CASE REPORT

Knee joint instability after total knee replacement in a patient with Ehlers-Danlos syndrome: the role of insert changes as practical solution

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SUMMARY

Treatment of knee joint instability in patients with hypermobile Ehlers-Danlos syndrome (EDS) can be challenging. A 53-year-old woman with hypermobile EDS underwent bilateral total knee replacement (TKR) due to valgus osteoarthritis. During follow-up, she developed hypermobility of both knee replacements. Revision of the insert resolved the issue in her left knee; however, the right TKR required two insert exchanges and unfortunately instability persisted. Therefore, a revision to a constrained prosthesis was performed. Insert exchanges in an unstable TKR can give short-term benefits, but for the long-term, we would recommend a constrained prosthesis.

BACKGROUND

Orthopaedic problems occur in 80%–90% of patients with hypermobile Ehlers-Danlos syndrome (EDS), with the primary musculoskeletal manifestation being joint pain.^{1,2} Total knee replacement (TKR) for osteoarthritis of the knee in patients with EDS has a less satisfying outcome than in non-affected patients. This case describes a 53-year-old patient, who underwent bilateral TKR with a posterior-stabilised (PS) prosthesis and subsequently developed recurrent knee joint instability. Instead of having to undergo a significant revision, her instability was effectively treated for several years with an insert exchange to a thicker insert in the left knee, and two insert exchanges in the right knee. This case report aims to increase awareness of orthopaedic surgeons towards possible complications associated with TKR in EDS, as well as to provide a practical solution in case instability does occur.

CASE PRESENTATION

A 53-year-old postmenopausal woman with hypermobile EDS and a history of a partial lateral meniscectomy first presented to the outpatient clinic in September 2010 with right-sided knee pain since 1 year. Radiographically lateral osteoarthritis was seen and a valgus deformity of 10 degrees was noticed. The Beighton criteria were determined to evaluate the grade of hypermobility and she scored 7 out of 9 and therefore she was classified as 'generalised hypermobility'. Family history combined with information of the department of clinical genetics confirmed that the patient and all of her three sons

had hypermobile EDS, meeting the Villefranche guidelines.³ The patient had no other clinical signs of EDS except for the musculoskeletal hyperlaxity.

TREATMENT

The patient was initially treated with a varus unloader brace. In June 2011, she revisited the outpatient clinic because of persistent pain and was eventually listed for a TKR.

A PS TKR (LCS COMPLETE cemented (DePuy Orthopaedics, Warsaw, Indiana, USA), femoral component 5, tibial component 5, insert 10 mm) was placed in December 2011 (Kellgren and Lawrence: patellofemoral grade 2, medial component grade 3 (femur) and grade 2 (tibia), lateral component grade 4 (femur) and grade 3 (tibia)), with high initial patient satisfaction postoperatively (body mass index (BMI): 44.32). Because of satisfying results of the right knee, in May 2012, she opted to have a similar procedure to her left knee (insert 10 mm) (Kellgren and Lawrence: patellofemoral grade 2, medial component grade 3/4 (femur) and grade 3 (tibia), lateral component grade 3 (femur) and grade 3 (tibia)) (BMI: 43.87). In October 2012, she revisited the clinic because of instability of the left TKR, which was initially treated by a stabilising knee brace. Due to persistent instability, fluoroscopy-guided knee examination under spinal anaesthesia was performed which demonstrated significant medial and lateral instability. The decision was made to revise the insert to a thicker, 15 mm insert (BMI: 44.17). Postoperative satisfaction again was good.

In October 2014, she presented with instability of the right TKR. Because of the good result after revision of the left knee, she was listed for a similar procedure in the right knee (January 2015) (BMI: 44.02). The replacement was complicated by a granuloma, surgically treated by excision.

In June 2017, she revisited the outpatient clinic with recurrent progressive instability of the right knee. The left knee was still stable. During examination, the knee again proved unstable in both varus and valgus stress. A revision to a rotating hinged TKR was considered; however, due to the extensiveness of the operation, the high possibility of an infection and the young age of the patient, it was agreed that another insert exchange would be attempted. The re-revision of the liner was performed in July 2017 and a 20 mm liner was

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placed (BMI: 32.03). After 6 weeks, the patient again experienced increased stability; however, she did experience tenderness of the medial collateral ligament which was treated with a hinged knee brace which led to satisfactory function. At final follow-up after the last surgery, both knees were stable, on the left side with a 15 mm insert and on the right side a 20 mm insert.

OUTCOME AND FOLLOW-UP

In December 2017, the patient revisited the outpatient clinic because of persistent pain and progressive instability during mobilisation. Stress examination of the knee revealed painful medial and lateral collateral ligaments. The decision was made to revise the knee to a rotating hinge prosthesis, which was performed in January 2018 (BMI: 30.06). Postoperatively, she described a less painful knee with a greater sense of stability. She could mobilise safely without walking aids during submission of this report, although with a limited range of motion (90-20-0).

DISCUSSION

Patients with hypermobile EDS frequently develop early osteoarthritis. This is believed to be due to a complex interplay of multiple factors such as joint hypermobility, underlying collagenopathy and repetitive trauma that frequently occurs in hypermobile EDS.⁴

Recognising patients with EDS is important for clinical decision-making. Identifying patients with EDS however can be challenging since clinical presentation can be highly variable, even between family members.

The role of orthopaedic surgery in EDS is controversial. Due to tissue and skin fragility, postoperative complications are common in patients with EDS. In 1999, Weinberg *et al* reported that problems of surgical procedures in EDS may be high relative to other populations without connective tissue disorders. He reported wound complications in 44 patients undergoing 214 surgical orthopaedics procedures, the highest rate of 13% was found in surgical procedures to the knee.⁵ Rose *et al* reported 10 patients with EDS who underwent 12 TKR, with a follow-up of 65 months. The results of the study indicate that TKR can be an effective treatment for knee pain and instability in patients with EDS. Primary indications for surgery were tibiofemoral or patellar instability. All but one patient reported increased tibiofemoral stability after the procedure. Of all 12 TKRs, one patient developed a postoperative deep venous thrombosis, and two patients suffered perioperative superficial wound infection.⁶ Cermak *et al* reported good results after two-stage conversion of a previously fused knee to a constrained rotating-hinged total knee arthroplasty.⁷

Based on this patient with EDS, a prosthetic insert replacement can be a good option for instability after TKR. The surgery is less invasive compared with total revision of the knee prosthesis and could be repetitively performed to achieve total stability.

In this case, the PS LCS COMPLETE prosthesis was used. Rose *et al* described a series of four patients (five knees) in which PS designs were used⁵; in all cases, good knee stability was achieved. Good results by using PS prosthesis for revision surgery of osteoarthritic knees with flexion instability have also been reported by other investigators.⁸

Different constrained condylar knee (CCK) prosthesis designs exist to give greater stability in the face of ligamentous or bony deficiency.⁹ An insert of a constrained condylar knee prosthesis is characterised by the high and broad central post that fits closely against the femoral cam, providing increased stability.¹⁰ A constrained hinged design is indicated in case of

global ligamentous deficiency, but is associated with suboptimal outcomes.¹¹ Such a revision to a rotating hinged TKR or CCK TKR was eventually performed in our case. However, due to the extensiveness of the operation with high risk for perioperative complications, and the young age of the patient, insert exchanges were performed initially. The insert exchanges improved stability and gave satisfying results temporarily, but because of persistent pain and progressive instability, revision to a hinged prosthesis was necessary. We believe that insert exchanges could be a practical solution with short-term benefits in young patients with hypermobile EDS, either to improve stability and also to postpone or sometimes prevent revision to a constrained prosthesis. For the long-term, we would recommend a constrained total knee.

Patient's perspective

'Despite the pain of the medial collateral ligament after the last insert exchange, I would choose insert exchanges again. Postoperatively, the stability of both knees improved significantly and I felt much better, more stable, than before. I would recommend it to other EDS patients with instability after TKR. Unfortunately, due to instability a revision to a constrained prosthesis was necessary. After the revision my right knee felt stable and therefore I am really happy. Still I would recommend the insert exchanges in the first place, because it will give more stability that could last for several years, and because of the extensiveness of a revision operation. If conversion to a constrained prosthesis is necessary, I can tell it will give you the stability you were hoping for.'

Learning points

- A high incidence of orthopaedic problems is seen in patients with hypermobile Ehlers-Danlos syndrome (EDS).
- Prosthetic insert replacement is a good short-term option to improve instability after total knee replacement (TKR) in patients with EDS, or to postpone revision to a constrained prosthesis.
- It is important to distinguish EDS patients with osteoarthritis from patients with osteoarthritis due to other causes, since this influences prosthetic implant selection and facilitates discussing short-term and long-term patient expectations.
- TKR/revision should be performed only in highly motivated patients with EDS and should be performed by a highly experienced orthopaedic surgeon.

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