

## Case Report

# Bilateral Absence of the Anterior Cruciate Ligament

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**Abstract:** We report a rare case of bilateral absence of the anterior cruciate ligament in an adolescent. The clinical picture, the diagnostic and therapeutic approach, the arthroscopic appearance and the follow-up are described. The available literature is discussed. **Key Words:** Anterior cruciate ligament—Arthroscopy—Congenital absence—Adolescence.

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**A**genesis of the anterior cruciate ligament (ACL) is a rare congenital disease that may be responsible for instability of the knee joint. This condition is frequently associated with other anomalies of the lower limb such as hypoplasia of the tibial intercondylar eminence and of the intercondylar notch.<sup>1-3</sup> Although the monolateral condition is relatively well documented in the medical literature, we found only 2 other published reports of bilateral absence of the ACL.<sup>3,4</sup> To our knowledge the only epidemiologic data on a large population come from the study by Giorgi.<sup>5</sup> In a large series of 2,500 knee radiographs, he described the morphologic variations of the intercondylar eminence, and reported only 1 case of aplasia of the eminence associated with sagittal laxity of the knee joint. Because of the rarity of this condition, the diagnostic and therapeutic approach are not standardized.

We describe a rare case of bilateral congenital absence of the ACL associated with other congenital anomalies of the knee joint in an adolescent. We describe the clinical picture, the diagnostic and ther-

apeutic approach, the arthroscopic appearance, and the follow-up.

### CASE REPORT

A 16-year-old boy was referred to us for bilateral instability of the knee joint, prevalent on the left side, that had been present for some years. Nothing abnormal had been noted at the boy's birth and his early somatic development was normal. His mother reported no abnormal intake of drugs during her pregnancy.

At the age of 11, following a moderate left knee sprain, the patient had noticed bilateral knee instability, especially on the left side, where it was associated with pain, swelling, and several episodes of giving way during daily activities.

On physical examination, the left knee presented slight joint effusion, a 15° recurvatum, a marked anterior laxity (positive Lachman and pivot shift tests and anterior drawer sign) and an instability on the frontal plane (laxity at the valgus ++ and varus + stress).

Standing radiographs revealed 8° valgus. Magnetic resonance imaging (MRI) showed a tear of the posterior horn of the lateral meniscus, an osteochondral loose body, reduction of cartilage on the weight-bearing area of the lateral femoral condyle, marked alteration of the Hoffa fat pad, a scarcely pronounced intercondylar notch, and absence of the ACL (Fig 1A). Hypoplasia of the tibial intercondylar eminence (Fig

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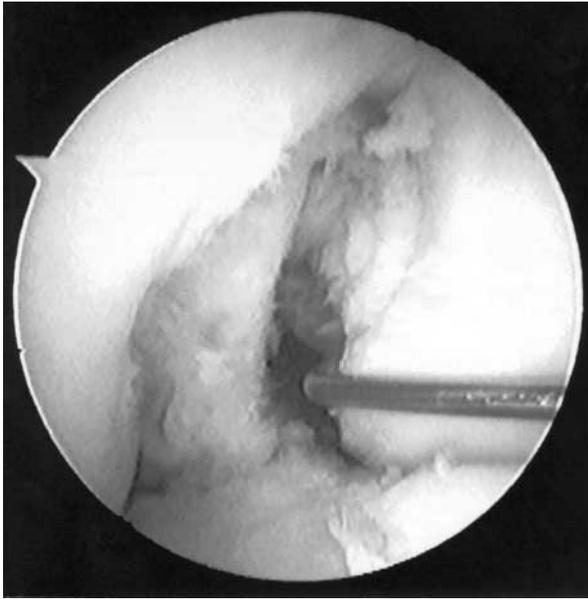


**FIGURE 1.** MRI of the left knee, gradient-echo sequences (TR = 500 msec, TE = 14 msec, FA = 90°). (A) Sagittal plane: absence of the ACL, patella alta, and hypoplasia of the intercondylar notch. (B) Coronal plane: hypoplasia of the tibial intercondylar eminence. (C) Axial plane: flattening of the trochlea femoralis.

1B) and flat trochlea femoralis (Fig 1C) with patella alta were also seen.

The right knee presented an anterior laxity that was

well tolerated. The MRI revealed a flat trochlea femoralis with a consequent lateral subluxation of the patella and absence of the ACL.



**FIGURE 2.** Arthroscopy of the left knee. The hypoplasia of the intercondylar notch and of the tibial intercondylar eminence are evident. The ACL is absent.

The decision to perform an arthroscopic examination of the left knee was taken to confirm the diagnosis and to treat the associated traumatic lesions. Arthroscopy confirmed the presence of a tear of the lateral meniscus extending to the popliteal hiatus and an osteochondral loose body of 1 cm of diameter, probably originating from the lateral femoral condyle where a cartilage lesion with the same shape and size was noticed. Hypoplasia of the medial meniscus, tibial intercondylar eminence, and the intercondylar notch, and the absence of the ACL (Fig 2) were also observed. A partial lateral meniscectomy performed and the loose body was removed.

The postoperative course was normal. On discharge, the patient was admitted to a rehabilitation program with isometric, isotonic, and isokinetic exercises, and the results were good overall. Two years later, pain is still absent, and there is no swelling or instability.

## DISCUSSION

Congenital absence of the ACL is a rare condition, first described in 1967.<sup>6</sup> This pathology may be associated with congenital absence of one or both menisci,<sup>4</sup> with lateral discoid meniscus,<sup>3</sup> with dysplasia of the intercondylar notch, of the femoropatellar joint,

and hypoplasia of the intercondylar tibial eminence,<sup>1-3</sup> or with a posterior cruciate ligament larger than normal.<sup>7</sup> This condition may also be associated with lower limb length discrepancy<sup>1</sup> and may predispose to sprains and dislocations of the knee joint.<sup>3</sup>

Congenital absence of the ACL is more frequently recognized during adolescence, even though studies have shown some cases discovered at an adult age.<sup>3</sup> Usually patients experience a giving-way sensation during daily activity (walking, playing) and sports and this is the reason why they come to the orthopaedic surgeon. Commonly, there is not history of trauma, but physical examination reveals joint laxity.

Our case history presents the onset of symptoms during adolescence. Joint laxity was well tolerated until a knee sprain provoked a meniscal tear and a chondral lesion of the left knee. After this episode, bilateral instability was reported. To explain the occurrence of the bilateral instability, we may hypothesize that the sprain broke a delicate equilibrium in which a major role was played by muscular control. The slight underdevelopment of the left thigh muscles from lack of use following the sprain presumably impaired the ability of the quadriceps muscle to counterbalance the laxity any longer. A second concomitant cause may be the altered mechanical force distribution on the right side.

The congenital origin of the condition we describe must be strongly suspected if not proved because of its bilaterality. The association with other joint anomalies, such as the lateral discoid meniscus and the dysplasia of the femoropatellar joint, the tibial eminence, and the intercondylar notch, as well as the complete absence of the ACL further proves it.

MRI showed both the absence of the ACL and its associated conditions. The advantages of MRI are the ability to obtain multiplanar sequences, to visualize hyaline cartilage, and to avoid the use of ionizing radiation.<sup>8</sup>

The therapeutic approach in the case of congenital absence of the ACL is not yet standardized. Some studies propose surgical treatment with ACL reconstruction alone<sup>1</sup> or associated with other extra-articular procedures to correct the associated anomalies.<sup>4</sup> Others suggest conservative treatment with good results.<sup>3,9</sup> Kaelin et al.<sup>10</sup> reported on 6 cases, 4 of which were treated conservatively and 2 surgically (1 a simple meniscectomy and 1 an extra-articular reconstruction according to Lemaire<sup>11</sup>). The authors reported a good result in the patient treated by meniscectomy alone and improved stability at clinical examination of the patient treated by ligament reconstruction. The

latter patient noticed no significant advantage after surgery. Tolo<sup>12</sup> described a case in which the major complaint was a femoropatellar instability in varus knee. The patient underwent a tibial valgus osteotomy followed by a patellar realignment. Ligament reconstruction was not required. Thomas et al.<sup>3</sup> described 10 patients, none of whom underwent a ligament reconstruction.

In our case, we treated only the intra-articular traumatic lesions because symptoms resulting from the meniscal tear and the cartilage lesion (i.e., pain, joint effusion) were prominent. Because the articular laxity had been well tolerated by our patient before the trauma, we maintained that adequate muscular reinforcement could be sufficient, once the causes of pain were eliminated. We are aware that anterior instability of the knee can induce a number of late lesions, such as meniscus tears and cartilage lesions,<sup>13</sup> and can also lead to the development of early arthritis. We are also aware that the meniscectomy, even if selective, can increase articular instability. To date, our patient had not reported any more episodes of giving way and carries out his day-to-day activities with no need of further treatment or braces. Even though studies suggest that joint instability resulting from the absence of the ACL can predispose to early arthritis,<sup>3</sup> there has been, to our knowledge, no prospective study regarding evolution of this condition.

We maintain that the therapeutic approach to this condition should take into account different elements, i.e., the patient's age, type and level of activity, and above all, the quality and amount of symptoms. We attribute a crucial role to the frequency and type of

giving-way episodes because we believe that a surgical approach should be reserved only for cases of relevant articular instability.

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