



Arthroplasty in patients with rare conditions

Spinopelvic Instability in Conversion Total Hip Arthroplasty: A Complicated Case of Loeys-Dietz Syndrome

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ABSTRACT

A 53-year-old patient with a history of Loeys-Dietz syndrome (LDS) presented with cutout after a right femoral neck fracture treated with a dynamic hip screw. This was treated with conversion total hip arthroplasty (THA), the second reported THA in a patient with LDS and the first in a post-traumatic reconstruction setting. The patient had 2 episodes of posterior hip dislocations within 2 weeks after the operation requiring a revision THA utilizing dual-mobility bearing to achieve stability. LDS is a connective-tissue disorder that is associated with joint hypermobility and spinal deformities, among other features. These factors can affect hip pathology, approaches to treatment, and outcomes. Patients with LDS should have a comprehensive musculoskeletal evaluation and history such as those with Marfan syndrome or Ehlers-Danlos syndrome, especially if undergoing THA. Further research on the implications of LDS on the hip and spine should be performed.

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Introduction

Loeys-Dietz syndrome (LDS) is a rare autosomal dominant connective-tissue disorder due to genetic mutations affecting the transforming growth factor β signaling pathway and the mothers against decapentaplegic homolog 3 protein [1]. It predisposes patients to aortic and arterial aneurysms or dissections, as well as congenital craniofacial and spine defects. The prognosis of LDS is variable and depends on the clinical disease phenotype along with treatment and early screening efforts.

LDS was first proposed by De Paepe et al. in 1996 as a subgroup of Marfan syndrome [2]. Patients with LDS can present with a variety of phenotypes and severities, often similar to those found in Ehlers-Danlos syndrome, Marfan syndrome, and Shprintzen-Goldberg syndrome. The differentiating features of LDS are craniofacial abnormalities (eg, easy bruisability and translucent skin), cleft palate or bifid uvula, and spine malformation and instability. Echocardiography and computed tomography angiography findings include aneurysms and increased vascular tortuosity.

Patients with LDS may initially present to the orthopaedic surgeon with upper cervical deformity, scoliosis, joint hypermobility, or talipes equinovarus [2]. Screening imaging may be required to identify atlantoaxial instability of the cervical spine, which can cause neurologic symptoms or signs of myelopathy and can progress to paralysis or death [3]. Regarding the hip, previous studies have shown an association with hip dysplasia and femoral head necrosis [4].

This article presents a rare case of LDS in a patient who underwent conversion total hip arthroplasty (THA) for failed femoral neck fracture fixation, with subsequent instability requiring revision THA. The musculoskeletal manifestations of LDS and the role of LDS in THA stability have not been explored in current literature.

Statement of informed consent

The patient was informed and consented to allow photographs and case data to be submitted for publication.

Case history

A 53-year-old male radiologist and avid cyclist presented as referral for screw cutout 6 weeks after dynamic hip screw (DHS) fixation for right femoral neck fracture. His past medical history included aortic root aneurysm and thoracolumbar kyphoscoliosis

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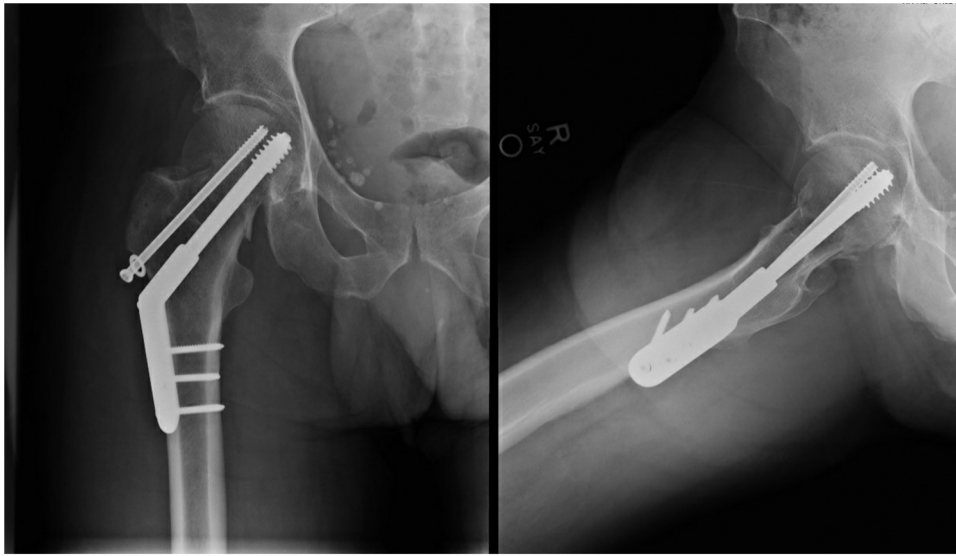


Figure 1. Radiograph of the patient's right hip demonstrating screw cut out from the femoral head.

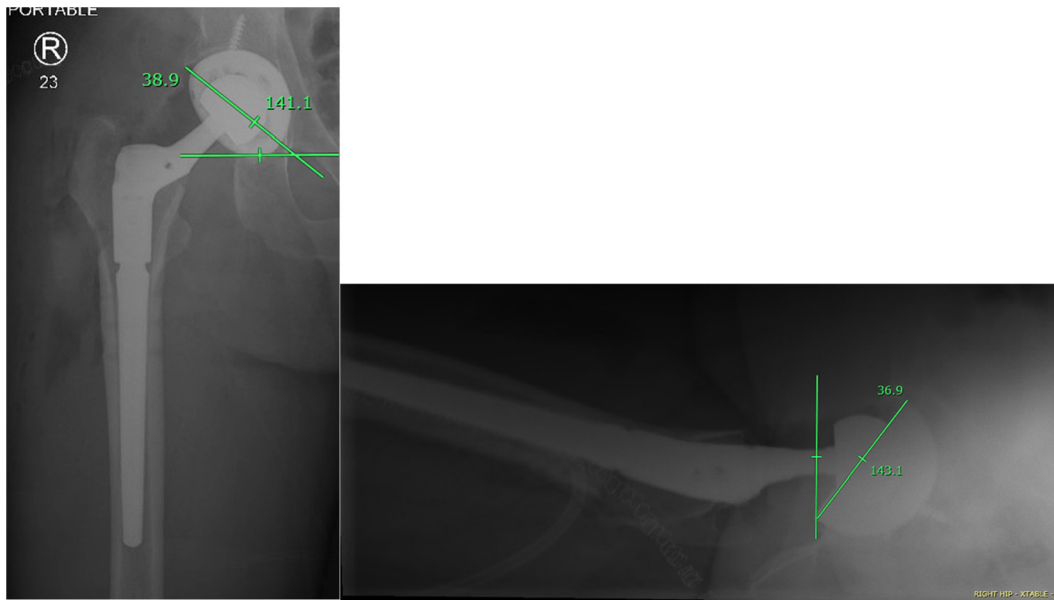


Figure 2. Lateral and AP radiographs of conversion THA.

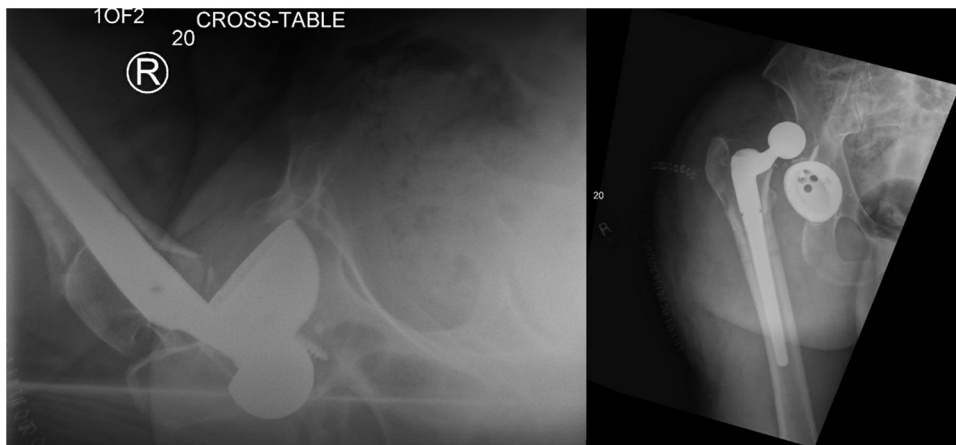


Figure 3. Lateral and AP radiographs showing posterior superior dislocation of right THA. The acetabular socket from the conversion THA is intact.

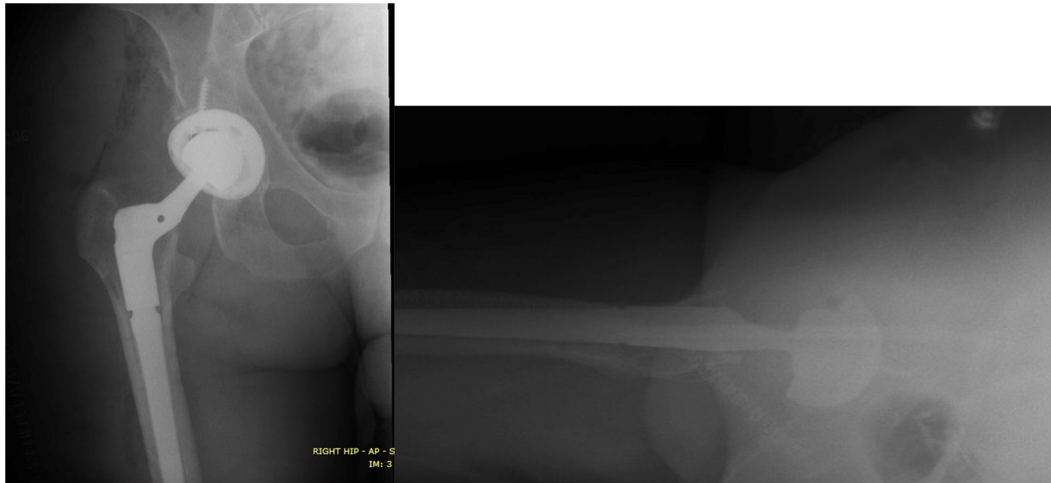


Figure 4. AP and lateral radiographs showing interval reduction of dislocation THA. There is slight subsidence compared with initial postoperative radiographs. The acetabular component retains the same position.

secondary to a genetic disorder, which was not elaborated on during initial presentation, and a prior L1 burst fracture. Past surgical history included a right femoral neck open reduction internal fixation, an aortic root aneurysm repair, and T12-L2 posterior spinal fusion with pedicle screws for the L1 burst fracture. His medications included aspirin and metoprolol.

Right hip imaging at that time showed DHS cut out into the right hip joint and subchondral bone (Fig. 1). Given his desire to return to high-level cycling, this fixation failure was treated with right conversion THA 45 days after his open reduction internal fixation. The THA was performed through a posterior approach with a modular, high-offset, tapered stem, acetabular component with porous

plasma spray coating and a ceramic on polyethylene bearing surface (Fig. 2). The abduction angle was well within the safe range of 30–50 degrees [5]. The cup was slightly overanteverted compared with the safe range of 5–25 degrees [5]. The hip was taken to 90 degrees of flexion, adduction, and internal rotation to the extreme of motion and judged to be stable intraoperatively. There were no reported sites of impingement. Layered closure was performed with #5 Ethibond (Ethicon, J and J Medical Devices, Somerville, NJ) for the short external rotators, a polydioxanone spiral tissue control suture for the iliotibial band closure, Vicryl ((Ethicon, J and J Medical Devices, Somerville, NJ) suture for subcutaneous tissue, and staples for skin. After a successful session with

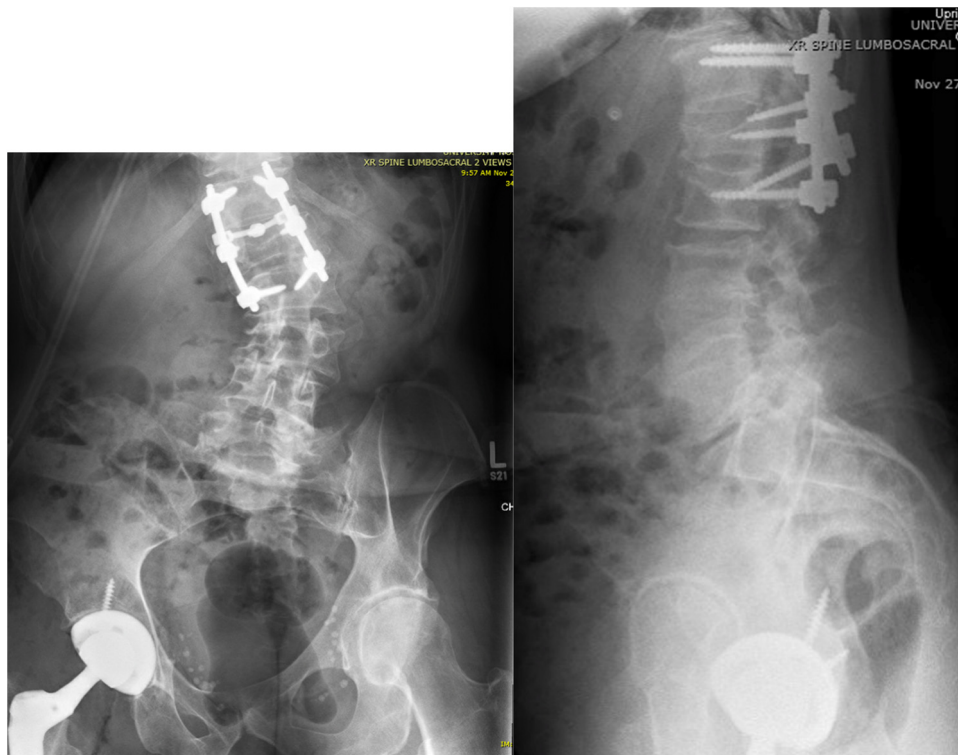


Figure 5. Standing AP and lateral lumbar spine radiographs demonstrating posterior instrumentation from T12 to L2. There is degenerative kyphoscoliosis believed to be a result of his prior L1 burst fracture and genetic condition combined.

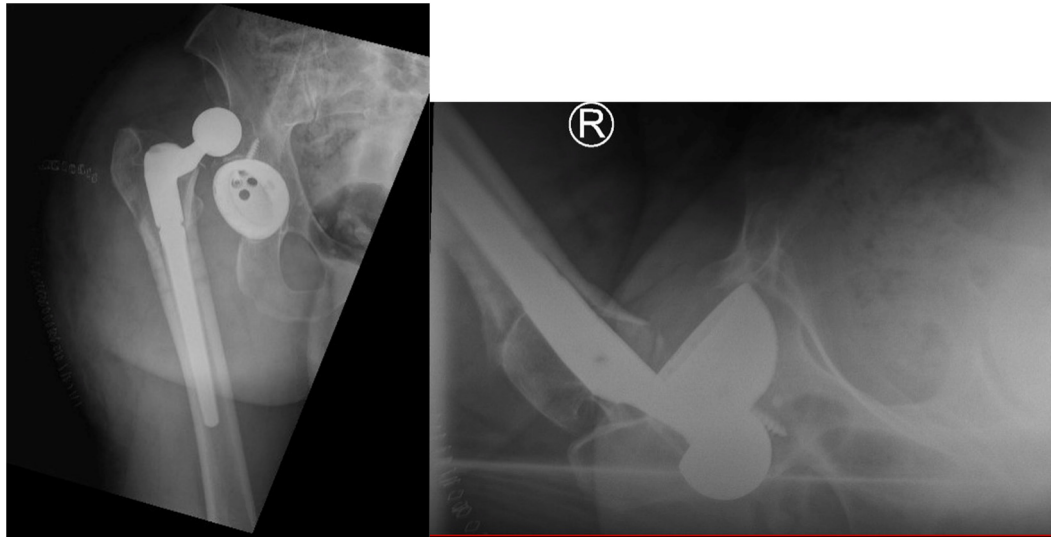


Figure 6. AP and lateral radiographs demonstrating the second posterior superior dislocation of the THA.

physical therapy, the patient was discharged on the first postoperative day.

In the evening on his first postoperative day, the patient attempted to arise from a chair which resulted in a posterior dislocation (Fig. 3) that was closely reduced under procedural sedation in the ED (Fig. 4). The postreduction imaging revealed slight subsidence of the femoral component. In discussion with the operative surgeon, the decision was made to proceed with strict posterior precaution education. The decision was made thinking that the dislocation may be a one-time event and not necessarily related to stem subsidence. The patient was placed into a knee immobilizer after reduction and admitted, allowing additional instruction on strict posterior hip precautions by physical therapy. Standing lumbar spine radiographs were obtained before discharge to evaluate the spinopelvic relationship with his known history of lumbar spine deformity (Fig. 5).

On postoperative day 12, the patient attempted to get into a lower sitting car, which resulted in a second posterior dislocation. Radiographs with anteroposterior (AP) and lateral views of the right hip after his second dislocation are shown in Fig. 6. This second dislocation was again successfully closely reduced under sedation as shown in Fig. 7.

The patient was placed in an off-the-shelf hip abduction orthosis, and discussion with an operative surgeon led to discharge and evaluation in clinic with plan for revision THA. On in-depth review of the lumbar spine films, dislocation films, and the patient's history, the unknown genetic condition was revealed as LDS. In the setting of 2 dislocations less than 2 weeks after his conversion THA, the degenerative lumbar kyphoscoliosis, and LDS, the decision to perform a revision THA was made to achieve long-term stability. Before proceeding, sitting and standing lumbar spine radiographs were performed to further assess the thoracolumbar scoliosis (Fig. 8).

Intraoperatively, the modular tapered stem was upsized one size after intraoperative assessment revealed the stem to be slightly subsided and loose. The acetabular component was not revised because it was felt to have appropriate anteversion and abduction based on prior measurements before and after dislocation events. The bearing was changed to a 46-mm dual-mobility liner and 46-mm polyethylene head with an inner 28-mm ceramic head. Intraoperative reduction was more difficult, and the hip was stable through flexion to 90 degrees, adduction, and internal rotation

without impingement or dislocation through extremes of motion. The postoperative images are shown in Fig. 9.

The patient was discharged home the next day and was followed up with physical therapy. The patient was followed up 2 weeks later and was noted to be doing well. Radiographs taken during the follow-up appointment showed the hip remained reduced (Fig. 10).

At the 6-week follow-up visit, the patient had no complaints of instability (Fig. 10). Clinically, the patient's right leg was 1 cm longer on the right, which was unchanged from the postoperative



Figure 7. Interval reduction of THA dislocation; no cross-table lateral available for comparison.

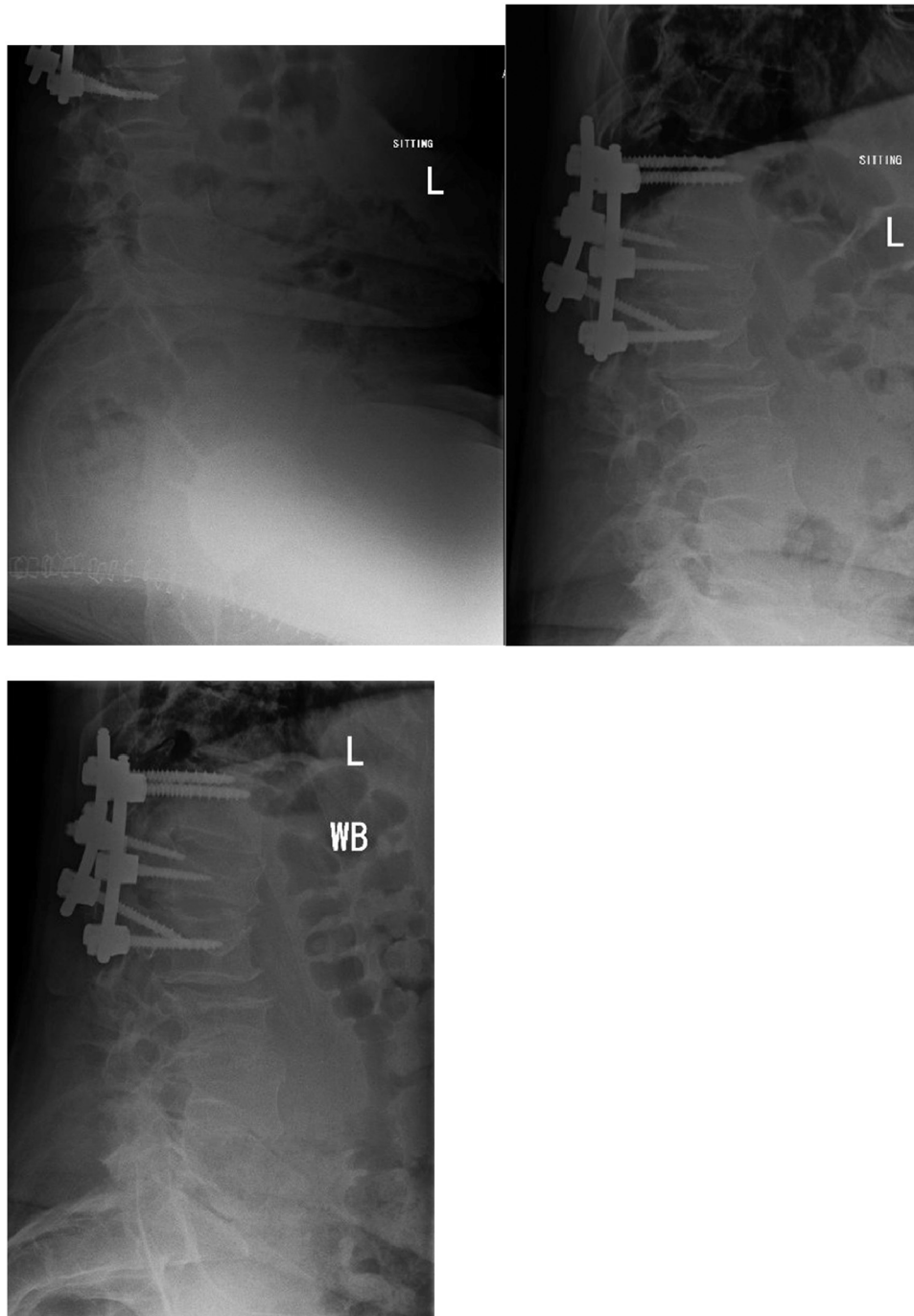


Figure 8. Sitting and standing lateral lumbar spine radiographs reveal thoracolumbar kyphosis with apex at L1 and spondylosis throughout lumbar spine.

measurement. The patient stated he continued to ride a stationary bike 50 miles a day with no pain or complaints.

He was also cautioned about bike riding and informed about the risk of increased prosthetic wear as well as how the extremes of motion of his hip while riding a bike could predispose him to dislocation.

Discussion

This case presents complicated management of a dislocated hip prosthesis in a patient with thoracolumbar kyphoscoliosis

secondary to prior trauma and LDS. The long-term effects of this connective-tissue disorder have not been explored in current orthopaedic literature. To the authors' knowledge, this case is the second reported THA in a patient with LDS and the first in a post-traumatic reconstruction setting [4].

We believe our patient's 2 dislocations after conversion THA were the result of multiple factors: inherent joint hyperlaxity from LDS, coexisting spine deformities, and early subsidence of femoral component. The choice to perform a revision THA allowed for further evaluation of the patient's biomechanics intraoperatively, as well as addressing of the tendency for posterior dislocation.

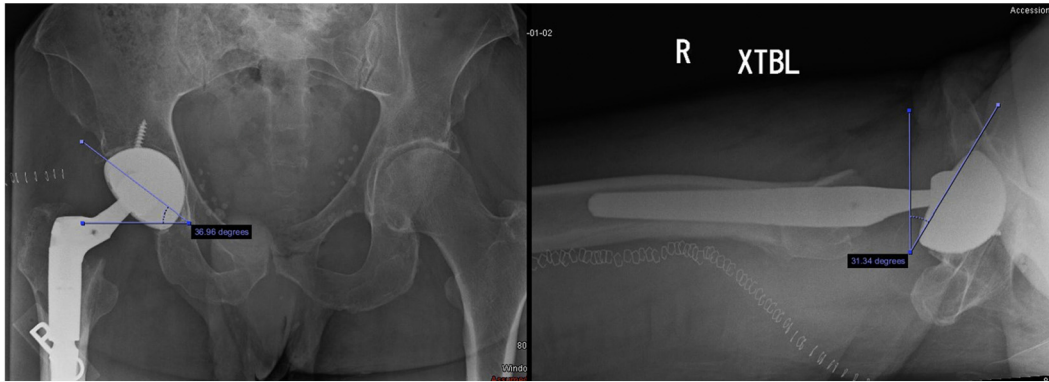


Figure 9. After revision THA, the acetabular component anteversion and abduction angles were 31.34° and 36.96°, respectively.

LDS is characterized by joint hyperlaxity and connective-tissue abnormalities, and this affects inherent hip stabilizers such as the abductor muscles, which when tensioned appropriately contributes to hip stability after THA. We hypothesize the baseline connective-tissue disorder was a contributor to instability after his initial THA.

In addition, this patient's prior thoracolumbar kyphoscoliosis made it difficult to appreciate the affect the sagittal and coronal balance of the lumbar spine may have had on hip stability after

THA. The relationship between sagittal balance in the spine and hip stability is a complex topic that is still under much research and is best evaluated full-length standing scoliosis films, which were not obtained during the treatment course. The C7 plumb line is a line drawn on a full-length standing scoliosis film from the center of the C7 body straight down. That line should pass within 2 cm of the posterior superior corner of the S1 vertebral body. The sagittal vertical axis (SVA) is the horizontal offset from the posterior superior corner of the S1 body to the C7 plumb line. Positive balance is

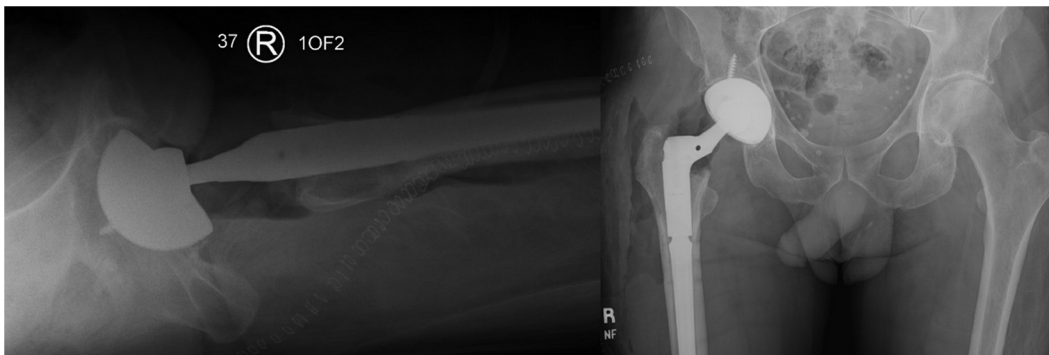


Figure 10. Radiograph of the pelvis showing unchanged alignment of the hardware components and no evidence of complications on postoperative day 8 after revision THA.

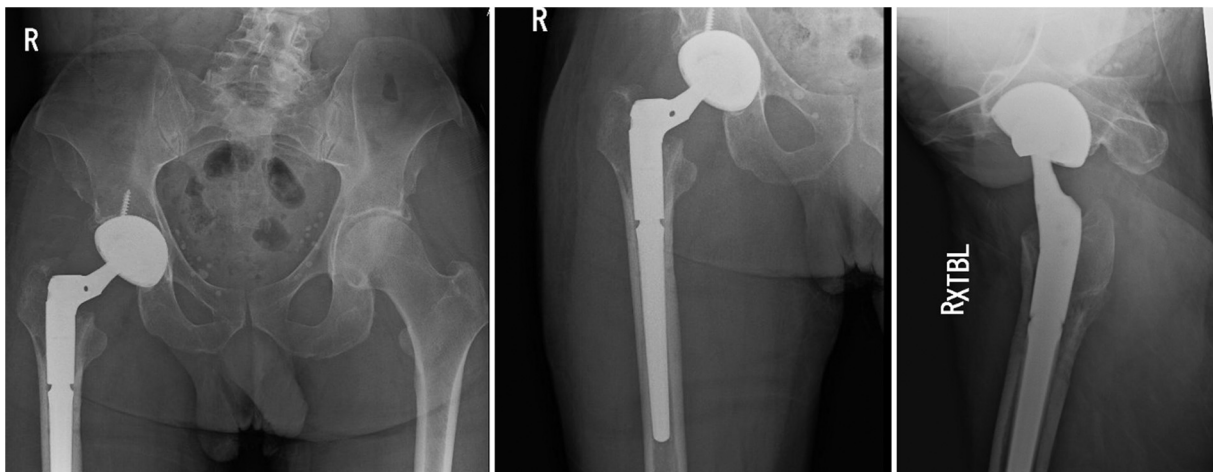


Figure 11. Radiographs of the pelvis at 6-week follow-up demonstrate continued alignment of hardware components and no evidence of failure, despite the patient cycling 50 miles daily.

Table 1
Normal ranges for selected spinopelvic measurements.

Sacral slope	42 ± 8°
Pelvic tilt	13 ± 6°
Pelvic incidence	55 ± 10°
Lumbar lordosis	Males: 41 ± 11° Females: 46 ± 11°

if the C7 plumb line is more than 2 cm in front of the SVA. Negative balance is if the C7 plumb line is more than 2 cm behind the SVA [6]. Our patient had a loss of lumbar lordosis from this prior L1 burst fracture measuring 22°. As the lumbar lordosis decreases, and the back becomes flatter, the tendency is to have a forward shift in the C7 plumb line creating positive balance. To compensate for this, the pelvis can adopt a retroverted position while standing to try to restore balance to the sagittal alignment [6]. This is another possible explanation for how the combined effects of LDS on the spine and prior trauma could have created an environment to dislocate.

Previous literature has elucidated the utility of radiographic parameters on spinopelvic stability [7]. The patient's sacral slope was measured at 39.1°, pelvic tilt at 18°, and pelvic incidence at 57.1°. Generally accepted normal values of these parameters are listed in Table 1 [8]. The patient's slightly decreased sacral slope implies the pelvis is more retroverted, which makes dislocation more likely upon standing or sitting. The patient's measured pelvic tilt was high normal, which is also consistent with a retroverted pelvis.

A high value of pelvic tilt, which indicates a more retroverted pelvis, can increase the risk of dislocation or impingement after THA by affecting the position of acetabular components in the sagittal plane. Although our patient's pelvic tilt was on the normal to high range, it may not have been the sole factor contributing to instability but may have been one of the factors involved. Spinopelvic parameters have therefore influenced 'safe zones' used for cup placement during surgery [9]. Steff et al. showed that stiff hips secondary to spinal fusion often lead to higher positioned acetabular components [10].

Consequences of reduced spinopelvic mobility include impingement after THA. If impingement is severe enough to overcome constraints of the implant, hip capsule, and inherent muscle tension, then dislocation can occur [11]. It is theorized that LDS had already caused baseline deficits in hip capsule and muscle tension in our patient. Some studies have also shown significantly higher rates of THA dislocations in patients with lumbar spinal fusions [12].

The last factor that may have contributed to dislocation is the early subsidence of the femoral component. Although intraoperatively the hip was stable through a range of motion as described earlier, with early subsidence, the stem may have been undersized. In a biomechanical study by Fottner et al., it was shown that undersized femoral components have reduced canal fill and increased micromotion at the implant-bone interface [13]. With subsidence, the tension on the abductors that was seen intraoperatively is lost, and in a patient with theorized tissue laxity, this may have been enough to lead to dislocation. Upsizing the stem and increasing offset allowed for increased tension on the abductors to help restore stability.

Dislocation after THA is a complication that can have many factors. Our patient had LDS, which a complex and rare connective-tissue disorder that may have affected the soft-tissue tension that contributes to stability in THA. Our patient also had thoracolumbar kyphoscoliosis, a result of LDS and prior trauma, which may have also contributed to stability after surgery. Finally, an early subsided

femoral component, leading to loss of abductor tension, may have also created an environment for dislocation. Given that the solution to achieve stability was to upsize the femoral component, increase offset, and use a dual-mobility bearing, it is difficult to tell exactly which factor led to the dislocation. It is very likely that a combination of factors contributed to dislocation. In patients with LDS, or any other disorder that can lead to complex spinal deformity and tissue laxity, we recommend thorough evaluation of all the factors that could contribute to instability.

Summary

This case informs surgeons about LDS, a rare connective-tissue disorder with complex spinal manifestations, and presents a complicated clinical course after conversion THA.

KEY POINTS

- Loews-Dietz syndrome is a rare connective-tissue disorder characterized by craniofacial abnormalities, spine malformation, and joint instability.
- Radiographic spinopelvic parameters (eg, sacral slope, pelvic tilt, pelvic incidence, and lumbar lordosis) can be used to fully evaluate how spinal deformity may affect position of the hip in the sagittal plane.
- Patients with complex connective disorders and spinal deformity should be thoroughly evaluated before total hip arthroplasty.

Conflict of interest

The authors declare there are no conflicts of interest.

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