

CASE OF CONGENITAL ABSENCE OF THE PATELLA AND ITS TREATMENT

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On going through the literature (see the bibliography given by Hindse-Nielsen, together with the additional references given here), but very little is found as to the treatment of this defect, and most often without any observation period given for the individual cases.

In most of the cases reported the absence of the patella has been combined with other defects as dislocation of the knee, genu recurvatum, defects of the femur and fibula, club-foot and dislocation of the hip (or various defects of the upper extremities, including often the functionally insignificant absence of the nails of the thumbs).

In these cases the treatment depends primarily on the chief defect as dislocation of the knee, etc., the patellar defect being a minor matter. In most of the cases of unilateral or bilateral absence of the patella without any other essential defect the function of the knee has been normal (thus Hindse-Nielsen has collected 27 cases; to these may be added the case reported by Starup and another one described by Holldack).

About half of the cases reported were observed in children; and in several of these children the patella has later developed (the cases described by Kjeldgaard) when the function of the knee-joint and the quadriceps had become normal. The symptomatic patellar defects as a rule show large femoral condyles and a large tibial tuberosity in compensation for the patellar action

as a hypomochlion, besides a vigorous quadriceps muscle and tendon which slides in the intercondyloid fossa. In a few cases, however, the quadriceps fails on deep flexion of the knee, and the patient tires readily. In one case (Heine) the quadriceps tendon was dislocated laterally on flexion of the knee; but on deep flexion of the knee this tendon shifts medially and the patient breaks down at the knees.

That absence of the patella may cause no particular inconvenience as long as the quadriceps is vigorous is evident, for instance, from the cases reported by Tscherning, Knode and Kudlek who have removed the patella without any functional defect.

Of cases without other essential defects and in which the treatment is described, the following have been reported:

Kjeldgaard: Child, 1 month old, with congenital hyperextension in the knee-joint, moving from 10° normal flexion to 90° forward flexion, and bilateral absence of the patella. Treatment: Massage and passive motions abolished the hyperextension in 26 days and rendered flexion of 45° possible. The child learned to walk at the age of 18 months.

At the age of 6 years complete flexion practicable in the left knee (95° flexion in the right). A normal patella is present in both extremities and the gait is normal.

Mayer: Boy, 10 years old with absence of the patella on the left side and a small patella on the right. Mayer recommends treatment with stretching directed against the extension defect and tendon plastic against the atrophy of the quadriceps.

Silfverskiöld (also reported by Schanz). Woman, 19 years old, otherwise normal. The left knee shows an active and passive extension defect of 20° . The right knee shows an extension defect of 90° and pes equinus. Both patellae are absent. The action of quadriceps is fairly good; insignificant lateral looseness of the knee-joint. Rather pronounced bilateral flatfoot. Treatment I: Supracondylar osteotomy of the femur and extension in order to straighten the knees. Treatment II: Arthrodesis of the right ankle, with pes equinus position for balancing of the shortness

of the leg. Treatment III: Plastic operating on the quadriceps, on both sides, by means of the sartorius and biceps.

9 months later the patient is able without support to ascend stairways. Both knees capable of full extension and active flexion to 90° . The sartorius and biceps feel like good extensors. The patient is always wearing a supporting splint on the right extremity, sometimes on the left too. No later observation reported.

Kofmann: In one case a patella was made operatively of a disc of periosteal-covered bone. The result was a movable patella on both sides and the patient learned to stand on his feet with support. No subsequent observation reported.

Kirmisson, Phocas & Potell: Good results have been obtained by exercise of the muscles with massage and electricity.

Kirmisson: Girl, 6 years old, with absence of both patellae and with flexion contracture in the knees. Treatment: Bilateral supracondylar osteotomy of the femur with straightening of the knees; the flexors of the knee moved more anteriorly. 6 months later, able to walk with crutches; the extensors of the knee appeared normal. No subsequent observation.

Holldack: Boy, 8 years old, with bilateral absence of the patella is provided with protective knee bandages, as in falling he bruises the knees worse than other boys, owing to the absence of patellae. Otherwise he is free from symptoms.

Hohmann: 5-year-old normal boy with bilateral absence of the patella, tiring readily, and knees giving way. The quadriceps, tendon and the inferior ligamentum patellae are not palpable. Medial femoral condyle markedly protruding; no genu valgum. Flexion complete. Slight defect in extension, with compensatory pes equinus. Knees flattened.

Treatment: Incision, 25 cm. in length, made from the transition between the middle and upper third of the femur to the tibial tuberosity. Quadriceps, muscle and tendon normal. But the greater part of the tendon goes down into the lateral capsule of the knee. The tendon is freed and attached together with the freed sartorius to the medial aspect of the tibial tuberosity. On reexamination, 2 months later, the gait is normal, without pes

equinus and without the knees giving way. Now the knees are capable of full extension except for 20° . Subsequent observation wanting.

Writer's Case.

Here an account will be given of the case of a patient who has become symptom-free through the operative treatment and in whom a fairly long observation period has shown the favorable result to be permanent.

The patient is a girl with a family history of good health and a past history of perfect health except for her knee affection. She had always been walking poorly. In 1936, when she was 8 years old, she was referred to this hospital where bilateral congenital absence of the patella was ascertained, together with lateral displacement of the quadriceps tendon and complete loss of active extension. Flexion contracture of 25° on the right side, 15° on the left.

Plaster bandages were applied to both knees in the best correction possible. After 2 weeks the knees were almost straight. Then operation (Bentzon) was performed: Plastic operation on the left quadriceps together with transplantation of the semitendinosus and semimembranosus.

An arcuate incision, medially convex, was made over the anterior aspect of the left knee, baring the lower part of the quadriceps and its insertion. The patella was absent but a rather vigorous tendinous streak was found, corresponding to the inferior patellar ligament. In addition, laterally a vigorous fibrous streak was found corresponding mostly to the vastus externus, which above turned into this streak and a rudimentary rectus femoris; below and laterally to the patellar ligament it apparently continued directly into the proximal part of the anterior group of muscles of the leg. This fascia element was detached from the upper part of the tibia, and for a distance of 12-15 cm. above; then it was shifted medially and sutured to the patellar ligament. Now a slight flexion of the knee showed that these muscle-tendon portions still had a tendency to lateral displacement on flexion of the knee.

For this reason, the semitendinosus and semimembranosus were detached (as in quadriceps plastic) and pulled forwards to the extensor apparatus of the quadriceps and knee, where it was fixed. Thus a new extensor apparatus was made and established securely in the midline, in relation to the knee.

One month later a grossly similar operation was performed on the right side. A medioconvex arcuate incision was made on the anterolateral aspect, from the greater trochanter to the tibial tuberosity, bearing the outer anterior aspects of the extensor apparatus. The patella was absent, the quadriceps somewhat hyperplastic and terminating distally in tendon elements which were densely interwoven with the knee capsule, ran like arcuate streaks on the lateral aspects, over the lateral femoral condyle (cf. Hohmann's case). These tendon elements were connected with the bone so tightly that mobilization of this extensor apparatus had to be given up. On this account, at a higher level, a mobilized muscle snip was placed under the medial part of the quadriceps, and at the same time the connection of this muscle with the aforementioned lateral dislocated tendon streaks was broken off. Quite distally, at the attachment to the tibia, a little plastic was performed on the tendinous tissue too, with medial shifting, in order, as far as possible, to create a tendinous streak running from the quadriceps musculature to the tibia in the midline. The elements of which this streak consisted were not very vigorous. Still they were judged to be strong enough for the musculature with which they were connected. Through a medial-posterior incision the semitendinosus and gracilis were shifted over to the extensor apparatus in the usual manner. The contracture of the knee was then redressed gently. The extremity was put in plaster cast. One month later the patient was given night bandages, massage and active exercise.

The patient was discharged from the hospital 3 months after the operation. At this time both knees and, especially, the extensors were functioning excellently, with complete extension and very good strength. The gait was normal and secure; on both sides, capacity for flexion of 110°.

25 months after the operation the record says: Right qua-

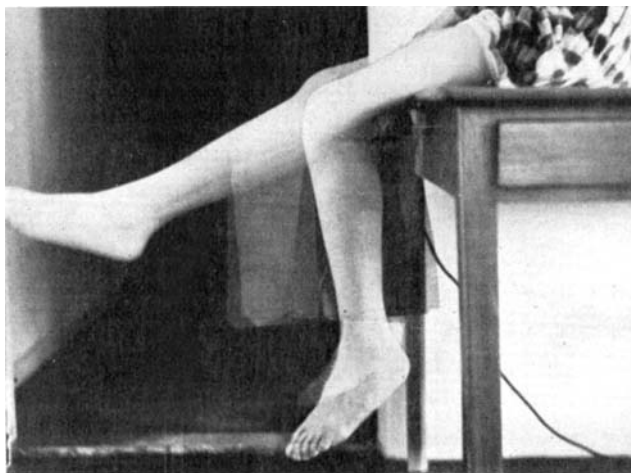


Fig. 1.

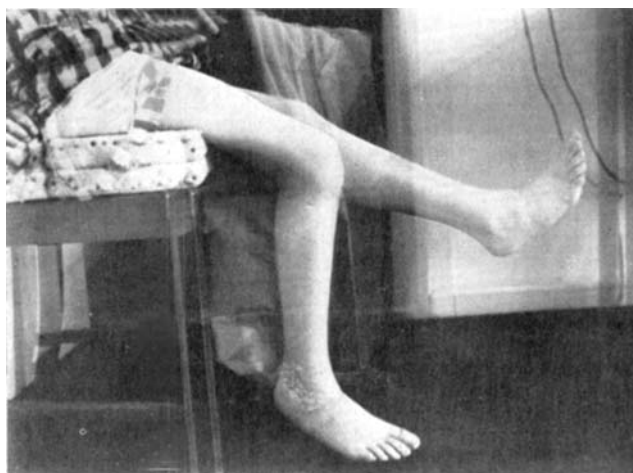


Fig. 2.

The photos are exposed twice to show the capacity of active knee-extension.

driceps shows a surprisingly natural form. The patient is able easily to extend her right leg and has sufficient reserve power to keep it in the horizontal plane (as against an extension defect

of 10°, use of sandbag). On the left side, where the "material" was more delicate, the extensors are weaker, and the patient is able to keep the leg extended against its own weight only for a short time.—The lateral preponderance of the transplanted extensor apparatus on the left side is so pronounced as to give



Fig. 3.

a tendency to rotatory subluxation under the extension. As there are two good inward-rotating flexors, one of them can be shifted forwards.

28 months after the operation the vigorous semitendinosus of the left extremity was transplanted to the extensor apparatus of the knee.

Reexamination, 3 years after the operation: Gait normal, without tiredness, "she is able to walk any distance; and to ride 30-40 kilometer on her bicycle daily."

Physical exam.: "She walks rapidly and freely, though with slight stiffening of the knees." No difference between the two

extremities. The knees are extended fully; slight valgus position of both knees. Intermalleolar distance 4 cm. (see Figs. 1 and 2).

X-ray exam. (Fig. 3) of both knees shows slight configuration, the lateral condyle being considerably broader than the medial. But there is no sign of any affection of the joint cartilage or other symptoms of arthrosis. No patella can be made out.

Reexamination, 7 years after the discharge: The patient is now 15 years old. Her condition is satisfactory. She says she is now able to "swim, ski and skate."

Physical exam.: Gait normal. When lying on her back she is able to elevate both extremities straight. The extensors of the knee are good and apparently equally vigorous on both sides. Flexion and extension normal on both sides. With knees together, the malleolar distance is 3 cm.

In this case, then, the operative treatment has resulted in a function which has to be characterized as quite normal; and the relatively long observation period (7 years) offers some guarantee that the result may permanent.

SUMMARY

Report is given of a case of congenital bilateral absence of the patella with marked lateral dislocation of the extensor of the knee. Through plastic operation of the extensor of the knee, combined with transplantation of medial flexors to the quadriceps tendon, it has been practicable to obtain a normal function of the joint. Observation period after the operation: 7 years.

ZUSAMMENFASSUNG

Es wird ein Fall von angeborenem, doppelseitigem Fehlen der Patella mit gleichzeitiger stark lateraler Dislocation des Streckmuskels des Knies mitgeteilt. Durch plastische Operation an dem Streckmuskel, kombiniert mit einer Transplantation medialer Flexoren zur Quadricepssehne hat man vollkommen normale Funktionen erzielt. Observationszeit nach der Operation: 7 Jahre.

RÉSUMÉ

Il est communiqué un cas congénital où les deux rotules manquent en même temps qu'il y a une forte dislocation latérale des muscles extenseurs des genoux. On a obtenu une fonction entièrement normale en pratiquant une opération plastique des muscles extenseurs, combinée avec la transplantation de fléchisseurs médiaux. La période d'observation qui s'est écoulée depuis l'opération s'étend sur sept ans.

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