Case Report

Neglected surgically intervened bilateral congenital dislocation of knee in an adolescent

Jaswant Kumar, Ish Kumar Dhammi, Anil K Jain

Abstract

Neglected bilateral congenital dislocation of knee is unusual. A 12 year old boy presented with inability to walk due to buckling of the knee. The symptoms were present since the child learnt walking. He preferred not to walk. Bilateral supracondylar femoral osteotomy was done at the age of 6 years. Patient had a fixed flexion deformity of both knees, 30° in the right (range of flexion from 30° to 45°) and 45° fixed flexion deformity in left knee respectively (range of flexion from 45° to 65°) when presented to us. The radiological examination revealed bilateral congenital dislocation of knee (CDK). No syndromic association was observed. He was planned for staged treatment. In stage I, the knee joints were distracted by Ilizarov ring fixators and this was followed by open reduction of both the knee joints in stage II. A bilateral supracondylar extension osteotomy was done 18 months after the previous surgery (stage III). The final followup visit at 4 years the patient presented with range of motion 5-100° and 5-80° on the right and left knee respectively with good functional outcome. The case is reported in view of lack of treatment guidelines for long standing neglected CDK in an adolescent child.

Key words: Congenital dislocation of knee, neglected congenital dislocation of knee, surgical treatment

Introduction

Congenital dislocation of knee (CDK) is a rare orthopedic birth anomaly and was first described by Chatelaine in 1822 (quoted by Shattock in 1891).[1] It represents a spectrum of hyperextension of knee to translation of femur over tibia. CDK is around 100 times less common than congenital hip dislocation and is diagnosed prenatally by ultrasonography.[2,3] Developmental dysplasia of hip and clubfoot are the commonly associated anomalies of CDK and represent about 70% and 50%, respectively.[4,5] Anomalies of upper limb, face, genitourinary system and gastrointestinal system are uncommon association with CDK.[6]

Various etiological factors have been proposed for CDK, such as abnormal fetal positioning, quadriceps fibrosis/atrophy, patellar hypolasia, contracture of iliotibial band, lack of formation or atrophy of suprapatellar pouch, lack or hypoplasia of cruciate ligaments, ligament laxity syndrome, arthrogryposis and spinal dysraphism. Bilateral CDK is almost always syndromic, most commonly associated with laxity syndromes, Ehlers–Danlos syndrome, Larson's syndrome, or Beal's syndrome.[6,7]

Treatment of CDK is either nonsurgical or surgical. The nonsurgical treatment includes manipulation, bracing and splinting and surgical treatment includes quadricepsplasty with or without femoral shortening osteotomy, with or without anterior cruciate ligament (ACL) reconstruction.[7]

The management of neglected CDK has been described only in a single case report.[8] Surgically intervened neglected bilateral CDK has not been described in literature. We discuss here one such case managed by staged surgical procedures.

Case Report

A 12-year-old male presented with inability to walk even a few steps due to buckling of knee and striking of both knees while taking a few steps, which was present since the child learnt walking after birth. The child preferred not to walk for these reasons. His mother noticed deformity immediately after birth. She, being illiterate, could not describe the deformity. There was no history of birth trauma and the child was born of normal vaginal delivery.

Department of Orthopaedics, Guru Teg Bahadur Hospital and University College of Medical Sciences, Delhi, India

Address for correspondence: Dr. Jaswant Kumar,
Department of Orthopaedics,Guru Teg Bahadur Hospital and University College of Medical Science, Delhi - 110 095, India.
E-mail: drjaswant20@gmail.com

Access this article online	
Quick Response Code:	Website: www.ijoonline.com
	DOI: 10.4103/0019-5413.125524

He was the first sibling of three brothers. The parents were apparently healthy and there was no family history of congenital, metabolic, or neurological disorders. The child developed his milestones normally. There was no evidence of any other congenital anomaly and the child was advised about the need for surgery at the age of 5 or 6 years by a village doctor. At the age of 6 years, the patient underwent surgery at the level of lower thigh, presumably bilateral supracondylar femoral osteotomy. The child continued to have inability to walk even after surgery. He consulted this institute at the age of 12 years.

General physical examination revealed stunted growth with no joint laxity. There were no other associated congenital anomalies; local examination revealed a fixed flexion deformity of 30° with further range of flexion of 15° on the right side and a fixed flexion deformity of 45° on the left side with further range of flexion of 20°. There was prominence anteriorly confounding to tibial plateau bilaterally. There was a surgical scar of around 8 cm on the lateral aspect of both thighs at the level of the upper two-third and lower one-third junction. The popliteal area was full and the contours of femoral condyles were palpable in this area. There was wasting of quadriceps muscles.

Plain radiograph showed hypoplastic femoral condyles displaced posteriorly in relation to upper end of tibial. The upper tibial surfaces were convex. The patellae were hypoplastic, located above and anterior to femoral condyles [Figure 1a, b]. Magnetic resonance imaging (MRI) of both knees revealed medial bowed femur, trumpet shaped metaphysis and small epiphysis, posterior subluxation of knee, splayed capsule and attenuated ACL and posterior cruciate ligament (PCL) [Figure 2a, b].

An informed consent was obtained from the parents of the patient for publication of the data. The surgical correction was planned in stages. First we applied Ilizarov ring fixators in order to distract the soft tissue around knee joints to avoid surgical shortening of femur. The construct consisted of one full and one half rings in femur and two full rings in tibia with hinges at knee joint. The distraction of 1 mm/day was performed till the overlap of tibia and femur was corrected which took 3 weeks. The deformity on the right side was corrected from 30° to 25°, whereas on the left side was corrected from 45° to 30°. The surgery was performed by the senior surgeon (AKJ) [Figure 3a]. In the second stage, open reduction of both femoral and tibial condyles was performed. Both the tibial condyles were convex. The ACL and PCL were markedly attenuated and elongated. The menisci were attenuated, but shaped like tibial condyles. The open reduction could be achieved easily and was stabilized by two cross K-wires [Figure 3b]. The limbs were immobilized with above knee plaster-of-Paris (POP) cast for 6 weeks. The K-wires were removed at 6 weeks and knee mobilization was started. The patient started walking at 6 months postoperatively with bilateral knee ankle foot orthosis (KAFO) and axillary crutches. He continued to walk for the next 1 year. At 1 year followup, the patient had 45° fixed flexion deformity on the right side with further range of motion till 100° and 25° fixed flexion deformity on the left side with further flexion range of motion till 90°. On followup X-rays at 1 year, the convex surfaces of upper end of tibia remolded to concavity [Figure 3c]. The patient was upright, doing his activities of daily living with brace.

A bilateral supracondylar femoral osteotomy (extension osteotomy) was done after 18 months of previous surgery to change the arc of motion (third stage) [Figure 4a, b]. The arc of motion was improved from 5° to 100° on the right side and from 5° to 80° on the left side. At 4 years followup visit, the patient had range of motion of 5°–100° on the right and 5°–80° on the left. More or less, the knees were found to be stable [Figures 5 and 6]. The patient was able to walk with a KAFO. His activity of daily living had improved and was able to drive bicycle. He gained in height also.

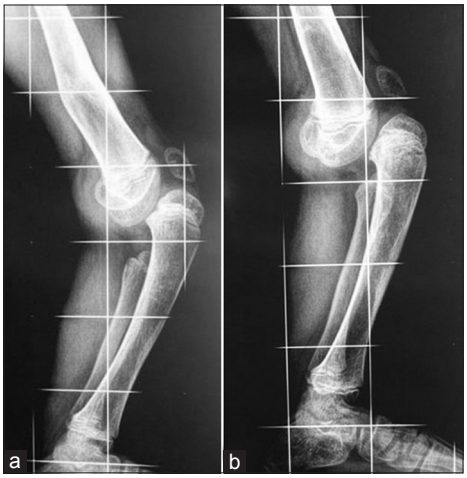

Figure 1: Preoperative x-rays (lateral view) (a) right knee joint (b) left knee joint showing grade III dislocation of both the knee joints

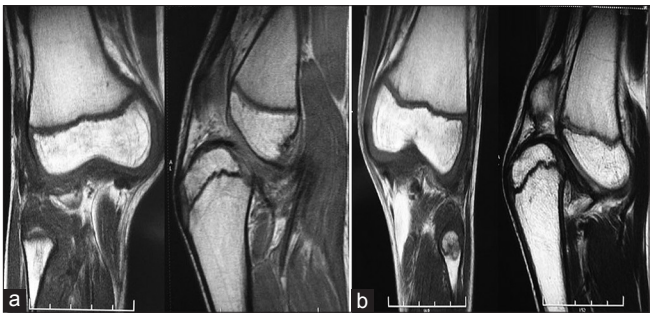

Figure 2: MR images T2WI (coronal and sagital views) (a) right knee joint (b) left knee joint showing small epiphysis, trumpet shaped metaphysis, attenuated ACL and PCL

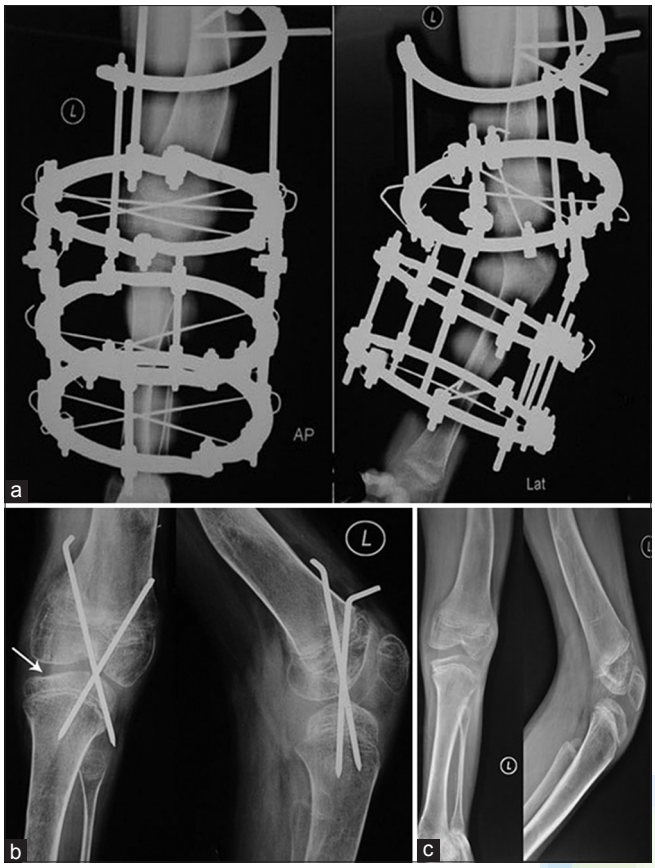

Figure 3: Postoperative x-rays (anteroposterior and lateral views) (a) left knee joint with ilizarov ring fixators showing distraction and alignment of knee joint (stage I) (b) Postoperative x-rays (anteroposterior and lateral views) of left knee joint after open reduction and stabilization with cross k-wires (stage II) (c) Postoperative x-rays after K-wrire removal at 1 1/2 years followup showing remodelling of tibia.

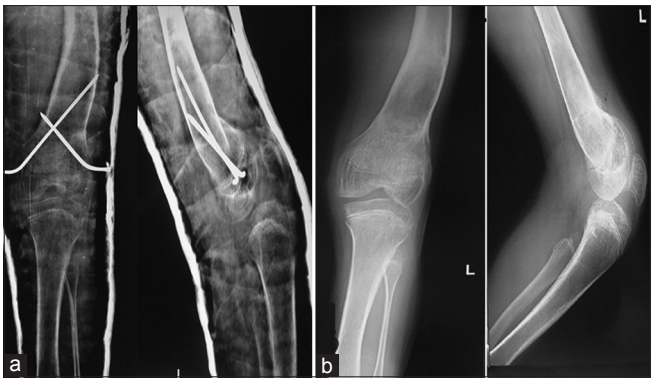

Figure 4: Immediate post operative x –rays (a) (anteroposterior and lateral views) of left knee after 3rd stage surgery "supracondylar extension osteotomy" (b) At 1 year followup x- rays after 3rd stage surgery of left knee showing sound union at osteotomy site

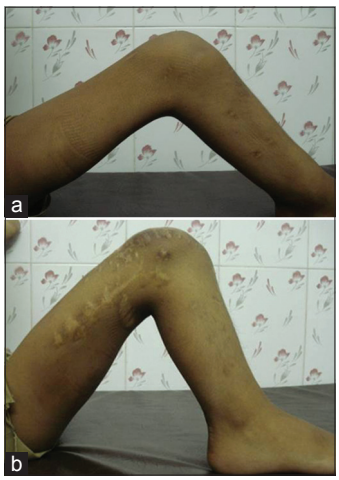

Figure 6: Clinical photograph showing (a) Range of flexion – left knee. (b) Range of flexion – Right knee

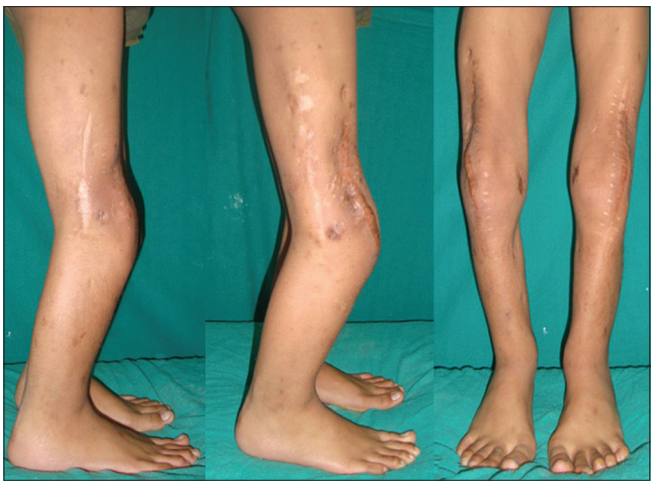

Figure 5: Clinical photograph of patient in standing position showing stable knee joints and almost full extension of knee joints

Discussion

The prevalence of CDK is less than 1 in 1000 live births and it is associated with other congenital anomalies and syndromes, especially when it presents bilaterally.[2,4-6] Therefore, patients should be thoroughly examined to look for such anomalies. In the case reported here, the patient had no other congenital anomaly. Early detection and treatment of CDK provides maximum potential to remodel the articular cartilage. Primary treatment in neonatal period is gentle manipulation combined with splinting and casting. Forceful manipulation is contraindicated because of the risk of fracture separation of epiphysis and insult to physis of proximal tibia.[9-13] In severe quadriceps contractures, which prevent reduction of knee joint, femoral nerve block or botulinum toxin type A can be effective to paralyze the quadriceps and allow the gradual stretching of quadriceps with corrective cast.[6,7] If patient is not responding to nonoperative treatment, operative treatment is indicated. Surgical treatment has been advocated for infants as early as 6 months of age, although good functional result may be achieved even up to 2 years of age.[5,14,15] If the treatment is delayed and the knee remains dislocated, it leaves the patient severely handicapped and will also affect the future growth of the child.

Classically, surgical treatment of CDK is V-Y quadricepsplasty with mediolateral arthrotomy to mobilize anteriorly placed ligamentous structures. Redundant posterior capsule and ACL deficiency were not considered important structures in the past. Unfortunately, quadricepsplasty results in weakness and fibrosis of quadriceps, which leads to extension of lag and limitation of knee flexion.[4,7,14] Redundant posterior capsule and ACL deficiency along with complication of quadricepsplasty may lead to recurrent anterior instability and even redislocation of reduced knee joints.[7] Femoral shortening is done to minimize the extensive quadriceps release. Posterior capsulorrhaphy with or without ACL reconstruction is advised mainly in patients of laxity syndrome as instability is common in such patients.[7] Consideration should be given to shortening of hamstrings along with capsulorrhaphy.[4,5,9]

Only one case of neglected CDK has been reported in a 16-year-old girl with ligament laxity. A flexion shortening osteotomy at the distal femur above the trochlear level was performed on her right knee. The fragment was fixed with 95° condylar blade plate. One month postoperatively, the patient was able to put weight on the right lower limb. Four months postoperatively, the left lower limb was operated in a similar manner. Three years postoperatively, a femoral supracondylar osteotomy was performed on the right lower limb to correct the range of motion arc and valgus.[8]

In the case reported here, the patient had previous bilateral supracondylar osteotomy, but knee joints were still dislocated with gross instability. Our patient was of short stature with deformed lower femora due to previous surgery, hence, we did not do femoral shortening. Once the femoral and tibial articular surfaces were brought at the same level following distraction by ring fixators, an open reduction was performed. Knee joints were allowed weight bearing with KAFO, so that the tibial surfaces which had become rounded secondary to absent previous loading could develop concave contour to articulate with femur. Once the knee joint surface attained parallelism, the supracondylar osteotomy was performed to alter the arc of motion of knee joint. We planned staged surgery instead of femoral shortening; we did soft tissue lengthening and later open reduction. Eighteen months after the second surgery, we performed a bilateral supracondylar femoral extension osteotomy in order to improve the arc of motion. At 2½ years followup, his knees were found to be stable.

In this rare case, we have described our staged plan in the correction of a difficult neglected bilateral CDK in a 12-year-old boy who had previous unsuccessful surgical management.

References

1. Katz MP, Grogono BJ, Soper KC. The etiology and treatment of congenital dislocation of the knee. J Bone Joint Surg Br 1967;49:112-20.
2. Charif P, Reichelderfer TE. Genu recurvatum congenitum in the newborn: Its incidence, course, treatment, prognosis. Clin Pediatr (Phila) 1965;4:587-94.
3. Elchalal U, Ben Itzhak I, Ben-Meir G, Zalel Y. Antenatal diagnosis of Congenital dislocation of the knee: A case report. Am J Perinatol 1993;10:194-6.
4. Bensahel H, Dal Monte A, Hjelmstedt A, Bjerkreim I, Wientroub S, Matasovic T, *et al.* Congenital dislocation of the knee. J Pediatr Orthop 1989;9:174-7.
5. Johnson E, Audell R, Oppenheim WL. Congenital Dislocation of the Knee. J Pediatr Orthop 1987;7:194-200.
6. Johnstone CE 2nd. Congenital deformities of the knee. In: Scott WN, editor. Insall and Scott surgey of the knee. 4th ed. Philadephia, PA: Chuchill, Livingston/Elsevier; 2006. p. 1191-206.
7. Herring JA. Disorders of the knee. In: Herring JA, editor. Tachdjian's Pediatric Orthopaedics. 4th ed. Philadelphia, PA: Saunders/Elsevier; 2008. p. 919-26.
8. Kazemi SM, Abbasian MR, Hosseinzadeh HR, Zanganeh RF, Eajazi A, Besheli LD. Congenital dislocation of the knee in a 16-year-old girl. Orthopedics 2010;33:356
9. Fernández-Palazzi F, Silva JR. Congenital dislocation of the knee. Int Orthop 1990;14:17-9.
10. Ko JY, Shih CH, Wenger DR. Congenital dislocation of the knee. J Pediatr Orthop 1999;19:252-9.
11. Muhammad KS, Koman LA, Mooney JF 3rd, Smith BP. Congenital dislocation of the knee: Overview of management options. J South Orthop Assoc 1999;8:93-7.
12. Haga N, Nakamura S, Sakaguchi R, Yanagisako Y, Taniguchi K, Iwaya T. Congenital Dislocation of the Knee Reduced Spontaneously or with Minimal Treatment. J Pediatr Orthop 1997;17:59-62.
13. Simonian PT, Staheli LT. Periarticular Fractures after Manipulation for Knee Contractures in Children. J Pediatr Orthop 1995;15:288-91.
14. Curtis BH, Fisher RL. Congenital hyperextension with anterior subluxation of the knee. Surgical treatment and longterm observations. J Bone Joint Surg Am 1969;51:255-69.
15. Ferris B, Aichroth P. The treatment of congenital knee dislocation. A review of nineteen knees. Clin Orthop Relat Res 1987;216:135-40.

How to cite this article: Kumar J, Dhammi IK, Jain AK. Neglected surgically intervened bilateral congenital dislocation of knee in an adolescent. Indian J Orthop 2014;48:96-9.

Source of Support: Nil, **Conflict of Interest:** None.