



Case Report

Congenital dislocation of the patella – clinical case[☆]



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ABSTRACT

Congenital patellar dislocation is a rare condition in which the patella is permanently dislocated and cannot be reduced manually. The patella develops normally as a sesamoid bone of the femur. This congenital dislocation results from failure of the internal rotation of the myotome that forms the femur, quadriceps muscle and extensor apparatus. It usually manifests immediately after birth, although in some rare cases, the diagnosis may be delayed until adolescence or adulthood. Early diagnosis is important, thereby allowing surgical correction and avoiding late sequelae, including early degenerative changes in the knee. A case of permanent dislocation of the patella is presented here, in a female child aged seven years.

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Luxação congênita da patela – caso clínico

RESUMO

A luxação congênita da patela é uma patologia rara, em que a patela se encontra permanentemente luxada e manualmente irredutível. A patela desenvolve-se normalmente como um osso sesamoide do fêmur. A luxação congênita da patela resulta da falência da rotação interna do miótomo que forma o fêmur, músculo quadricípite e o aparelho extensor. Usualmente manifesta-se imediatamente após o nascimento, embora em alguns casos raros o diagnóstico possa ser adiado até a adolescência/idade adulta. O diagnóstico precoce é importante, permite a correção cirúrgica, evita as sequelas tardias, notadamente alterações degenerativas precoces do joelho. É apresentado um caso de luxação permanente da patela, numa criança de sexo feminino, com sete anos.

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Palavras chave:

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Introduction

Congenital dislocation of the patella comprises a pathological condition of permanent lateral dislocation of this bone. It is impossible to reduce it through manual maneuvers. It may arise in isolation, associated with lower-limb malformations, or within the context of a polymalformative syndrome.¹

It is generally diagnosed at birth. These infants present genu valgum and contracture of the flexed knee, in association with external rotation of the tibia. When these deformities are not present, this pathological condition may not be diagnosed until adulthood is reached.²

Radiological examinations, especially X-rays (XR), computed tomography (CT) and magnetic resonance imaging (MRI) are essential for identifying and characterizing lesions associated with permanent patellar dislocation (trochlear dysplasia or chondral lesions). However, the diagnosis for this pathological condition is essentially clinical.³

This condition can only be corrected through a surgical procedure. Several options exist. Early correction is important for avoiding sequelae.⁴

Case report

The patient was a seven-year-old white girl without any relevant antecedents who was examined in an external pediatric orthopedics consultation due to deformity of the left knee. She did not have any previous history of trauma. Her parents said that there had not been any initial episode of patellar dislocation.

Upon objective examination, she presented irreducible lateral dislocation of the left patella. It was observed that, during active and passive movements, her extension and flexion did not present any amplitude deficits and were not painful. She presented grade IV strength deficit during extension of the left lower limb.

Knee XR was performed under conditions of weight-bearing (Fig. 1) and found lateral patellar dislocation. The lateral XR on the left knee did not show dislocation (Fig. 2). The axial XR on the left patella again showed lateral dislocation of the patella, in association with trochlear dysplasia (Fig. 3). MRI on the left knee (Fig. 4) confirmed that in addition to the lateral dislocation of the left patella, there was dysplasia of the trochlea, but without other associated lesions. The patient was referred for a pediatric consultation in order to rule out polymalformative syndrome.

Surgical correction of the dislocation was performed, using the technique described by Stanisavljevic, and it followed an uneventful course. Plaster-cast immobilization from the lower leg to the foot was used for six weeks.

Currently, the patient is being followed up through external pediatric orthopedic consultations. The patella is now centered, without any episodes of dislocation so far, and the strength of the left lower limb has recovered (grade V). The leg presents flexion of 140° and an extension deficit of 5°.

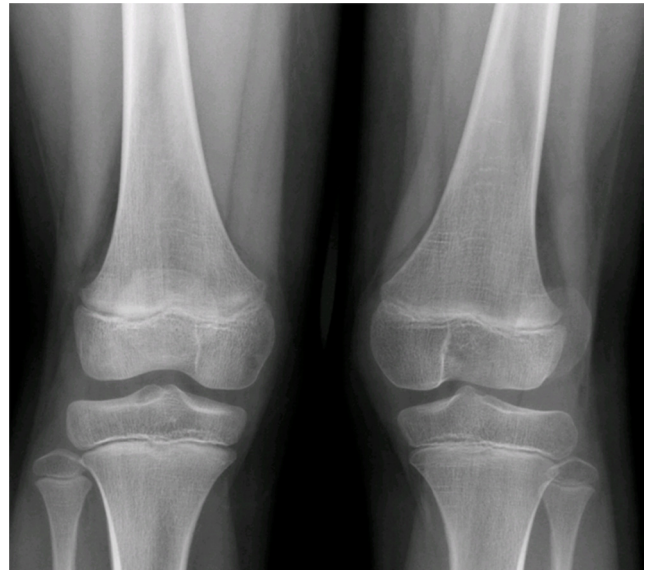


Fig. 1 – X-ray with weight-bearing on both knees.

Discussion

The congenital abnormalities of the patella include its absence, hypoplasia and permanent dislocation.

Permanent congenital dislocation is a pathological condition in which the patella remains constantly dislocated, even when the leg is extended. The patella remained fixed on the lateral face of the femoral condyle. The dislocation is irreducible, unless surgical techniques are used. It is a rare pathological condition of unknown incidence,⁵ which is generally detected in the first decade of life. It usually affects both legs and in most cases is associated with polymalformative



Fig. 2 – X-ray of the left knee in lateral view.

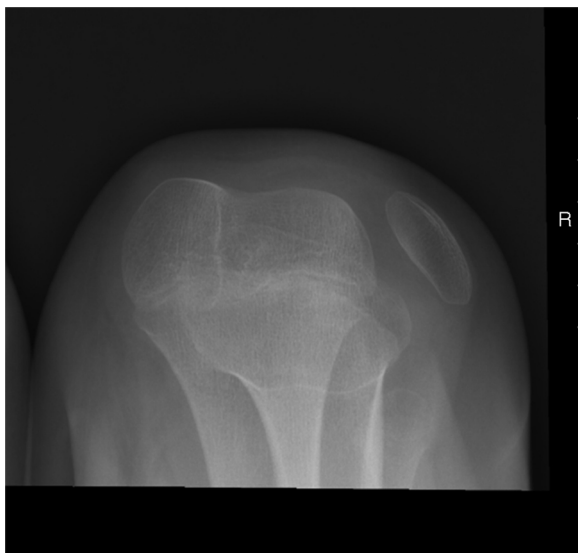


Fig. 3 – Axial X-ray of the left patella.

syndromes, such as nail-patella syndrome, Rubinstein-Taybi syndrome and William-Beuren syndrome.⁶

Between the eighth and tenth weeks of embryo development, internal rotation of the myotome responsible for the formation and orientation of the extensor apparatus of the lower limb takes place. Non-rotation is presumed to be the cause of congenital dislocation of the patella.⁷

This pathological entity may be manifested with a varying spectrum of clinical presentations. When it is not diagnosed at birth, objective examination shows the presence of contracture when the leg is flexed, valgus that is more accentuated in the affected lower limb and an associated deficit of extension. It is also observed that the intercondylar chamfering is empty, with a palpable patella on the lateral face of the knee.⁵ Apparent gait imbalance may also be seen, caused by incompetence of the extensor apparatus.⁸

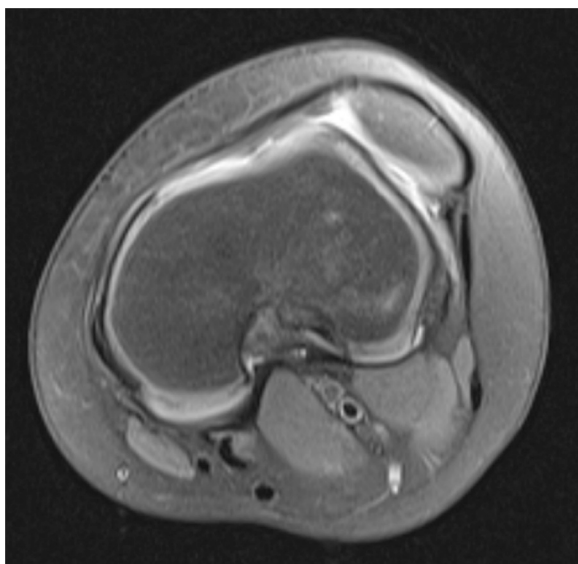


Fig. 4 – Magnetic resonance imaging of the left knee.

The diagnosis of congenital dislocation of the patella is made through XR,⁷ if the child is more than 3-5 years of age, given that ossification of the patella starts at this age range.⁹ The anteroposterior view of the knee makes it possible to view the size and position of the patella, along with other alterations associated with this pathological condition, such as hypoplasia of the lateral femoral condyle, diminution of the joint interline (later cases) and the position of the tibia in relation to the femur. The axial view of the patella also makes it possible to assess the position, size and shape of the patella, and also the intercondylar groove.⁷

Echography is important, especially for younger children whose patellar ossification has not yet occurred. This makes it possible to view the cartilaginous part of the patella, along with the cartilage of the femur and tibia and their relationships. It also enables viewing of the knee ligament insertions and can be used as a dynamic examination, making it possible to demonstrate the position of the patella in relation to the femur at different degrees of flexion.¹⁰⁻¹² CT supplies detailed information on the bone components of the knee, but because of its use of ionizing radiation, its use for evaluating children's knees is not recommended.¹³ MRI has the capacity to effectively distinguish between the cartilages of adjacent joint structures,¹⁴ which makes it essential for pre-operative assessments. The appropriate technique includes producing thin slices in order to avoid a false diagnosis of patellar agenesis and to identify the size and position of the quadriceps muscle and the bone insertions of the quadriceps and patellar tendons, thereby aiding in surgical planning.¹⁵

The natural history of undiagnosed and untreated congenital dislocation of the patella follows a course with many alterations to the knee, including flattening of the lateral femoral condyle, with worsening of the valgus deformity and external rotation of the tibia. The dislocated patella remains hypoplastic and, because of the unequal load distribution, an early decrease in the lateral joint interline of the knee is seen, with appearance of subchondral cysts. Thus, it is important to recognize and surgically treat this condition during childhood.¹⁶

Surgical treatment of congenital dislocation of the patella has the objective of realigning the extensor apparatus. Several surgical procedures exist. Stanisavljevic's procedure is among the most popular methods today.⁶ In this procedure, the incision is started 5 cm below the greater trochanter and then extends distally, makes a curve at the base of the patella and ends medially below the medial condyle of the tibia. The patella is displaced and a flap from the fascia lata is excised and conserved in saline solution. The joint capsule is opened laterally and the quadriceps is released and placed in its anatomical position. The patellar tendon is divided longitudinally and its lateral portion is sutured as medially as possible. The gap in the lateral portion of the thigh is closed using the flap from the fascia that was previously excised.⁷

Congenital dislocation of the patella is a rare pathological condition that is generally present at birth. It is extremely important to diagnose it and implement surgical correction early on, so as to avoid sequelae.

Conflicts of interest

The authors declare no conflicts of interest.

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