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Case report

Congenital idiopathic knee dislocation: A clinical case useful for defining an etiological hypothesis, times and methods of treatment

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Abstract. Congenital knee dislocation (genu recurvatum) is a rare condition, occurring in about 1 in 100,000 live births. It can be associated with genetic syndromes or neurological disorders, but also occurs as an isolated condition. Factors such as oligohydramnios, breech presentation, and other limb abnormalities are often related. This case report details the presentation of a newborn female delivered at 37 weeks by emergency cesarean section, who exhibited congenital left knee dislocation without any other abnormalities. Upon birth, the left leg was observed to be hyperextended, with the foot positioned toward the body and slight cyanosis on the plantar skin. Initial treatment involved non-pharmacological pain management, followed by a successful manual reduction of the dislocation and application of a splint. The joint remained stable after 60 days of treatment, with no recurrence noted. Genetic testing and other screenings returned normal results. The discussion suggests an idiopathic nature of the dislocation, as the absence of a prenatal diagnosis points to a potential late-acquired condition in pregnancy. Hypotheses include limited uterine space and fetal movements that could have contributed to hyperextension. Pressure exerted on the small fetal patella likely facilitated lateral displacement, allowing the dislocation to occur. Early intervention proved crucial to prevent long-term complications, though the rarity of this condition makes future prognosis difficult to predict (www.actabiomedica.it).

Key words: knee dislocation, fetal movements, patella, manual reduction

Introduction

Congenital dislocation of the knee, also known as "genu recurvatum," is a rare condition with an incidence of approximately 1 in 100,000 live births (1). It is characterized by posterior displacement of the femoral condyle in utero, with upward displacement of the proximal articular surface of the tibia, beyond simple hyperextension. Currently, there is no established consensus on its classification (2). The condition may be associated with genetic syndromes and neurological disorders, including Larsen syndrome, Desbuquois dysplasia, Marfan syndrome, Ehlers-Danlos syndrome, congenital arthrogryposis, and myelomeningocele. It can also present as an isolated condition, often linked to oligohydramnios, breech presentation, and other lower limb abnormalities such as congenital hip dysplasia and clubfoot (1, 2).

Case Report

The patient was a female newborn, delivered at 37 weeks of gestation by emergency cesarean-section due to suspected placental abruption, presenting with a congenital dislocation of the left knee. The

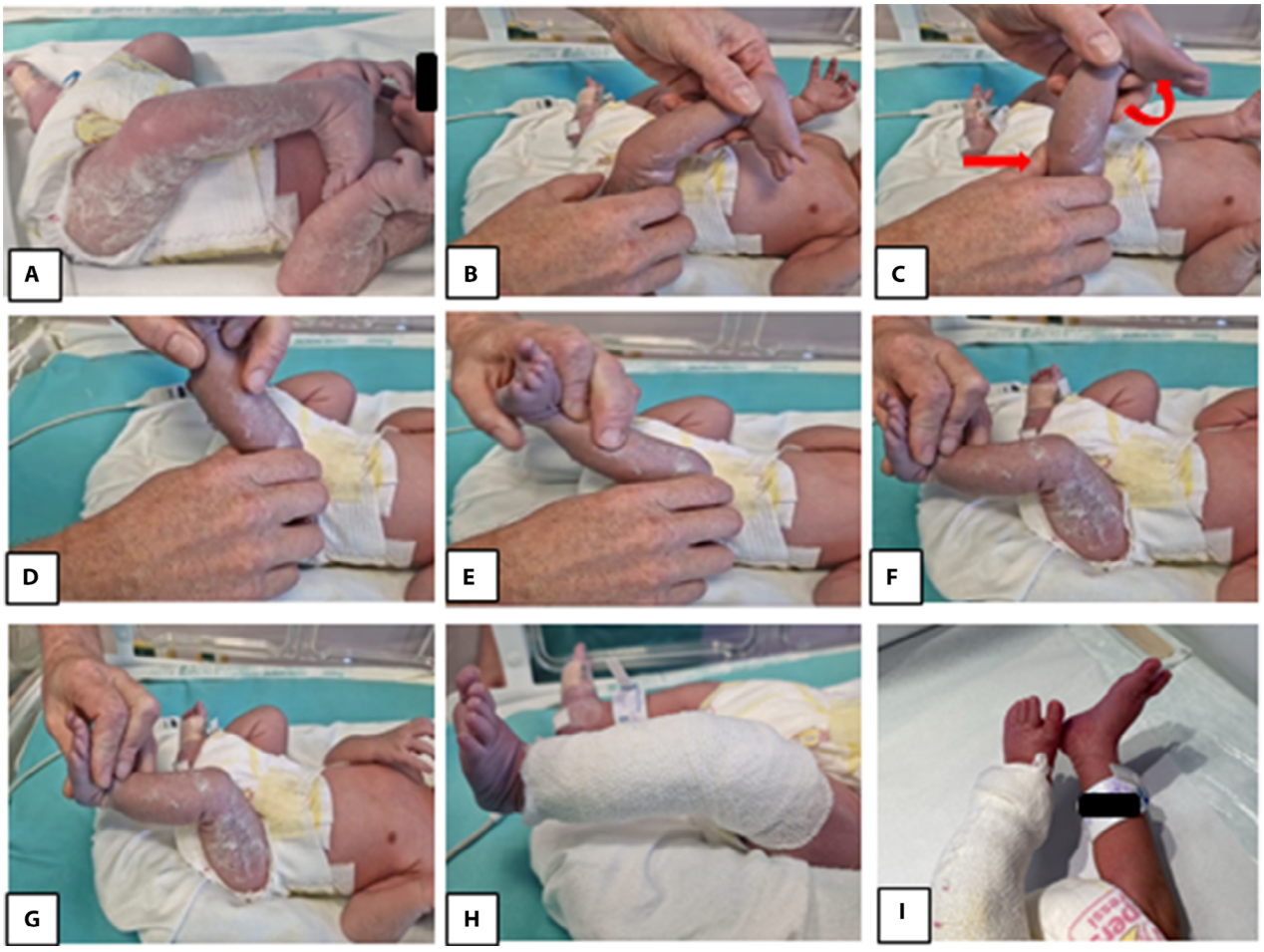

Figure 1. Treatment sequence: 1A condition at birth, from 1B to 1F sequence of reduction maneuvers, 1H, and 1I splint, and reduction of foot cyanosis.

newborn exhibited no clinical abnormalities, malformations, or associated limb deformities. During the pregnancy, three ultrasound scans were performed at the recommended intervals according to national guidelines, and no dislocation was detected. Immediately after delivery, the midwife noted an abnormal position of the left lower limb: the leg was hyperextended, forming a 60-degree angle with the thigh, with the foot positioned towards the body. The foot appeared mobile and free of deformities, though cyanosis signs were noted on the plantar skin. The leg remained fixed in this position, with deep skin folds at the knee (Figure 1A). The newborn was placed in an incubator immediately after birth. About two hours later, following the stabilization of cardio-pulmonary adaptation, the orthopedic procedure to reduce the dislocation was initiated. Non-pharmacological pain management techniques were employed, including containment, warmth, sucking, and oral administration of 24% sucrose solution. The reduction maneuver was performed with the infant lying on her back, with the operator positioned at her feet. The operator flexed the infant's hip, creating a 90-degree angle between the femur and the surface (Figure 1B). Holding the foot with one hand, the operator applied gentle traction and rotated the foot while stabilizing the thigh with the other hand, using the thumb to fix the popliteal area (Figure 1C). The leg, initially hyperextended, was progressively realigned to a neutral position. The dislocation was reduced by palpating the

femoral condyles and applying pressure towards the hip. Additional gentle traction and flexion ensured the tibia and femur were properly aligned, completing the reduction (Figure 1D and 1E). The femoral condyles were no longer palpable after the procedure, and the patella was felt in the correct position (Figure 1F and 1G). Following reduction, a splint was applied to stabilize the joint (Figure 1H). Cyanosis of the foot resolved quickly (Figure 1I).

An X-ray examination confirmed the correct position of the joint components, showing a slight widening of the shape of the distal femur and a mildly blurred metaphyseal line (Figure 2). The splint was maintained for 15 days, then changed every 20 days. After approximately 60 days, the splint was removed permanently. The joint remained stable, with no deficits in mobility or stability. A slight reduction in muscle mass due to the cast was noted, but this resolved within three months. There were no skin lesions, and the baby's growth was normal and symmetrical. No motor deficits of the limb and no alteration of knee stability were present at the end of the follow-up period that lasted for six months. Array Comparative Genomic Hybridization (CGH) revealed no associated genetic alterations, and various screenings (metabolic, EKG, Color Doppler ultrasound, transfontanellar ultrasound, ultrasound of the hips, audiometric test, eye exam, and blood tests) all returned normal results.

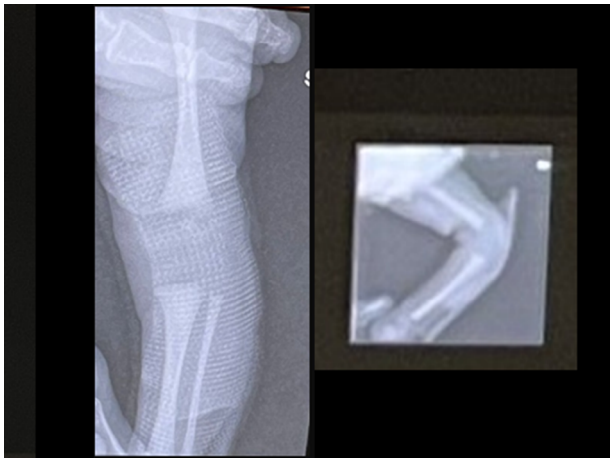

Figure 2. X-ray of the knee after reduction.

Discussion

The absence of a prenatal diagnosis suggests that the dislocation may have occurred later in pregnancy, although it remains idiopathic. There are different treatment options. Due to the rarity of the condition, the scientific literature often examines small sample sizes and diverse clinical scenarios, including isolated dislocations and those associated with other limb defects or genetic disorders (3,4). As a result, there are no definitive guidelines regarding the optimal intervention. However, as some authors suggest, early conservative treatment within the first day of life is preferable, as it is generally effective, though some cases may still require further intervention (4,5). In the absence of other malformations or associated limb abnormalities, we believe that prompt reduction of the dislocation is advisable. Delayed treatment may inadvertently cause harm during routine newborn care, while prolonged dislocation can lead to increasing quadriceps contracture and pain over time. As with other congenital limb anomalies, such as developmental dysplasia of the hip, the observation of skin folds is a valuable diagnostic tool. Some authors suggest that the number and depth of skin folds can indicate the duration of the condition (1). We believe they are also useful in assessing the success of the reduction maneuver. In our case, the skin folds became symmetrical from asymmetrical after the reduction of knee dislocation. Isolated idiopathic genu recurvatum is distinct from cases associated with arthrogryposis multiplex congenita, Larsen syndrome, or congenital clubfoot (6,7). Recurrence is rare after proper early treatment, suggesting that significant ligamentous laxity alone is unlikely to cause dislocation (6). The search for associations with genetic diseases and syndromes is always necessary (2,6). Genetic, endocrine, and organ-related causes were ruled out based on the tests performed. No general dysmorphic signs and no other limb malformations were associated with the dislocation. The pregnancy history did not reveal any risk factors. If the dislocation occurs in the third trimester of pregnancy and it is isolated, it may not be detected at prenatal ultrasound diagnosis and therefore be unexpected at birth (7,8). Some authors hypothesize that idiopathic genu recurvatum may result from congenital laxity or hypoplasia of the anterior cruciate

ligament (8). However, in congenital idiopathic cases, recurrence is typically absent after proper treatment, which suggests that an isolated ligament laxity significant enough to cause dislocation at birth is unlikely. When knee dislocation is associated with other orthopedic conditions, such as clubfoot or arthrogryposis congenita, diagnosis and definitive treatment are more difficult (9). Given the small stature of the mother, we consider it plausible that the limited space in the uterine cavity could have caused the fetus's foot to press against the uterine wall (8). As fetal movements continued, the leg could have slipped forward, leading to hyperextension and dislocation, especially if the lateral displacement of the patella occurred simultaneously. This would allow the tibia to dislocate anteriorly (Figure 3). In neonates, the patella is still small and cartilaginous, limiting its stabilizing function against dislocation. Once the knee is reduced, the patella returns to its position and resumes its stabilizing function. Effective management includes the serial application of splints, allowing the knee to return to a normal shape and function within a few weeks, as observed in this case (10). Previous studies suggest that early conservative treatment, including serial casting, is highly effective in congenital knee dislocations and may prevent the need for surgical intervention (5,7,12). Other authors also suggest the use of a splint with traction (4). Based on our experience, we recommend replacing the splint every 15–20 days during the first two months. This approach allows for regular assessment of joint stability, gradual adjustment of flexion angles if needed, and prevention of compression-related complications due to growth. From our observations, we have developed a treatment protocol for future cases. This protocol includes an initial clinical evaluation to assess the limb's neurovascular status and identify any associated congenital anomalies. Treatment begins immediately after diagnosis with the application of progressive casting. Routine X-rays are not performed unless there is diagnostic uncertainty. The necessity of radiographic imaging remains a topic of debate. Some authors consider X-rays essential for staging and monitoring purposes (1,2,3,11), while others argue that they are not mandatory, particularly in cases where the dislocation is clearly reducible at birth (5). We did not obtain an X-ray before starting treatment, because reducibility was evident at birth. Instead, we considered it more beneficial to perform an X-ray following treatment to confirm success, demonstrate the absence of bone complications, and minimize radiation exposure for the newborn. The newborn's gradual development of motor skills, from crawling to walking, allows time for the joint to heal without weight-bearing stress, supporting full recovery. Surgical intervention is considered in cases where excessive quadriceps contracture prevents successful reduction, when knee flexion remains limited to less than 30 degrees, or if conservative treatment fails. As recommended by

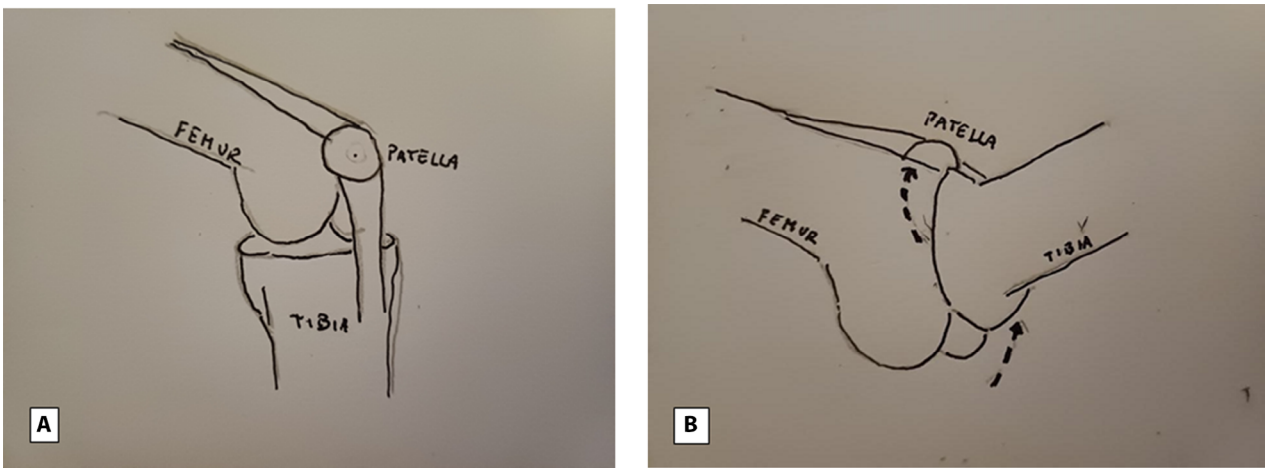

Figure 3. A) The normal relationships between the articular structures B) The modification of the relationships in the dislocation.

other authors, surgical management may involve techniques such as external fixation, percutaneous quadriceps lengthening, or quadricepsplasty, depending on the severity of contracture and joint stiffness (1,2,5,8). Early surgical intervention may be necessary in cases of severe congenital knee dislocation to achieve stable and functional outcomes (7,8). External fixation has been reported as an effective approach for maintaining gradual correction, while quadricepsplasty or tenotomy with or without arthrolysis can help release excessive soft tissue contractures and improve knee mobility (5). Surgical interventions always represent a complex challenge even in post-neonatal ages (12).

Conclusion

The diagnosis is primarily clinical, with gentle manual reduction as the first treatment approach recommended in the literature. Early reduction helps prevent stress on the muscles, tendons, and vascular structures around the knee. While these tissues are initially elastic enough to tolerate strain, prolonged tension could lead to damage and local inflammation. Joint cartilage may be similarly affected. Screening for associated organ anomalies and genetic alterations is recommended, even in isolated cases. Prompt intervention is crucial, and multidisciplinary collaboration is essential for accurate diagnosis and to evaluate how the condition might impact the individual's overall development, not only in terms of walking ability. However, due to the rarity of such cases and limited data, it is challenging to predict whether the reduced knee will develop early-onset osteoarthritis or undergo normal development. Standardized protocols for diagnosis and treatment should be developed to optimize outcomes and effectively address the emotional impact and concerns of families.

Founding: None.

Ethics Approval and Consent to Participate: Not applicable to this study. The typology of the study (case report) does not require the local ethics committee's approval. All procedures performed in this study were followed with the ethical standards of the institutional and national research committee and with the 1964 Helsinki Declaration and its later amendments or comparable ethical standards.

Consent for Publication: Written informed consent was obtained from the parents. The manuscript follows the Italian Privacy Law.

Availability of Data and Materials: All relevant data are within the paper. All supporting data is available from the corresponding author and Tiziana Corsini, U.O.C of Orthopaedics, Fidenza Hospital, AUSL of Parma, according to Italian Privacy Law.

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Declaration on the Use of AI: None.

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