FAMILIAL JOINT LAXITY AND RECURRENT DISLOCATION OF THE PATELLA

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Recurrent dislocation of the patella probably has several causes. In many patients abnormal laxity of the knee is present and this may predispose to dislocation (De Palma 1954). A family has come to our notice in which three members suffered recurrent dislocation of the patella and a fourth had probably sustained a single dislocation. All the former had abnormally lax knees and most other joints showed similar hypermobility. Five other living members of the family were also "double jointed" but had not suffered dislocation.

The particular interest of the patients we shall describe lies in the association of familial joint laxity with recurrent dislocation of the patella.

CASE HISTORIES

The pedigree of this family is shown in Figure 1.

The boy, M. S. (IV 3 in the pedigree), who drew attention to the family was six years old when first seen by Mr G. Lloyd-Roberts at the Hospital for Sick Children. He had suffered from recurrent dislocation of the right patella about once a month since the age of four years, there being no history of injury. On examination the right patella showed abnormal lateral mobility; the medial and lateral ligaments of the knee were abnormally lax and allowed no less than 30 degrees of abduction and 10 degrees of adduction on straining. There was genu recurvatum of 20 degrees. The left knee showed similar, but less marked, instability. There was no fixed knock-knee deformity. Other joints showed an abnormal laxity, most evident in the ankles, wrists and metacarpophalangeal joints (Fig. 2) but present also in the shoulders, elbows and interphalangeal joints. Radiographs of the knee showed no abnormality of bone structure.

FIG. 1

Pedigree of the family with recurrent dislocation of the patella and joint laxity.
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At operation the medial capsule and patellar retinaculum were markedly atrophic, and this appeared to be the only significant abnormality. To overcome the recurring dislocation the patella was held in place by taking a strip from the quadriceps expansion at the medial side, looping it round the rest of the quadriceps expansion and stitching it to itself; the semitendinosus was also transplanted to the medial border of the patella. The patellar ligament was then split and the lateral half reattached to the tibia medial to its other half. Six months after the operation there had been no further dislocation and an almost full range of movement had been regained.

The mother of the boy, Mrs S. (III 2 in the pedigree), aged twenty-six, had also had recurrent dislocation of both patellae since childhood, which occurred spontaneously about once a month until recently when the frequency diminished. The clinical findings were very like those in her son, the patellae being unduly mobile with a tendency to subluxate laterally on flexion of the knee. She also showed the excessive lateral mobility of the knees (Fig. 3) and genu recurvatum of 15 degrees. Both lateral femoral condyles appeared underdeveloped, and this was confirmed by tangential radiographs of the patello-femoral compartment. She had no other lax joints.

Mr M. (II 3 in the pedigree) was the grandfather of M. S. and the father of Mrs S. Born in 1901, he gave no history of dislocation of the patella, except possibly once on the left, twenty years ago. He, like M. S. and Mrs S., had laxity of both knees. The left knee showed osteoarthritic changes with a small effusion. Radiographic examination was not possible.

Three other living members of the family have a history of weak knees. These are III 6, a brother of Mrs S.; II 5, a brother of Mr M.; and III 9, a son of II 5. On examination, each

FIG. 2

Figure 2—An illustration of the remarkable joint laxity in the index case, of the boy M. S., aged six.

FIG. 3

Figure 3—Strain radiograph of the mother of the boy in Figure 2 which shows the gross lateral instability of the knee.
of these was found to have genu recurvatum and laxity of the lateral ligaments of both knees. III 9 had bilateral talipes equinovarus. All other living members of the family were examined and one more, II 7, was found to have laxity of the ligaments of both knees. II 5 and III 6 also had significant laxity of the interphalangeal and metacarpo-phalangeal joints. None of these men, however, had had any history of dislocation of the patella. In addition two deceased members of the family, the great-grandfather, I 2, and a great-aunt, II 1, were probably affected. The great-grandfather was said to have "knees which stuck out at the back like ours," and the great-aunt was said to have had the weakest knees in the whole family. "She was a fat woman whose knees gave way every time she laughed." Towards the end of her life she was able to walk only with support, and the account given by the sisters was so characteristic of dislocation of the patella in association with joint laxity that we regard this diagnosis as established.

**DISCUSSION**

In a study of fifty-four patients with recurrent dislocation of the patella, De Palma (1954) found that "many cases disclosed some lateral instability of the joint arising from generalised relaxation of all the ligaments about the joint." The primary abnormality in the family we report is joint laxity, always affecting the knee, but often other joints as well. Of the six affected members of the family, only one, Mrs S. (III 2), presented any of the other known predisposing causes of dislocation, in this case hypoplasia of the lateral femoral condyles.

The pattern of inheritance in this family is that of a dominant gene, arising as a mutation probably in the great-grandfather (I 2) of our patient M. S. (IV 3). Similar joint laxity with a dominant pattern of inheritance is seen in osteogenesis imperfecta, Marfan's syndrome (arachnodactyly) and Ehlers-Danlos syndrome (McKusick 1956). Patellar dislocation may occasionally occur with these syndromes, but no other signs of any of these conditions are present in this family. Families with joint laxity alone and the pattern of inheritance of a dominant gene have been described by Key (1927) and Sturkie (1941). In Key's family (in which the author is in error in calling the condition sex-linked, since a pair of affected members are father and son) all the joints were affected, and one of the five affected had recurrent patellar dislocation; and it is interesting also that four had talipes equinovarus (like III 9, in our family). In Sturkie's two families there was no dislocation. The family we report belongs to this group of families with joint laxity due to dominant genes.

In spite of the frequency with which lax knee joints are found in association with recurrent dislocation of the patella, this condition is rarely found in more than one member of a family. Further, in addition to joint laxity, there are probably other causes of recurrent dislocation that may on occasion be genetically determined. De Palma (1954) described one patient in his series of fifty-four, a girl whose mother was similarly affected; he did not mention any laxity of the ligaments in this patient. During the past ten years 120 patients with recurrent dislocation of the patella were treated at the Royal National Orthopaedic Hospital; no special family inquiry has yet been made for these, but examination of the notes showed that three (two in the same family) had a relative affected. These three patients were specially examined for joint laxity but none was found. In another girl, whose mother was similarly affected, seen at the Hospital for Sick Children, neither had joint laxity. Lange (1951) has described a Eurasian family with recurrent patellar dislocation; his patients were a girl and her mother, but two sisters of the mother and a sister of the girl were also said to be affected. He made no mention of joint laxity. In the family reported by Moore (1930) the underlying abnormality may have been like that of the family we describe. His patient had recurrent dislocation of both patellae, associated with "a lax joint capsule," and both knees were treated successfully by operation. Nine other members of this family were reported to be similarly affected, but were not examined.
SUMMARY AND CONCLUSIONS

The family we record draws attention to an association between recurrent dislocation of the patella and joint laxity, which is not confined to the knee. This may pass unrecognised if specific inquiry is not made. In this and other families reported, the joint laxity is inherited, as though due to a dominant gene, but some only of those affected suffer recurrent dislocation of the patella. It is probable that there are other genetically determined causes of recurrent patellar dislocation. In three other families we have seen more than one subject of patellar dislocation, but none had lax ligaments, and two other families have been recorded with no mention of associated joint laxity.

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REFERENCES