BILATERAL CONGENITAL DISLOCATION OF THE PATELLA

P. CARPINTERO, M. MESA, A. CARPINTERO

We report the case of a six-year-old boy with Down syndrome who presented with flexion contractures of both knees and genu valgum. The diagnosis of bilateral congenital dislocation of the patella was established by clinical and xray examination. The patient was treated surgically with a satisfactory result.

Keywords : Congenital dislocation ; patella ; Down syndrome.
Mots-clés : luxation congénitale ; rotule ; syndrome de Down.

INTRODUCTION

Congenital dislocation of the patella (CDP) is a very rare condition which occurs bilaterally extremely infrequently. Twenty-eight cases have been reported so far, only four of them bilateral (3). To consider a dislocation of the patella as congenital, several criteria must be fulfilled, such as permanent dislocation, absolute inability to extend the knee ; unimpaired passive mobility of the knee joint and absence of the patella from the throchlear groove at birth (2).

CASE REPORT

J.O.B., a six-year-old boy with Down syndrome, was seen in December 1989, referred by the pediatrician because he presented with a gait disorder : knee and hip contractures as well as external rotation of both legs. This had been a problem since he began to walk at the age of 18 months. He had previously been seen in another hospital, where his gait had been attributed to the characteristic hypotonia seen in patients with Down syndrome. There was neither a history of intramuscular injection into the thigh nor a family history of patellar disorder.

Physical examination showed the characteristic facies of Down syndrome, generalized laxity, flat feet and normal mobility of both hips. The knees were in 20 valgus, with a range of motion of 0° / 30° / 145°, and the patient could not actively or passively extend the knees. Both tibiae were in external rotation, the left one more so than the right. Both patellae could be palpated on the lateral aspect of the femoral condyles. On x ray examination (figs. 1, 2) the hypoplastic patellae (the right one more than the left) were located at the lateral aspect of the lateral femoral condyles. A bilateral congenital dislocation of the patella was diagnosed, and the patient was operated in January 1990.

A release and realignment of the extensor mechanism was performed together with a release of the iliotibial band. The distal portion of the vastus lateralis was detached from the rest of the quadriceps and from the capsule proximally. The knee capsule was divided laterally and the medial articular capsule was reinforced. The patellar ligament was then divided longitudinally ; its lateral half was detached from the tibial tubercle, placed under the medial portion of the ligament, and attached to the medial side of the tibia (fig. 3) (1). In the postoperative period, both knees were immobilized in a plaster cast at 20° flexion for

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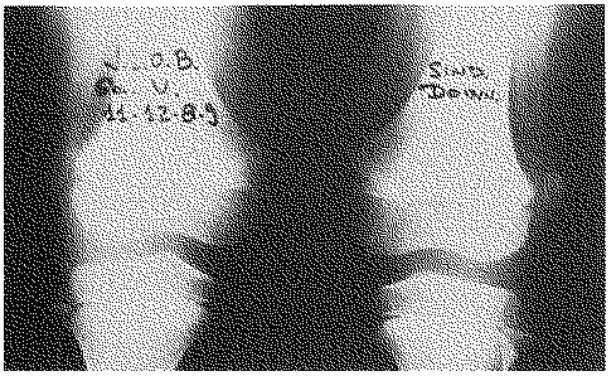


Fig. 1. — AP view of both knees, showing lateral rotation of both patellae and patellar hypoplasia (more marked on the right side).

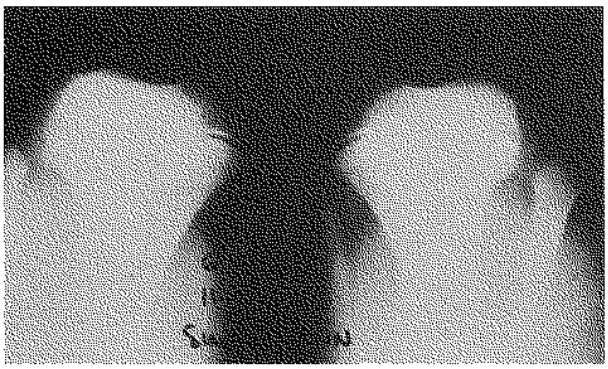
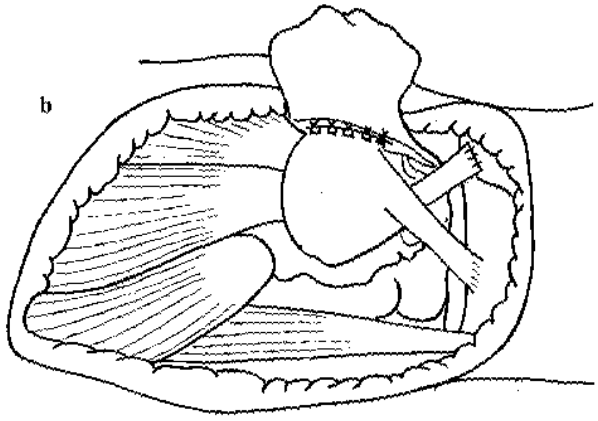


Fig. 2. — Sunrise view, showing lateral dislocation of the patella and patellar hypoplasia (especially on the right side).

a



b

Fig. 3a and b. — Release of the extensor mechanism of the patella with partial transposition of the patellar ligament and reinforcement of the medial articular capsule.

five weeks, and the patient was subsequently referred for physical therapy. At present, over four years later, the child shows active mobility of 0°/10°/140° in both knees; a discrete genu valgum of 10° persists, together with external rotation of both tibiae. CT scan of the femoropatellar joints shows the presence of a subluxing dysplasia of both patellae; this is more marked in the right knee (fig. 4).

DISCUSSION

The etiology of congenital dislocation of the patella is unknown, although a genetic background is suggested by its frequent association with other congenital disorders, such as dislocation of the radial head, congenital absence of the fibula, congenital cardiac anomalies, talipes equinovarus and carpal fusion (5). Among the cases published, five were patients with Down Syndrome (1, 3), whereas in other cases no associated disease has been found. Therefore, it may

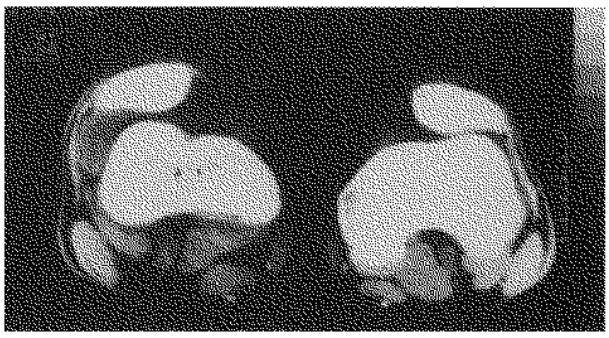


Fig. 4. — Postoperative CT scan.

be an epiphenomenon within a pattern of multiple malformations or an isolated occurrence.

It is very difficult to establish the diagnosis at birth, although the disorder is present at that time, but early diagnosis is important in order to avoid the secondary deformities which appear with growth. Although the most practical age for open reduction has not been established, the most rewarding seems to be between 2 and 3 years of age, because neglected cases require more extensive release of contractures to obtain surgical reductions and more difficult and complicated surgical reconstructions to ensure patellar stability (2, 3, 5). Consequently, this clinical diagnosis must be considered in any newborn who presents with a flexion contracture or a limited extension of the knee, once other disorders have been discarded (3). Nevertheless, the diagnosis is usually made when the child begins to walk with a permanent flexion of the knee, which is quite remarkable in the rare cases with bilateral dislocation. Once the patella has become ossified, at about the age of five, the diagnosis is easier as it may be demonstrated on xray examinations. However, ultrasonography may be useful when the child is younger.

Surgery has been considered the treatment of choice allowing relocation of the patella in its anatomic position (3, 4, 5).

We present a case of congenital dislocation of the patella in a young patient with Down syndrome : this supports the theory that in some cases, a genetically determined disorder may be the cause (5).

We suggest that in spite of its rarity, one must consider this diagnosis when faced with a newborn presenting with a flexion contracture of the knee, once arthrogryposis or other gross congenital anomalies have been discarded.

We confirm the good results of the surgical treatment through release of the patella and partial transposition of the patellar ligament (1, 4), although the prognosis in a case of Down syndrome is generally poorer than it may be in other cases (5).

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SAMENVATTING

P. CARPINTERO, M. MESA, A. CARPINTERO. Aangeboren bilaterale patella luxatie.

De auteurs beschrijven het geval van een 6-jarige jongen lijdend aan een Downsyndroom, met bilaterale flexiecontractuur van de knieën en genu valgum. De diagnose van bilaterale luxatie van de patella steunde op het klinisch en röntgenologisch onderzoek. Heelkundige behandeling met een bevredigend resultaat voor gevolg.

RÉSUMÉ

P. CARPINTERO, M. MESA, A. CARPINTERO. Luxation congénitale bilatérale de la rotule.

Les auteurs décrivent un cas rare de luxation congénitale bilatérale de la rotule chez un enfant avec syndrome de Down. Le patient présentait un genu valgum associé à un flexum permanent et irréductible. Le traitement chirurgical a permis d'obtenir un résultat satisfaisant.