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CASE REPORT

Unilateral congenital dislocation of the knee and hip : a case report

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We report our experience with a unilateral congenital dislocation of the knee associated with a developmental dysplasia of the hip on the same side. Our case is a good example of congenital dislocation of the knee caused by abnormal intrauterine pressure leading to this type of congenital postural deformity. To our knowledge this is the first case of congenital dislocation of the knee reported after cervical cerclage of an incompetent cervix to prevent a pre-term delivery. The Pavlik Harness was used to treat the knee and the hip at the same time with a satisfactory result after 20 months of follow-up.

Keywords : congenital dislocation of the knee ; congenital dysplasia of the hip ; intra-uterine pressure.

INTRODUCTION

Congenital dislocation of the knee (CDK) includes a spectrum of hyperextension disorders of the knee and is characterised by the forward displacement of the proximal tibia on the femoral condyles (*4,12,14,17*). This rare but clinically challenging disorder was first described by Chanssier (*in 22*) in 1812 and later by the Swiss physician Chatelaine in 1822 (*in 10*). The estimated incidence is 1:100 000 live births (80 times less than Developmental Dysplasia of the hip (DDH)). It can be diagnosed just after birth by the hyperextension of the knee and palpation of the prominent femoral condyles in the popliteal space. The classification of CDK as described by Laurence (*14),* includes grade I hyperextension without tibiofemoral dislocation, grade II subluxation with anterior displacement of the tibia, and grade III dislocation of the tibio-femoral joint with the tibia anterior and superior to the femur. When diagnosed it is very important to rule out any associated abnormalities such as DDH and clubfoot. It is crucial to know whether the CDK is part of a musculoskeletal syndrome or not (*21*).

The true aetiology of CDK remains unknown. In some cases hereditary and genetic influences (*5,17*) are well demonstrated but this is not applicable to the majority of cases. Amongst aetiologic contributing factors, the following have been described : absent or hypoplastic anterior cruciate ligament (ACL) (*12*), quadriceps contracture, absence of the suprapatellar pouch, lack of intrauterine space, trauma to the mother and the breech position.

Aetiologic factors can be described as being extrinsic or intrinsic. The extrinsic type is the result of mechanical factors mainly due to abnormal

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intrauterine pressures, leading to an intrauterine malposition. It usually results in a hyperextension or subluxation of the knee. Reduction is easy to achieve, even spontaneous reduction has been mentioned (*7*). Conservative therapy is the treatment of choice, with a good prognosis (*7,13*).

The intrinsic type is related to certain syndromes (arthrogryposis, achondroplasia, Larsen syndrome), genetic abnormalities and/or neuromuscular imbalance (*2,7,9*). It is usually a subluxation or dislocation of the knee. The reduction can pose a challenge and if reduced, 90° flexion is not always achievable. Conservative therapy is the treatment of choice, but in case of failure, open surgery is needed. The prognosis depends on the severity of CDK and the associated syndromes (*2*).

Both the extrinsic and the intrinsic types are often associated with DDH and talipes equinovarus. Other associated anomalies mentioned in the literature and less frequently reported include congenital dislocation of the elbow, cleft palate, chest cage deformities, fibular hypoplasia, hydrocephalus and spina bifida (*1,12*). Nogi *et al* (*18*) mentioned that, in case of associated DDH, the knee should be treated prior to the hip , because improvement of knee flexion allows better treatment of the hip (*19*). Extrinsic CDK associated anomalies can usually be treated more easily and with better results (*18*) than the anomalies associated with the intrinsic CDK, which are often difficult to treat (*7,9*).

The pathologic findings reported were derived from spontaneously aborted foetuses (*22*), preoperative arthrography (*19*) and from corrective surgery of CDK after failure of conservative treatment (*22*). Obliteration of the suprapatellar pouch or fibrosis of the quadriceps muscle (*21*), shortening of the quadriceps muscle mainly the vastus intermedius with or without fibrosis, a tight anterior joint capsule (*16,20*), anterior displacement of the hamstrings on the medial side changing their function to extensor muscles, anomalies of the ACL (absence, elongation) (*12*), flattening of the femoral condyles, and changes to the posterior slope of the tibial-plateau (*17*) have all been described in the literature (*22*). Today it is quite clear that the above mentioned pathologic findings are secondary changes to the CDK (*2,22*).

Fig. 1. — Congenital dislocation of the knee and hip, left side. Picture taken immediately after birth.

CASE REPORT

A full-term baby girl (weight : 3.59 kg, Apgar score 5/5) born by normal cephalic delivery, presented with a unilateral CDK and DDH on the left side. During the 23rd week of pregnancy, there was high risk of pre-term delivery without amniotic fluid leakage, because of an incompetent cervix. Taking into account the premature delivery of the couple's first child, a cervical cerclage was performed to prevent premature delivery. We examined the child ten minutes after birth. It seemed that, *in utero*, the child's left leg had been positioned over the stomach towards the right side with a hyperflexion/hyperendorotation of the hip and hyperextension of the knee (Fig. 1). With the anterior side of the left leg lying over the stomach it is not clear whether the left foot had been locked below the chin or in the axilla as mentioned in the literature (*17*). We believe that the leg has been locked between the child's stomach and the uterus. Furthermore a transverse crease was present in the skin over the anterior aspect of the knee just above the patella (*5*).

We carefully performed a reduction maneuver and reduced the subluxed (Grade II) knee. The reduction was easily felt and achieved with a maximum flexion of 25°. More flexion was not possible, probably due to a shortened quadriceps mechanism. The radiograph taken immediately after confirmed the knee reduction (Fig. 2). Further examination showed a DDH on the same side, confirmed with ultrasound (Fig. 3a). No other associated anomalies were found. Ultrasonography of the left knee, performed one day after birth and compared with the normal right knee showed a normal quadriceps

Fig. 2. — The first radiograph (A/P and lateral) after reduction. The white arrow marks the tranverse crease above the knee on the lateral view.

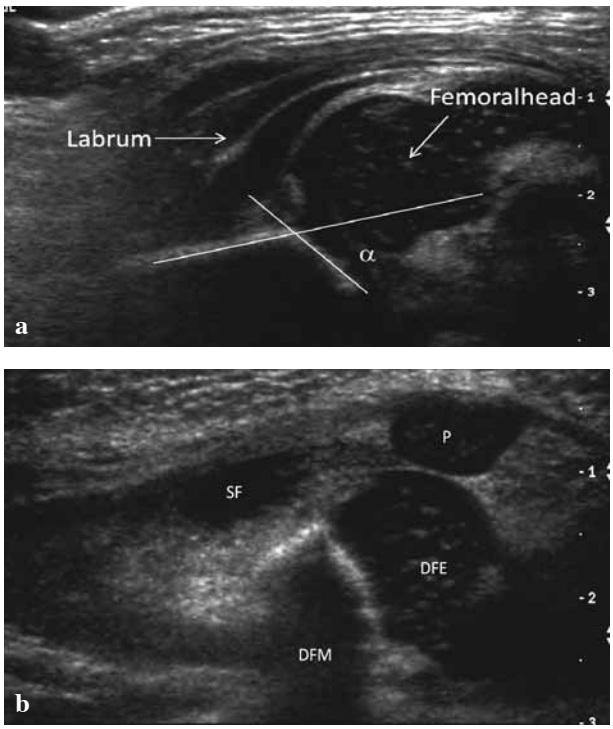

Fig. 3. — (a) Sonogram of the left hip : Graf type IIIa, eccentric hip, α = 49°. (b) Sonogram of the left knee, DFE : distal femoral epiphysis, DFM : distal femoral metaphysis, P : patella, SF : suprapatellar pouch fluid.

muscle and tendon, presence of a suprapatellar pouch with small amounts of fluid and presence of both cruciates (Fig. 3b). No fibrosis of the distal quadriceps as mentioned by Kamata *et al* (*11*) was seen.

Taking into account the reducibility of the hip and knee joint, we decided to apply a Pavlik harness (*8*) to treat the knee and the hip at the same time. In 4 days we achieved a flexion of 50° to 60° at the knee joint. We had no difficulty at all in reducing the hip despite the CDK. After 3 months of treatment the child had a stable hip and knee joint with a good ROM for both joints. The knee did not dislocate but there was a slight posterior laxity, also mentioned by Niebauer *et al* (*17*). We decided to apply the Pavlik harness for another 2 months but it did not have any influence on the posterior laxity. After 20 months we saw no problem in walking but when standing a slight recurvatum could be noted. The left knee still has a slightly greater laxity as compared to the right knee, especially when performing the drawer tests (Fig. 4 & 5).

DISCUSSION

All extrinsic type CDK can be treated conservatively if treated early ; treatment consists of early manipulation followed by splinting, casting or Pavlik harness until 90° of flexion is achieved (*6,13, 15,19*). Usually 2 to 3 months of treatment is enough. The emphasis of the treatment in the extrinsic type CDK is, in our opinion, the prevention of extension. Our newborn patient is a clear case of extrinsic CDK due to abnormal intra-uterine pressure . The reducibility of the CDK, the reducibility of the associated DDH, the absence of any musculoskeletal syndrome and the satisfactory result of our treatment confirm this diagnosis. We advise the use of ultrasound to evaluate the CDK. The absence of quadriceps fibrosis and the presence of the supra-

Fig. 4. — Follow-up after 20 months

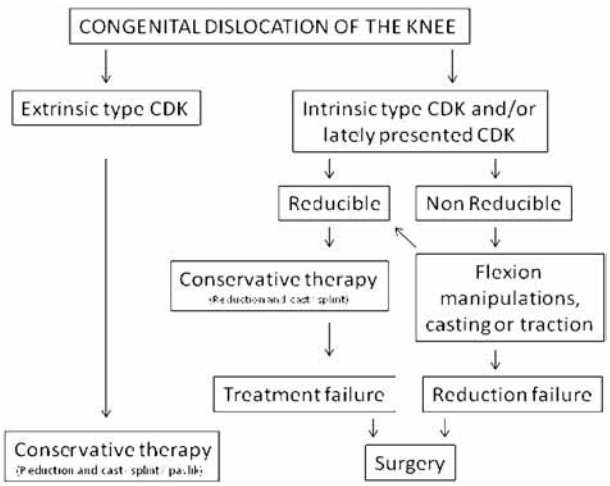

Fig. 6. — Treatment of congenital dislocation of the knee

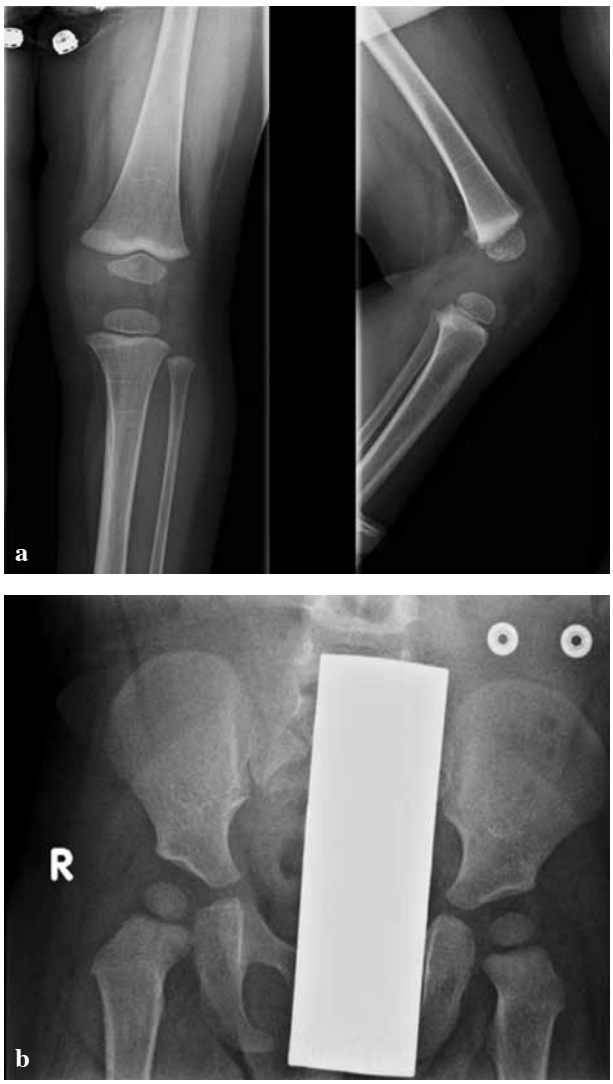

Fig. 5. — Radiograph of the left knee and the pelvis (left hip) after 20 months.

patellar pouch seen on the ultrasound are important findings. Regarding our case we can postulate that fibrosis of the quadriceps tendon or absence of the suprapatellar pouch is the result of CDK and not the cause, as mentioned before (*17,22*). Presence of the suprapatellar pouch is a sign of good prognosis and that the conservative treatment will succeed. Absence of the suprapatellar pouch has been one of the repeated findings in those patients in whom conservative treatment failed (*10*).

In case of the intrinsic type CDK (or CDK presented in a later stage (*1,17*)) one should first evaluate the reducibility of the knee. If reduction is not achievable then a period of manipulation and casting or a period of traction (1 to 2 weeks) may be necessary. It is at this moment of time that one must

be very cautious not to damage the physis. The emphasis here, in our opinion, is on the reduction and the maintenance of reduction. If reduction of the CDK cannot be achieved despite manipulative casting and/or despite a period of traction , surgery is indicated (*6,19,20*). This is also the case if reduction is possible but a 90° flexion cannot be achieved despite conservative treatment of 3 months. Some of the surgical interventions are : Z-plasty of the quadriceps mechanism, V-Y advancement, Achilles tendon allograft (*21*) and release of the anterior capsule and intra-articular adhesions (*1,3,19,20*). According to Ooishi *et al* (*19*) the best time for surgery is approximately one year after birth and before the child starts to walk (*4*) (Fig. 6).

The basic findings in CDK are : shortening of the quadriceps tendon, tight anterior capsule and absence of the suprapatellar pouch (*19*). These findings have been described for both the extrinsic and intrinsic type CDK. This was not seen in our case, except for the shortened quadriceps tendon. This may be because the CDK initiated in the last stages of the pregnancy. So the timing in which CDK initiates *in utero* could explain the severity of the secondary changes seen at the knee joint.

We emphasize clear diagnosis, exclusion of other anomalies and early treatment. Whether cervical cerclage was an associated factor in the aetiology remains a question. Further research is needed to understand whether cerclage of an incompetent cervix does increase the risk for congenital postural deformities.

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