation.

Materials and Methods

➤ A 14-year-old girl with Beals syndrome underwent a posterior release (Ponte technique) and fusion (T4-L4, pedicle screws only construct) for a sever lumbar scoliosis with hyperkyphosis.



Results

Lumbar scoliosis was corrected from a preoperative value of 78° to 30° after surgery (61.5% of correction), kyphosis from 78° to 39° after surgery.

Postoperative recovery was initially uneventful.

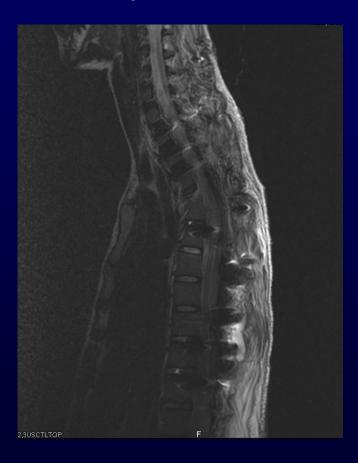
However, 10 days after surgery, a spontaneous anterior T3 dislocation occurred, causing a complete spinal cord lesion.





Results

A revision surgery, consisting of wide laminectomy, T4 corporectomy and extension of fusion to T2 was performed.





Three years after the event the patient is still paraplegic.

Conclusions

There's a growing literature about proximal junctional failures after posterior instrumented fusion for spinal deformities.

However, there's only one report about spontaneous proximal spinal dislocation after scoliosis surgery (Prader Willi syndrome).

Our report describes a similar event, with complete spinal cord lesion without neurological recovery after revision surgery.

Risk factors are not well understood. The muscular hypoplasia can have played a role, but is not confirmed.