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Case Report

Approach to Manage Congenital Absence of Anterior Cruciate Ligament in a 13 years old Patient – A Rare Case Report of Eastern India

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ABSTRACT

Congenital absence of anterior cruciate ligament is highly uncommon occurrence. It has since been documented as a standalone anatomical entity or, more frequently, in conjunction with other congenital anomalies. Surgical treatment for this patient population has only been reported in very few cases. In this article, we share our experience in managing a case of unilateral congenital deficiency of anterior cruciate ligament (ACL) in a 13 years old female patient by physeal sparing arthroscopic ACL reconstruction, using All-inside technique.

Introduction

With an incidence of only 0.017 per 1,000 live births (1), congenital anterior cruciate ligament absence is a very uncommon disorder. In 1956, Giorgi's radiography investigation was the first to characterize it (2). Anterior cruciate ligament (ACL) absence at birth may occur alone, or more frequently in combination with other congenital conditions affecting the lower extremities (3). In 1997, Park et al. published the first case with an absent anterior cruciate ligament (4,5). The authors of that study didn't undertake any reconstruction; they only did arthroscopic assessment. Considering the rarity of the problem, there is no agreement on the best therapeutic strategy, while the majority of research have found that non-surgical methods are the best care even in the early stages of degenerative joint disease. There are few reports of this patient population receiving surgical therapy. Congenital ACL deficiency is caused by a genetic disorder or a birth defect. The exact cause of congenital ACL deficiency is not fully understood, but it is thought to be caused by a defect in the genes that control the development of the ACL, whereas traumatic ACL injury is caused by a sudden

twisting or pivoting motion of the knee. This can happen as a result of a fall, sports injury, or car accident. The symptoms of congenital ACL deficiency and traumatic ACL injury are similar. These symptoms include knee instability, pain, swelling, and locking or catching of the knee. However, the symptoms of congenital ACL deficiency may be present at birth or early childhood, while the symptoms of traumatic ACL injury usually occur after a sudden twisting or pivoting motion of the knee. Congenital ACL deficiency is usually present at birth or early childhood. Traumatic ACL injury can occur at any age, but it is most common in young adults. Non-surgical treatment may be an option for some people with congenital ACL deficiency. This treatment may include physical therapy and bracing. However, surgery is usually necessary to reconstruct the ACL in people with congenital ACL deficiency. Surgery to reconstruct the ACL is also usually necessary for people with traumatic ACL injury. The prognosis for people with congenital ACL deficiency and traumatic ACL injury is good in most cases with appropriate treatment. With surgery and rehabilitation, most people are able to return to their pre-injury level of activity. It is important to note that there is some overlap between the symptoms of congenital ACL deficiency and traumatic ACL injury. For example, a person with traumatic ACL deficiency may also experience knee instability. However, the presence of knee instability in a young child or adolescent should raise suspicion of congenital ACL deficiency.

The reconstructed ACL in a congenital ACL deficiency patient can affect the knee biomechanics in the presence of other associated congenital anomalies in a number of ways. First, the reconstructed ACL may not be as strong or as effective as a normal ACL. This is because the tissues used to reconstruct the ACL are not as strong or as elastic as the native ACL. As a result, the reconstructed ACL may be more likely to tear or rupture, particularly if the patient participates in high-impact activities. Second, the reconstructed ACL may not be able to fully compensate for the other congenital anomalies that are present in the knee. For example, if the patient also has ligament laxity or malalignment, the reconstructed ACL may not be able to prevent the knee from becoming unstable. Third, the reconstructed ACL may not be able to fully restore the normal biomechanics of the knee. This is because the reconstructed ACL is not attached to the same bones as the native ACL. As a result, the reconstructed ACL may not be able to provide the same level of stability and control as a normal ACL. Overall, the reconstructed ACL in a congenital ACL deficiency patient can affect the knee biomechanics in a number of ways. The severity of these effects will depend on the severity of the deficiency, the presence of other associated congenital anomalies, and the level of activity of the patient. Several studies have showed that ACL reconstruction significantly improved knee stability in patients with hypoplastic femoral condyles. However, the reconstructed ACL did not fully restore the normal biomechanics of the knee. Patients with hypoplastic femoral condyles continued to have increased stress on the medial compartment of the knee, even after surgery. Here are some specific examples of how the reconstructed ACL can affect the knee biomechanics in the presence of other associated congenital anomalies: In patients with congenital genu valgum (knock knees), the reconstructed ACL may not be able to prevent the tibia from rolling in during weight bearing. This can lead to increased stress on the medial compartment of the knee, which can increase the risk of osteoarthritis. In patients with congenital genu varum (bowlegs), the reconstructed ACL may not be able to prevent the tibia from rolling out during weight bearing. This can lead to increased stress on the lateral compartment of the knee, which can increase the risk of osteoarthritis. In patients with congenital ligament laxity, the reconstructed ACL may not be able to provide enough stability to prevent the knee from becoming unstable (17). This can increase the risk of knee pain and injury. These findings suggest that ACL reconstruction can be an effective treatment for instability in patients with hypoplastic femoral condyles. However, patients may still be at increased risk of osteoarthritis in the medial compartment of the knee. Here, we present a case of unilateral congenital ACL absence treated with physeal sparing arthroscopic ACL reconstruction, using All-inside technique in a 13-year-old girl. We also address radiological evidence that may help explain this absence and potential surgical challenges.

Case Report:

A 13-year-old female patient presented to our outpatient department, with pain and instability around the left knee joint. She never had any history of injury. Her pain was sporadic, happening around every one to two months. Onset happened when playing sports, climbing stairs, or walking on uneven terrain. The range of motion was between 0 and 140 degrees, and a physical examination revealed no tenderness or swelling. She had tested positive for grade 3- Lachman's test and anterior drawer test, pivot shift test and unaltered varus and valgus stability. There is no discernible difference in limb length. Left lateral radiographic view revealed a reduced posterior tibial slope, hypoplasia of the left lateral tibial spine and lateral femoral condyle, and a narrow intercondylar notch on anterior-posterior simple radiographs.

Standing anteroposterior and lateral radiographs demonstrate decreased posterior slope and hypoplasia of the left tibial spine. Hypoplastic lateral femoral condyle and a depressed lateral tibial plateau, Magnetic resonance imaging (MRI) reveals non visualization of the anterior cruciate ligament (Figure 1-3). The lateral femoral condyle appeared to be relatively smaller, the intercondylar notch was narrower and smaller in both height and width, the tibial spines were hypoplastic, and the intercondylar notch was quite stenotic. Buckling of posterior cruciate ligament was noted. All the other ligaments and both menisci of the left knee seemed normal.

The coronal image reveals narrow intercondylar notch and hypoplasia of the medial femoral condyle. Sagittal image of MRI scans of the left knee reveal the absence of the anterior cruciate ligament.A grade 3 Lachman test and a 2+ pivot shift test were found during an examination conducted while the patient was under anesthesia for planned cruciate ligament reconstruction surgery. A negative posterior drawer was present. Stability of the collateral ligaments was intact.Patellar hypermobility was present. An severely stenotic intercondylar notch, hypoplastic tibial eminences, and the absence of ACL were all seen during arthroscopic examination. The

cartilage in all three compartments was healthy. To expand the unusually narrow femoral intercondylar notch, notch plasty was used. For physeal-sparing ACL reconstruction procedure, a quadrupled semitendinosus autograft was used. Fluoroscopy was performed to check that the pediatric ACL femoral guide was positioned entirely within the epiphysis and was aimed at the center of the ACL femoral footprint (Figures-4-7).

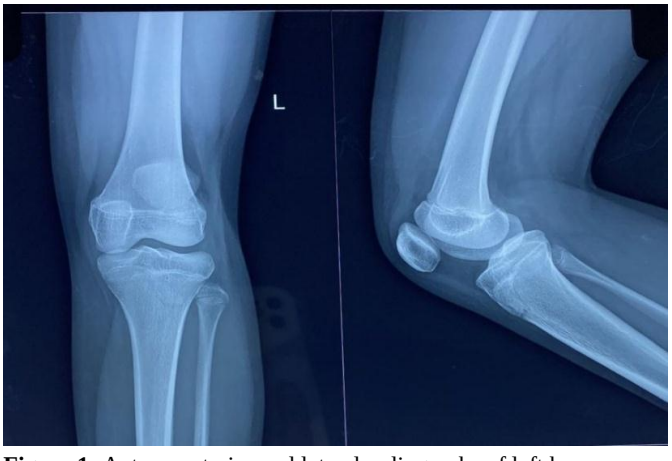


Figure 1: Anteroposterior and lateral radiographs of left knee

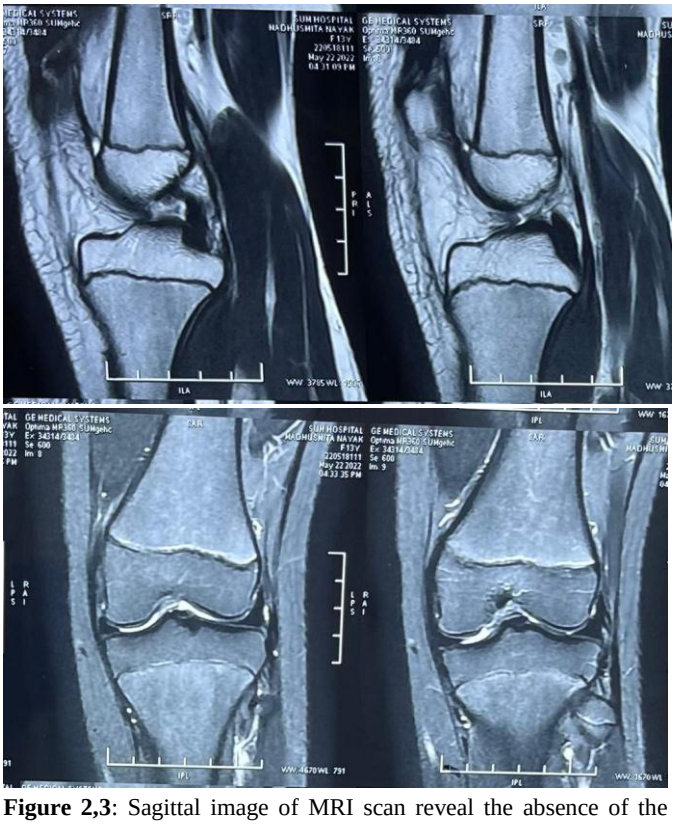
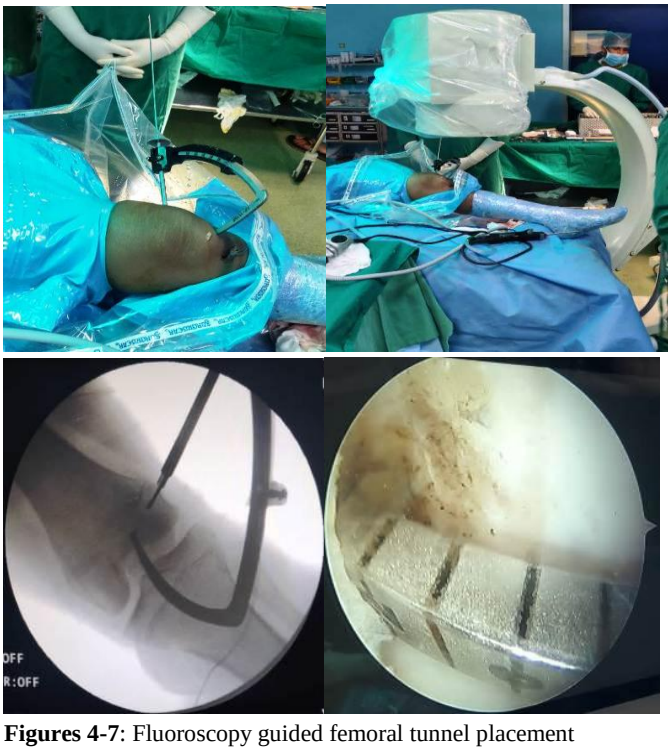


Figure 2,3: Sagittal image of MRI scan reveal the absence of the anterior cruciate ligament, coronal image reveals narrow intercondylar notch and hypoplasia of the medial femoral condyle



Figures 4-7: Fluoroscopy guided femoral tunnel placement

A 20mm retrograde hole was drilled into the femoral tunnel, and a suture was then inserted to allow subsequent graft transit. The tibial tunnel was drilled in the middle of the ACL tibial footprint using a conventional ACL guide. The angle of the guide was extended to roughly 70 degrees. By creating a vertical tunnel through a more central piece of the tibial physis by using the tibial guide at this high angle, less of the physis cross section was affected. On the tibial footprint of the ACL, a 10 mm-diameter tibial footprint was made. The knee was almost fully extended when the tibial fixation was finished. After the knee joint's stability and normal range of motion were established, the reconstruction was finished (Figure-8).

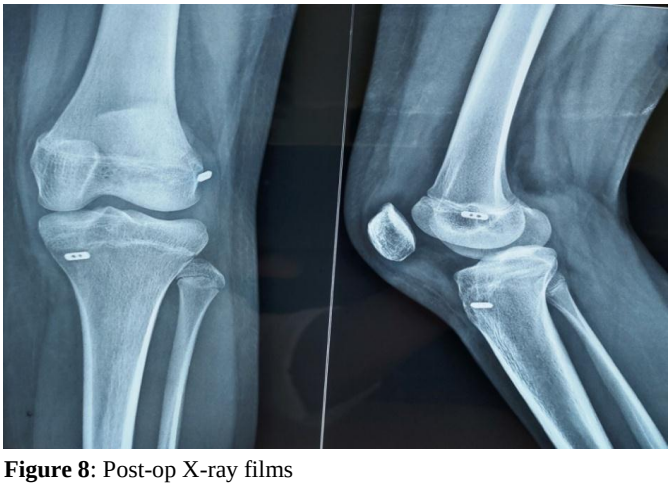
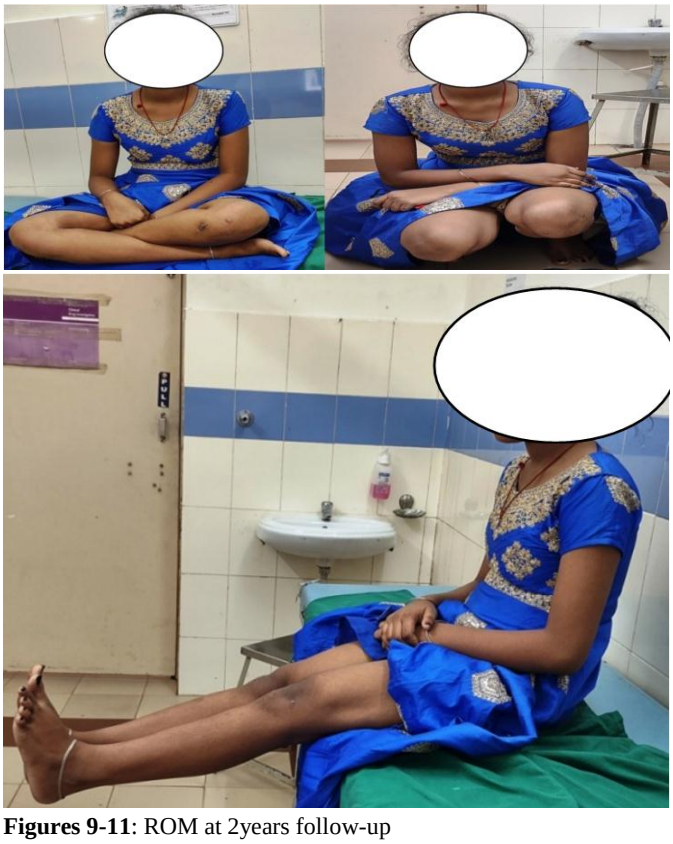


Figure 8: Post-op X-ray films

The patient underwent postoperative rehabilitation that was structured. For four weeks following surgery, the patient was required to wear a knee brace to limit knee flexion from 20° to 70° (for 24 hours a day except the hours dedicated to restore motion). After surgery, range-of-motion exercises were started both actively and passively. Within 3 weeks, full range of motion was recovered. For the first six weeks following surgery, patients followed a program of gradual weightbearing while using crutches as support. There was overall improvement without any indications of instability at the last follow-up. She had acheived complete range of motion (Figures-9-11).



Figures 9-11: ROM at 2years follow-up

The pivot shift and Lachmen were both negative. Postoperative radiographs show a vertical transphyseal tunnel in the tibia and a physeal-sparing tunnel in the lateral femoral epiphysis. The semitendinosus autograft has an adjustable loop button for both femoral and tibial fixation.

Discussion

Congenital cruciate ligament absence is an extremely uncommon disorder. It's been described as either a singular anomaly or as a component of a syndrome complex. The blastemal tissue of the interchondral disc directly condenses and differentiates to create the structures of the knee during the seventh to tenth week of intrauterine development (6).The knee resembles that of an adult by around the eighth week (O'Rahilly stage 22) (7). According to statistics, there are 0.017 cases of dysplasia andor ACL absence for every 1000 live births (1).It's important to remember that no individual PCL deficiencies have been documented (6).The intercondylar notch and tibial spines do not develop in the absence of the ACL because they are not provided with growth stimulus (8).Manner et al classified ACL dysplasia into three main types (9). Although the cause of this condition is unknown, genetic factors are probably a component because monozygotic twins (10) and numerous family members of same family (11) have been documented to have it.Despite displaying positive results on objective instability tests, these patients frequently learn to live with their congenital knee problem and rarely complain about the same (12).According to a study published in the journal The American Journal of Sports Medicine, 80% of patients with congenital absence of the ACL had stable knees without surgery. The factors that were most associated with stability were the strength of the muscles around the knee and the shape of the knee joint.It is important to note that not all congenitally deficient ACL knees are stable. Some patients may experience instability, pain, or other problems, even if they do not have surgery (18).

The most well-known and readily visible of the radiographic findings that could point to a diagnosis of congenital absence of the anterior cruciate ligament is hypoplasia of the tibial spine (13-15).The intercondylar notch's height and width narrowing, which can be verified on a tunnel view radiograph or Magnetic resonance imaging , is the second crucial indicator (9,13).Only in cases where the patient experiences symptomatic instability,anterior cruciate ligament reconstruction is recommended. For patients who have symptomatic knee instability and congenital ACL absence, Gabos and colleagues recommended reconstructive surgery (14). Because of the changed architecture of the knee, reconstruction of anterior cruciate ligament in these patients become more challenging (15).It is debatable how best to handle ACL injuries in people who have true skeletal immaturity. Previous reports of surgical procedures using extraarticular or direct suture techniques had dismal outcomes (16). Despite the development of numerous techniques, debate about the approach that offers the patient the least danger while still producing the best results persists. The reconstruction technique described here aims to offer graft stability and isometry without running the risk of negative physeal effects. During early follow-up, this intraarticular physeal-sparing technique permits graft fixation at femoral and tibial sites, enabling anatomic reconstruction while lowering the risk of growth abnormalities in skeletally immature patients (19-24).

Conclusion

Congenital absence of anterior cruciate ligament is an extremely rare anomaly that is frequently misdiagnosed because it can go for a long time without showing any symptoms while also showing signs only following minor trauma. Conservative treatment is gold standard for majority of the patients, and reconstruction of anterior cruciate ligament is planned for those with symptomatic instability. We come to the conclusion that in carefully chosen patients with congenital ACL deficit, reconstructive surgery is an effective choice for restoring knee stability and function. It is likely that obtaining stability with an ACL reconstruction that spares the physis at a younger age, before the skeleton has fully developed, may result in less degenerative changes in the knee. In the current article we share our experience in managing a patient whose symptoms were relieved following arthroscopic ACL reconstruction. Long-term monitoring

of this patient is necessary to chart her clinical progress and look for signs of instability or the onset of osteoarthritis.

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Conflict of Interest

None

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None

Ethical Approval

As this is a case study so no ethical approval was required but written consent was approved from the patient and patient's family to publish this work.

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