

Congenital Dislocation of the Patella-A Case Report and Literature Review

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Abstract

Congenital Dislocation of the Patella (CDP) is an extremely rare condition characterized by a permanent, irreducible patella dislocation associated with a fixed flexion contracture of the knee. It should always be differentiated from other more frequent conditions. Several techniques have been described for the surgical treatment, which is mandatory to achieve patella reduction and a normal knee function.

We report the case of a one-year-old girl who was diagnosed at birth by magnetic resonance imaging after a suspicious physical examination. No other medical conditions were identified. She underwent surgical treatment with a modified Langenskiöld technique, aiming for a less invasive approach with no peri or post-operative complications. Six months after surgery she was able to stand and walk properly for her age with knee full extension achieved.

Keywords: Congenital Dislocation; Patella; modified Langenskiöld Technique

Abbreviations

CDP: Congenital Dislocation of the Patella; ROM: Range of Motion; MRI: Magnetic Resonance Imaging

Introduction

Congenital Dislocation of the Patella (CDP) is permanent and irreducible. It is typically associated with a fixed flexion contracture of the knee, genu valgum and sometimes with foot deformities [1-3]. As it is an extremely rare condition it should be distinguished from other patellar instability pathologies due to congenital or genetic causes (as Down's syndrome and nail-patela syndrome) [1]. Treatment is only achievable by surgery [4,5]. We report a case of a toddler with unilateral CDP who underwent surgical treatment with a modified Langenskiöld technique [6-8].

Case Report

A female infant was born at 39 weeks of gestation by eutocic delivery weighting 2890g. After birth a right knee and left foot deformity were noted. She had no other medical conditions. There was no family history of genetic or musculoskeletal disorders. At physical examination the right patella was impossible to find at the anterior aspect of the knee and careful inspection revealed the

patella lying laterally to the lateral femur condyle. The right knee presented flexion contracture at 20°, with extension and reduction of the patella impossible to achieve either actively or passively (Figure 1). The left lower limb presented a clubfoot deformity. CDP was suspected and an MRI was performed which confirmed the diagnosis (Figure 2). Surgical repair was proposed. Regarding left clubfoot deformity, treatment with the Ponseti method was instituted.

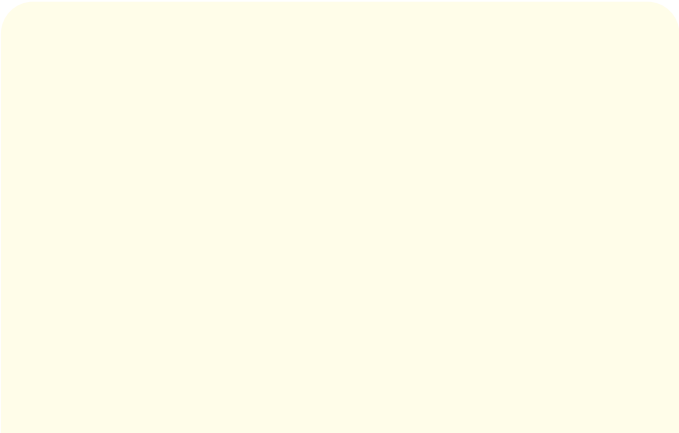


Figure 1: The patella was lying laterally to the femoral condyle of the right knee (a) and associated with a 20° flexion contracture (b).

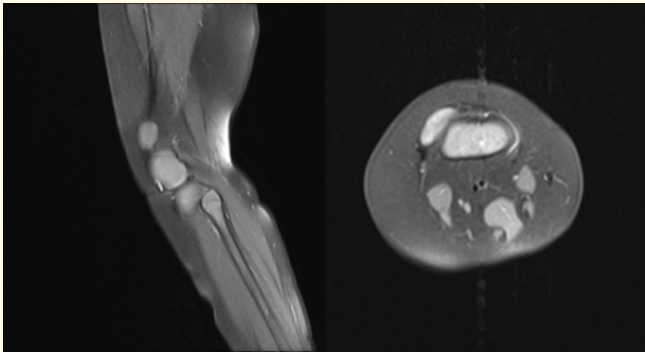


Figure 2: MRI image of the right knee showing the dislocation of the patella.

At one year old she was submitted to surgical treatment of the CDP according to a modified Langenskiöld technique introduced by Paley as part of the surgical management of congenital femoral deficiency [6].

A 6 cm anterior knee incision was made starting proximally to the femur trochlea towards the inferior pole of the tibial tubercle (Figure 3). Blunt dissection was used to isolate the patellar retinacula, the patella, the patellar tendon and the quadriceps femoris. The iliotibial band and the tendinous part of the vastus lateralis and the posterior capsule were released. The synovial layer of the knee was dissected, and the patella remained attached only to the quadriceps tendon and the patellar ligament (Figure 4). The hole in the synovium layer was sutured in a longitudinal direction, and a new longitudinal line was generated on the synovium at the center of the desired position for the patella. An incision was created at this new position, and the patella was secured circumferentially to the synovium with a running absorbable suture. The medial capsule with the vastus medialis was advanced over the patella toward the lateral border and sutured, while the lateral capsule was left

open. After releasing the tourniquet, the knee joint was moved to ensure the patella was stable till 90° range of motion (ROM). A 5° flexion contracture persisted which we consciously accepted (Figure 5). Superficial layers were sutured.

Figure 3: A 6cm anterior approach was made. Patella is dislocated laterally.

Figure 4: The patella was completely isolated from the capsule and only remained attached to the quadriceps and patella tendons.

Figure 5: The medial capsule with the vastus medialis was advanced over the patella toward the lateral border and sutured, while the lateral capsule was left open.

The knee was immobilized in full-extension and serial long leg cast were made during the first 6 weeks. Then, the cast was removed, progressive range of motion and weight bearing started. Six months after surgery she was able to stand and walk properly for her age with knee full extension achieved (Figure 6).

Discussion

There has been significant confusion regarding CDP. It should be distinguished from other types of patellar instability conditions that are recurrent, habitual or permanent reducible dislocations [1,5]. In those, patella dislocation occurs after birth, during childhood or adolescence and slowly becomes permanent and irreducible. It might be present in Down syndrome, nail-patella syndrome, Kabuki syndrome, and Ellis-van Crevald syndrome [5,9]. In opposition, CDP which is extremely rare, is an embryological malformation as described by Stanisavljevic, *et al.* It occurs within the first trimester of intrauterine life and consists of a failure of the internal rotation of the myotome containing the quadriceps femoris and patella [3].

CPD manifests at birth as a permanent, irreducible dislocation associated with a flexion contracture. Physical examination is challenging due to the small size of patella at early age. However, the absence of the patella should be noticed in the anterior aspect of the knee and careful inspection should obviate its presence lying laterally to the lateral femur condyle [10,11]. In our case, dislocation of the patella was noted immediately after birth and all major syndromes were excluded before establishing diagnosis.

Ossification of the patella does not become radiographically evident till 2-3 years of age. Ultrasonography has become an useful technique to locate the unossified patella and confirm the diagnosis [10-13]. However, as we reported in our case, MRI not only confirms the diagnosis, but also clearly identifies the location of the patella and provide significant information on anatomic and soft-tissue anomalies being recommended pre-operatively [5,11,12].

The patella cannot be repositioned without surgery in CPD. However, serial casting and bracing have been described as effective for the minimization of flexion contracture [14].

Optimal timing and treatment strategy is yet to be defined. Some advocate surgical treatment should be performed as soon as possible [14,15]. Nonetheless the size of the structures at an early age might hamper the surgical technique and increase the risk of intraoperative complications which is why we decided to delay the surgery until the patient was 1 year-old. Numerous technical procedures and results are reported in the literature. Ganhem, *et al.* reported good results with an extensive quadriceps release and

derotation on 8 cases with an average follow-up of 6.9 years. All the patients were able to walk on their operated limb, and the patella was centered in the trochlea in all the cases [14]. Wada, *et al.* reported 6 patients with an average age of 2.1 years treated with a combination of lateral release, medial plication, V-Y lengthening of the quadriceps, medial transfer of the lateral patellar tendon and posterior release of the knee.

Improvement in the flexion contracture of the knee was achieved in five of seven knees by serial casting and/or a brace. Genu valgus and external tibial torsion improved after surgery in all knees. The operated knee was mobile in all cases, with less than 10° flexion contracture of the knee [5]. Camathias, *et al.* reported poor outcomes with an extensive release procedure essentially involving medializing the entire lateral quadriceps and medial soft tissue stabilization was performed as described by Stanisavljevic [16]. The failure rate was up to 80% in a 20 knees series with recurrent dislocation of the patella or CDP, a mean operative age of 12.8 years and a mean follow up of 7.5 years.

A modified Langenskiöld procedure which consists of patella medialization within the synovium, distal patellar tendon realignment without detaching the distal insertion of the patella, VMO advancement, lateral release and in the case of congenital patella dislocation with external rotatory subluxation of the tibia on the femur, lengthening of biceps femoris and iliotibial band, was performed by Ramos, *et al.* The authors reported 18 knees in 13 patients with a mean age of 15,8 years and diagnoses of recurrent (6 patients, 8 knees), chronic (4 patients, 6 knees) and congenital (3 patients, 4 knees) dislocation of the patella. There was no recurrence in recurrent and congenital groups. All patients achieved full extension postoperatively [8].

Mo, *et al.* also described excellent results with this technique, performed on 11 patients with 16 knees enrolled and a mean age at the time of operation of 3.1 years. There were no recurrent dislocations at a mean follow up of 37.8 months, and all patients had full extension postoperatively [7].

In our case, we chose to perform the modified Langenskiöld technique. In an attempt to be less invasive, we made a smaller incision and minimized the release of the knee structures. Distal patellar tendon realignment was not made. We believe that it is not a mandatory procedure to achieve a full ROM and a good knee function in early ages. However, we still don't know if further surgeries will be needed. It seems obvious that an extensive mobilization of the knee structures will implicit knee disfunction and degenerative alterations over the years. We believe that by a conservative ap-

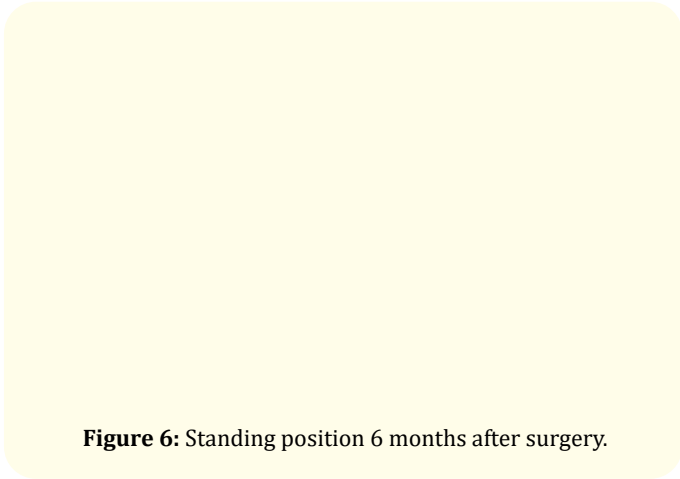


Figure 6: Standing position 6 months after surgery.

proach we would preserve the long-term knee dynamics and function. We tolerated a 5° flexion after surgery that was fully corrected by serial casting and physical therapy at the end of 3 months.

Conclusion

CPD is a very rare condition that should be differentiated from other more frequent conditions. The surgical treatment is mandatory, and we believe that modified Langenskiöld technique seems to be reliable and reproducible. We highlight the possibility of being less invasive in order to avoid future complications without compromising outcomes of this technique.

Conflict of Interest

The authors have no conflicts of interest to declare.

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