

# Total knee arthroplasty after former knee fusion in a patient with Ehlers Danlos syndrome

Katerina CERMAK, Bruno BAILLON, Dimitri TSEPELIDIS, Michel VANCABEKE

From Erasme University Hospital, Université Libre de Bruxelles, Brussels, Belgium

A 46-year-old female patient with Ehlers-Danlos Syndrome had undergone fusion of her right knee 25 years before presentation. This markedly affected her quality of life. She underwent a two-stage conversion to a constrained rotating-hinge total knee arthroplasty. She regained a satisfiing range of motion and she has a painfree, mobile and stable knee at 42 months follow-up. Conversion of knee fusion to TKA is a demanding procedure that should only be performed by experienced knee surgeons, in selected cases and on highly motivated patients. Complication and revision rates are reportedly very high but global satisfaction is surprisingly good. Ehlers-Danlos Syndrome was not shown to be a contraindication for such surgeries.

**Keywords** : knee arthrodesis ; fusion ; total knee arthroplasty ; Ehlers-Danlos Syndrome ; Achilles tendon allograft.

### **CASE REPORT**

A 46-year-old female patient with type III Ehlers-Danlos Syndrome (EDS) had undergone a right knee fusion with an external fixator 25 years before presentation. The indication was a major knee instability with recurrent spontaneous episodes of femorotibial and femoropatellar dislocation. In her past orthopaedic history, she also had a cervical fusion for an unstable spondylolisthesis, resulting in diffuse muscular weakness which required displacement in a wheelchair. Her main complaint was the position of her fully extended fused knee in the wheelchair, resulting in a bulky horizontal inferior limb. The patient's request was either to have a knee flexing to 90° or to undergo amputation. Clinical and radiological evaluation revealed a painless fully extended knee without cutaneous adhesions and with femorotibial and femoropatellar fusion (Fig. 1).

## **INTRODUCTION**

Conversion of knee fusion to total knee arthroplasty (TKA) is a rare procedure. The postoperative complication rate is reportedly very high (55-100%) (3,5,7). Ehlers-Danlos patients tend to have joint laxity and wound healing problems. We report a case of a knee fusion takedown in an Ehlers-Danlos woman, with a very good outcome.

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- Katerina Cermak, MD, Resident.
- Bruno Baillon, MD, Deputy Director.
- Dimitri Tsepelidis, MD, Intern.
- Michel Vancabeke, MD, Deputy Director.
  Department of Orthopaedic Surgery, Hopital Erasme, Université Libre de Bruxelles, Brussels, Belgium.
   Correspondence : Dr Cermak Katerina, Department of

Orthopaedic Surgery, Hopital Erasme, Université Libre de Bruxelles, 808, Route de Lennik, 1070 Brussels, Belgium.

E-mail:katia.cermak@hotmail.com

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Fig. 1. – Preoperative radiograph showing total knee fusion

After multidisciplinary discussion, it was decided to suggest a two-stage conversion of the fused knee to a total knee arthroplasty. She was informed of the surgical risks. Because of her coexisting femoropatellar fusion, the first surgical stage consisted in releasing the femoropatellar fusion and resurfacing the patella with a prosthetic component. This applied excessive tension on the flimsy patellar tendon which ruptured. Tendon reattachment with a staple and a wire loop was then carried out (Fig. 2). No complication occurred after this first stage of the conversion. The patient underwent a rehabilitation program to strengthen her quadriceps.

Three months later, the femorotibial fusion was converted to a fully constrained rotating-hinge cemented prosthesis (RT Plus (Smith & Nephew<sup>®</sup>, Baar, Switzerland). As expected, the patellar tendon had not healed and its length after prosthetic implantation was insufficient. An Achilles tendon allograft was then used as a patellar tendon plasty, with an osseous bloc fixed with a screw on the anterior tibial tuberosity. It was decided to accept a high position of the patella and a rather lax extensor complex in order to ensure the main goal of the procedure wich was knee flexion in a wheel chair, considering active extension and «locking» as secondary objectives for this patient (Fig. 3). There was no orthopaedic complication.

Active extension was forbidden for 6 weeks. Immediate weightbearing was allowed with rigid knee brace support for the first 6 weeks.



*Fig. 2.* — Postoperative radiograph after the first surgical step : Takedown of femoro-patellar fusion.

The patient was evaluated 42 months after surgery. She had an active flexion of  $120^{\circ}$ , a lack of active extension of  $10^{\circ}$  and full passive extension. A 2.8 cm length discrepancy in favour of the left side was noted. The patient was pain free, had an optimal desired position in her wheel chair, was very assertive and painless for bed to chair transfer and globally very statisfied.

#### DISCUSSION

Knee arthrodesis has been recognised to be an adequate salvage procedure for various indications including major instability (1). However, complications such as pain, cutaneous healing impairment, sepsis recurrence, hardware failure, lack of fusion (15% of cases) or considerable restrictions in daily life mobility may occur. Even a painless, strong, successfully fused knee can be associated with such disabling functional limitations and such a poor quality of life, that patients may ask for amputation as an end solution to their social and functional limitations. Conversion of a previous knee fusion to a TKA may be an alternative in such situations (2).

Indications for conversion of a fused knee to TKA can thus be made when confronted with severe deficit of function, pain or discomfort of the fused knee, poor quality of life, or sometimes when a patient is enthusiastic about a successful contralateral TKA.



Fig. 3. - Postoperative radiograph after the second surgical step : Total knee arthoplasty with Achilles tendon allograft for patellar tendon plasty.

The main goal of conversion of a fused knee to TKA is to recover a range of motion. The knee must become mobile but must also remain stable and painless. The indication must be carefully thought through. It is mandatory that surgeons performing such conversion procedures be experienced in TKA. On the other hand, patients involved in a desarthrodesis surgery must be prepared for the potential complications and long rehabilitation program following this procedure.

Contraindications to arthrodesis conversion to TKA are young age, neuro-muscular disease, poor bone stock, a past history of severe regional infection, immune impairment potentially reactivating infection, poor skin coverage and-poorly motivated patients (7,8).

Tips and tricks of such conversion procedures are numerous. Extended surgical approaches are usually needed and anterior tibial tubercle osteotomy must be performed in 45 to 100% of the cases (8)].

If ligamentous stability is achieved intraoperatively, a semi-constrained implant can be used. However, care should be taken not to use polyethylene thicker than 18 mm, as it has been shown to increase patellar instability and to complicate skin closure (3). If collateral ligaments are not reliable, a constrained prosthesis should be preferred (43% to 48% of the cases) (3,7,8).

Our patient presented type III Ehlers-Danlos Syndrome, with functionally debilitating ligament laxity (6,12). We thus chose not to trust our patient's ligaments for stability and opted directly for a constrained implant.

Anterior overstuffing is a common problem after knee fusion conversions, resulting in increased tension on the soft tissues, substantial difficulties for wound closure and poor postoperative function (3). The femoral prosthetic component must therefore be positioned as posterior as possible. Specific femoral implants with very thin anterior edge and with a deep trochlea are available. Some authors recommend not resurfacing the patella (3). We did not encounter any anterior overstuffing in our patient.

The quality of the extensor mechanism is an important factor for postoperative outcome. The patella can be absent in up to 17% of the cases (3,7,9). If still present, the patella may be fused to the femur (from 23 to 94% of the cases (3,7,9)). Patellar osteotomy is usually done in the same surgical stage as femoro-tibial fusion takedown (3,7,8,9). We chose to perform a two-stage conversion, starting with patellofemoral fusion takedown, in order to reinforce the quadriceps muscle between the two surgical stages.

Retraction of the extensor apparatus is always present in a fused knee. Adhesiolysis is one of the most common revision procedures needed after takedown of a knee arthrodesis (7). Various techniques can be used to release, reinforce or reconstruct the extensor mechanism with rather poor functional results : V-Y quadricepsplasty (7,9), patellar expansion and a synthetic ligament (3), and even wire reinforcement of the weak patellar tendon have been described (3). We used for our patient an Achilles tendon allograft as a patellar tendon plasty. To our knowledge, tendon allograft had not been reported in combination with a knee arthrodesis takedown.

The skin condition is crucial for arthrodesis conversion to TKA. Kim et al have reported a very high rate (50%) of necrosis of the skin edges after such procedures (9). Some authors report simple skin grafting to cover a small area of cutaneous necrosis (9) or vascularized gastrocnemius flap, which has been associated with poorer functional outcomes (3,7). Other authors recommend a two-stage conversion using soft tissue expanders as a first surgical step (3,4,10).

Ehlers-Danlos syndrome is usually associated with a hyperextensible skin (12). One might think that this extensible cutaneous condition is an advantage when confronted with skin coverage problems ; however, the skin in Ehlers-Danlos patients is also fragile and susceptible to dystrophic scarring. A wound complication rate of 13 to 17% has been reported after various types of knee surgery in Ehlers-Danlos patients (11,13). Fortunately, our patient did not develop any wound healing problems.

Postoperative pain after TKA conversions can be difficult to control initially, especially because most patients were preoperatively painless. The largest study available (36 cases) reports only 61% of patients completely pain free at 7.7 years follow-up (9). Our patient reports no pain 42 months after surgery.

Initial range of motion can be disappointing, even a few months after surgery. This insufficiency is often caused by adhesion and arthrofibrosis encountered in up to 40% of cases (3,5,7). However, the range of flexion improves with time and exercise during the first year to achieve long term satisfactory results, ranging from 77.7° to  $100^{\circ}$  (3,4,5). Extension lag is also frequent and ranges from 0° to  $40^{\circ}$  (4,5,8,9). Immediate postoperative mobilisation, when possible, is required. Our patient had a very satisfying 120° active flexion, an active extension lag of 10° and no passive extension lag.

Other reported postoperative complications include ligament laxity, prosthetic loosening, polyethylene wear, infection, extensor tendon contracture or ruptures, capsular myositis ossificans or peroneal nerve paresis.

Surgical revision rate may be as high as 62% (3,5). Some patients (11 to 28.5%) have to undergo a knee re-fusion (3,5,7) for chronic ligament instability or sepsis reactivation.

Surprisingly, despite the high rate of complications and revision surgery, a high proportion of patients are very satisfied and prefer their mobile knee (3,9).

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