Section: Radiodiagnosis

Case Report

BILATERAL CONGENITAL ABSENCE OF THE ANTERIOR CRUCIATE LIGAMENT ASSOCIATED WITH DEVELOPMENTAL DYSPLASIA OF THE HIP AND DYNAMIC KNEE INSTABILITY: A MULTIMODALITY IMAGING CASE REPORT (DOUBLE CRUSH SYNDROME)

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ABSTRACT

Congenital absence of the anterior cruciate ligament (ACL) is an exceptionally rare developmental anomaly. We report a rare case of an infant presenting with bilateral knee instability associated with developmental dysplasia of the hip. Multimodality imaging including radiographs, dynamic ultrasonography, and magnetic resonance imaging demonstrated bilateral congenital absence of the ACL with characteristic secondary morphologic features. Early diagnosis is crucial to ensure appropriate conservative management and to avoid misdiagnosis as traumatic ACL injury.

Keywords: Congenital ACL absence; Knee instability; Pediatric orthopedics; MRI; Dynamic ultrasound.

INTRODUCTION

Congenital absence of the cruciate ligaments is an uncommon developmental anomaly, with an estimated prevalence of approximately 0.017 per 1000 live births.[1] During normal fetal development, the cruciate ligaments arise from the articular interzone between 7 and 10 weeks of gestation. The posterior cruciate ligament (PCL) typically develops first,[2] followed by formation of the anterior cruciate ligament (ACL).

This condition is frequently associated with other developmental abnormalities of the lower limb. These may include femoral shortening, underdevelopment of the intercondylar notch, hypoplasia of the tibial intercondylar eminence, and the presence of a discoid meniscus.[3-5]

In this report, we describe a case of congenital absence of the ACL associated with developmental dysplasia of the hip and dynamic instability of the knee, along with a brief review of the relevant literature.

Case Report

K/c/o congenital dislocation of right hip pavlik harness given for management. Patients parent c/o left knee instability for 1 week. Advised for x-ray of both knees and right knee shows posterio-lateral subluxation of femur in relation to tibia (figure 1). left knee joint xray shows mild posterior subluxation of femur in relation to tibia (figure 2). And for further investigation USG was done at SRMC. On static ultrasound right knee shows (figure 3) distal femur and proximal tibia are in alignment and on dynamic ultrasound repersentative still frame from dynamic ultrasound showing transient subluxation of proximal tibia., not seen in alignment with distal femur. on static ultrasound (figure 4) distal femur and proximal tibia are in alignment and on dynamic ultrasound repersentative still frame from dynamic ultrasound showing transient subluxation of proximal tibia. ., not seen in alignment with distal femur. Bilateral hips shows no subluxation. Hence patient came for MRI knee.

On MRI right knee shows (figure 5) on sequences in sagittal pdfs / t2wi images absence / non

visulaisation of ACL fibres, empthy intercondylar notch and no continuous low signal band. Figure 6 on sequence T2W in coronal plane (confirmatory view) no oblique ACL band, intercondylar notch anatomy, tibial spine morphology and coronal T2 image confirms non-visualization of ACL. Figure 7:- axial plane on T2/PDFS shows (notch and tibial spine) very important in congenital abcenmce shows hypoplastic intercondylar notch, abscent ACL footprint, tibial eminence changes, demonstarate shallow intercondylar notch. Figure 8:- secondary signs of PCL in seq: T2WI / PDFS in sagittal plane shows hypoplastic medial and lateral meniscus.

On MRI left knee joint shows there is significant posterior subluxation of the femur in relation to the tibia. The distance between the posterior margin of the medial femoral condyle and medial tibial condyle is 1.5 cm. Mild lateral subluxation of the distal femur in relation to the tibia also noted. The anteroinferior aspect of the medial femoral condyle is seen to abut the posterosuperior aspect of the medial tibial condyle. There is significant reduction in the joint space in between the anteroinferior aspect of the lateral femoral condyle and posterosuperior aspect of the lateral tibial condyle. The distal femur, proximal tibia and fibula and the patella show normal signal Intensities. No marrow edema identified. The growth plates show normal signal intensities. The anterior cruciate ligament is not identified. The posterior cruciate ligament is visualized, and is aligned vertically. The posterior horn of the lateral meniscus appears deformed with altered signals. The anterior horn and body of the lateral meniscus appear unremarkable. The medial meniscus is hypoplastic.

The knee joint capsule is thickened. The collateral ligaments cannot be commented upon due to the thickened joint capsule, which seem to hinder their visualization. Minimal joint effusion seen. The bulk of the muscles posterior to the distal thigh and knee joint is reduced.

Left knee joint MRI (Figure 9 and 10) in sagittal plane with PDFS/ T2WI, coronal T2 and axial PDFS / T2WI sequences shows absent ACL and vertically oriented PCL. Sagittal PDFS (figure 11) Shows hypoplastic / absent medial meniscus and deformed posterior horn of medial meniscus.

Figure 1: X-Ray Knee: AP / Lateral View of Left Knee Shows Posterio-Lateral Subluxation of Femur in Relation to Tibia

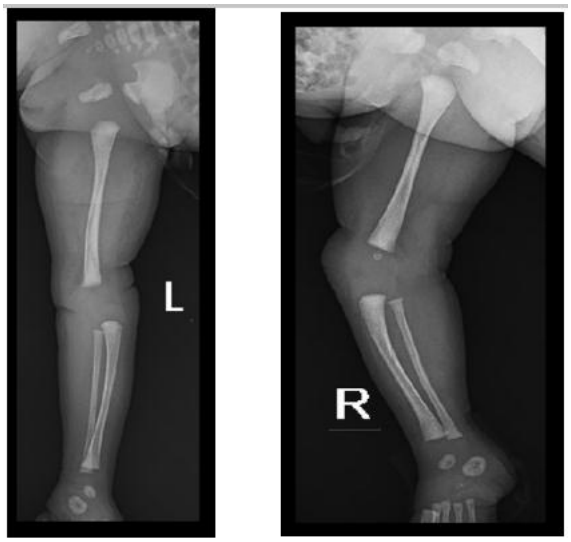
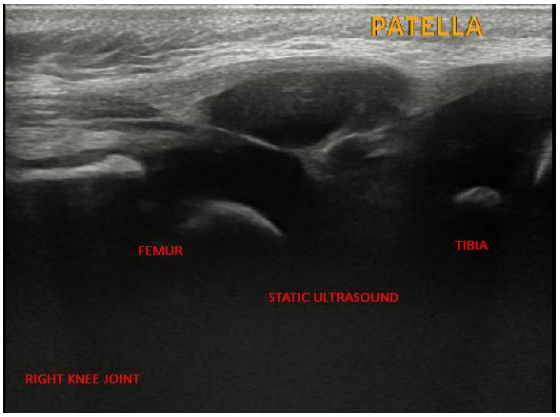


Figure 2: X-Ray Knee: AP / Lateral Right Knee Shows Mild Posterior Subluxation of Femur in Relation to Tibia

ULTRASOUND



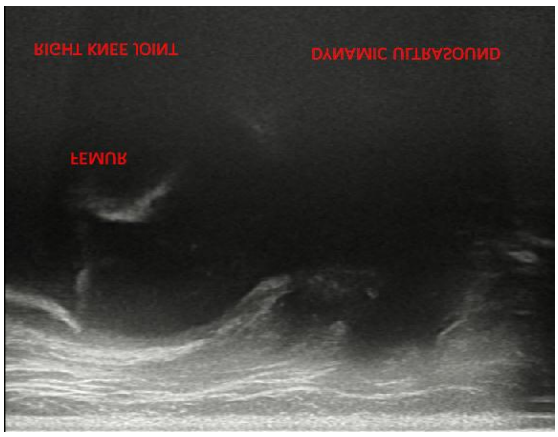


Figure 3: Right Knee

Static ultrasound image: distal femur and proximal tibia are in alignment.
Dynamic ultrasound image: repersentative still frame from dynamic ultrasound showing transient subluxation of proximal tibia. Not seen in alignment with distal femur

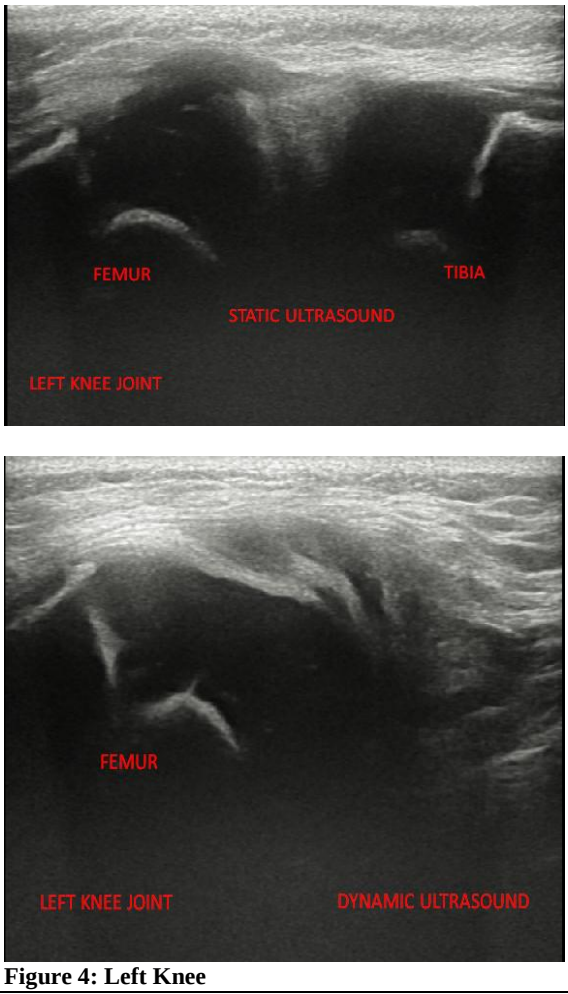


Figure 4: Left Knee

Static ultrasound image: distal femur and proximal tibia are in alignment.
Dynamic ultrasound image: repersentative still frame from dynamic ultrasound showing transient subluxation of proximal tibia. ., not seen in alignment with distal femur.

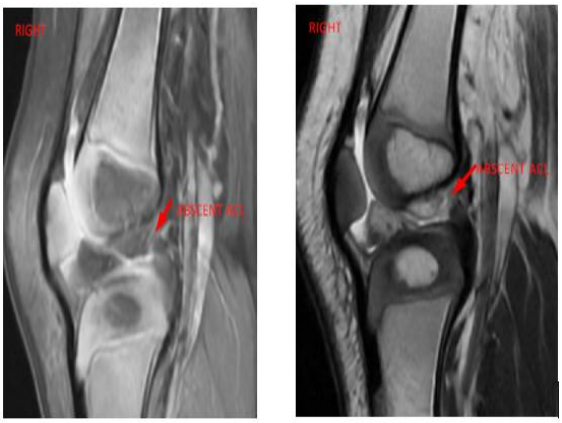


Figure 5: Right Knee MRI with Sequences: Sagittal Pdfs / T2wi Images Show: Absence / Non Visulaisation of ACLFibres, Empthy Intercondylar Notch and No Continuous Low Signal Band

Figure 6: Coronal Plane (Confirmatory View) Right Knee: With Sequences: T2wi Shows: No Oblique ACL Band, Intercondylar Notch Anatomy, Tibial Spine Morphology and Coronal T2 Image Confirms Non-Visualization of ACL

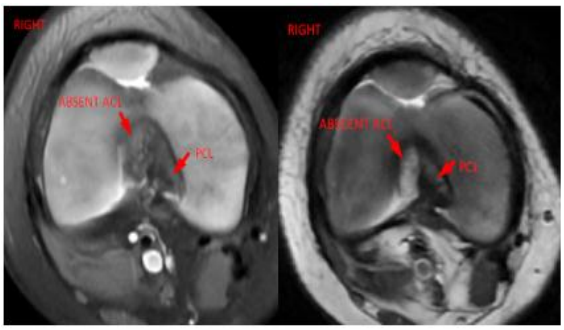


Figure 7: Axial plane (notch and tibial spine) right knee with seq: t2/pdfs sagittal shows: hypoplastic intercondylar notch, abscent acl footprint and tibial eminence changes. Axial t2 iamges demonstarate shallow intercondylar notch

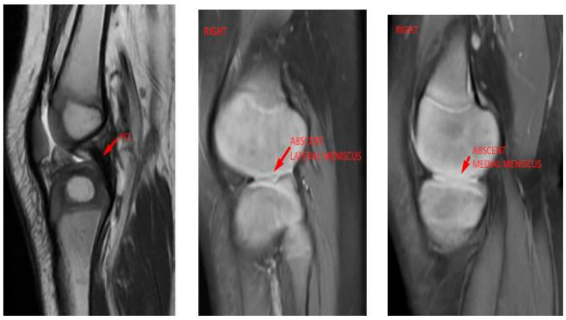


Figure 8: Right Knee MRI in Seq: T2wi / PDFS in Sagittal Plane Shows Secondary Signs of PCL Shows Hypoplastic Medial and Lateral Meniscus

Figure 9: Left Knee MRI in Sagittal Plane with Pdfs/ T2wi, Coronal T2 and Axial Pdfs / T2wi Sequences Shows Absent ACL and Vertically Oriented PCL

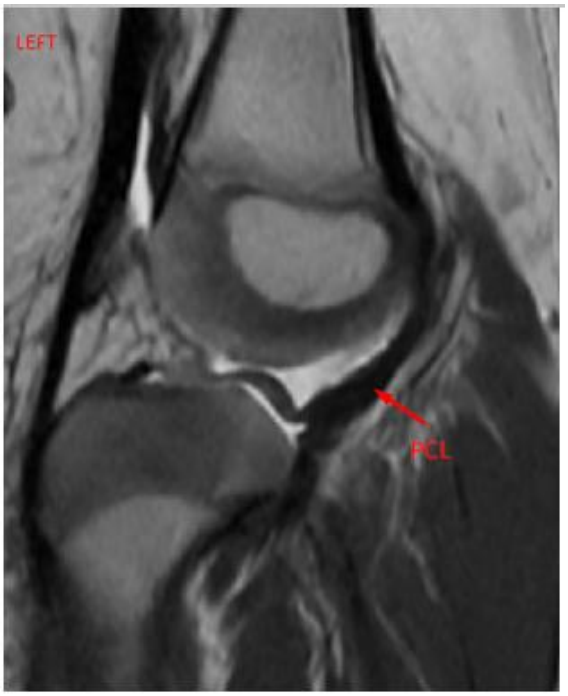


Figure 10: Left Knee MRI in Sagittal T2 Shows Vertically Oriented PCL

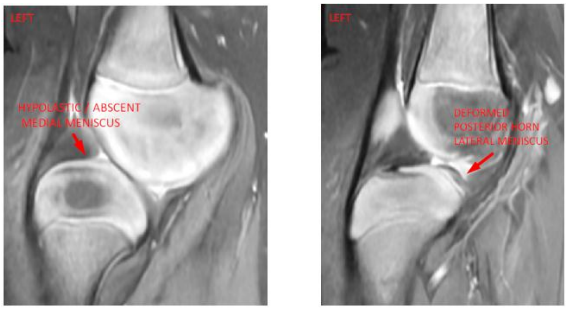


Figure 11: Left Knee MRI in Sagittal Pdfs Shows Hypoplastic / Absent Medial Meniscus and Deformed Posterior Horn of Medial Meniscus10

DISCUSSION

Congenital absence of the anterior cruciate ligament (ACL) is a rare condition that was first described by Niebauer and King in 1960.[6] Since then, multiple cases have been documented in the literature. In 2006, Manner et al. proposed a radiological classification based on findings in 31 patients, dividing the condition into three types.

Type I includes cases with hypoplasia or absence of the ACL, accompanied by a partially narrowed femoral intercondylar notch and underdeveloped tibial spines. Type II is characterized by complete absence of the ACL along with hypoplasia of the posterior cruciate ligament (PCL), with more pronounced narrowing of the femoral notch and further underdevelopment of the tibial spines compared to Type I. Type III represents the most severe form, with absence of both ACL and PCL, complete absence of the femoral intercondylar notch, and aplasia of both tibial spines.

This classification system does not take into account the presence or severity of osteoarthritis, as most reported cases were identified before skeletal maturity. However, in older patients, degenerative changes and joint instability play a significant role in determining management strategies, suggesting the need for an updated classification that incorporates these factors.

Additionally, associated hip abnormalities such as developmental dysplasia or congenital dislocation, if undiagnosed or untreated during childhood, may progress to advanced osteoarthritis. Therefore, evaluation of the hip joint is important in older individuals presenting with congenital ACL deficiency. Similarly, clinicians should remain vigilant for possible knee involvement in patients with established hip osteoarthritis.

Management of congenital ACL absence remains controversial, with both conservative and surgical approaches being considered. In pediatric patients, treatment decisions are primarily based on skeletal maturity and the degree of knee instability. Many patients tolerate the condition well during childhood, and surgical intervention is often deferred until skeletal maturity to avoid damage to growth plates. However, conservative management

may lead to suboptimal outcomes in some cases. On the other hand, ligament reconstruction has shown favorable results in selected patients.[7-9]

Persistent anterior knee instability increases the risk of meniscal and cartilage damage, ultimately leading to early osteoarthritic changes. Surgical intervention may also be considered because restricting physical activity in children is often impractical and poorly accepted.[10-12]

CONCLUSION

Multimodality imaging plays a vital role in diagnosing congenital ACL absence and guiding appropriate management.

Clinical Message

Congenital absence of the ACL should be considered in infants with knee instability. Early diagnosis using MRI and dynamic ultrasound prevents misdiagnosis and unnecessary surgical intervention.

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