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Congenital Knee Dislocation: Challenges in Management in A Low Resource Center

Osakwe Onyebuchi Gregory[1],Asuquo Joseph Effiong[2],Abang Innocent Ebegi[3], Eyong Micheal[4], Udosen Anthony Martin[5]

[1]*Dept. Of Orthopedics And Traumatology, University Of Calabar Teaching Hospital, Calabar*
[2]*Dept. Of Orthopedic Surgery And Traumatology, University Of Calabar, Calabar.*
[3]*Dept. Of Orthopedic Surgery And Traumatology, University Of Calabar, Calabar*
[4]*Dept. Of Pediatrics, University Of Calabar, Calabar.*
[5]*Dept. Of Orthopedic Surgery And Traumatology, University Of Calabar, Calabar*

Abstract
Background: *Congenital knee dislocation (CDK) is a rare deformity often missed by young and upcoming orthopedic surgeons practicing in poor resource centers. The aim of this article is to document our challenges in the management of this deformity in the face of socio- cultural beliefs.*
Patients And Method: *This is a case series. The clinical information of all patients was retrieved from our patient journal system from September 2011 – September 2016. All the patients were seen and managed at the University of Calabar Teaching Hospital.*
Results: *Three patients with five deformities were managed during this period. Two of the patients had bilateral deformity with associated bilateral clubfeet in both and spinal bifida in one. None of them had associated developmental dysplasia of the hip. The remaining was an isolated case with unilateral left knee involvement. All patients were females. None had positive family history. Two had breech delivery. None of the mothers reported maternal illness during pregnancy or ingestion of un-prescribed drugs. The case with spinal bifida was abandoned by the parents in the special care baby unit. All cases were started initially on conservative management with good initial outcomes. Only the isolated unilateral deformity completed treatment and is currently being followed up with good results while the others were lost to follow up.*
Conclusion: *Congenital knee dislocation is a rare deformity with sporadic occurrence. Early presentation and treatment could yield good results. Socio-cultural believes could be a hindrance to successful treatment in a resource poor setting.*
Keyword: *Congenital knee dislocation, challenges, clubfoot, sporadic, serial manipulation*

I. Introduction

Congenital knee dislocation (CDK) is a rare malformation characterized by a spectrum of deformities ranging from hyper-extension (recurvatum), subluxation, dislocation and limitation of flexion of the knees in newborn with or without associated musculoskeletal abnomalities. It has a worldwide incidence of 1 / 100,000 live birth which is 1% of developmental dysplasia of the hip. Forty to hundred percent of patients with CDK have associated musculoskeletal abnormalities which include developmental dysplasia of the hip and especially talipesequinovarus. Others are spinal bifida, cleft palate, Larsen syndrome, arthrogryposis, fibula hypoplasia, dislocation of the elbow and chest cage deformities. It was described by a Swiss physician Chatelaine in 1822.[1,2, 3]

The exact etiology remains unknown. It has been associated with certain factors including extrinsic factors such as intrauterine packaging disorders, breech presentations and intrinsic factors like genetic malformation but most cases are sporadic. Neuromuscular imbalanceshave also been implicated. The most common theory is changes that occur in the quadriceps muscle and tendon which can be seen in all patients. The deformity may be unilateral or bilateral and mostly affect girls but some literature report equal distribution.[2, 3, 4]Depending on severity it has been divided into three grades by Laurence viz: grade 1, congenital hyperextension; grade 2, congenital hyperextension with anterior subluxation of tibia on femur and grade 3, congenital hyperextension and anterior dislocation of tibia on femur.[5, 6]

Diagnosis of CDK is usually simple. Usually recognized by, the presence of genu recurvatum,, dimple or deep crease over the anterior aspect of the knee and absence of suprapatellar pouch. Management is started early with conservative method comprising serial manipulation and casting or splinting. Surgery is reserved for cases of failed conservative treatment or cases who present late, say after one year of age and options include V-Y Quadriceplasty and percutaneous Quadriceps resection. Bracing is used as adjunct to maintain reduction. Despite all the various treatment modalities, some controversy exists as to when to start treatment or which type will benefit from surgical therapy ab nitio. [4, 7, 8, 9] In Nigeria only one article has documented the management of

this deformity. [10] We present the management of three sequential cases with the challenges encountered in our center.

II. Case 1

A month old infant first seen by the pediatricians who were confused of the diagnosis and referred to the pediatric surgeons who subsequently referred the baby to us for our opinion in the management. The parents complained of the abnormal orientation of the knee, legs and deformity of the feet from birth. She was delivered by spontaneous vaginal delivery with breech presentation and prolonged labor by a traditional birth attendant. Birth weight could not be ascertained but was a term baby. Physical examination revealed bilateral knee dislocation (grade 3) with an angle of -80° on the left and -50 on the right. Both were rotated to about 40° medially. Passive flexion was 5° - 10°. See fig. 1 – 3. The hip ultrasonography was normal. Apart from bilateral Congenital TalipesEquinovarus, no other concomitant musculoskeletal abnormalities were detected.

Figure1. Bilateral congenital knee dislocation with bilateral clubfeet

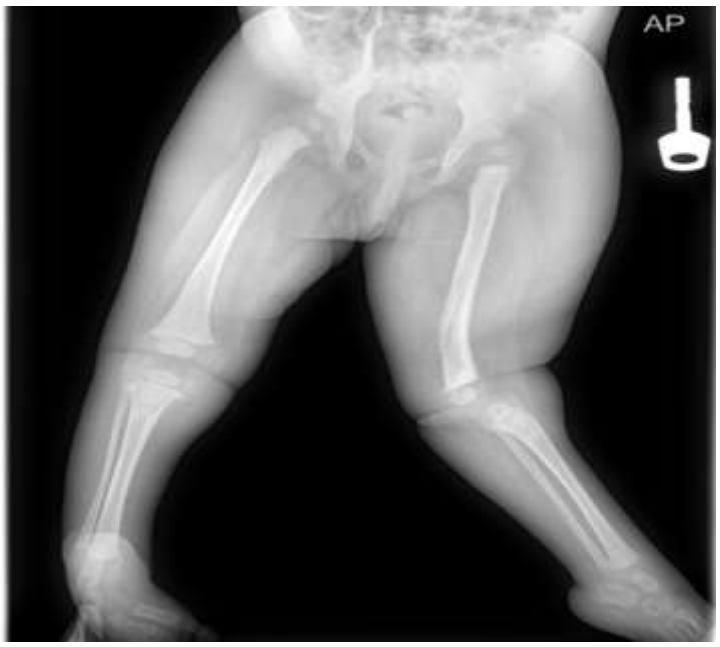

Figure 2

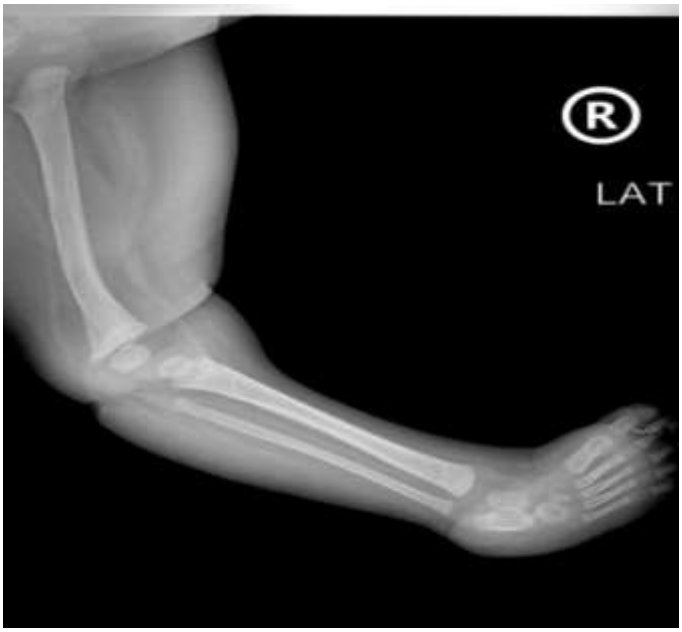

Figure 3

We commenced management with serial manipulation and casting. The left knee had remarkable improvement with up to 80° flexion being achieved after about five sessions of casting. Parents had financial challenges and believed so much on reincarnation. They were lost to follow before further treatment could be planned.

III. Case 2

A week old neonate delivered through cesarean section due to obstructed labor resulting from breech presentation and subsequently admitted into the special care baby unit. The gestational age was 39 weeks. We were invited to see by the pediatricians due to knee and foot deformity. Physical examination revealed knee hyperextension of -45° bilaterally (grade 2), bilateral clubfeet, bilateral duplicated 5thfinger digit and spinal dysraphism (high type) (See fig. 4). The child had multiple congenital malformations which did not fit any syndrome. We counseled the parents for serial manipulation and casting because there was passive flexion of the knee was between 10° – 15°. We initially planned for serial manipulations and casting but the parents never believed in any form of orthodox remedy. They abandoned the child in the Hospital leaving no means of contact before some relatives eventually came and took the child away and she was lost to follow up.

Figure 4

IV. Case 3

This was a two weeks old neonate referred from the pediatric out-patient to pediatric orthopedic clinic. The child was a term baby delivered through spontaneous vaginal delivery. Mother complained of left knee deformity. Physical examination revealed isolated left knee congenital hyperextension (grade 1) with greater than 70° passive flexion at the knee. No positive family history and no any associated musculoskeletal malformation. Developmental dysplasia of the hip was ruled out with Barlow and Ortholani test, corroborated with hip ultrasonography.The parents gave consent and we applied serial casting of the limb with knee initially at extension then gradual flexion two weekly till we achieved full flexion. The result was encouraging and parents were advised on the use of pediatric knee brace. She is still being followed up in clinic.

V. Discussion

Congenital dislocation of the knee (CDK) or congenital genu recurvatum is still a rare condition as exemplified by our series over a five year period where we saw 3 patients. This is not different from that reported by other series. Either it is under diagnosed or under reported. All our patients were females. This is similar to that reported by other series which showed a female preponderance.[2, 6, 11, 12, 13]It could be unilateral or bilateral and >50% of the cases are bilateral. In our series we saw more bilateral cases than unilateral although cases were few and not enough to draw statistical conclusions. There is a 50 – 70% association with developmental dysplasia (DDH) of the hip and clubfoot. We had 2 patients with associated clubfoot bilaterally but DDH was ruled out while the remaining was an isolated case. Some researchers report similar findings of clubfoot and DDH as a common association with CDK but same cannot be said about clubfoot being associated with CDK. There is an association with neurologic anomalies like spinal dysraphism (meningomyelocoele) as in one of the patients in our series.

Exact etiology is unknown but both extrinsic factor and intrinsic factors have been proposed. However, three theories have been postulated. Mechanical theory prescribed the origin of the deformity from abnormal fetal intrauterine posture or "intrauterine packaging disorder". There is association with breech delivery as in our series where 2 (66%) out of 3 had breech presentation. Primary embryonic theory which explains the other defects associated with the deformity. Mesenchymal theory, this easily explains the mesenchymal defect which presents as fibrosis of the quadriceps muscle. The intrinsic factor includes genetic abnormality but most cases are sporadic as reported by the 3 cases and demonstrated by some researchers.[1-2, 12, 14-17]

"Leveuf and Pais classification is the most popular classification and categorizes the deformity into three subgroups. Grade 1 is the most common type and not a true dislocation and accepted as congenital hyperextension. Nearly 15 to 20° of hyperextension can be detected and passive range of flexion is maximum of 90°. At Grade 2, congenital subluxation with joint incongruency is seen. Passive flexion of the knee is im-possible and 25 to 40° of hyperextension can be achieved. At Grade 3 cases there is no contact between the joint surfaces of tibia and femur. Proximal tibial epiphysis is located anterior to femoral condyles". [17-18]The three cases reported fell into all grades. Outcomes are variable and depends on the grade and associated congenital abnormalities.

Tarek and Shady introduced a modified grading system based on range of passive knee flexion with better suitability for choosing treatment option. Grade 1, $>90^0$ range of passive flexion. Grade 2, 30 – 90° range of passive flexion. Grade 3, $<30^0$ range of passive flexion. Treatment is dependent on presentation and associated anomalies and could be conservative or surgical. Many modalities can be used for conservative treatment including the use of serial casting to increase knee flexion, the Pavlik harness for posturing of the knee in further flexion, skin traction and skeletal traction.[2, 19]*Haga et al*[20] suggested that it is advisable to wait 1 month for spontaneous reduction of CDK in cases not associated with clubfoot, Arthrogyroposis or Larsens syndrome. Most authors recorded successes with conservative treatment of most cases. *Nogi and MacEwen*[21] had successful treatment with manipulation in 23 out of 27 patients while *Johnson etal*[12] had improvement in ROM in 10 out of 17 patients with manipulation and casting. *Omololu et al*[10] in Ibadan treated 30 patients with manipulation and serial casting and had excellent results in 20 patients following 2 years follow up. He and other authors recommended early conservative treatment for most cases.

For patients with associated DDH and CTEV, it was advocated that the Knee be manipulated first before the foot or the Hip. Also primary anomalies like Spinal dysraphism should be treated concurrently.Surgical options of V-Y quadriceplasty (VYQ), percutaneous quadriceps recession (PQR) and other soft tissue releases were reserved for resistant cases and for late presentation especially in older children. Compared to PQR, VYQ is associated with increased morbidity due to a long incision with scarring, adhesions and wound breakdown as well as blood loss. However it is more successful in achieving and maintaining reduction in severe and resistant cases.

Tarek et al[9] developed the following protocol of management – serial casting is performed in patients with grade 1; in grade II CDK in Neonates, serial casting is started. If a range of flexion >90° is achieved after 4

attempts several casting is continued while if less than 90° they proceed to PQR. VYQ is indicated in patients with grade III CDK or in recurrent cases.

In our series, we had improved ROM in all our cases with serial manipulation and casting. Two of our patients had challenges with funds (which is common in our environment) and couldn't follow up their treatment adequately. One of the two above (patient 1) had delayed manipulation and also absconded for 6months.When last seen, she was Tarek grade 3 and being planned for V-Y Quadriceplasty before being finally lost to follow up. The other two patients had early manipulations with greatly improved range of knee movements after few visits.

VI. Conclusion

Congenital knee dislocation is a rare deformity, most cases are sporadic. Poor parent compliances and sociocultural believe may hamper treatment in a low resource center. Health education campaigns along with Insurance coverage is essential in ameliorating this disorder in such centers.

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