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CONGENITAL COMPLETE DISLOCATION OF PATELLA CAUSING SERIOUS DISABILITY IN CHILDHOOD: THE OPERATIVE TREATMENT

By

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While the congenital predisposition to recurrent dislocation of the patella in its various forms has been dealt with quite extensively in the literature (*De Palma* 1954, *Kapel* 1936), the author has been unable to find any description of the complete, congenital, irreducible dislocation of patella as an ailment *per se* since that of *Mouchet & Durand* (1921).

However, the condition may well be more common than formerly believed, and may possibly be the underlying cause in some cases of permanent, total dislocation of patella in adults, because the malformation for years may give rise to very few symptoms and thus escape diagnosis during childhood. At a later stage, secondary arthrosis may develop in the knee, but is inconstant. Cases are known where the patient with a total dislocation of patella has not been bothered by the disorder throughout a long span of life.

As in cases of congenital, complete dislocation of patella which have been reported earlier, the disorder was also seen in the two present cases together with other congenital abnormalities. But here the progressive disability caused by the patellar dislocation completely dominated the clinical picture. The author was unable to find any description of the characteristic clinical findings in similar cases. The rarity of such cases prevents the presentation of a larger number of cases:

Case 1. Marit L., born 1956, girl, was admitted to the hospital at the age of five. She had an osteotomy performed on both femurs (probably a rotation osteotomy) in another hospital at the age of one year. The osteotomies had united in some varus

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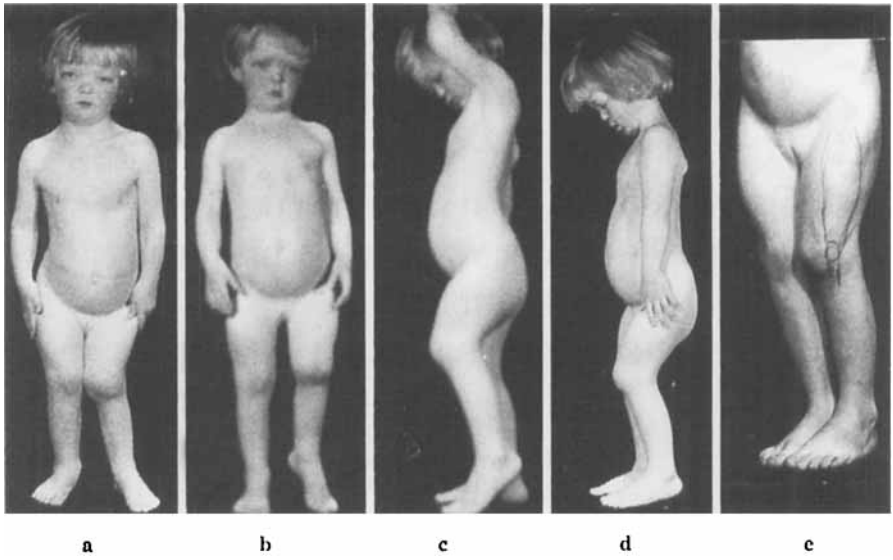


Fig. 1.
Marit L., before operation. Demonstrates the typical clinical findings: Flexion contracture, valgus of the knee, outward rotation of the leg. Apparent, but no real shortening of the limb. The foot kept in equinus when walking. To the right the laterally dislocated extensor apparatus with the patella has been drawn on the skin.

position. Roentgenologically and clinically, both hips were normal. She had several congenital malformations (a non-syphilitic saddle-neck nose and moderate malformations of the external genitals). The actual disorder was located in the left lower limb. She displayed a marked limp on this side, walking with the left foot in equinus, which was caused by a 45° flexion contracture of the knee. From this position she could only bend the knee another 10° (Fig. 1). There was no real shortening of the limb, only a relative shortening due to the flexion contracture, and she lowered the left iliac spine when walking. In order to elevate the iliac spine to a normal level, she increased the plantar flexion of the foot accordingly. When asked to bring the heel to the ground, she did this by flexion of the other knee. In addition a 20° valgus was found in the knee, with some outward rotation of the leg. These factors made walking extremely difficult and deformed her posture (Fig. 1). The underlying cause of this flexion contracture of the knee was found to be a complete lateral dislocation of the patella (Figs. 2 A–B). The lateral patellar position of patella with the quadriceps muscle and the infra-patellar ligament are drawn in the right photographic picture in Fig. 1. As may be seen, the extensor apparatus of the knee has no possibility of normal function.

Case 2. Berit D., born 1953, girl, had arthrogryposis multiplex with pathological changes of both hips and right-sided club-foot. Her club-foot had earlier been treated elsewhere, and she was admitted to the Coast Hospital for treatment of a relapse, which was done by operation. Upon control examination it was noted that she

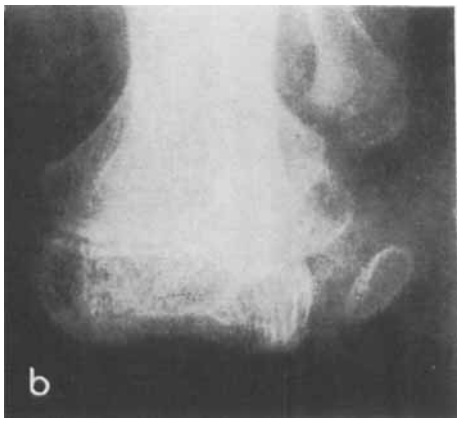


Fig. 2 b. Marit L. Roentgenogram of left knee before operation, tangential view: patella in its lateral position.

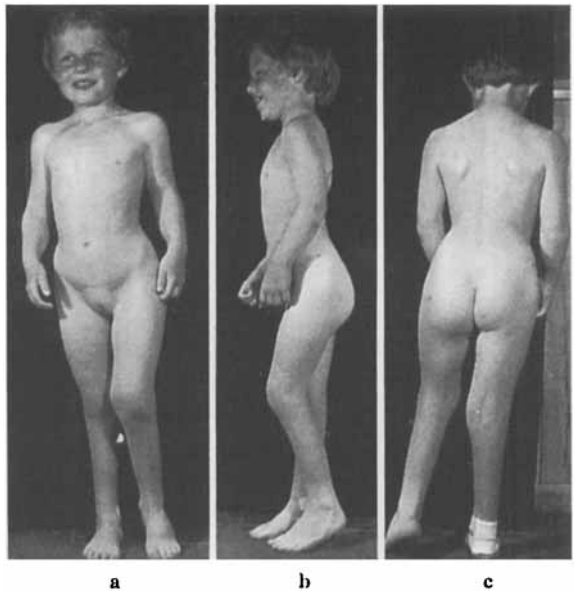
Fig. 2 a. Marit L. Roentgenogram of left knee before operation: patella lateral to the lateral femoral condyle. A wide gap is seen in the medial inter-articular space resulting from valgus deviation of the leg. Fibula projected behind tibia demonstrates outward rotation of the leg.

walked with her left knee in flexion and in valgus and with some outward rotation of the leg. She also kept her foot in equinus contracture or other pathological changes of this foot (Fig. 3). A closer examination of the knee disclosed a flexion contracture of 10°, a valgus of 30° and an outward rotation of the leg. The clinical picture was analogous to that seen in the first case, and the cause was found to be the same: a total dislocation of patella, which could be felt at the lateral aspect of the lateral femoral condyle. This finding was confirmed by the roentgenogram, which also gives an explanation of the pronounced valgus of the knee: a marked gap in the medial part of the joint (Figs. 4 and 5).

TREATMENT

As the operation in these cases is more complicated than in even the more severe cases of irreducible patellar dislocation of the common type, the technique will because of that be described in detail.

The operation was performed in two stages:



a b c

Fig. 3.
Berit D., before operation: the same typical clinical picture as in the first patient: Flexion contracture and valgus of the knee, outward rotation of the leg, compensatory equinus of the foot when walking.

I. stage: Correction of the flexion contracture: through an ordinary bayonet incision in the popliteal fossa, all flexor tendons of the thigh were lengthered, together with incision of the posterior fibrous capsule. In the first case, where the flexion contracture was most marked, it became necessary also to loosen the femoral origin of the gastrocnemical heads to achieve full correction. After the operation a plaster-cast was worn for six weeks, the first four weeks with the knee in slight flexion to avoid injury to nerves and vessels by too strong tension. The last two weeks the patient was allowed to walk about with a plaster-cast with the knee in full extension. During the following six weeks no plaster-cast was worn, and active exercises of the knee were performed.

II. stage: Twelve weeks after the first operation: transposition of the extensor apparatus (Figs. 6, A–B). This procedure necessitated a rather radical technique. A modified Krogius operation was combined with moving of the tibial tuberosity in medial direction. The rectus muscle had to be dissected free in a proximal direction to a considerably larger extent than in the usual Krogius operation, in order to place the muscle in a straight line with the patella after this had come into a normal

Fig. 4.

Marit D. Roentgenogram of left knee before operation. The picture similar to that of the first case. Note the wide gap in the medial part of the joint due to the valgus of the knee. Outward rotation of the leg. Patella is laminated.

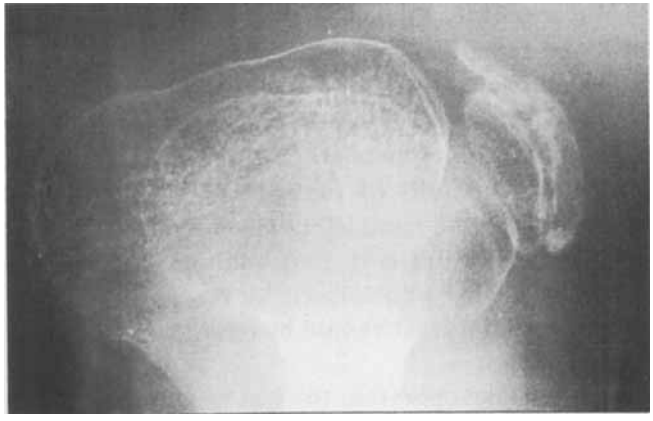


Fig. 5.

Berit D., roentgenogram of left knee before operation, tangential view: note extreme lateral, almost dorsal dislocation of patella.

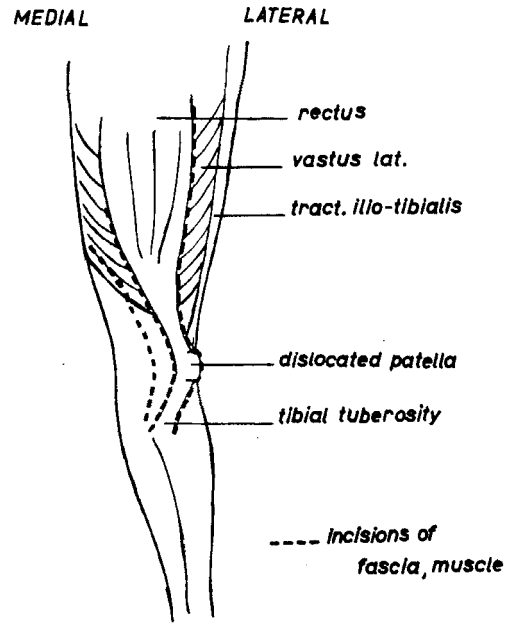


Fig. 6 a.

Diagram showing the situation before operation. Dotted lines: incisions in fascia and muscles to free the rectus with tha patella, and to prepare a medial flap to be transposed to cover defect on lateral side (see Fig. 6 b).

position. To obtain this normal position of patella, the tibial tuberosity had to be moved medially.

The rigid and markedly thickened tractus ilio-tibialis, which was exposed through the same incision, was incised at the beginning in order to correct the valgus of the knee. In both cases the valgus and the medial gap in the joint were spontaneously corrected when the ilio-tibial tract was cut (see Fig. 4, before operation, and Fig. 11, after operation).

The defect created laterally by the transposition of the extensor apparatus was to large to be completely covered by the flap which was transposed from the medial side, and additional cover was obtained from the distal, pedicaled flap formed by the cut ilio-tibial tract. For this reason, the ilio-tibial tract should be cut at a sufficiently high level (Fig. 6-B).

After the second stage operation the hip was included in the plaster-cast, and was immobilized in slight flexion to release tension of the rectus muscle. The cast was removed after four weeks and replaced with a plaster-cast allowing the patient to be up and about. After an-

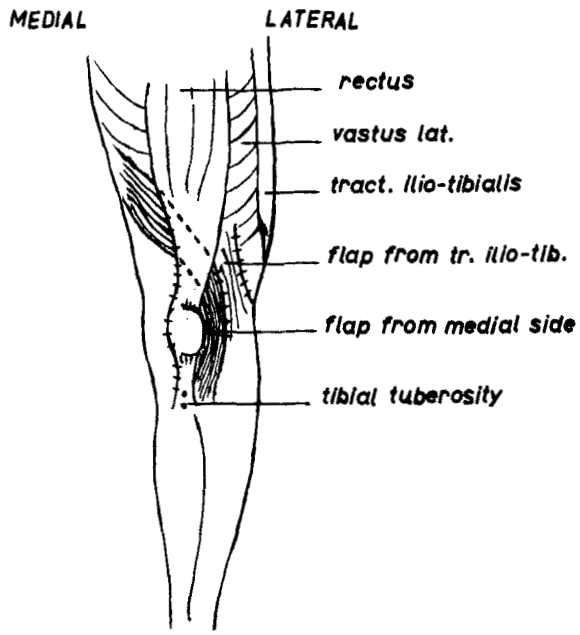


Fig. 6 b.

Diagram showing the situation after operation: The rectus muscle and patella have been brought into a normal position. The tibial tuberosity has been transposed more medially and fixed with two nails. Defect on lateral side is covered with flap from the medial side (pulled under the rectus tendon) and with part of tractus ilio-tibialis.

other three weeks the cast was removed, and moving exercises were started.

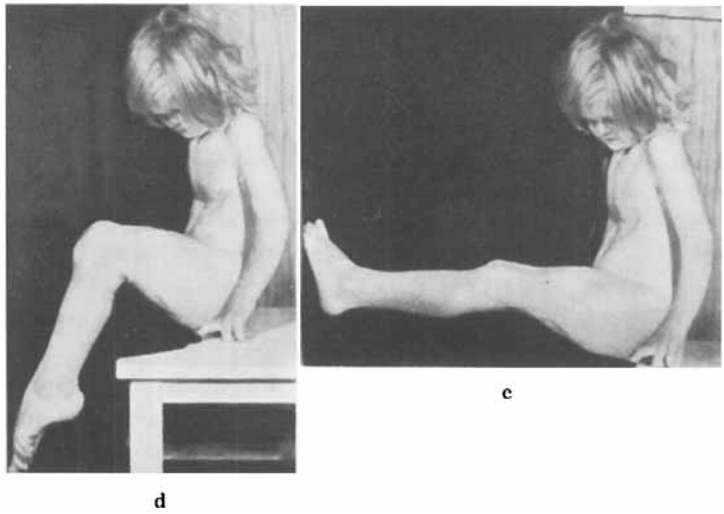
Upon control examination four months p.o. of case 2, the patella appeared to be located in front of the lateral condyle, and six months later a correcting operation was done, when the tibial tuberosity was moved another centimeter in a medial direction. A new medial strip, two centimeters wide, was prepared and transposed to the lateral side to cover the new defect. X-ray control now showed the patella in the mid-line.

RESULTS

Case 1. Examination three years after the operation: the patient walks and runs normally without a limp. She is able to bend her knee 90° + 20° (Fig. 7). She can also extend her knee fully from a hanging position of the leg against some resistance. In the prone position, flexion to more than 90° is performed with good force. The knee shows a straight

a b c

Fig. 7.
Marit L. Three years after operation: patella in normal position. No valgus of the knee. Both lower limbs equally long. Flexion of the knee 90°, full active extension with unsuspended leg.



d c

axis with no valgus. Patella is felt in front in the mid-line (Figs. 8, A-B-C).

Case 2. Examination 1½ years after the operation reveals a slight limp, which was attributed to her dysplastic hips (Trendelenburgs symptom slightly positive). However, the pronounced limp and equinus which she had before the operation were no longer present. She could bend her knee 90° + 5° and had full active extension on a hanging leg. The valgus was eliminated on the operated side (Fig. 9). A tangen-

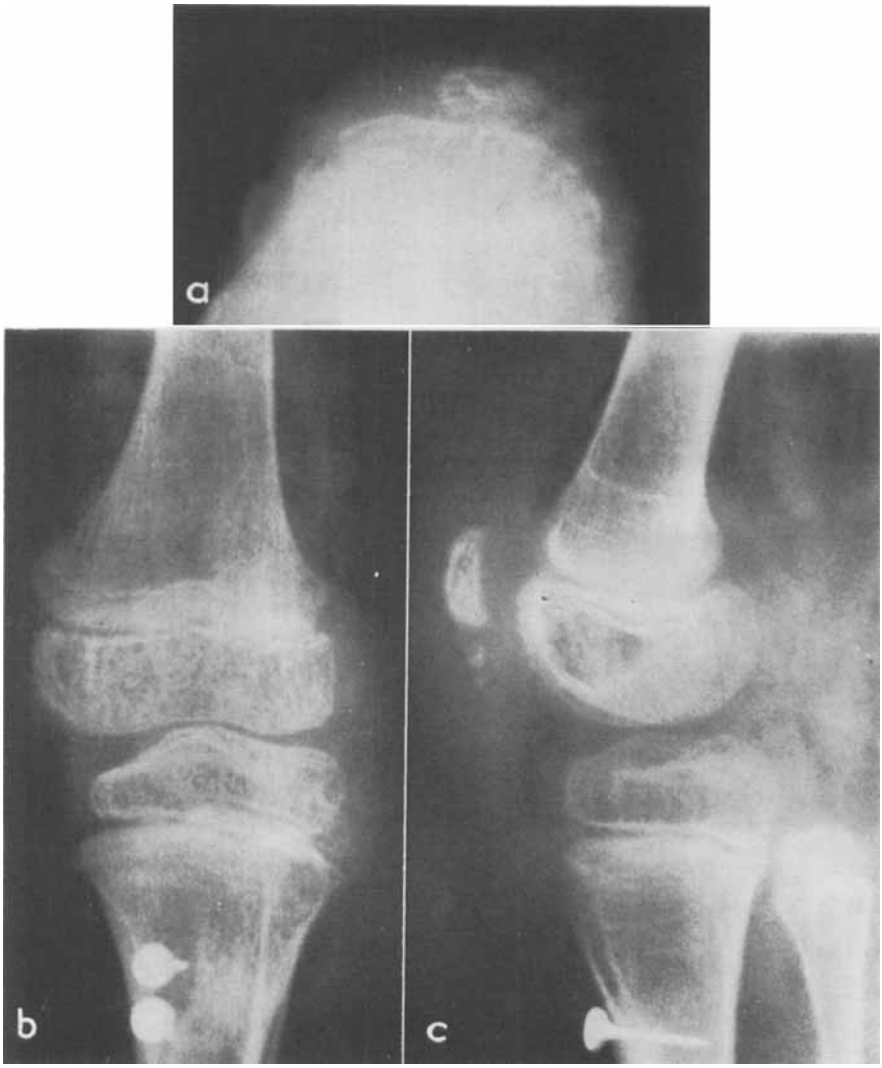


Fig. 8.

a. Marit L. Roentgenogram of left knee after operation, tangential.view: patella in normal position.

b. Marit L. Roentgenogram of left knee, three years after operation. No medial gap of the joint, no valgus. Patella in the mid-line.

c. Marit L. Roentgenogram of left knee, three years after operation, lateral view: patella in normal position. The fragmentation and calcification seen at the lower pole may be seen normally. Note: dorsal position of fibula is still present.

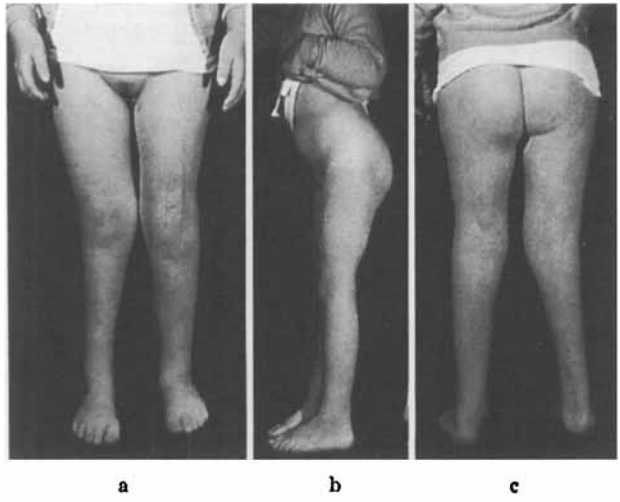


Fig. 9.
Berit D., The operated left knee as seen 1½ years after operation. Patella in normal position. No valgus. Flexion to 90° with good force. Full active extension.

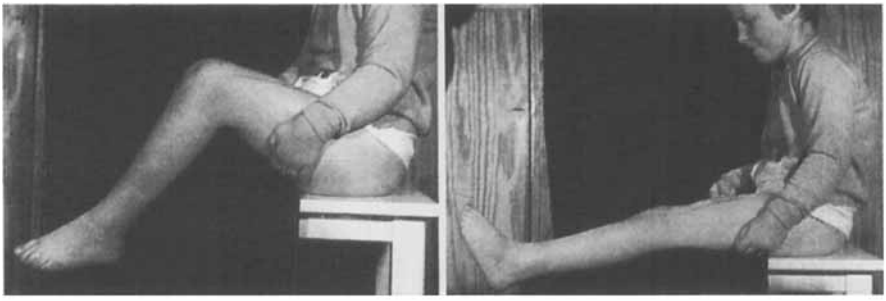


Fig. 10.
Berit D. Roentgenogram of left knee, 1½ years after operation, tangential view: patella in normal position.

Fig. 11.
Berit D. Roentgenogram of left knee, 1½ years after operation: no medial widening of the joint, no valgus, patella in the midline.

tial roentgenogram showed the patella in the midline (Fig. 10), and the medial gap in the joint was no longer present (Fig. 11).

SUMMARY

Congenital, complete, irreducible dislocation of patella is a rare condition, but may be less rare than formerly believed. It is suggested by the author, that some cases of permanent, complete dislocation of the patella in adults may be of congenital origin. The author has observed a patient who had gone through a long span of life with a total, irreducible dislocation of patella almost without symptoms from her knee. Admittedly, in this case symptoms from the knee may have been overshadowed by a concommitant bilateral, rather painful disorder of the hips, but it was felt that the patellar dislocation was of congenital origin.

Although the condition thus may give no considerable symptoms

throughout life, it may in some cases lead to pronounced and progressive disability already in childhood, as shown by the two cases reported in the present paper, and therefore the recognition of the disorder is important.

The disability is due partly to the inefficiency, partly to the abnormal function of the extensor apparatus of the knee. The resulting typical clinical picture is: flexion contracture and valgus of the knee, and outward rotation of the leg.

These cases necessitate a more radical operation than even severe cases of irreducible, complete patellar dislocation of the common type, and the operation has to be performed in two stages:

Stage I: Correction of the flexion contracture.

Stage II: Radical transposition of the extensor apparatus with the patella. Correction of the valgus position is obtained by incision of the ilio-tibial tract. The operative procedure is described.

The author has operated two girls, five and eight years old. The results in both cases were peermanent and excellent as judged by clinical and roentgenological examination after three and 1½ years, respectively.

RESUME

Une dislocation congénitale, totale et irréductible de la rotule est une maladie rare, peu traitée dans la littérature. Elle est cependant plus fréquente qu'on l'avait supposé autrefois, étant donné que certains cas de luxation totale permanente chez les adultes peuvent être congénitaux. L'auteur a ainsi observé le cas d'une malade qui avait eu pendant la plus grande partie de sa vie une luxation totale irréductible bilatérale de la rotule, sans que la maladie l'ait notoirement gênée. Peut-être était-ce parce qu'elle souffrait simultanément d'une maladie congénitale bilatérale de la hanche, très douloureuse, que les ennuis provenant des genoux étaient restés dans l'ombre.

Bien qu'il puisse donc arriver que la luxation patellaire congénitale totale donne peu de symptômes, l'auteur montre qu'elle peut déjà dans la plus tendre enfance provoquer une invalidité croissante et qu'il est important de poser à un stade précoce le diagnostic et de pratiquer l'operation.

L'auteur décrit les symptômes caractéristiques qui n'ont pas été décrits antérieurement dans la littérature. Par ailleurs, il décrit la méthode opératoire. L'intervention doit avoir lieu dans ces cas en deux temps et elle est beaucoup plus radicale que celle habituellement appli-

quée, même dans le cas graves de luxation patellaire totale et irréductible.

L'auteur a opéré deux fillettes âgées respectivement de 5 et 8 ans. Après une période d'observation de 3 ans et de 1 an et demi, les résultats ont pu être considérés comme très satisfaisants.

ZUSAMMENFASSUNG

Kongenitale, nichteinrenkbare Totalluxation der Patella ist ein seltenes, in der Litteratur wenig besprochenes Leiden. Es ist doch wahrscheinlich ein häufigeres Leiden als angenommen, da gewisse Fälle von permanenter Totalluxation bei Erwachsenen einen kongenitalen Ursprung haben können. Der Verfasser hat zum Beispiel eine Patienten gesehen, die einen grossen Teil ihres Lebens mit einer doppelseitigen, uneinrenkbaren Totalluxation der Patella behaftet war, ohne dass das Leiden erwähnenswerte Beschwerden hervorgerufen hatte, möglicherwiese doch, weil sie gleichzeitig ein doppelseitiges, schmerzvolles, angeborenes Hüftleiden hatte, das eventuelle Beschwerden von seiten der Kniee in den Hintergrund drängte.

Obwohl es also vorkommen kann, dass die Totalluxation geringe Symptome gibt, weist der Verfasser nach, dass sie bereits im frühzeitigen Kindesalter zu einer zunehmenden Invaliditet führen kann und dass die Diagnose und Operation im Früstadium wichtig ist.

Der Verfasser beschreibt die charakteristischen Symptome, die bisher nicht in der Litteratur beschrieben worden sind. Weiterhin beschreibt er die Operation, die in diesen Fällen zweizeitig ausgeführt werden muss und die bedeutend radikaler ist als die, welche gewöhnlich selbst bei schweren Fällen von nichteinrenkbarer Totalluxation der Patella angewendet wird.

Der Verfasser hat zwei Mädchen im alter von 5, beziehungsweise 8 Jahren operiert. Das Ergebnis wird nach 3 und 1½ Beobachtungszeit vorgewiesen und kann als sehr gut bezeichnet werden.

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