Congenital dislocation of knee: A correctable deformity

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ABSTRACT

We report the case of a late preterm small for gestational age baby girl presenting with deformities of both knees at birth. Diagnosis by radiologic investigation revealed bilateral knee joint dislocation and ultrasonography revealed developmental dysplasia of the hip. Concomitant treatment of the congenital dislocation of the knee with serial casting and the hip with Pavlik harness provided satisfactory results. Cases with delayed presentation or which do not respond to conservative treatment need surgery. Delay in treatment may lead to long-term instability and stiffness. Emphasis should be laid on the immediate recognition and treatment of the condition.

Key words: Congenital dislocation, Knee, deformity

Congenital dislocation of the knee (CDK) is a very rare condition that comprises a spectrum of deformities from subluxation to complete dislocation. The incidence of CDK is estimated at 1/100,000 live births, i.e., 100 times rarer than developmental dysplasia of the hip (DDH). It is bilateral in one-third of the cases. It is commonly associated with breech delivery, oligohydramnios, congenital talipes equinovarus (35%), and DDH (45%) [1]. In general, the diagnosis established immediately after birth according to the position of the genu recurvatum and is confirmed by radiography. The treatment with conservative methods at an early stage is most likely to yield successful results.

CASE REPORT

A late preterm small for gestational age baby girl with birth weight 2.29 kg born by normal vaginal delivery to a primi mother presented with deformities of both knees at birth (Fig. 1). On examination, both knee joints were rotated and hyperextended. A passive flexion of both knees to an anatomically straight position could not be performed. The movements of the toes were normal. The rest of the newborn examination was normal.

X-ray of both knee joint revealed anterior tibial translocation on femur (Fig. 2). Ultrasonography of the hip showed DDH. Gentle manipulation followed by above knee POP cast and Pavlik harness (dynamic hip abduction brace) was applied. The cast was changed every week for 3 weeks. After 3 weeks, the knee adopted a normal shape. Ultrasonography of the hips was done and was found to be normal. The cast was removed and discontinued at 3 weeks of age. A follow-up at the age of 4 months (Fig. 3) showed normal position and range of motion of the knee and the DDH had recovered. Repeat X-ray of both knee joints was normal (Fig. 4). Currently, the baby is under follow-up and has attained all the development milestones for her age.

DISCUSSION

CDK was first described by Chatelaine in 1822. It is a rare condition that comprises a spectrum of deformities from subluxation to complete dislocation [2-11]. The incidence is nearly 1 in 100,000 live births without any difference between right and left knees. Studies and case reports have not shown any predominance in sex distribution. The main etiologic factor is not known yet. The essential anatomical abnormality is a short quadriceps muscle together with subluxation of the hamstring muscles which lie anterior to the axis of knee flexion.

There are numerous hypotheses concerning the etiology of CDK, and both intrinsic and extrinsic causes have been suggested. The intrinsic causes are genetic abnormalities, whereas the extrinsic causes are mechanical factors [9]. In a review of 200 cases by Provenzano, 7 families had a history of CDK [6]. MacFarland [5] reported a case of a family in which a mother and her three children from three different fathers had CDK. Curtis and Fisher have described “heritable congenital tibiofemoral subluxation” [4], a genetically transmitted syndrome where CDK is combined with some abnormalities of the face and spine. The familial occurrence suggests a possible genetic basis for CDK, whereas a non-genetic dysplasia etiology is supported by the sporadic occurrence of most of the cases. The latter etiology is more in keeping with the present series of patients, who all lacked a positive family history.

The extrinsic causes of CDK are considered to include a lack of amniotic fluid, lack of intrauterine space, malposition of

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the fetus, fibrotic contracture of the quadriceps, and traumatic dislocation during birth [4,7,10,11]. Abnormal intrauterine positioning is implicated by observations of hyperextended knees being associated with a breech presentation. Fetal molding due to oligohydramnios or extended breech position was suggested as a cause by Shattuck and supported by Niebauer and King [10]. The incidence of breech positioning at delivery was 21% in our patients. Other factors proposed as contributing to CDK include quadriceps contracture and hypoplasia of the anterior cruciate ligament [7,11].

In our case, the exact etiology for CDK is not known; however, X-ray of both knee joints revealed anterior tibial translocation on the femur, probably secondary to anatomic abnormality as stated earlier. As literature states, DDH is the most common association of CDK [1]. CDK contributes, at least in part, to the development of DDH, with the contractive quadriceps femoris muscle and dorsally displaced hamstrings potentially rendering the hip joint unstable [9]. In our case, we could not find any other association other than DDH. In the follow-up a clinic, DDH was being corrected with the application of Pavlik harness.

All infants with hyperextension of the knee should have a radiographic examination to differentiate genu recurvatum from true dislocation of the knee. In congenital genu recurvatum, the tibial and femoral epiphyses are in proper alignment except for the hyperextension. In the subluxed knee with dislocation, the tibia is completely anterior or anterolateral to the femur. The tibia is shifted forward in relation to the femur and is frequently lateral as well [12].

Serial manipulation and casting are recommended in newborns. Casting should be done in full flexion position as knee allows. Forced flexion is not advised because of potential problems such as fracture, epiphyseal damage, and impaired circulation. The cast should be changed once in 1 or 2 weeks until the accurate reduction is achieved. The early conservative treatment is recommended as the therapy of choice and is successful with gentle manipulation, strapping and serial casts if carried out early, i.e., within 2-3 months [3,13].

Cases with delayed presentation or which do not respond to conservative treatment need surgery. Surgical treatment involves lengthening of the quadriceps tendon by V-Y plasty [14]. Prognosis is mostly favorable in unilateral cases [15] and when surgery is performed before 2 years of age. Delay in treatment may lead to long-term instability and stiffness [13].
should be laid on the immediate recognition and treatment of the condition.

CONCLUSION

Early detection and timely intervention of congenital dislocation of the knee could save the child from long-term disabilities and would help them to attain normal development milestones as in our case.

REFERENCES