Congenital Knee Dislocation at Birth - An Extraordinary Case of Spontaneous Reduction*

Luxação congênita do joelho no nascimento - Um caso extraordinário de redução espontânea

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Abstract

Congenital knee dislocation (CKD) is a rare malformation characterized by hyperextension deformity of the knee with anterior tibia displacement, present at birth. Rarely reported, CKD might occur as an isolated deformity or commonly associated with musculoskeletal abnormalities, with the most common ones being developmental dysplasia of the hip (DDH) and clubfoot. The etiology is unknown, but CKD has been associated with certain intrinsic and extrinsic factors. Treatment with conservative methods at an early stage is most likely to yield successful results. We report here a rare case of successful spontaneous reduction of CKD in an infant within 24 hours of life.

Keywords
- joint dislocations/ congenital
- joint dislocations/ therapy
- remission, spontaneous
- infant, newborn

Resumo

A luxação congênita do joelho (LCJ) é uma malformação rara caracterizada por deformidade de hiperextensão do joelho com deslocamento anterior da tíbia, presente ao nascimento. Raramente relatada, a LCJ pode ocorrer como uma deformidade isolada ou comumente associada a anormalidades musculoesqueléticas, sendo as mais comuns a displasia do desenvolvimento do quadril (DDQ) e o pé torto congênito (PTC). A etiologia é desconhecida, mas a LCJ foi associada a certos fatores intrínsecos e extrínsecos. O tratamento com métodos conservadores em um estágio inicial tem maior probabilidade de produzir resultados bem-sucedidos. Relatamos aqui um caso raro de redução espontânea bem-sucedida de LCJ em um bebê nas suas primeiras 24 horas de vida.

Palavras-chave
- luxações articulares/ congênitas
- luxações articulares/ terapia
- remissão espontânea
- recém-nascido

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Introduction

Congenital knee dislocation (CKD) was first described by Chanssier in 1812 with a reported incidence of 0.017 in 1,000 live births with indistinguishability between the right and left knees and affecting mostly girls. Congenital knee dislocation may be an isolated deformity; however, from 40 to 100% of patients have associated musculoskeletal abnormalities, with the most common being developmental dysplasia of the hip (DDH) and clubfoot. Others are spinal bifida, cleft palate, Larsen syndrome, arthrogryposis, fibula hypoplasia, dislocation of the elbow and chest cage deformities.[1] The pathognomonic feature of CKD is fibrosis and shortening of the quadriceps muscle with hypoplasia of the suprapatellar pouch.,

The etiology is yet to be known; however, CKD has been associated with certain factors, including extrinsic factors such as intrauterine packaging disorders, breech presentations, neuromuscular imbalances, and intrinsic factors like genetic malformation. Nonetheless, three different theories have been described with regards to this scarce entity.

The first theory, the primary embryologic theory explains other added deformities accompanying the knee problem. The second theory, which is the mechanical theory, explains that the deformity originates from abnormal fetal posture during the intrauterine period of life. The third and final theory is the mesenchymal defect theory, according to which the mesenchymal defect causes fibrotic quadriceps muscle.[2]

Leveuf and Pais classified CKD into three grades.[3] Grade 1, the most common type and represents congenital hyperextension of the knee joint at birth. From 15 to 20 degrees of hyperextension can be detected, and passive range of flexion is at a maximum of 90 degrees. Grade 2, congenital subluxation with joint incongruency. Passive flexion of the knee is impossible, and hyperextension of 25 to 40 degrees is achieved. Grade 3 represents total dislocation of the tibial epiphysis anterior to the femoral condyles with no contact between the joint surfaces of tibia and femur.

The diagnosis of CKD is established at birth by the classical position of genu recurvatum. A deep transverse skin fold is found anteriorly with femoral condyles prominently palpable in the popliteal fossa.[4] The patella is deeply placed and usually not palpable or maybe absent. There will be apparent external rotation of the limb and valgus deformity of the tibia. Prenatal ultrasonographic examination may identify CKD.

Babies with CKD should be diagnosed and treated immediately after birth. Management is primarily conservative with direct reduction and gentle, persistent manual traction with serial casting in increasing flexion. Forced flexion is strictly prohibited as it may lead to iatrogenic fractures, epiphyseal damage and compromise the vascularity of the limb. The cast should be changed serially every 2 weeks until reduction is achieved. If and when casting fails, surgical management consists of quadriceplasty, mini open quadriceps tenotomy, iliotibial (IT) band and capsular release, and anterior cruciate ligament (ACL) advancement.[5] Nevertheless, it is important that in concomitant associated musculoskeletal anomalies, the knee must always be manipulated and treated first, before the foot or the hip.

Case Report

A newborn infant was referred by the pediatrician to the orthopedics team for a hyperextended left knee after being delivered via an emergency lower section caesarean surgery, at a gestational age of 37 weeks, due to maternal preeclampsia and fetal distress in labor. The baby was delivered head first, and there were no intraoperative complications reported. Her birth weight was 2.18 kgs and she was born with an Apgar score of 8/9.

She exhibited typical features associated with CKD over her left knee. Physical examination revealed knee hyperextension of $\sim$ 70 degrees with anterior knee crease (►**Fig. 1**). Passive flexion was 5 to 10 degrees. The child did not exhibit any other congenital malformations such as club feet, DDH, or spina bifida. The X-rays of the left knee showed the tibia displaced anterior to the long axis of the femur (►**Fig. 2**). On palpation of the left knee, the patella was deeply placed and noted to be smaller in comparison with the right. The child was diagnosed with a grade II unilateral (left knee) CKD.

We counselled the parents for serial manipulation with casting and commenced with initial splinting of the limb in 5 to10 degrees flexion. She was then monitored closely

Fig. 1 Showing hyperextension of the left knee of 90 degrees.

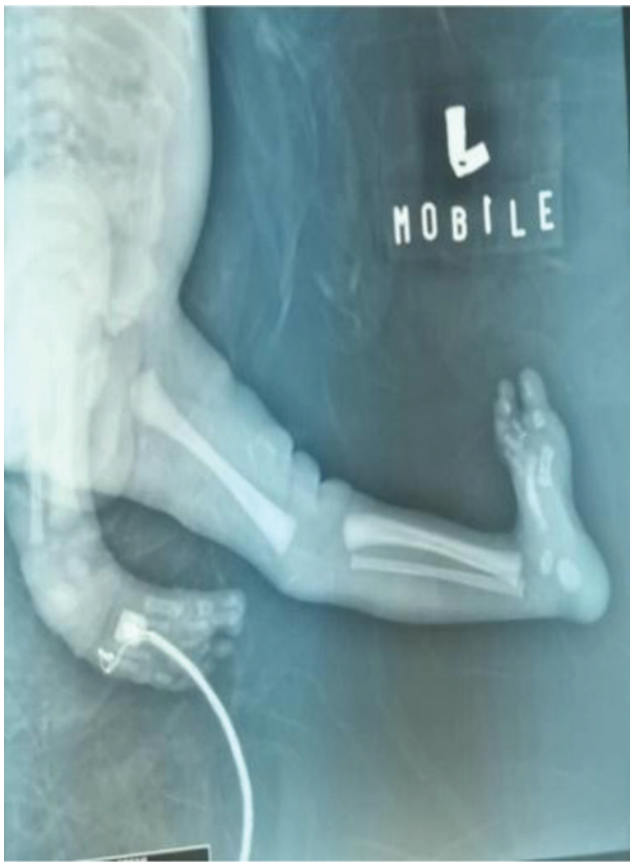

Fig. 2 Showing anteroposterior view of the left lower limb.

Fig. 3 Showing spontaneous reduction of the left knee within 24 hours of birth.

in the neonatal intensive care unit (NICU) with strict circulation charting. An hour later, the left lower limb was noted to be blueish, cool to the touch, and the oxygen saturation (SpO2) was not detectable over the toes of the left foot. Both dorsal pedis artery (DPA) and posterior tibial artery (PTA) pulses were not palpable. A decision was then made to remove all forms of external splinting and bandages and to observe the limb. The pulses returned and the limb was once again well perfused after removing the splint.

The following day, during examination, the left knee was noted to be in a normal position. The patient had an active range of movement from -10 degrees to 120 degrees. The pulses of the limb were strong and equal to those of the contralateral limb. The left knee was spontaneously reduced with a period of 24 hours (►**Fig. 3**). We decided to abandon our initial plan of manipulation and casting. The child was put on a physiotherapy regime for range of movement exercise of the left knee. No further imaging was repeated.

We reviewed the child after a week, and, at 8 months of age, there was no evidence of recurrence. The child, however, had a lax left knee with a range of movement of -10 to 120 degrees. She is still currently under our routine follow-up.

Discussion

Congenital knee dislocation is a rare musculoskeletal anomaly (with a ratio of 17 in 1 million live births) most often associated with other pathologies and syndromes. Diagnosis of associated conditions and syndromic involvement helps in proper management and in predetermining the outcome. With other associated deformities of the hip and foot, the overall treatment becomes more arduous, and reduction of knee dislocations should always be given priority.

The mainstay of treatment for CDK is early non-operative reduction. One must refrain from using any force that can lead to tibial deformity, fractures, physeal separation and compromise the distal circulation of the limb.

The literature advocates that simple non-syndromic CKDs respond well to conservative management with serial casting and usually have a better prognosis in comparison to CKDs with associated anomalies. Importantly, the family should always be aware of foreseen complications of CKD, such as early arthrosis, extension lag, extension weakness, or potential knee problems in the future. Early diagnosis of this deformity is vital, as initializing treatment as early as possible promises a good outcome.

Conflict of Interests
The authors have no conflict of interests to declare.

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