Isolated congenital aplasia of the anterior cruciate ligament treated by reconstruction in a 5-year-old boy

Agénésie isolée du ligament croisé antérieur traitée par ligamentoplastie à l’âge de 5 ans


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RESUMÉ

L’agénésie isolée du ligament croisé antérieur est une entité rare. Elle est exceptionnellement symptomatique. Nous rapportons le cas d’un garçon de 5 ans dont l’instabilité du genou a été traitée par ligamentoplastie selon la technique de Clocheville. À 5 ans de recul, le résultat fonctionnel est satisfaisant, le patient est asymptomatique et pratique le sport. Il n’a présenté aucune complication hormis une limitation de la flexion à 130°. Une IRM a permis de montrer la bonne intégration du greffon. L’instabilité antérieure du genou chez l’enfant entraîne une limitation des activités physiques. Elle est à l’origine de lésions méniscales et d’une arthrose précoce. La technique de Clocheville est adaptée au jeune enfant qui présente une instabilité du genou, qu’elle soit d’origine traumatique ou congénitale.

Mots clés : Ligament croisé antérieur, agénésie, enfant, autogreffe.

ABSTRACT

Agenesia of the anterior cruciate ligament is a rare entity that is exceptionally symptomatic. We report a case observed in a 5-year-old boy who presented an unstable knee, treated by Clocheville ligamentoplasty. At 5 years of follow-up, the functional outcome was satisfactory. The patient was symptom-free and participated in sports activities. He did not develop any complications except flexion limited to 130°. Magnetic resonance imaging demonstrated good integration of the autograft. Anterior instability in the child limits physical activity and can cause meniscal injury and early osteoarthritis. The Clocheville technique is adapted for pediatric patients with knee instability caused by a congenital defect or injury.

Key words: Anterior cruciate ligament, aplasia, child, autograft.
INTRODUCTION

Aplasia of the anterior cruciate ligament (ACL) is rare in children. Thomas et al. [1] have reported the largest series found in the literature: 12 cases. Generally speaking, an associated malformation is found, a congenital short femur or a congenital luxation of the knee. Isolated absence of the ACL is exceptional. In 1956, Giorgi [2] found a single case of anomalies of the intercondylar eminence visible on x-ray in a series of 2500 knees.

The incidence of isolated aplasia is unknown since most of the time there is little clinical evidence. This malformation is well tolerated and therefore there is little need for surgery in childhood. Furthermore, it is often preferable to wait until skeletal maturity before proposing ligamentoplasty so as to prevent epiphysiodesis.

We report the exceptional case of isolated aplasia, symptomatic beginning at the age of 3 years, which required early treatment with ligamentoplasty.

OBSERVATION

The patient was a boy who had consulted at the age of 3 years 8 months for his left knee giving way, causing frequent falls. He presented no personal or familial history and the examination revealed no sign of constitutional hyperlaxity. The clinical exam showed a positive Lachmann test with displacement and an anterior drawer greater than 10 mm compared to the contralateral side. The lower limbs were not of equal length, with a difference of 10 mm, confirmed by a knee x-ray (involved side longer). MRI demonstrated complete agenesia of the ACL, the anterior tibial spine, and hypoplasia of the medial meniscus (fig. 1A and fig. 1B).

Given the invalidating unstable knee, we indicated surgical treatment. The child was operated on at the age of 5 years and 2 months and then was followed up regularly.

SURGICAL TECHNIQUE

We performed a Clocheville ligamentoplasty developed by C. Bonnard in 1991 and adapted to arthroscopy by Robert and Bonnard [3].

The graft was harvested from the central third of the patellar tendon with a widened tendon-periosteal band on the anterior tibial tuberosity and a widened tendon-periosteal band on the patella. The femoral tunnel was approached from outside to inside using a 90° arthroscopic sight placed in the posterior part of the intercondylar groove, above the femoral growth cartilage. A guide-pin was placed with arthroscopic and lateral radiological guidance. Drilling was done with a 6-mm auger cutting device. A groove was made on the anterior intercondylar area of the tibia by medial parapatellar arthrotomy.

The graft had been tacked beforehand with Vicryl 20 thread and was then introduced into the joint through the femoral tunnel from outside to inside. It was then positioned in the tibial groove. An Acufex RCI® 7 × 25-mm interference screw was used for femoral fixation (fig. 2A and fig. 2B). Tibial fixation was provided by two absorbable Mitek G2® suture anchors and three Vicryl 20 reinforcement sutures on the cartilaginous layer of the anterior intercondylar area.

The intraoperative stability and isometry were satisfactory. The flexion obtained was 140° with complete and symmetrical extension. An antalgic femoral catheter was placed at the end of surgery.

Postoperative support was provided by a resin knee brace with the knee flexed at 15° for 1.5 months. No rehabilitation was necessary.

The patient was examined at the 5-year follow-up. Physical activities were resumed at the 5th month after surgery at a higher level than before surgery. The patient participated in sports at school as well as playing tennis and basketball. He presented no pain or signs of instability.

He presented limited knee flexion compared to the contralateral side on the order of 10° (130 versus 140°). The Lachman test showed a delayed stop of 5 mm compared to the contralateral side. No complication was noted in terms of the graft harvest.

The Tegner activity score was 7/10 and the Lysholm was 99/100. The knee was classed IKDC C because of the flexion deficit. Laxity was compared with the Lerat device using a 3-kg weight (12% of body weight) [Lerat et al. [4]]. It found a 4-mm anterior drawer in the medial compartment versus 1 mm on the healthy side, and a 7-mm anterior drawer in the lateral compartment versus 3 mm on the healthy side. Therefore, there was a 3-mm medial differential and a 4-mm lateral differential. Weightbearing front and lateral plain films were normal (fig. 3A and fig. 3B). The knee x-ray showed 10 mm lengthening on the operated side, particularly in the femur (8 mm). This inequality had not progressed in 5 years. It was well tolerated and was not corrected with insoles. The mechanical femorotibial axis was normal and symmetrical. The follow-up MRI done 4 years after surgery showed complete LCA reconstruction with graft integration on the femur and tibia (fig. 4A and fig. 4B). No chondral or meniscal lesion was found.

**DISCUSSION**

Aplasia of the anterior cruciate ligament is rare and its paucity and frequent lack of symptoms are mentioned in all publications on this topic. This abnormality is one of the true malformations (occurring during the period of
embryogenesis) in contrast with congenital deformations (taking place during the fetal period) [Dejour et al. [5]].

From an embryological point of view, the ACL is a formation derived from intra-articular mesenchyma that appears, depending on the authors, between the 7th and 10th week of fetal development by condensation and differentiation [Dejour et al. [5], Ellison et Berg [6]]. It appears in the anterior position and migrates to the posterior position at the same time that the femoral condyles differentiate. The two cruciate ligaments and the menisci develop from the same mesenchyma. They appear well before the development of the joint space. Migration of the cruciate ligaments participates in the development of the articulatory surfaces. Therefore, the development of the spinous processes of the tibia ceases when there is not traction from the ACL, which explains that in cases of agenesia, the tibial intercondylar eminence is absent. According to this principle, the later movement of the articulatory surfaces is apparently predetermined by the early appearance of cruciate ligaments. Its function as a central pivot and the shape of the femoral condyles are closely related. A simple rotational movement is converted into a much more complex movement because sliding and rolling are associated.

These notions in embryology explain the articulatory anomalies observed in the case described herein (agenesia of the anterior spinous process of the tibia and hypoplasia of the medial meniscus).

Agenesia of the ACL is often associated with other lesions such as a congenital short femur [Johansson and Aparisi [7], Kaelin et al. [8]] or a congenital luxation of the knee [Katz et al. [9], Ferrone [10]]. However, the high incidence of this association is debated [Thomas et al. [1]]. The familial origin of isolated agenesia of the ACL has recently been discussed [Frikha et al. [11]].

The functional repercussions of isolated ACL agenesia are exceptional. Indeed, these subjects, first as children and then as adults, for the most part have a normal life and participate in sports. Noble [12] described the case of an autopsy discovery in an 81-year-old subject with regular medical follow-up. He had never presented an episode of knee instability or pain. Likewise, in the study conducted by Johannsson and Aparisi [7], a female patient was followed until the age of 60 years with no functional complaints. Kaelin et al. [8] conducted a clinical, radiological, and arthroscopic study on six patients with unequal lower limb lengths associated with clinical knee instability. They discovered four cases of ACL agenesia. None of the patients complained of instability. Five patients out of six presented hypoplasia of the intercondylar groove and flattening of the tibial eminence. Frikha et al. [11] reported the cases of five patients. Three of them presented osteoarthritis progression, two of whom required two valgus tibial osteotomies and one total knee replacement.

Since the majority of patients presenting ACL agenesia have few or no symptoms, the number of cases of ligamentoplasty reported in the literature is low [Dejour et al. [5], Kaelin et al. [8]].

Conservative treatment for ACL absence or tear was the norm in children because of the possibility of femoral or tibial epiphysiodesis [Lipscomb and Anderson [13], Kocher et al. [14], Koman and Sanders [15]]. However, conservative treatment of traumatic ACL tear can end in poor results, as shown by many studies. Unstable knee symptoms are frequent and require patients to limit their physical activities. Furthermore, anterior instability foretells meniscal

Fig. 4. – a) Postoperative MRI at 10 years of age. Axial view. Graft in place. No epiphysial cartilage anomalies. b) Sagittal view. Graft in place.
and chondral lesions. It leads to early osteoarthritic degeneration of the knee [Anderson [16], Kannus and Jarvinen [17], McDaniel and Dameron [18], Mizuta et al. [19]]. Janarv et al. [20] report a series of 28 patients from 10 to 15 years of age with total rupture of the ACL due to injury treated conservatively with 3 months of rehabilitation. Eight of them chose this treatment because they were symptom-free, but at the cost of a reduction in physical activities. Another study, conducted by Kannus and Jarvinen [17], showed poor results stemming from the conservative treatment in total traumatic ruptures of the ACL. At a mean of 8 years of follow-up, all patients had invalidating instability and four out of seven presented signs of degeneration on x-ray.

From an experimental point of view, crossing the tibial or femoral physis exposes the patient to a risk of partial epiphysiodesis proportional to the area of the growth plate crossed [Makela et al. [21]]. An over-the-top position presents a risk of valgus misalignment because it involves the perichondral membrane located around the growth cartilage, as demonstrated by Seil et al. [22] on sheep. Extra-articulatory reconstruction was used with the objective of restoring stability without damaging the epiphysial cartilage. It seems that the isometric defect of this technique can be the source of instability episodes and meniscus lesions [McCarroll et al. [23]]. In addition, the risk of valgus seems high because of lateral hyperpressure [Kocher et al. [14], Koman and Sanders [15]]. McCarroll et al. [23] presented ten patients treated with lateral tenodesis using a band of fascia lata. Nine patients later presented episodes of instability of the knee and four required later surgical revision with intra-articulatory ligamentoplasty.

Several ACL intra-articulatory reconstruction techniques that prevent epiphysial cartilage perforation have been described. Parker et al. [24] reported excellent results in positioning the pes anserinus tendons over the top of the femur and in a groove formed in the tibial epiphysis. The anterior position of the graft in the tibia presents a risk of conflict with the intercondylar groove increased by the position under the medial meniscus horn. Brief [25] proposes reconstruction with the semitendinosus and gracilis tendons on the tibia with no bone drill holes, passed under the intermeniscal ligament and attached on the lateral cortex of the femoral head. Anderson [26] attaches the four hamstring ligaments in an epiphysial tunnel of the anterior intercondylar area of the tibia and in a femoral epiphysial tunnel. This technique seems to be the most anatomical and does not cross the epiphysial cartilage.

Fixing the graft with an interference screw in the tibial tunnel presents a major risk of epiphysiodesis [Kocher et al. [14], Koman and Sanders [15]]. Therefore, this tibial fixation is most often provided by an extracortical device (button, staple, or suture supported by a screw). This type of fixation can be the source of residual laxity.

The Clocheville technique uses a graft harvested from the patellar ligament. This technique is anatomical for the tibia but not for the femur. The residual laxity in our patient can be partially explained by this isometric defect. Robert et al. [27] showed the results of 15 children aged from 9 to 13 years who presented ruptures resulting from injury to the ACL treated with this technique, with good clinical and radiological results. Slowed growth in the lateral half of the femoral physis was noted, which resulted in two cases of valgus knee, 4º and 7º, at the end of growth, with no functional consequences. One case of 7º tibial varus was observed, related to lateral hyperpressure. Finally, a loss of tibial slope was noted in two cases, in relation with the technical circumstances and the use of staples, with no clinical repercussions. Two cases of limited knee flexion were also reported. In our series, we had knee flexion limited by only 10º compared to the contralateral side.

We believe that there is a surgical indication in children and adolescents who present chronic anterior unstable knee, whether it be congenital or the result of injury, as long as it is symptomatic and invalidating in daily life. Reducing physical activities in children is difficult to observe and is difficult for the child to accept. In our case, the indication for surgery was chosen because of the functional discomfort in daily activities and the risk of progression to degeneration.

The objectives of this early surgery in the child are the same as for adults: stabilization of the knee and protection of the meniscus and cartilage. However, it presents risks of specific complications because of the presence of the growth plates. In the literature, we found no reports on cases of ACL agenesia treated with surgery in such a young child.

**CONCLUSION**

Isolated aplasia of the ACL is rare, most often symptom-free. Surgical treatment should be reserved for children presenting invalidating knee instability and/or meniscal complications for which orthopedic treatment is not adapted. The Clocheville technique is adapted to the young child because it does not cross the growth cartilage.

**References**


