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Case reports

Single time angular deformity correction and treatment of knee instability in congenital fibular hemimelia. A case report

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ABSTRACT

Background: Fibular hemimelia is the most frequently occurring congenital anomaly of long bones. These patients, among other deficiencies, have a poor development of the anterior cruciate ligament (ACL). Unless it causes clinically assessed instability of the knee, nonsurgical treatment is given. When surgical treatment is required, correction of angular limb deformity must be realized prior to ACL reconstruction.
Methods: We present the case of a 16-year old patient with congenital fibular hemimelia. Physical examination showed genu valgum, anteromedial rotatory instability and recurvatum of the right knee. We decided to perform surgical correction of the angular deformities and ACL reconstruction in the same surgical time.
Results: Twelve months after surgery, the patient had no evidence of clinical instability, with a range of motion from −5°–110° of the right knee. No claudication or gait instability was found. The KT-1000 arthrometer showed a difference of 2 mm between both knees.
Conclusion: The ACL reconstruction and corrective osteotomies of angular deformities performed in a single surgical procedure had a good clinical result in a 12 month follow up-period, restoring stability of the knee and allowing a normal gait cycle.

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1. Introduction

Fibular hemimelia is the most common congenital deficiency of long bones [3,11]. Clinical presentation is variable and can range from hypoplasia of the fibula to its complete absence [8]. The therapeutic options for congenital fibular hemimelia include either amputation with early prosthetic fitting or tibial lengthening.

Patients with fibular hemimelia may additionally have poor development or absence of ACL, among other deficiencies [4,5,11]. The literature suggests that a congenital disorder of ACL not causing functional instability of the knee during normal daily activities must be treated conservatively. In the presence of significant knee instability which interferes with daily activities, reconstruction surgery of ACL should be considered. Previously, corrective osteotomy of angular deformities must be completed. Both procedures are traditionally performed in two separate surgical times [3–5,9]. We present the case of a 16-year old woman with congenital fibular hemimelia, associated to genu valgum, recurvatum, anteroposterior and medial instability of the right knee, who underwent corrective osteotomies for angular deformities and ACL reconstruction in one single surgical procedure.

2. Case report

Sixteen year old female with congenital fibular hemimelia of the right lower limb underwent tibial lengthening in 2006, leading to symptomatic genu valgum and recurvatum after surgery. In September 2009, the patient consulted at our center because of anteroposterior and medial instability of the right knee. Physical examination evidenced genu valgum of 24° with axial load and recurvatum of 24° with a range of motion of −10°–110° (Fig. 1). Preoperative physical examination showed a complete instability of the knee. The patient presented joint failure, positive anterior drawer and Lachman test. However, positive pivot shift test was only assessed under anesthesia. Medial instability due to persistent valgus deformity was also present. Full-length radiographs confirmed genu valgum of 24° and recurvatum of 24° (Fig. 2), and an MRI of the right knee revealed complete absence of ACL (Fig. 3). The final diagnosis of our patient was congenital fibular hemimelia associated to genu valgum and recurvatum secondary to tibial lengthening and anteromedial instability of the right knee due to the absence of ACL.

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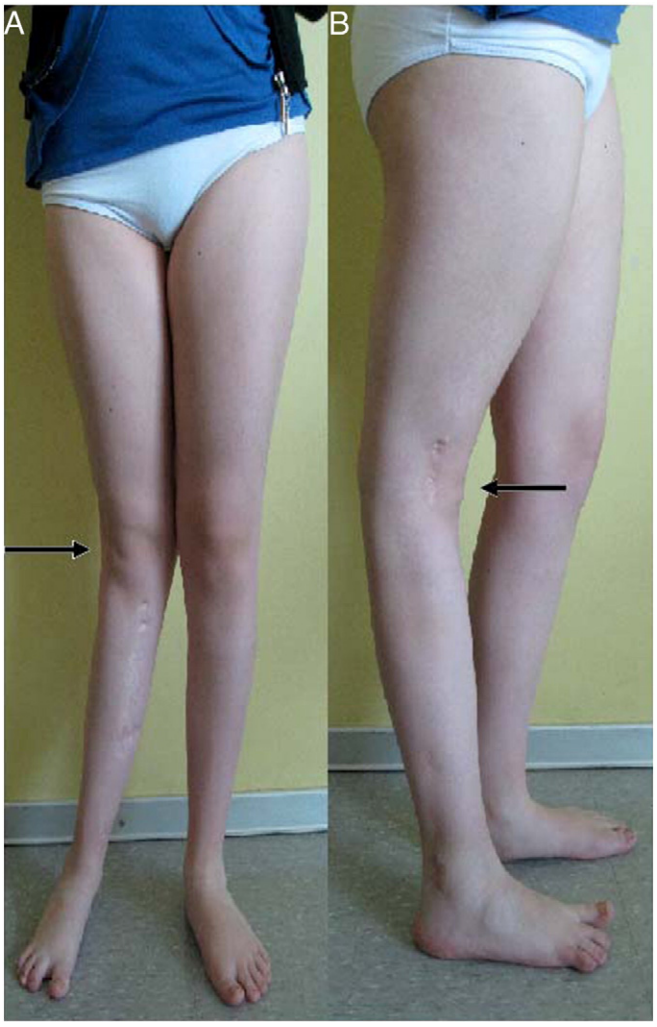

Fig. 1. Preoperative images. A. Genu valgum (arrow) of 24°. B Recurvatum (arrow) of 22°.

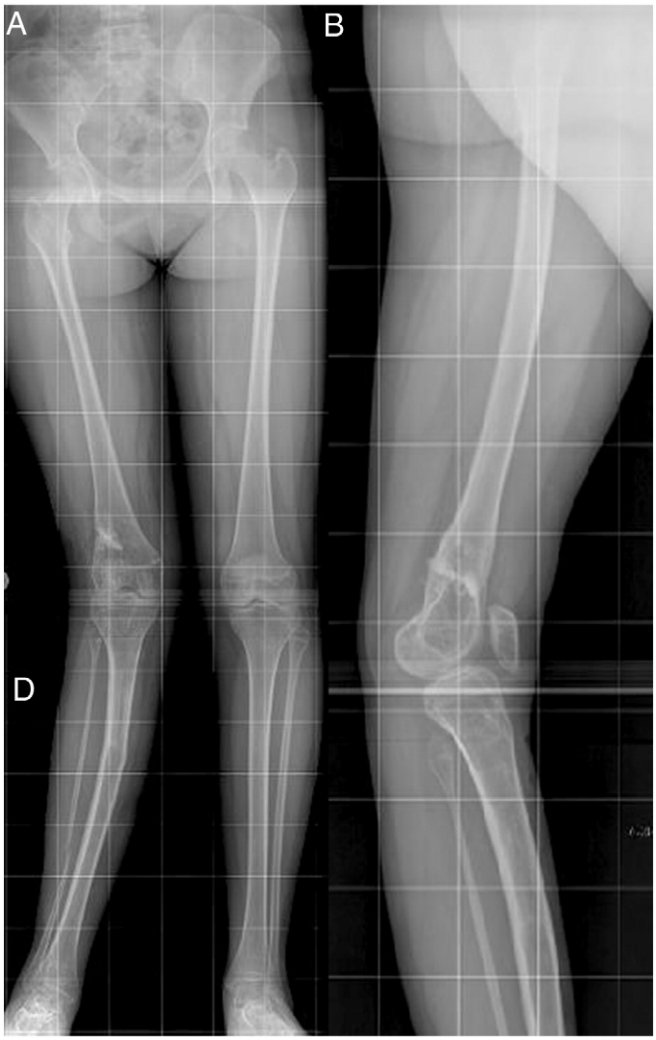

Fig. 2. A. Preoperative full length radiograph which demonstrates genu valgum of 24°. B. Preoperative lateral projection of right lower limb showing 24° of recurvatum.

3. Surgical technique

Preoperative planning basically included a misalignment test [10] and the respective CORAs were identified. The osteotomies were planned in order to correct the lateral distal femoral angle (LDFA), posterior distal femoral angle (PDFA), medial proximal tibial angle (MPTA) and posterior proximal tibial angle (PPTA). External fixation allowed progressive intraoperative correction in order to ensure perfect alignment and stable internal fixation with early weightbearing was achieved with an intramedullary nailing system. Percutaneous osteotomy of the femoral metaphysis was performed correcting valgus and recurvatum and stabilized with a retrograde unreamed femoral nail (UFN). Subsequently, metaphyseal–diaphyseal tibial osteotomy was realized to correct tibial valgus and stabilized with an antegrade unreamed tibial nail (UTN). Finally, and in the same surgical procedure, ACL reconstruction was performed with a quadriceps tendon allograft. Proximal bone block was performed with an interference titanium screw and distal fixation with a bioabsorbable scew. The tunnels were 10 mm wide, all arthroscopically guided, and drilled after intramedullary nailing. No special tips are mentioned, since the orientation of the femoral tunnel is oblique, from medial to lateral and away from the central notch were the nail lies. We could not recognize any anatomic structure compatible with an ACL. A very narrow notch was identified, which had to be opened. The physis was partially closed.

Immediately after surgery the knee was placed into a continuous passive motion (CPM) device. The patient was discharged with crutches and non weight-bearing was indicated for 30 days. During rehabilitation exercises were carried out to regain articular range of motion according to tolerance. After a month, weight-bearing was tolerable and isokinetic closed chain exercises were initiated. In the last postoperative control 12 months after surgery, clinical stability was confirmed along with a range of motion of −5–110°, and normal gait was evidenced without claudication nor clinical instability (Figs. 4 and 5). The measurement of ligament laxity using KT-1000 arthrometer showed an average difference of 2 mm compared with contralateral healthy limb.

4. Discussion

Fibular hemimelia is the most common occurring congenital disorder of the long bones [3,12], and is associated frequently with a partial or complete absence of ACL. ACL reconstruction in these patients is a rare procedure and according to different authors, surgical treatment should only be considered in patients with symptoms that interfere with their daily activities [4,6,7]. Congenital absence of ACL may lead to complications such as meniscal tears, clinical knee instability, tendency for knee subluxation [1,2] or even dislocation after tibial lengthening for fibular hemimelia, which was the case of our patient.

Fig. 3. Preoperative MRI of the right knee demonstrates complete absence of ACL, suggesting a congenital disorder.

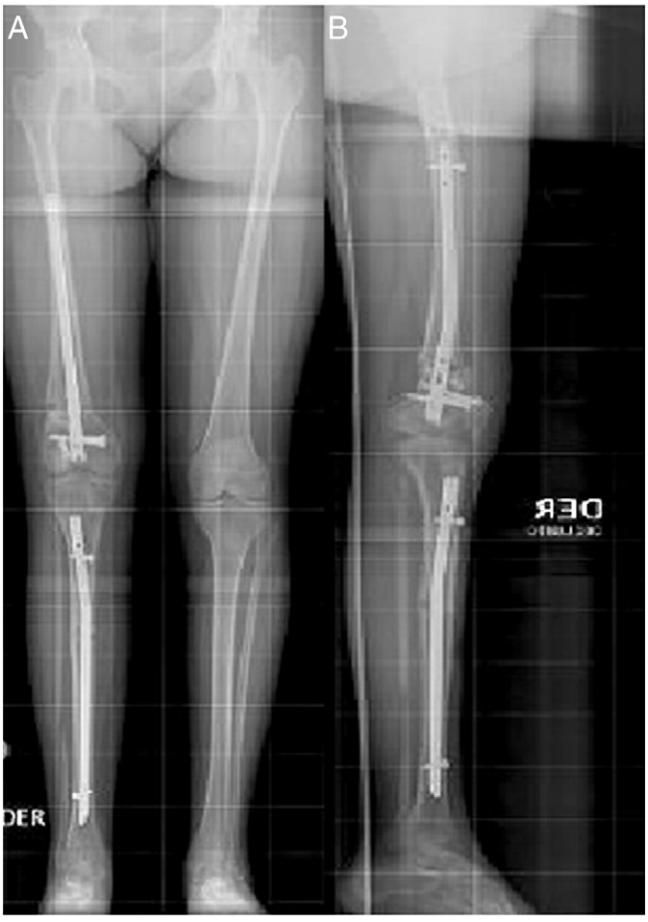

Fig. 4. Full-length postoperative radiographs demonstrate angular deformity correction in both AP (A) and lateral (B) projections.

Most studies in orthopedic literature involving congenital disorders of lower limbs associated to ACL deficiency give more emphasis to angular deformity correction and limb lengthening rather than referring to knee instability and ACL reconstruction. Katz et al. [7] report five cases of congenital dislocation of the knee in which ACL was reconstructed in three patients. Kaelin et al. [6] describe extra-articular reconstruction of ACL in one of six patients with congenital aplasia of ACL and leg length discrepancy. Gabos et al. [4] describe 4 patients with symptomatic congenital ACL deficiency who had undergone ACL reconstruction using allografts. Most authors agree on correcting angular deformities prior to ACL reconstruction, given better results over time [4]. Nevertheless, no study has published corrective surgery for angular limb deformity and ACL reconstruction in the same surgical procedure, which is what we decided to perform in our patient. The patient's symptoms — anterior and medial instability of the affected knee, associated to the angular deformities lead us to surgery. We believe that both procedures can be performed in a single time surgery without the risk of technical difficulties if done in the right order. Corrective osteotomy for angular deformity was done first, followed by ACL reconstruction. Both procedures were realized without difficulties, and the first step did not interfere with the second. The last clinical evaluation of our patient occurred 12 months after surgery, which evidenced a normal knee extension and 110° of flexion. The patient had normal gait cycle without signs of instability or claudication. KT-1000 arthrometer 6–12 months after the operation showed a difference of 2 mm compared to the other leg.

Few studies in orthopedic surgery describe clinical follow-up of patients with congenital disorders of the lower limb submitted to ACL reconstruction surgery. In the study of Gabos et al. [4], four patients were followed for an average period of 31 months, showing normal flexion, one patient had a 10° extension lag, and KT-1000 results indicated differences of 2–4 mm compared to the unoperated side [4]. These results are similar to those of our patient but differ in follow-up time. One of the weaknesses of our study is the short follow-up time. Since degenerative changes cannot be assessed 12 months after surgery, further evaluations must be made in our patient in order to obtain more certain results over time.

5. Conclusion

ACL reconstruction and corrective osteotomies for congenital fibular hemimelia performed in a single time procedure showed a good clinical outcome, restoring stability, function and a normal gait pattern in our patient. However, longer follow-up time is necessary to determine whether ACL reconstruction plays a role in preventing early onset of degenerative changes in the evolution of these patients.

6. Conflict of interest

Each author certifies that he has no commercial associations (e.g., consultancies, stock ownership, equity interest, patent/licensing arrangements, etc.) that might pose a conflict of interest in connection with the submitted article.

Financial disclosure: none.

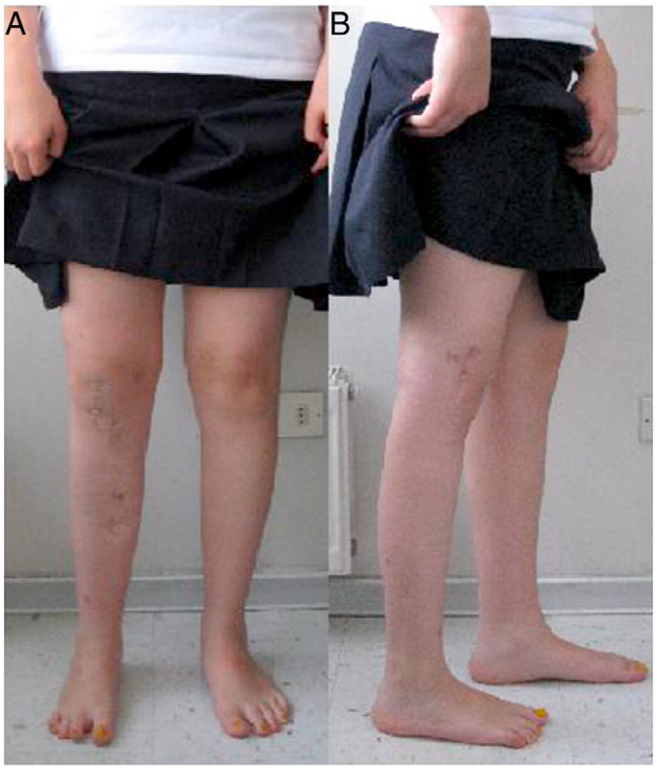

Fig. 5. Postoperative image. Both angular deformities, genu valgum (A) and recurvatum (B) of the right lower limb have been corrected.

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