Anterior Cruciate Ligament Reconstruction With Achilles Tendon Allograft in a Patient With Ehlers-Danlos Syndrome

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Ehlers-Danlos syndrome (EDS) is a heritable connective tissue disorder characterized by varying degrees of joint hypermobility, vascular fragility, and skin hyperextensibility.[22,26] It affects 1 in 5000 live births, and orthopaedic manifestations are common, especially those involving the knee and shoulder joints.[5,26] The diagnosis of EDS is based primarily on clinical criteria. The principal clinical features of EDS are easy bruising, skin hyperextensibility, delayed wound healing with atrophic scarring, and joint hypermobility.[10] Easy bruising or bleeding from minor trauma is a common initial manifestation.[6] Although hematologic studies are typically within normal ranges, capillary fragility can be confirmed with a Rumpel-Leede (or tourniquet) test. EDS can cause recurrent dislocation and subluxation owing to ligamentous and capsular laxity, leading to bone, cartilage, and soft tissue damage.[20,22]

In the setting of anterior cruciate ligament (ACL) reconstruction, a variety of grafts are available for surgical reconstruction, including autografts, allografts, or synthetic materials. The most popular autografts used for ACL reconstructions are the patellar tendon and 4-strand semitendinosus-gracilis autograft.[3,21,24] The most popular allografts for ACL reconstruction are the bone–patellar tendon–bone and Achilles tendon–bone allograft. Allografts offer several benefits: shorter operating time, no donor site morbidity, availability of larger grafts, smaller incisions, less pain, better cosmetic result, easier rehabilitation, a lower incidence of postoperative arthrofibrosis, and no weakening of extensor or flexor apparatus.[1,8,9,14,15,18,19] However, as compared with autografts, allografts are associated with a higher risk of aseptic revisions and lower rates of graft survival in the young and active patient population.[11,12,16,17] For patients with EDS, autogenous grafts pose a problem attributed to the impaired intrinsic mechanical properties of the tissue in these patients, which may lead to stretching and recurrence of instability and failure after surgery.[26]

There have been 3 related case reports where nonautograft methods were used in patients with EDS, including an Achilles tendon allograft for chronic shoulder instability,[2] an allograft for reconstruction of a chronic patellar tendon rupture,[7] and patellar tendon reconstruction with allograft and synthetic augmentation.[13] However, no studies have described surgical considerations for ACL reconstructions in patients with EDS, particularly the advantages of using allograft and perioperative considerations.

We present a case of a 25-year-old man who elected to have ACL reconstruction with undiagnosed EDS prior to seeing the senior author (X.L.). The purpose of this report is to emphasize the importance of recognizing the signs and symptoms of EDS and conducting preoperative testing among patients suspected of having EDS. Special considerations in this patient population should focus on graft choice and establish appropriate intra- and perioperative management to lower the surgical and failure risks of patients with EDS and ACL injuries.

CASE REPORT

Our patient was a 25-year-old man who worked as a physically active security site supervisor. He sustained a left

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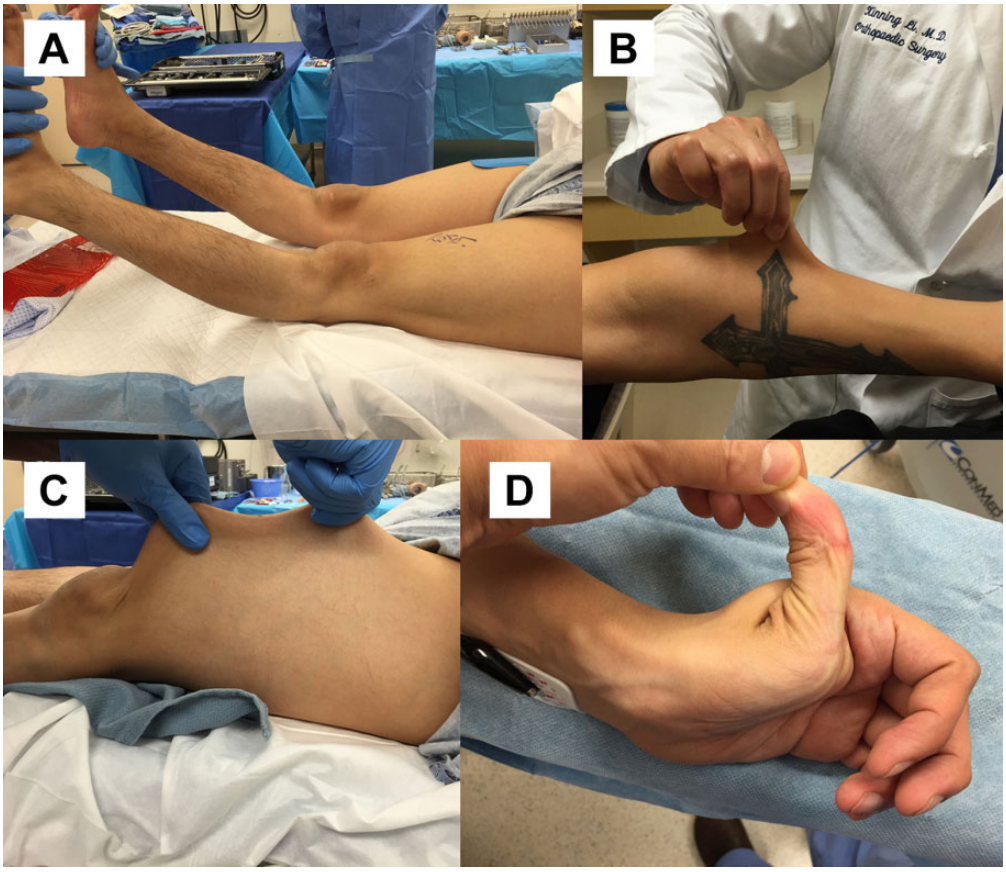

Figure 1. (A) Intraoperative examination under anesthesia demonstrates bilateral knee hyperextension, which is symmetrical between the anterior cruciate ligament–ruptured knee (left) and normal knee (right). (B) Skin on the forearm is very elastic on examination in the clinic. (C) Elastic skin is also seen on the thigh under anesthesia. (D) Thumb is double-jointed on hyperextension in the operating room under anesthesia.

femur fracture and left knee injury when a car hit him 2 years previously. He subsequently underwent placement of a retrograde nail and recovered well postoperatively. However, he continued to complain of instability and posterior medial knee pain. He could walk with normal gait but reported occasional limping and buckling of his knee with long walks. He could not run or pivot with sports activities for 2 years and felt that his knee moved in different directions with ambulation.

Physical examination of the left knee showed full range of motion but revealed a posterior sag sign, grade 2 positive posterior drawer test, and positive Lachman test with 3B translation and a positive pivot shift. Physical examination of the right knee also revealed a posterior sag sign with a grade 2 positive posterior drawer test with a negative Lachman examination result and normal pivot shift. Of note, both knees presented with 30° of hyperextension in the operating room under anesthesia (Figure 1A), and the patient's skin was very elastic on the arm and thigh (Figure 1, B and C). He also had double-jointed thumb on examination (Figure 1D). On the basis of the preoperative clinical examination, the senior author suspected a diagnosis of EDS.

Radiographs of the left knee showed anatomic alignment and no fracture. The joint spaces were preserved. An intramedullary rod with interlocking screws of the femur was visualized without any perihardware lucency or fracture. Magnetic resonance imaging of the left knee was obtained, revealing a complete rupture of the ACL, posterior cruciate ligament attenuation, and tear of the posterior horn of the medial meniscus (Figure 2).

After discussion, the patient elected for arthroscopic ACL reconstruction and meniscus repair. Autograft and allograft options were both discussed with the patient. Although the patient did not have a diagnosis of EDS in his preoperative medical history, it was later confirmed with genetic testing. After a long discussion with the patient, the senior author had concerns about using an autograft-native tissue, given its intrinsic laxity, and instead opted for an allograft to ensure long-term stability with the ACL reconstruction procedure.

The ACL reconstruction was performed with anteromedial drilling with flexible reamers and anatomic femoral tunnel positioning (Figure 3). The tibial tunnel was drilled with a rigid reamer (Figure 4A). A metal screw was placed in the femur for the graft fixation (Figure 4,

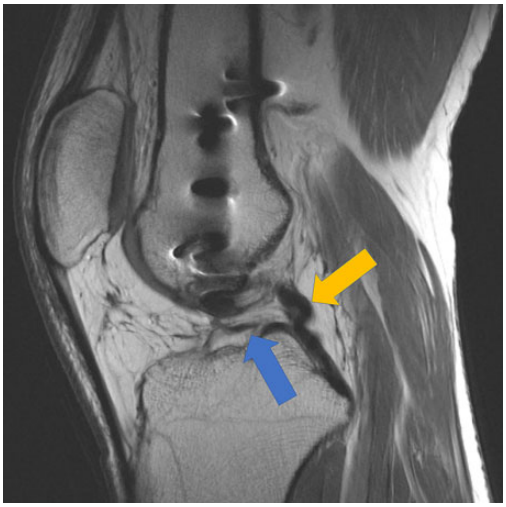

Figure 2. Magnetic resonance image demonstrates anterior cruciate ligament rupture (blue arrow) and attenuation of the posterior cruciate ligament (yellow arrow).

B and C); a biocomposite screw was placed in the tibial tunnel; and the graft was subsequently backed up with a 4.75-mm SwiveLock anchor (Arthrex) into the tibia distal to the drill hole. Tensioning of the graft was done in 20° of knee flexion with slight posterior drawer. Final ACL reconstruction is shown in Figure 4D and Figure 5.

The patient was also referred to a geneticist for possible EDS. With extremely hypermobile joints, hyperelastic skin, some wide and atrophic scars, and a family history of dislocations, joint discomfort, and migraines, he was diagnosed with classic type II EDS. He did not show aortic dilation or lens dislocation.

Postoperative recovery was unremarkable. The patient's knee range of motion was 0° to 130° at 6 weeks postoperatively. Regarding outcome measures, preoperatively his modified Cincinnati score was 52 and his International Knee Documentation Committee (IKDC) score was 55.2. Six months postoperatively, his scores were 98 and 87.4, respectively. At postoperative 9 months, he returned to normal exercises and activities without pain or swelling. Upon last visit at postoperative 2 years, the patient continued to participate in regular preinjury activities and had a Cincinnati score of 100 and an IKDC of 100. His Lachman examination was 1A with positive pivot glide on examination; however, the patient reported no subjective feelings of instability. Our patient consented to this case report publication.

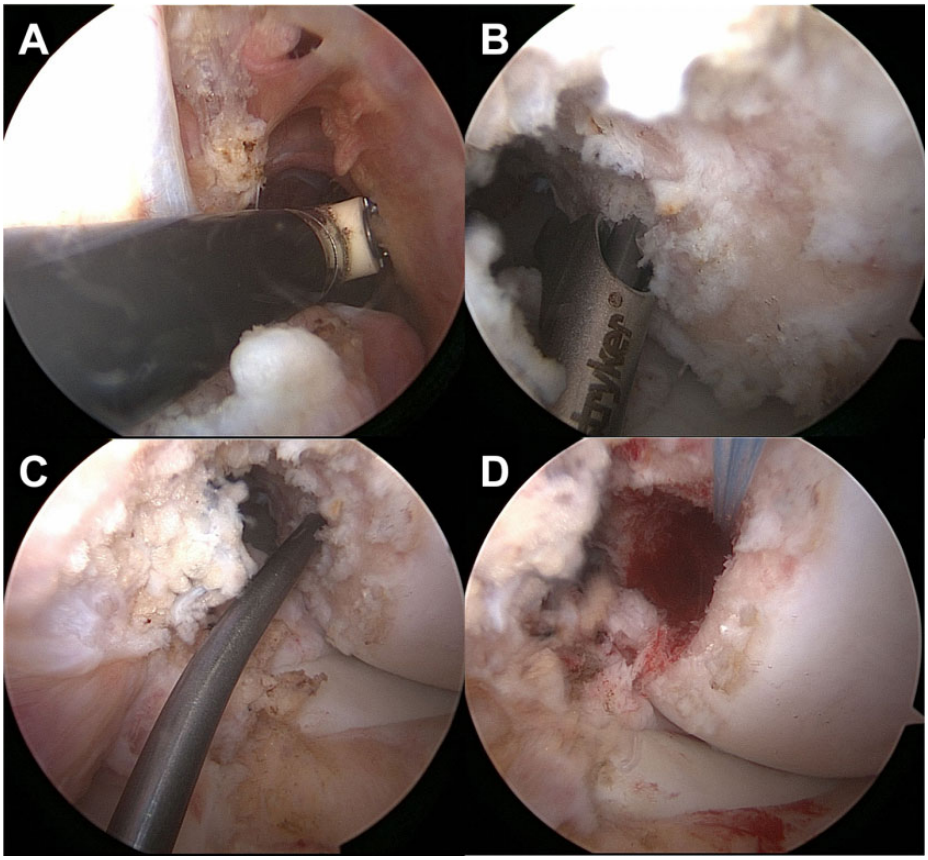

Figure 3. (A) Empty femoral wall sign with complete anterior cruciate ligament rupture. (B) Over-the-top guide for the flexible guide pin is placed anatomically on the femoral insertion. (C) Guide wire is placed into the femur. (D) Tunnel (10 mm) is drilled with a flexible reamer.

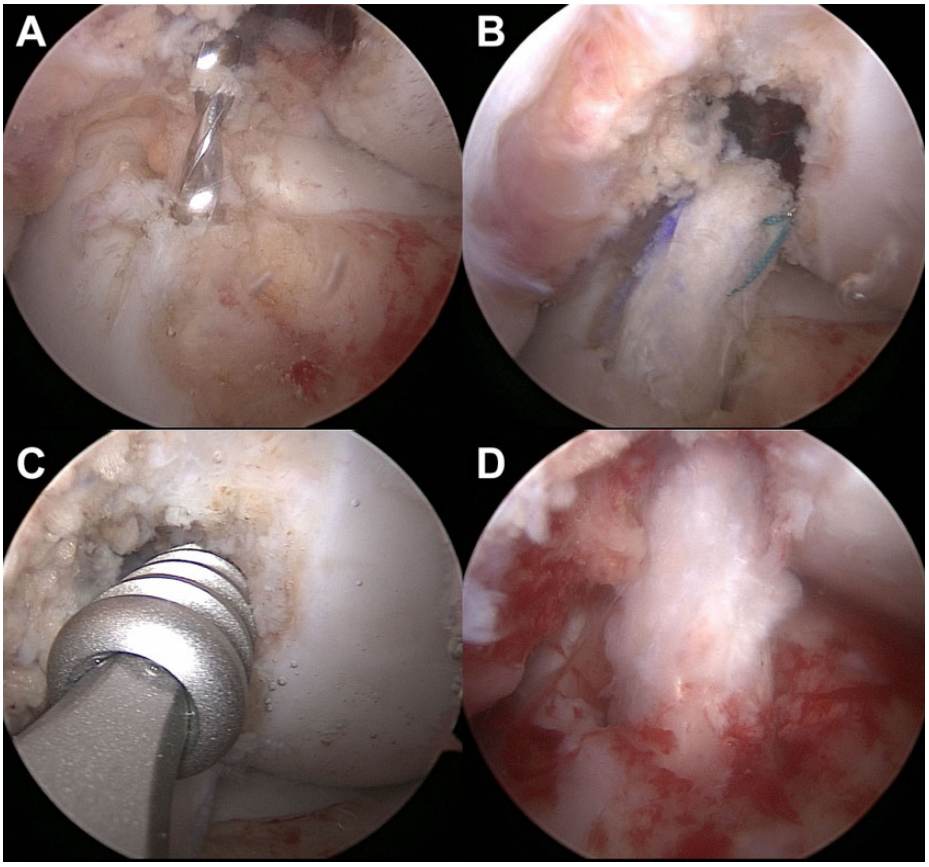

Figure 4. (A) Tibia rigid guide pin is drilled on the anatomic tibial insertion anterior cruciate ligament footprint. (B) Achilles allograft is shuttled from the tibia to the femur. (C) Metal screw is used for the femoral fixation. (D) Final anterior cruciate ligament reconstruction with Achilles tendon allograft.

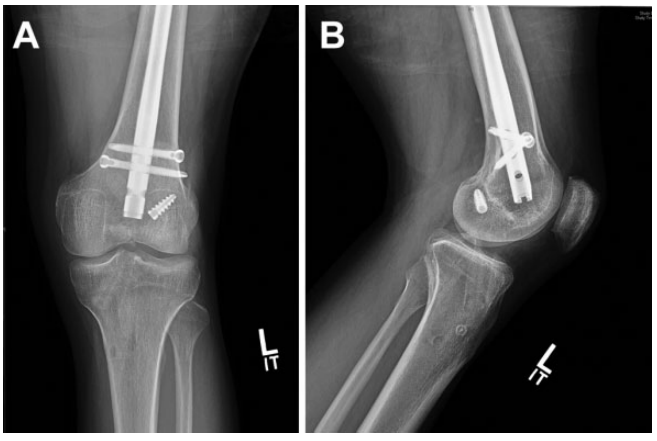

Figure 5. Postoperative radiograph of the knee: (A) anterior-posterior and (B) lateral views.

DISCUSSION

Despite extensive studies on the management of ACL injuries and optimal reconstructive technique, very little literature exists regarding the optimal treatment strategy for ACL reconstruction in patients with EDS. To the best of our knowledge, there is no study reporting ACL reconstruction with allograft tissue in patients with EDS. But there is 1 case report of a patient with EDS who had ACL reconstruction with an autograft and synthetic ligament augmentation, as well as 3 related case reports where nonautograft methods were used for ligamentous injuries other than ACL rupture. Williams et al[26] detailed an ACL reconstruction using an autograft augmented with a synthetic ligament in an 18-year-old patient with a known diagnosis of EDS, with good outcomes. Chaudhury et al[2] described the use of bilateral Achilles tendon allografts to provide anterior and posterior shoulder stability in a patient with EDS who had undergone previous unsuccessful stabilization attempts. Iacono et al[7] and Matziolis et al[13] discussed successful reconstruction of a patellar tendon rupture using an allograft alone and an allograft with synthetic ligament augmentation, respectively.

Hypermobile joints and hyperextensive skin were assessed on our patient during the preoperative clinical examination in the office and also under general anesthesia. Joint hypermobility (see Figure 1A) in patients with EDS can manifest as frequent joint dislocations and can be classified with the Beighton score or criteria. A Beighton score is obtained by assessing forward flexion of the trunk with fully extended knees, hyperextension of elbows and

knees beyond 10°, passive apposition of thumbs to the flexor aspect of forearms, and passive dorsiflexion of little fingers beyond 90°. A Beighton score ≥5 of 9 defines hypermobility for adults and ≥6 for prepubertal children. Skin hyperextensibility is tested at a neutral site, such as the volar surface of the forearm, by pulling up the skin until resistance is felt (see Figure 1, B and C). Atrophic papyraceous scars are indicative of delayed wound healing and can be found on patients with EDS.[6,23] Therefore, preoperative clinical examination plays a critical role in diagnosing a patient with EDS and allows for shared decision making with the patient regarding graft choice. With a high suspicion of EDS and additional confirmation of the syndrome with genetic testing, the senior author decided to reconstruct the ACL using allograft tissue in lieu of autograft, given concerns with stretching of the native autograft over time.

Clinical identification of EDS is also pivotal for optimal perioperative anesthetic management, as special consideration is required throughout the procedure based on the clinical manifestations of EDS.[6] The cervical spine should be evaluated for any atlantoaxial instability from ligament laxities, and extra care must be taken with movement and positioning of the patient during intubation or laryngeal mask insertion to prevent bruising and joint dislocations.[23] Fiberoptic intubation may be considered to minimize trauma, as simple laryngoscopy can significantly damage the mucosa and airway.[10] Temporomandibular joint luxation is also possible during mask ventilation, and care is required to prevent this from occurring.[25] Tissue fragility poses another substantial risk for perioperative complications. Therefore, external shear forces and tissue pressure should be minimized with additional padding, especially to the eye to protect from retinal detachment and globe rupture. The propensity for tissue to bleed in these patients can also cause problems, such as hemorrhage and hematoma formation. Therefore, the use of tourniquets, the number of needle pricks, and the amount of time that catheters are placed should be minimized.[4] In addition, all patients should be blood typed and cross-matched in case a blood transfusion is needed.

For the patient with suspected EDS, it also is important to set up a consultation with a geneticist to confirm the diagnosis and further characterize the type of EDS. The patient should also be referred to appropriate specialists to undergo cardiovascular and ophthalmologic examinations. Patients with EDS have a significantly increased risk of developing cardiac valvular and vascular disease and aortic dilatation, as well as retinal detachment, glaucoma, and scleral fragility. Therefore, multidisciplinary management and follow-up care are essential for such patients.

CONCLUSION

We recommend that surgeons be aware of the signs and symptoms of EDS when evaluating patients preoperatively for orthopaedic surgery. If a patient is suspected of EDS, it is crucial to obtain a proper diagnosis as soon as possible, as management of this condition demands additional perioperative, intraoperative, and anesthestic considerations. In patients with EDS, autografts and allografts could each be considered an option for ACL reconstruction; however, the senior author decided to proceed with allograft reconstruction owing to the concern of compromised integrity of the patient's native tissue, which may ultimately lead to stretching and possible surgical failure. An appropriate surgical management plan should be established preoperatively, including selection of the appropriate graft as a shared decision with the patient and ensuring that all possible steps are taken to minimize perioperative complications. A multidisciplinary team is essential in the treatment of patients with EDS, as they require orthopaedic surgical intervention during the pre-, peri-, and postoperative periods.

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