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Unilateral congenital knee dislocation of the female neonate: A rare case report

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Abstract

Congenital dislocation of the knee (CDK) is a hyperextension deformity of the knee at birth rarely encountered. It occurs in isolation or in combination with other musculoskeletal anomalies. Treatment of CDK is serial plaster of Paris casting. Those not responding to conservative management are treated with percutaneous quadriceps recession (PQR) and V-Y quadricepsplasty (VYQ). It may occur as an isolated deformity, it may be associated with musculoskeletal anomalies such as developmental dysplasia of the hip and clubfoot, or it may occur as part of a syndrome such as arthrogryposis multiplex congenita or Larsen syndrome, or it may occur in paralytic conditions such as meningomyelocele. We describe a 12-day female neonate presented with hyper-extended right knee and was managed conservatively by closed reduction and serial above knee corrective pop splinting followed by above knee cast for a total duration of one and half months. At 11 months follow up no residual deformity was seen.

Keywords: Congenital Knee Dislocation; Closed Reduction; Serial Corrective Above Knee Casting

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Introduction

Congenital dislocation of the knee (CDK) is abnormal hyperextension of the knee at birth which is a rare grotesque condition. It was the first recognized by Chateline, with an incidence of 1:100,000 with a female to male ratio of 3:1 [1]. Congenital dislocation of the knee is divided into three grades based on radiographic evidence of the femorotibial relationship that is simple recurvatum, subluxation, and dislocation.

Grade I: Simple Recurvatum

Grade II: Subluxation (anterior subluxation of the tibia on the femur)

Grade III: Dislocation (anterior dislocation of the tibia on the femur).

Although the precise cause of CDK is unknown, both intrinsic and extrinsic factors have been suggested. Intrinsic factors are genetic anomalies and neuromuscular imbalances, whereas extrinsic factors are aberrant intrauterine pressure that results in intrauterine malposition. Some rare associations are with Congenital absence or hypoplastic anterior cruciate ligament, quadriceps muscle changes, Syndromic like Larsen's syndrome and Arthrogyriposis Multiplex Congenita (AMC) or paralytic conditions such as meningomyelocele. The Associated conditions are 31% with CTEV (Congenital Talipes Equines Varus) and 50% with DDH/CDH (Developmental Dysplasia of Hip/Congenital Dislocation of Hip) [2,3]. Management starts with closed reduction and cast or splint [4]. Surgical reduction is indicated when conservative measures fail or when the child is referred after the age of one year. In this case report, we present a neonate girl with unilateral CDK OF the right knee and its management.

Case Presentation

A 12-day-old girl presented with unilateral abnormal hyperextension in the right knee since birth in Paprola Ayurvedic Medical College (Himachal Pradesh) (Figure 1). Clinical examination revealed unilateral hyperextension of the right knee with a range of movement between 10° to 30° in full extension. Passive flexion was not possible, and deep skin grooves above the anterior aspect of the right knee was observed. The radiological evaluation of the right knee reveals anterior dislocation of the tibia on the femur (Figure 1b) which reveals Grade-III CDK. Further clinical and radiological examination of bilateral hip or ankle joints was normal. The dislocation was closely reduced by gently pressing the femur from posteriorly with pressing the tibia from anteriorly then applying an above knee splint in 0-degree knee extension initially then performed check x-ray which reveal normal reduction followed by above knee plaster of Paris (POP) slab (shown in Figure 2,3) applied in 0-degree knee extension for 10 days followed by POP cast for 2 weeks. At one and half months, no hyperextension deformity of the right knee was observed and the range of movement of the right knee achieved was >90 degrees in passive as well as active knee flexion. Follow-up after 11 months shows no abnormal hyperextension (Figure 4).

Detailed management was as follows

- On 1st day: closed reduction with long leg splint applied for 1day.
- On 2nd day: A/K pop slab applied in normal knee extension (0°) for 10 days.
- After removal of A/K pop slab and observation for any abnormal clinical presentation was done.
- Finally, A/K pop cast applied in knee flexion in 90 degree for 4 weeks followed by crepe bandage application. (Figure 1-4)
- Follow up after 11 months without any abnormal clinical presentation.



Figure 1:

- a. Hyperextension of the right knee.
- b. X ray of right knee lateral view - showing anterior dislocation of the tibia on the femur.



Figure 2:

- a. Above knee plaster of Paris slab.
- b. Xray showing reduced knee joint



Figure 3:
a. Above knee plaster of Paris cast in 90-degree
b. Crepe bandage application



Figure 4: Picture of normal knee extension on standing shown from front at 12 months (a) & back (b).

Discussion

Congenital dislocation of the knee is hyperextension of knee with varying degrees of anterior displacement of tibia. Three theories are described in literature about aetiology of CDK.

- Mechanical theory - Due to abnormal intrauterine position
- Primary embryologic theory - Due to embryonic defect
- Mesenchymal theory - Due to quadriceps contracture

Majority of CDK cases are sporadic with Incidence 1 in 100,000 live births [1,2]. Conservative management with closed reduction and corrective serial casting is performed in early infancy with CDK. A maximum of four to five weekly manipulations and castings are attempted. If a range of flexion $>90^\circ$ is achieved, serial casting is continued. If the range of flexion remains $<90^\circ$ then recommended surgery or in late infancy if nonoperative modalities fail surgical reduction is recommended i.e., percutaneous quadriceps recession (PQR) and V-Y quadricepsplasty (VYQ) [5].

A review of the literature revealed that the results of surgery seem to be most favorable when performed before the age of 2 years. The most common findings during surgery are tight anterior capsule, along with quadriceps fibrosis and contracture, especially in the inferior and lateral parts. Although most authors have reported satisfactory results with surgery in childhood, Johnson et al believe that the results are poorer in older children with CDK managed by V-Y advancement or Z-plasty of the quadriceps; they concluded that prognosis is most favorable in unilateral cases, and when surgery is performed before the age of 2 years [6]. Further, Oetgen et al stated that the most striking observation in patients with CDK is the high incidence (78%) of knee instability [7]. Bell et al, believe that the cruciate ligament abnormalities are secondary to the delay in treatment [8]. In contrast, Katz

et al proposed that cruciate ligament abnormalities can be primarily observed in older children [9]. Currently, conservative management of CDK in 12 days female neonates with closed reduction and serial corrective casting yields very good results. Follow-up after 11 months shows no abnormal clinical presentation in late infancy also and parents are quite satisfied with the result achieved after treatment, they are advised to consult us if any abnormal clinical presentations are observed in the child in the coming days.

Conclusion

Congenital dislocation of knee is a rare entity. Timely diagnosis and conservative treatment can result in excellent outcomes as in our case report. Surgery is required in patients who fails in conservative treatment, and those who presents late.

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