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CASE REPORT

Congenital bilateral knee dislocation - A case of spontaneous reduction

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Abstract

Congenital knee dislocation (CKD), or genu recurvatum, is a disorder that occurs in approximately 1 in every 100,000 live births, with nearly equal distribution between sexes, defined by hyperextension of the knees present at birth. Involvement can be unilateral, which is more frequent, or bilateral, which is rare and accounts for about one-third of cases. The etiology of CKD is still uncertain, but it is believed that there is an association between factors such as breech fetal presentation, oligohydramnios, neuromuscular imbalances, and genetic malformations. Treatment of CKD includes conservative management, physical therapy, and surgical intervention, depending on the degree of dislocation classification. This report presents a newborn with bilateral knee dislocation, of positional etiology, diagnosed in the delivery room and later confirmed by X-ray, with no other associated musculoskeletal or genetic abnormalities found. The patient received conservative treatment, achieving spontaneous and complete resolution of the condition.

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INTRODUCTION

Congenital knee dislocation (CKD) is a rare deformity of knee hyperextension.[1] It was first described by Chanssier in 1812 but officially cited only in 1891 by Shattock.[2] Its etiology is still unknown. CKD has been associated with extrinsic and intrinsic factors[3] and is strongly related to other musculoskeletal malformations, including developmental dysplasia of the hip (DDH) and congenital clubfoot and genetic conditions.

X-ray images must be ordered for individuals with genu recurvatum at birth to confirm the diagnosis of CKD.[4] [5] Treatment varies according to the degree of dislocation, ranging from conservative management to surgical reduction. [5]

This report presents the case of a female newborn with bilateral CKD treated conservatively since the first hours of life, who recovered completely.

This report illuminates a rare condition in which diagnostic confirmation and early treatment are fundamental for a better prognosis. It also provides the medical community with input to support the recognition and management of patients with CKD and stresses the success of conservative treatment in improving patient quality of life.

CASE REPORT

During birth, a full-term white female newborn delivered by cesarean section required resuscitation maneuvers without positive pressure ventilation. Physical examination found she had two hyperextended knees without active or passive flexion, making it impossible to perform the Barlow and Ortolani maneuvers. The patient had significant cyanosis in the feet and legs without involvement of the thighs. She had palpable peripheral pulses bilaterally, with no temperature difference between them. The primitive reflexes were present and symmetrical, and no other alterations were seen on ectoscopy. The patient was referred to rooming-in, and an X-ray of the legs and an assessment by an orthopedist were requested for diagnosis and management. Six hours after birth, the patient's knees were still hyperextended, with symmetrical reflexes and improvement in cyanosis. At 12 hours, the patient could slightly flex her legs when actively stimulated, although her knees were still hyperextended when she was in a passive position. The pediatric orthopedics team evaluated the patient 24 hours after birth (Figure 1). Her condition improved, with the legs now predominantly flexed, without pain on mobilization, with hyperextension only on manipulation. The Barlow and Ortolani maneuvers were performed, with a negative result for both. The X-ray report described bilateral knee dislocation and clinical improvement after birth, with a diagnosis of Grade 3 CKD of positional etiology.

The image shows the patient in a passive position with hyperextended knees at birth, a condition that remained for the first 24 hours after birth.

Conservative management and outpatient follow-up commenced one week after birth. The patient was discharged 48 hours after birth and referred to the pediatric and orthopedic outpatient clinics. She was taken to the pediatric orthopedic outpatient clinic one week after birth (Figure 2) and, on physical examination, presented active and passive flexion of the legs, with hyperextension only during manipulation. The Barlow and Ortolani maneuvers were negative. Reflexes were present and symmetrical, cyanosis resolved, and she had palpable peripheral pulses. The patient



Figure 1. Bilateral dislocation of the knees. Personal file.
Fonte: The image elucidates the passive position in hyperextension of the knees seen at birth and which was maintained during the first 24 hours of the patient's life.



Figure 2. Passive position in semi-flexion of the knees. Personal file.
Fonte: The image shows the result of conservative treatment of Congenital Knee Dislocation of positional etiology. After 1 week of life, it is possible to notice the passive flexion position of the lower limbs that was present after the child's 3rd day of life.

was diagnosed with Grade 1 CKD and prescribed conservative management. Her parents brought her to the clinic 60 days later, and since she presented a spontaneous reduction, the patient was discharged with a childcare follow-up plan.

The image shows the outcome of conservative management prescribed to a patient with congenital knee dislocation of positional etiology. Passive flexion of the legs was noticeable one week after birth, with signs first noted three days after birth.

DISCUSSION

Congenital knee dislocation (CLK) is a deformity of knee hyperextension with anterior displacement of the tibia over the femur and hyperextension of the toes, which can reach the mouth. Known as genu recurvatum, its incidence is approximately 1 in 100,000 live births.[6] Initially, it was believed to be more frequent in girls, but today, it is known that the distribution between the sexes is practically equal.[7] There is no preferential involvement of the left or right knee. Bilateral involvement occurs in a third of the cases.

The pathogenesis of CKD is unclear, but an association between extrinsic factors, including breech birth, oligohydramnios, and neuromuscular imbalances, and intrinsic factors, such as genetic malformations, is believed to exist.[7] Three theories attempt to explain it: the primary embryological theory, which explains the additional deformities that accompany the knee condition; the mechanical theory, which describes a deformity caused by abnormal fetal position during intrauterine life; and the mesenchymal defect theory, which describes a fibrotic quadriceps muscle and its consequences. Forty to one hundred percent of patients with CKD have associated musculoskeletal abnormalities such as developmental dysplasia of the hip and congenital clubfoot.[8] Associations with spina bifida, cleft palate, Larsen syndrome, arthrogryposis multiplex congenita, meningomyelocele, Down syndrome, Marfan syndrome, and Ehlers-Danlos syndrome have been described.[8]

Individuals with CKD present contractures of the extensor part of the quadriceps and a tight anterior capsule of the knee joint. The entire anatomy of the knee is altered: the suprapatellar bursa may be obliterated by adhesions of the rectus femoris tendon, the collateral ligaments may be displaced forward, the hamstring muscles may be subluxated forward and function as knee extensors, the iliotibial band may be hypertrophied, the patella may be hypoplastic, absent or displaced outwards, and the cruciate ligaments may be altered or absent.

At birth, affected newborns adopt a genu recurvatum position. Clinical signs include, in addition to hyperextension of the knee, an increase in the transverse skin folds on the anterior surface of the knee and protrusion of the femoral condyles into the popliteal fossa. CKD can be classified into three degrees: grade 1, genus recurvatum, hyperextension greater than 15°, with preserved full flexion ability; grade 2, hyperextension with anterior subluxation of the tibia on the femur, greater than 15°, with limited flexion of the knee from the neutral position, resistance or instability to flexion; grade 3, hyperextension with anterior dislocation of the tibia on the femur, usually without flexion and unstable.[8] This classification helps guide prognosis and treatment.

Only 26% to 66.7% of the cases of CKD are diagnosed during prenatal care.[9] In 1986, the first in-utero diagnosis of CKD was performed based on X-ray images. Ultrasound-based diagnosis was first performed in 2003.[10] In most cases, diagnosis occurs immediately after birth, in the first physical examination, based on the presence of genu recurvatum. Attention is due to the number of anterior cutaneous grooves in the knee since many grooves indicate a more likely origin of recent intrauterine dislocation, which is less severe. In contrast, the absence of grooves suggests a more severe, long-term dislocation.[10]

Diagnostic confirmation is based on simple X-ray images (anteroposterior and lateral views of the knee) after physical examination in the immediate neonatal period. These can confirm the displacement of the proximal tibia in the femoral condyles and may assist in diagnosing fractures.

Treatment must be initiated immediately and based on the grade of dislocation. Manual reduction and immobilization with a cast should be started within 24 hours after birth,

especially for grades 1 and 2, for four weeks in grade 1 and six to eight weeks in grade 2 cases, and followed by physiotherapy. If the splint cannot maintain the reduction, the knee must be immobilized in a plaster cast at 90° for three weeks. If this fails in CKD grades 2 and 3 patients, percutaneous resections of the quadriceps tendon and quadricepsplasty can be performed to obtain a 90° knee flexion. It is important to emphasize that forcibly flexing the limb more than the limb allows can trigger iatrogenic fractures, epiphyseal damage, and local vascular impairment.

Patients with unilateral involvement or undergoing surgery before two years of age have better outcomes. Individuals with dislocations not reduced immediately after birth, knee flexion of less than 50°, and without anterior skin grooves have worse outcomes. Treatment delays may cause joint instability and long-term stiffness. Therefore, early recognition and treatment are of great importance since they change the course of the condition and significantly affect the quality of life of affected children.

As in a third of the cases, our patient had both knees dislocated, but unlike most patients with CKD, she did not present other musculoskeletal or genetic abnormalities. She was diagnosed with genu recurvatum immediately after birth, with confirmation of CKD based on X-ray images. She was prescribed conservative treatment, which makes the case very interesting since her reduction was entirely spontaneous. Twenty-four hours after birth, our patient was able to flex her legs and presented hyperextension only with manipulation.

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