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Surgery of a sacral malformation in a Klippel–Feil Syndrome

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Introduction

Klippel–Feil syndrome is known as mostly a cervical spine congenital malformation with many different accompanying malformations (see the cervical spine case). One of the difficult accompanying malformations is located at the lumbosacral spine with transition anomalies, lack of segmentation, spina bifida, etc. Here we demonstrate a case of lumbosacral deformity which needed surgery due to progressive clinical problems, while leaving aside the severe cervical malformations.

Case description

The patient is a 43-year-old woman. She has suffered for 8 months from severe mechanical low back pain mainly in the area of the lumbosacral junction. The patient also suffers from a neurogenic bladder dysfunction and walking longer than 5 min is very difficult for her. The patient shows a Klippel–Feil syndrome Type III with multiple cervical malformations.

Surgical procedure

The goal of this surgery was not to correct anything but to stabilize the malformed lumbosacral junction which became symptomatic in terms of back pain and radicular L5 and sacral root syndrome. Since, the anatomy to place any stabilizing system at the lumbosacral junction in this specific case is very complex, a soft tissue sparing technique for an interbody fusion L5/S1 through a posterior approach has been chosen: with a fibular strut a sacral lumbar interbody fusion has been achieved and intraoperatively a relatively contained and stable L4/5 segment could be proven. The exiting sacral root and L5 have been decompressed through the same posterior midline approach. Two years after surgery, the patient is still satisfied with her results. Back pain is significantly reduced. Neurology is subjectively normal and a cervical surgery has not been necessary to date, in spite of a quite narrow upper cervical spine canal.

Discussion and conclusions

This patient's lumbosacral anatomy is quite complex, so that a posterior surgery with instrumentation, pedicle based, is very demanding at this very much horizontalized junction. Also an anterior approach to this region could be more than a relevant challenge due to the fact that the disc interspace of L4/5 as well as of L5/S1 is barely accessible. An alternative in this rare constellation is a posterior interbody fusion with a fibular strut, or another solid bone graft, plus minor screws through the sacrum into the body of L5 could be an alternative, as demonstrated in this case. If fusion becomes solid, the lumbosacral junction may be sufficiently stable without further surgical measures. A posterior decompression can be done without the risk to further destabilize the lumbosacral junction.

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