# Habitual Patella Dislocation in a patient with Osteogenesis Imperfecta

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#### Citation

Vaidya RK, Pradhan P. Habitual Patella Dislocation in a patient with Osteogenesis Imperfecta . *Kathmandu Univ Med J.* 2023;84(4):448-52.

# **INTRODUCTION**

Habitual dislocation of patella, also referred to as obligatory dislocation of patella, is a condition when patella dislocates every time the knee is flexed and relocates spontaneously in extension.<sup>1</sup> Unlike in case of recurrent dislocation, habitual dislocation is usually painless and is not accompanied by trauma.<sup>2</sup> It usually presents as child begins to walk. There is usually no flexion contracture as in case of persistent dislocation of patella.<sup>3</sup> However, with age, dysfunction and deformity might be noticeable in these children.<sup>1</sup> It is a rare entity among the adults. Moreover, its occurrence in patient with osteogenesis imperfecta (OI) has not yet been documented. Various surgical techniques have been described in the literature for habitual patella dislocation, but there is no single standard procedure shown to be effective.<sup>1</sup> Surgery, in case of its association with OI, is even more challenging, as bones are malunited and muscles are weak.

# ABSTRACT

Habitual patella dislocation is a rare entity and its occurrence with osteogenesis imperfecta (OI) has not yet been documented. Various surgical techniques have been described in the literature for its treatment, but no single standard procedure is shown to be effective. A 24-year-old girl, diagnosed with osteogenesis imperfecta, presented with complaints of pain and instability in both knees, more on the right. She was diagnosed to have right habitual patella dislocation. She had prominent quadriceps muscle wasting and X-rays confirmed patella baja. CT scan and MRI suggested trochlear dysplasia, Dejour type A and a normal Tibial tuberosity (TT) - Trochlear groove (TG) distance. She was operated with lateral release, medial plication and tibial tuberosity transfer. She had a full knee range of motion (ROM) and pain-free ambulation in four months. In one year follow-up, she had good functional outcome.

#### **KEY WORDS**

Habitual patella dislocation, Lateral release, Medial plication, Osteogenesis imperfecta, Tibial tuberosity transfer

# **CASE REPORT**

A 21 years old girl diagnosed with OI presented to Orthopedics outpatient department (OPD) with complaints of mild pain and instability in both her knees, especially on the right. Her patella dislocated every time she flexed the knee and spontaneously relocated on extension. Her left patella subluxated on flexion. She was diagnosed with right habitual patella dislocation.

Upon examination, she had features of OI: short stature, bluish sclera, dentinogenesis imperfecta, history of fracture of multiple bones and weak muscles (fig. 1A). There was also anterolateral bowing of her right femur due to malunion after fracture of femur 7 years back. Physical examination showed that she had prominent quadriceps and thigh muscles wasting. Although her patella remained in position during extension, it shifted laterally upon every flexion (fig. 1 B,C). Passive range of motion (ROM) was painless, but active ROM was painful during active extension. Stand up



Figure 1. Features of OI: Bluish sclera, dentinogenesis imperfecta B. Patella in position in extension C. Patella laterally dislocated in flexion

from a sitting position was painful and needed support of the hands on thighs. Q-angle on the right knee was found to be 20°. Craig's test performed showed increased anteversion angle on both sides, more on the right (fig. 2).



Figure 2. Craig's test showing increased femoral anteversion angle on either side

Plain radiographs obtained showed "patella baja" in lateral view (fig. 3) and completely dislocated patella in skyline view. Usually, patella alta is associated with patellar instability. Her long leg standing X-Ray showed anterolateral femoral bowing (fig 4). CT scan showed shallow trochlear groove with trochlear depth 1.12 mm, medial to lateral facet ratio 38%, trochlear angle 167° and TT-TG distance 9.3 mm (fig 5). Features were suggestive of trochlear dysplasia, Dejour type A. As per MRI, medial patello-femoral ligament was found to be elongated without any obvious tears.



Figure 3. Patella baja with A. Insall-Salvati index=0.6, B. Blackburne-Peel ratio=0.5, C. Caton-Deschamps index=0.6

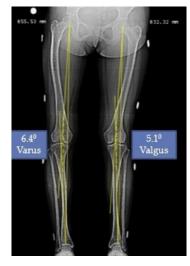


Figure 4. Long-leg standing X-ray

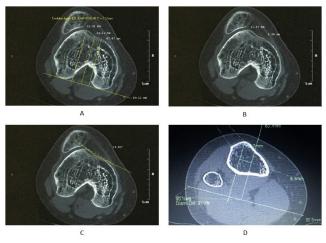


Figure 5. CT scan right knee A. Trochlear depth=1.12 mm, B. medial to lateral facet ratio=38%, C. Trochlear angle=167 degrees, D. TT-TG distance=9.3 mm



Figure 6. Surgical steps: lateral release, TT proximalisation and medialization, medial reefing

After preop rehabilitation, surgery was done under spinal anesthesia with patient in supine position. Lateral parapatellar incision around 15 cm was given extending proximally. Subcutaneous flaps were raised medially to explore the soft tissue around the patella. Ilio-tibial band was partially excised. Vastus lateralis obliquus and vastus intermedius were identified and released followed by lateral retinaculum graduated release. As she had patella baja, tibial tuberosity was transferred proximally and medially (to compensate for increased femoral anteversion although TT-TG distance was < 10 mm,) (fig. 6). Femoral derotation surgery was not performed. The TT was fixed with a 6.5 cm cancellous screw. Finally medial reefing was done. Soft tissue balancing was checked at every step to ensure proper patellar-tracking in full knee range of motion. Closure of the wound was done in layers with a



Figure 7. Post-op X-ray rt knee AP and Lat.

hemosuction drain and the operated leg was immobilized in a long posterior leg back slab.

Post-surgery, the knee was immobilized in the slab for 4 weeks and only touch-weight bear was allowed. Gradual knee ROM exercise and isometric exercises were started early in 3<sup>rd</sup> weeks post op to prevent stiff knee. Partial weight bearing was allowed from the 5<sup>th</sup> week and complete weight bearing after 12 weeks post-surgery. At 4 months follow-up, patient had full knee ROM and was able to perform all house-hold works without any difficulty. During 1-year follow-up, the operated knee was stable, painless and had full range of motion (fig. 8). There were no episodes or tendency to dislocation. In her post op one year X-ray, Insall-Salvati index was 1.1, Blackburne-peel ratio 1.2 and Caton-Deschamps index 1.2 (fig. 10).



Figure 8. Follow-up 1 year flexion and extension, patella in position during flexion



Figure 9. One year post-op skyline view

Figure 10. One year Post-op A. Insall-Salvati index=1.1, B. Blackburne-Peel ratio=1.2, C. Caton-Deschamps index=1.2

# DISCUSSION

There are no cases of habitual patellar dislocation in young adult with osteogenesis imperfecta described in the literature. The cardinal physical sign in habitual dislocation is that with patella in the midline, flexion is only possible upto 60-70 degrees and further flexion can occur only when patella dislocates.<sup>4</sup> Patella dislocation is primarily due to an imbalance of the 4 quadrant vectors around the patella, and this may be congenital or acquired. The principle pathophysiology is contracture of the soft tissues lateral to patella.5 In most of the cases, vastus lateralis contracture is the main pathology.<sup>6</sup> Lateral patellar retinaculum contracture, tight ilio-tibial band and/or tensor fascia lata, weak vastus medialis, elongated or torn medial patella-femoral ligament, hypertrophy of vastus lateralis, attenuation of medial patellar retinaculum, generalized ligamentous laxity, shallow trochlear groove, dysplastic lateral femoral condyle, patella alta, abnormal patellar facets, genu valgum, genu recurvatum are other predisposing factors.<sup>6</sup> In our case, however, patella baja was associated with dislocation which is a rare condition.

Due to the presence of multiple factors responsible for habitual dislocation, there are many surgical procedures advocated for its management.<sup>3</sup> No single procedure has shown to be effective in its treatment. A combination of various procedures involving proximal and distal reconstructions are advised.<sup>1,3,5,7</sup> As pathology is primarily proximal, proximal procedures are done first and distal procedures are added when necessary.<sup>5</sup>

Williams described the soft tissue realignment procedures in habitual dislocation of patella.<sup>8</sup> He suggested to begin with extensive lateral release followed by division of vastus intermedius or elongation of rectus femoris if required. If per-operatively, there is still tendency to dislocation, he advocated distal realignment.<sup>8</sup> Twenty years later, Williams suggested that proximal release was always necessary and quadriceps lengthening was an essential part of the treatment.<sup>2</sup> Bakshi suggested release of any superolateral contracture followed by lengthening of rectus femoris with or without vastus intermedius as per requirement.<sup>9</sup> He advocated pes anserinus sling procedure to be better that vastus medialis advancement to the lateral border of patella.<sup>9,10</sup> He also said that TT transfer might be useful in adults but could cause genu recurvatum in children. Gao

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et al. performed extensive lateral release, medial reefing and transfer of lateral half of patellar tendon and rectus femoris lengthening in few cases.<sup>11</sup> Joo et al. performed "Four-in-One" procedure (lateral retinacular release, medial vector augmentation, semitendinosus tenodesis and transfer of the patellar tendon).<sup>12</sup> Shen et al. performed combined proximal and distal procedures (lateral release, medial retinaculum advancement and anteromedial tibial tubercle transfer) in 12 patients with habitual dislocation of patella.<sup>13</sup> Benoit et al. performed distal advancement of the patella by complete mobilisation of the patellar tendon, lateral release and advancement of vastus medialis obliquus.14 Kwak et al. used corrective osteotomy of distal femur along with lateral release and medial reefing.15 Mittal et al. performed "modified Four-in-One" procedure (lateral retinaculum release, vastus medialis obliques advancement, partial patellar tendon transposition and reconstruction of medial patellofemoral ligament.<sup>3</sup>

As our patient was a known case of OI with history of multiple fractures and weak muscles, surgery was performed in a different way to minimize excessive bony or soft tissue procedures. Although there was significant anteversion of the femur, derotation surgery was not planned. As per the usual trend, we began with extensive lateral release. Surgical indication of tibial tubercle osteotomy (TTO) has been TT-TG distance of more than 20 mm as described by Dejour et al. and this has been the standard for TTO.<sup>16</sup> But as there was patella baja, we had to proximalise TT; and we believed medialization of TT can also compensate for the rotational factor. So, TT was medialized as well although TT-TG distance was around 9.3 mm. Intra-operatively, patella was checked for dislocation after medial plication. As there was no tendency for dislocation, MPFL reconstruction was not performed.

Habitual patella dislocation is an unusual case in adults and is due to contracture of one of the elements of quadriceps muscle. Operative correction usually involves quadricepsplasty followed by other proximal and distal procedures as per requirement and there is no standard protocol for its treatment. This case again implies on the surgical dilemma in treating habitual patella dislocation, specially complicated with other syndromes, and highlights the fact that per-operative decision on how much to intervene, does matter in the outcome.

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