CONGENITAL KNEE DISLOCATION: CASE REPORT

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ABSTRACT

Congenital knee dislocation of knee is a rare condition, with an uncertain incidence in sub-Saharan Africa. The present case report describes the care of a 14 day old female referred to our orthopaedic services with a congenital hyperextended knee deformity. The patient was managed non-operatively with serial manipulation and casting. At 6 months follow up the patient was able to achieve normal passive knee range of motion. The pathophysiology and treatment options of congenital knee dislocations are reviewed.

Key words: Congenital knee dislocation, Congenital deformity of the knee

INTRODUCTION

Congenital Knee Dislocation (CKD), also known as congenital Genu recurvatum, is a musculoskeletal deformity of the knee joint. It is very rare with a prevalence of 1 in every 100,000 newborns, seen most frequently in females (1). CKD is characterized by hyperextension of the knee with limited flexion and anterior curvature of the lower limb (2). It presents at birth and is often associated with additional musculoskeletal anomalies such as clubfoot, osteogenesis imperfecta, femorotibial subluxation, developmental dysplasia of the hip. CKD has been associated with a number of syndromes including Larsen syndrome, arthrogryposis multiplex congenita, myelomeningocele, spastic cerebral palsy, cervical myopathy, Marfan syndrome, Ehlers-Danlos syndrome, Down syndrome, and Turner syndrome (3).

There is limited reliable data regarding CKD in sub-Saharan Africa. As such prevalence and management of the condition remains obscure. The purpose of this case report is to document the management of a patient with CKD and describe the role of non-operative treatment for the correction of the deformity. It is in this regard that, via a clear description of how conservative management can be of invaluable benefit in less complicated cases, an effort to increase the current knowledge about the management of CKD remains the epicenter of our primary intent.

CASE REPORT

A 42-year-old woman; gravida 5; para 5 with no past medical history, was referred to the labour and delivery ward at 38 weeks complaining of back and pelvic pain. During her pregnancy, a triple test was performed determining the child was at low risk of trisomy 13, 18, and 21. Ultrasounds obtained through pregnancy did not find any abnormalities. The mother, however, had not been evaluated by her obstetric providers for over a month prior to presentation.

A spontaneous vaginal delivery was performed. A 3100g female baby with Apgar scores of 8/9 was delivered and presented with an obvious hyperextension deformity of her right knee. At 14 days old the patient presented to the orthopaedic clinic for initial evaluation. The prenatal and perinatal history was unremarkable. The physical exam revealed a healthy child with a right supra-patella deep skin crease. The right knee was held in 70 degree of hyperextension and she was able to achieve 30 degrees of passive flexion for a full arch of motion of roughly 100 degrees (Figure 1). She was diagnosed with type II CKD (Table 1).

Table 1
Tarek CKD grading system

Grade range of passive	Flexion	Radiology
GI	>90°	Simple recurvatum
GII	30–90°	Subluxation/dislocation
GIII	<30°	Dislocation

Figure 1
Initial patient presentation

Figure 2
Knee flexion obtained before the last cast application

The patient underwent conservative management given her young age and range of passive flexion. The management consisted of serial manipulation and casting of knee in maximum achievable flexion. We used above knee complete cast with the foot positioned in neutral plantigrade position. Completed cast changes with manipulation were performed weekly until we could achieve a passive knee flexion of 90 degree (Figure 2) after a total period of 4 weeks. After removal of the last cast the passive flexion on the affected knee was equal to the normal knee (Figure 3). At 6 months follow up the passive range of motion was 137° which was equal to the contralateral side (Figure 4).

Figure 3
Knee flexion at removal of the last cast

Figure 4
Comparison of both knees flexion on a follow up at 6 months of age

DISCUSSION

Congenital Knee Dislocation (CKD) is a rare condition with an incidence in 1 in 100,000 which presents with a fixed hyperextension deformity and anterior displacement of the tibia on the femur (4–6). Patients may be described as having a "backward" knee as the foot may be extended to the level of the mouth or shoulder. Our patient was a 14 day old female who presented with grade 2 unilateral right CKD.

According to Mehrafshan *et al*. (7) most of the patients present to the orthopaedic clinic within 30 days of birth and classified in grade 1. Our patient presented within expected time likely due to the obvious nature of the deformity. The prenatal and perinatal history concerning this patient was unremarkable.

CKD has been associated with other joint abnormalities including deformities at the hip, foot, elbow, and spine (8). Developmental dysplasia of the hip and clubfoot are common concurrent conditions (6,9). Abnormality of the anterior cruciate ligament and quadriceps may also be associated with the condition. Bilateral CKD is almost always associated with an underlying syndromic process such as hyperlaxity diseases. These include Larsen, Beals, and Ehlers-Danlso syndrome. Patients may present with ipsilateral hip dislocations in 70% of cases and clubfoot in 50% of cases. Unilateral cases may further be associated with neurologic spastic conditions. While neurological conditions often present with unilateral hyperextension deformities, they may present with a unilateral hyperextension deformity and a contralateral flexion contraction (9–12). A thorough physical exam is required in all patients to ensure no additional congenital injuries exist. However, our patient presented with an isolated CKD, without any other remarkable abnormalities.

The diagnosis of CKD can be made prenatally using prenatal ultrasound or just after birth by physical exam and radiography (3). The ultimate cause of CKD is thought to relate to abnormal fetal positioning and movement *in utero*, regardless of the associated deformities or conditions. Initial fetal molding is often due to oligohydramnios or breech position (8,9). Concurrent neurologic or hyperlaxity conditions prevent normal movement resulting in atrophy and fibrosis of the quadriceps and contractures of the iliotibial band (1,13).

Various classification systems have been described based on physical exam and radiographic findings of the femorotibial relationship. CKD can be classified into three grades using the Tarek CKD grading system (14): simple recurvatum, subluxation and dislocation (3,5).

CKD can also be classified into three grades using Leveuf and Pais classification, based solely on radiological views (15).

- Grade 1: No true dislocation but congenital hyperextension of 15 to 20° and passive flexion of up to 90°.
- Grade 2: Congenital subluxation with joint incongruency is observed. No passive flexion of the knee is possible and 25 to 40° of hyperextension can be achieved.
- Grade 3: Dislocation with no contact between the joint surfaces of tibia and femur.

If left untreated, CKD can result in a stiff unstable knee with a severe recurvatum and valgus deformity. Treatment with surgical or non-operative management is based on severity of deformity and age at presentation. Treatment should begin as soon as the deformity is identified as prognosis is adversely affected by delay in treatment (16). If additional musculoskeletal deformities are present, the knee should be treated first (1).

Our patient was treated with serial manipulation and casting over 4 weeks period with a successful result, which is the recommended initial management of patients with CKD. Non-operative treatment involves applying gentle traction and anteroposterior force on the tibia to allow for knee flexion. Care should be given not to apply forceful manipulations due to the risk of fracture or epiphyseal injuries. The knee should be immobilized in plaster of Paris and repeat manipulations should be performed until 90° of flexion is achieved. At this point the knee may be placed in a removable splint for several additional months to maintain correction. Conservative management is often successful if initiated prior to 12 months of age. In one of the largest case series to date, Silva *et al*. (17) described their treatment of 14 patients with 22 affected knees. All children presenting before the age of 12 months treated by skin traction until reduction was achieved. They then underwent serial manipulations with placement in plaster of Paris casts until 90°of flexion was attained. If after six months the conservative method had failed, or the patient first presented after the age of twelve months, surgical treatment was used (17). Using this approach a good result was obtained with conservative treatment if started before the age of three months. Complications of serial manipulation

and casting may include trauma to the proximal tibia physis which may result in tibia procurvatum as well as dislocations within the cast if not placed correctly. In our case, there were no complications during serial manipulation or casting.

Generally operative management is advised when non-operative treatment has failed or in late presentations. Surgery typically involves either a Percutaneous Quadriceps Recession (PQR) or a V–Y Quadricepsplasty (VYQ) (3,18). V-Y Quadricepsplasty may be performed in a single stage or over multiple stages in complicated cases (19). Patients with laxity syndromes may benefit from the addition of a posterior capsulorrhaphy as well as an ACL reconstruction if absent (20). This may prevent recurrent dislocations. Patients with severe genu valgum may also benefit from an opening wedge supracondylar osteotomy (7,8,16). Complications include quadriceps scarring and weakness, infections, adhesion formation, and wound breakdown.

CONCLUSION

Congenital Knee Dislocation (CKD) is a rare musculo-skeletal disorder characterized by obvious knee recurvatum at birth. We recommend a trial of non-operative management even in advanced grades of CKD in patients who present within the first 6-12 months (14). If conservative management fails, surgery should be performed prior to 12 months if possible. Either a Percutaneous Quadriceps Recession (PQR) or a V–Y Quadricepsplasty (VYQ) may be performed.

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