

Case Report

Congenital Dislocation of the Knee: a Surprise in the Delivery Room

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Abstract

Congenital knee dislocation (CKD), is an exceptionally rare condition (1 in 100.000) characterized by hyperextension of the knee evident at birth. Its exact etiology remains unknown, and it may manifest as an isolated finding or in conjunction with multiple genetic syndromes. Treatment strategies vary from conservative measures to surgical intervention, according to the severity of the deformity and degree of passive reduction. In this report, we present a case of a newborn female, delivered after an uneventful pregnancy with normal ultrasound findings, who exhibited an hyperextension of 90° at birth. Early treatment with castings and Pavlik harness was initiated. Follow-up at 3 months-of life revealed a resolution of hyperextension, with restored full flexion and mobility.

Keywords: Congenital anomalies; Knee Dislocation; Neonatology; Orthopedics; Pediatric

Introduction

Congenital knee dislocation (CKD) is a rare congenital condition, affecting approximately 1 in 100.000 live births [1,2] with a higher prevalence (2:1 ratio) among females [3]. There appears to be no discernible laterality in CKD, with approximately one-third of cases with bilateral knee involvement [3].

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Its main clinical finding is a hyperextension of the knee with forward displacement of the proximal tibia on the femoral condyles, and can be easily confirmed through a simple radiography [2].

Currently, the exact etiology and physiopathology of CKD remains unknown. However, particular extrinsic factors have been implicated as contributors, such as oligohydramnios, intrauterine packaging disorders, leading to fetal molding or extended breech position, which causes contracture of the quadriceps after feet malposition [4].

Additionally, intrinsic factors such as absent suprapatellar pouch, deficient/hypoplastic anterior cruciate ligament and quadriceps contracture may contribute to its development [4].

CKD may present as an isolated deformity, or in conjunction with other skeletal malformations: approximately 65% of patients with CKD exhibit associated congenital anomalies, including developmental dysplasia of the hip, club foot or myelomeningocele [2]. While most cases of CKD are sporadic, it can also manifest as part of multiple genetic syndromes, such as: arthrogryposis multiplex congenita, Larsen syndrome, characterized by an imbalance between flexion and extension; Marfan and Ehlers-Danlos, with a state of hyperlaxity; as well as Desbuquois or Collins Pope Syndrome [2].

The diagnosis is usually made at the time of birth in the delivery room based on the characteristic knee recurvatum position, with anterior foldings. Radiographic evaluation further helps confirm the diagnosis [5].

Classification systems based in clinical and radiological findings help dictate treatment options as well as prognosis [4]. Treatment should be performed as soon as possible. Non-surgical approaches have demonstrated to be effective in most milder cases, and are often preferred as initial treatment in the majority of cases [3].

With this interesting case report, we endeavor to raise awareness to this rare condition, facilitating informed discussions regarding treatment options and prognosis with parents and caregivers.

Case Presentation

The patient, a female, was born from a first pregnancy to a healthy non-consanguineous couple. The gestation period was uneventful, with regular antenatal care and no abnormalities detected on ultrasounds. Patient was delivered at 37 weeks' gestation, via assisted vaginal delivery with a vacuum extractor, with no complications. Apgar index score was 9/10/10, indicating a normal transition to extra-uterine life. Birth weight was 2145g, below the 10th percentile according to Fenton charts.

Immediately after the delivery, a left lower limb deformity was noticed, marked by hyperextension of the knee to 90°, along with anterior skin folds and grooves (figure 1). Although passive reduction of hyperextension to 0° was achieved with no tension, no knee flexion could be initially attained (figure 2). Manipulation of the limb was painless. No additional musculoskeletal abnormalities were found upon physical examination, including the evaluation of the hip, feet and spine. No distinctive dysmorphic features were observed.



Figure 1: Hyperextension of 90° immediately after delivery.



Figure 2: Passive reduction up to 0°.



Figure 3: Cast applied with 30° flexion.



Figure 4: Pavlik harness at 2 months-old.

Within the first 24h hours of life, a radiography was performed, with no signs of fractures. Subsequently, an orthopedic consultation was requested, and patient was reviewed on the fifth day of life after being discharged from the maternity center. The hyperextension was reduced to 30° of knee flexion, categorizing it as a grade II in the Tarek Classification.

Patient was placed in a cruropodal cast immobilization with knee flexion and remained in the initial cast for one week (figure 3). Upon reassessment, signs of active hyperextension were no longer present, and passive flexion reached 100°. A Pavlik harness was placed and the patient was monitored monthly (figure 4).

At three months of age, the harness was removed and the patient exhibited a full flexion of the knee and extension to 0°, accompanied by favorable radiographic alignment. Further clinical evaluation was normal, indicating an adequate neurodevelopment and the absence of motor deficits.

Discussion

CKD is a rare limb deformity, that presents itself as an unexpected challenge in the delivery room, as its prenatal diagnosis, although achievable, is even rarer, as reported by Cavoretto et al [6]. Its initial assessment mandates a comprehensive and thorough evaluation of both the newborn and the deformity itself. Determining the degree

of hyperextension and the feasibility to reduce it are pivotal to decide the adequate course of treatment, which may range from conservative measures to surgical intervention in severe cases.

The presence of anterior skin grooves serves as a significant prognostic indicator: the bigger number of grooves typically indicates a more recent dislocation and thus a less severe deformity, often more responsive to conservative treatment [5,7] It is also important to perform a detailed physical examination to rule out other associated anomalies or genetic syndromes, with particular emphasis on the hips, feet and spine [3].

Even though the appearance of CKD can be intimidating and alarming, its prognosis is generally favorable and predictable according to knee's mobility and range of motion. Thus, an early referral to an orthopedic specialist and a prompt treatment can significantly contribute to complete resolution of the deformity.

Treatment is often based on classification systems. Most are based on the radiographic findings of the femoro-tibial relationship. For instance, Abdelaziz and Samir proposed a classification system based on the initial range of passive knee flexion, dividing findings in three grades: grade I with >90°, grade II 30-90°, and grade III <30° [5,8]. Parents can be informed that type I and most type II dislocations are associated with good outcomes when promptly treated with casting or harnesses [4].

Factors associated with a poorer prognosis include genetic syndromes, absence of anterior groove, delayed treatment, inability to achieve >90° flexion after serial casting or recurrence of hyperextension [4,8].

Conservative treatment consists of reduction, gentle manipulation and serial casting, progressing from extension to gradual flexion as tolerated by the knee [9]. However, forced flexion is to be avoided, as it may cause iatrogenic fractures of the limb [9,10]. The aim is to achieve progressive increase in knee flexion and maintenance of reduction, with variable duration in treatment [4]. Casts are typically changed every one to two weeks over a period of two or three months. Once 90° of flexion is achieved, the use of a Pavlik harness is recommended [2,8]. Surgical treatment is indicated in Grade III hyperextension or when conservative measures fail to achieve adequate flexion. In cases where multiple musculoskeletal deformities occur, recommendations are for the knee to be treated first [11].

In this case, despite the absence of identifiable extrinsic risk factors, our patient presented favorable prognostic features, such as visible anterior grooves, CKD as an isolated deformity and no syndromic features. The early referral and treatment, with good response to initial casting enabled the transition to Pavlik harness, resulting in full recovery after 3 months of treatment.

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