Management of Congenital Neglected Dislocation of Patella: A Case Report

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Abstract: 14-year-old boy presented with deformity in the right knee joint since childhood, associated with a limp. On examination he was found to have laterally dislocated hypoplastic patella which was irreducible along with week quadriceps. The same was managed surgically by patella repositioning and reconstruction by lengthening the Quadriceps, transfer of tibial tuberosity medially, medial plication and distalisation of Vastus Medialis Obliquis (VMO). Additional stability was provided using semitendinosus. The key steps followed in the surgical reconstruction have been described in the article and reported. Such a presentation is rare and reconstructive procedures to be carried out vary based on age of the patient and findings that exist in each case.

Keywords: Congenital dislocation patella, Quadriceps mechanism, Realignment, operative treatment.

INTRODUCTION

The patellofemoral joint has a low degree of congruency. Stability depends on [1] bony structures – patellar and trochlear shape and [2] soft tissue structures – quadriceps muscle, in particular, the vastus medialis obliques, lateral retinaculum, medial retinaculum, medial patellofemoral ligament (MPFL), and medial patello-tibial ligament [1]. The major restraints in the initial knee flexion (<30°) are the soft tissue structures, but after 30°, patellar stability depends on bony anatomy [2]. Major instability factors as described by Dejour are patellar tilt, tibial tubercle–trochlear groove distance as measured by axial computed tomography (CT) or magnetic resonance images, patella alta, and trochlear dysplasia [3]. The secondary instability factors are excessive external femoral rotation, excessive external tibial rotation, genu recurvatum, and genu valgum. These instability factors predispose to injury of the MPFL which is the major soft tissue restraint which contributes an average 53% of the total force that prevents lateral displacement of distal knee extensor mechanism [4].

The MPFL is a distinct soft tissue structure within the medial retinaculum, which originates from saddle between the adductor tubercle and epicondyle and inserts at the superior two-thirds of medial border of patella, typically at the location where the perimeter of the patella becomes more vertical. It is approximately 55 mm long and its width ranges from 3 to 30 mm. This ligament is most taut in full extension, with the quadriceps contracted, and assists in guiding the patella into the trochlea during the early stages of flexion. The common attachment of the tendon of the vastus medialis muscle and the ligament to the superiomedial patella suggests that there may be a dynamic element for stability. Patellar tracking is significantly affected by a lateral force in the absence of the MPFL, but returns to normal following reconstruction [5]. In nearly all patella dislocations, there is damage to the MPFL. 70% is damaged at the patellar insertion while the remaining is damaged at the femoral origin. In all, however, there is also interstitial damage [6, 7]. MPFL reconstruction is most helpful for patients who have recurrent patellar dislocations associated with hyperelasticity and for patients who have obligatory patellar dislocations. Both of these scenarios are associated with incompetent medial retinacular tissue [8].

Congenital dislocation of the patella is a well-known but rare entity that occurs in 2 forms. The first form occurs as a permanent lateral dislocation that is irreducible without operative intervention and the second as a recurrent dislocation of the patella [9, 10]. The second form is more common, and symptoms begin later in childhood (at 11–12 years) compared with those of permanent congenital dislocation [9]. Patients with permanent congenital dislocation of the patella can be subdivided further into 2 groups. In the first, the patella may be dislocated irreducibly and hypoplastic at birth, generally present with flexion contracture. In the second group, the patella is located in the trochlear groove at birth and progressively becomes fixed in a laterally dislocated position by early childhood [9]. Children with congenital permanent dislocation of the patella frequently are not diagnosed until they have begun to walk; this probably is due to the normal-appearing gait as well as the fact that the patella does not begin to ossify until 3 years of age. In addition, the patella is usually hypoplastic. Congenital dislocation of the patella has been reported as being associated with...
syndromes such as Down’s syndrome and nail-patella, diastrophic dysplasia, dislocation of the radial head, fibular hemimelia, talipes equinovarus, carpal fusion, and cardiac anomalies [9-15]. On physical examination, full active extension always is limited, and there is marked genu valgum as well as external rotation of the tibia [9-11, 13-15].

However, the treatment of older patients with a fixed lateral patella dislocation can be challenging. Most patients are surgically treated in a timely manner with appropriate conservative or surgical management to maximize their overall function. We present a case of a 14 years old boy with a history of congenital persistent dislocation of patella and underwent delayed surgical reconstruction.

CASE REPORT

A 14 year old male child presented with deformity in the right knee and difficulty in walking due to weakness and muscle wasting. He developed pain over right knee on weight bearing for the past 6 months. Developmental milestones were normal and no relevant family history. On examination, gait was somewhat dragging with patella displaced laterally and high riding. (Figure 1, 2). No limb length discrepancy was noted. Physical examination revealed painless movements of the knee with limitation in terminal degrees. Movement of ankle and hip was normal. No neurovascular deficits. No other congenital anomalies noted. Blood count and biochemical profile were within the reference ranges. Radiographs showed laterally displaced and high riding patella with hypoplasia and shallow trochlea. (Figure 3).

Under General Anaesthesia, application of tourniquet and adequate preparation a midline incision of 30 cm was made over the right knee joint extending proximally up to mid-thigh and distally below tibial tuberosity. (Figure 2). Lateral release was performed (Figure 4) by dividing the quadriceps from the fibrous adhesions to the iliotibial band and the lateral intermuscular septum. The lateral capsule was incised laterally to the dislocated patella and along the lateral border of the patellar tendon to the tibial tuberosity. The medial capsule and retinaculum were incised medially to the patella. Lateral release performed, quadriceps V-Y plasty done and patella mobilised centrally over trochlea groove. (Figure 5). Trochlea groove found to be hypoplastic and shallow with absence of median ridge. Patella was found to be hypoplastic. Next, tibial tuberosity was osteotomised, shifted distally and medially along with patella tendon in order to align patella in patellar groove. (Figure 7, 8). Vastus medialis advanced to the superiomedial pole of patella to enforce medially. Position of patella checked in flexion and found to be unstable at 90° flexion. This was taken care of with the help of Semitendinosus which was divided proximally, withdrawn at its insertion, looped around superior pole of the patella and sutured on itself. This stabilised the patella at 90° flexion of knee (Figure 5 &6). Post-operative x rays (Figure 8, 9) was taken and the limb was protected with above knee cast at about 30° flexion for 3 weeks, non-weight bearing and then gradual weight bearing as tolerated was allowed after 4 weeks. Review was done thrice in the first post-operative year and found to be good.
DISCUSSION

Congenital dislocation of the patella often is familial and bilateral. Occasionally, it is accompanied by other abnormalities, especially arthrogryposis multiplex congenita and Down syndrome. It is persistent and irreducible and usually accompanied by abnormalities of the quadriceps mechanism. The vastus lateralis may be absent or severely contracted, and the patella may be dislocated laterally and attached to the anterior aspect of the iliotibial band. Often the patella is small and misshapen and in an abnormal location in the quadriceps mechanism. Genu valgum and external rotation of the tibia on the femur commonly develop. The capsule on the medial side of the knee is stretched, the lateral femoral condyle is flattened, or the insertion of the patellar tendon is located more laterally than normally [9-15].

Eifert noted that two clinical syndromes have been described in the literature: congenital dislocation of the patella or fixed lateral dislocation of the patella and habitual dislocation of the patella, which he suggested should be more accurately termed “obligatory dislocation” of the patella. Persistent Dislocation means Patella is dislocated lateral and persistent in that location. When associated with generalized syndrome, Knee flexion contracture is present, which produces functional disability and early surgical correction is necessary for good functional outcome. Both static and dynamic forces tend to displace the patella laterally [16].

Brattström first described the Q angle as an angle formed by the line of pull of the quadriceps mechanism and that of the patellar tendon as they intersect at the center of the patella. Clinically, this angle is represented by the intersection of a line drawn from the anterior superior iliac spine to the center of the patella with a second line drawn from the center of the tibial tuberosity to the center of the patella. For this measurement to be accurate, the patella must be centered on the trochlea by flexing the knee 30 degrees. In males, the Q angle normally should be 8 to 10 degrees; in females, the normal angle is 15 degrees ± 5 degrees. This valgus angle gives a lateral force vector to the patellofemoral joint as the knee is extended. The factors that can increase this Q angle are genu valgum, increased femoral antversion, external tibial torsion, a laterally positioned tibial tuberosity, and a tight lateral retinaculum. Any of these factors that increase the Q angle can be a contributing factor in recurrent dislocation of the patella. The shape of the patella itself also can be a contributing factor. An abnormally small or dysplastic patella can interfere with the smooth gliding motion of the patella in the intercondylar groove. A high-riding patella (patella Alta) is considered by many authors to be the most common cause of recurrent dislocation or subluxation of the patella. Dysplasia of the femoral condyles also can be a contributing factor. Patients with a hypoplastic lateral femoral condyle have lost a bony support that prevents lateral subluxation of the patella from the intercondylar groove [17].

The diagnosis of congenital dislocation of the patella is difficult to make before the patient is 3 to 4 years old because of lack of ossification of the patella. MRI can show the cartilaginous patella lying lateral to the femur and can confirm the diagnosis when congenital lateral patellar dislocation is suspected. Several authors have described the use of ultrasound to define the position of the cartilaginous patella. Because the severity of the deformity is directly related to the length of time that the deformity is allowed to remain uncorrected, surgery can be done as soon as the diagnosis is made to try to prevent a valgus, flexion, or external rotation deformity of the knee.

In a group of 35 patients with congenital or habitual dislocation of the patella, GAO, Lee, and Bose found the underlying pathological condition to be contracture of the quadriceps mechanism in all; the contractures were more severe in patients with congenital dislocations. Operative techniques vary according to the extent and degree of these operative
findings. The primary objective is release of the contracted structures on the lateral side of the patella (the lateral capsule, iliotibial band, and lateral portion of the quadriceps) to allow reduction of the patella. Medial plication of the lax capsule is necessary to stabilize the reduced patella [18].

Gao et al.; obtained satisfactory results in 88 patients with extensive lateral release, medial plication, and transfer of the lateral half of the patellar tendon [18]. Langenskiöld and Ritsilä reported successful treatment of 18 congenitally dislocated knees with lateral release and medial transfer of the patellar tendon [19]; Gordon and Schoenecker treated 10 patients with 13 involved knees using lateral release and vastus medialis obliquus advancement and entire patellar transfer in skeletally immature patients and medial transfer of the tibial tubercle in skeletally mature patients. At an average 5-year follow-up, all patients reported pain relief and a marked increase in activity tolerance [20].

CONCLUSION
Congenital dislocation of the patella should be suspected in every child with knee flexion contracture, genu valgus, external tibial torsion, foot deformity and delayed walking. Better results are seen when the child is operated at younger age however well planned and meticulously executed surgical procedures at maturity also give satisfactory results.

ABBREVIATIONS
VMO-Vastus Medialis Obliquus, MPFL-Medial patellofemoral ligament, CT-computed tomography, GA-General Anaesthesia, Q angle-Quadriceps Angle.

REFERENCES