PROGRESSIVE FIBROSIS OF THE VASTUS INTERMEDIUS MUSCLE IN CHILDREN*  
A Cause of Limited Knee Flexion and Elevation of the Patella

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In the nine years from 1951-59 we have observed twelve children—eight girls and four boys—in whom a marked limitation of flexion of the knee joint developed in a relatively short time. The children were between one and seven years old (Table I). The parents of all of them stated that at the beginning the knee movements were restricted only very little and that it was only in the course of time that they grew more and more limited. In no case was there a previous history of trauma or of inflammation of the joint or of neighbouring tissues; the knee was never swollen, warm or painful. In the youngest children the limitation of movement was usually noticed because the child could not sit on the chamber pot in the normal way; in older children insufficient flexion prevented them from squatting. When walking, the children did so with an extended knee even if movement of the knee was still partially preserved.

The initial diagnoses in most cases were those of trauma or of inflammation, although all the usual symptoms of these conditions were missing. In one case, probably because the patellar tendon reflex was absent, the condition was thought to be a post-polio myelitic contracture. The children were initially treated by plaster splints or, more often, by manipulation and physiotherapy. In no case did such treatment effect improvement; in fact, mobility steadily decreased.

All the children were first examined by us after having been treated in different ways. We also attempted once more to mobilise the knees by manipulation, splints, massage, electrotherapy and so on, but we did not succeed even temporarily in improving the flexion. On the contrary, we found that children who were temporarily discharged from our clinic and who returned some months later, always had even a more limited flexion than during their first hospitalisation (Cases 1, 3, 4, 9, 10, 11, 12).

CLINICAL PICTURE

In eight children the condition affected only one knee and in four children there was a limitation of flexion of both knees. In one girl (Case 3) so affected there was also partial rigidity of some muscles of the leg and foot like that seen in an incomplete syndrome of arthromyodysplasia (arthrogryposis). This case is therefore rather different from the others in its suggestion that there may be a connection between arthromyodysplasia and our cases of progressive fibrosis of the vastus intermedius. There is of course the difference that in

* Submitted for publication January 1960.
### Table I: Analysis of Material

<table>
<thead>
<tr>
<th>Case number</th>
<th>Sex</th>
<th>Side affected</th>
<th>Other congenital anomalies</th>
<th>Knee flexion contracture (degrees)</th>
<th>Age at onset (years)</th>
<th>Time range of operation (years)</th>
<th>Movement after operation (degrees)</th>
<th>Investigations</th>
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<tbody>
<tr>
<td>1</td>
<td>F</td>
<td>Left</td>
<td>—</td>
<td>1 (left knee flexion 120 degrees)</td>
<td>6</td>
<td>3</td>
<td>90</td>
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<td>M</td>
<td>Right</td>
<td>—</td>
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<td>3</td>
<td>80</td>
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<td>1 (left knee flexion 135 degrees)</td>
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<td>2</td>
<td>90</td>
<td>Electromyography, Electromyography, Electromyography, Histology</td>
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<td>Right</td>
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<td>2 (right knee flexion 135 degrees)</td>
<td>9</td>
<td>2</td>
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<td>Congenital dislocation of hip</td>
<td>3 (left knee flexion 120 degrees)</td>
<td>9</td>
<td>2</td>
<td>90</td>
<td>Electromyography, Electromyography, Electromyography, Histology</td>
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<td>F</td>
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<td>4 (right knee flexion 135 degrees)</td>
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<td>5 (left knee flexion 135 degrees)</td>
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<td>6 (right knee flexion 135 degrees)</td>
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<td>90</td>
<td>Electromyography, Electromyography, Electromyography, Histology</td>
</tr>
</tbody>
</table>

**Notes:**
- Right and left indicate the affected side.
- Congenital dislocation of hip refers to the type of congenital anomaly.
- Knee flexion contracture and knee flexion indicate the degree of contracture.
- Electromyography, Electromyography, and Electromyography refer to the type of investigation.
this patient the condition of the knee joint constantly deteriorated, which does not occur in arthromyodysplasia.

The appearance of the affected knee was not markedly different from that of the other one. In older children the patella was situated somewhat higher and more laterally than normal (Figs. 1 and 2). In the older children, where the pathological process had been going on for some time, there was wasting of the thigh of about half an inch (1 to 1.5 centimetres). There was no wasting of the calf, except in the case of the girl (Case 3) with obvious affection of the muscles of the lower limb.

On the front of the thigh there could be felt a tendon-like structure attached to the patella. This was most easily felt when the knee was flexed as far as possible. There were no changes in the lateral parts of the extensor apparatus of the knee.
The patellar reflexes were diminished or even absent, and the knee flexor reflexes were weaker than normal. All the other reflexes of the lower extremity and reflexes elsewhere in the body were normal.

The knee movements were limited in a characteristic way. During an active or passive attempt at flexion the movement was always stopped exactly at one point, and quite suddenly, as if a solid obstacle were in the way. Movement was not limited by pain and even forcible attempts at flexion were not painful. It seemed that the block was caused by the middle part of the tendon of the quadriceps muscle. As extension of the hip joint was not limited, it appeared that flexion of the knee was limited by some condition affecting the vastus intermedius rather than the rectus femoris.

![Histological appearances](image1)

**Fig. 5**

Histological appearances. Figure 5—Increase in number of endomysial and sarcolemmal nuclei. (Haematoxylin and eosin, ×270.) Figure 6—Proliferation of endomysial and sarcolemmal nuclei with invasion of sarcoplasm. Degeneration of muscle fibres with partial loss of striation. (Haematoxylin and eosin, ×600.)

**Fig. 6**

**RADILOGICAL FINDINGS**

Radiographs did not show any appreciable changes, but in two children who were treated with plaster splints shortly before radiological examination there were indications of a slight decalcification. In children old enough to have a centre of ossification in the patella this was somewhat smaller than on the normal side. The patella was usually situated rather higher than on the healthy side (Figs. 3 and 4).

**ELECTROMYOGRAPHIC FINDINGS**

In most children electromyography was done. The results were generally similar: the rectus femoris and vastus intermedius did not show any activity; the vastus medialis and lateralis showed a decrease of spikes to a transient state (the action potential activity was mono- or biphasic), a decrease of spikes and a reduction of frequency. In the knee flexors the pattern was either normal or showed a slight lowering of the peaks with sometimes a decrease
of potentials. The peaks were however of normal shape. (This investigation was carried out by Dr I. Lesný and Dr VI. Janda in the laboratory of our clinic.)

Chronaximetric investigation was carried out in three children. The results were practically within normal limits. (This investigation was carried out by Dr I. Lesný.) During routine electrodiagnostic investigation the reaction of degeneration was never found.

**FINDINGS AT OPERATION**

Because of the failure of conservative treatment operation was advised in all cases. Ten children were operated on at the Second Orthopaedic Clinic and one was operated on elsewhere. The last child returned to her own country and nothing is known of her later course.

![Incision over medial border of rectus femoris. The vastus intermedius displayed and the line of division of its fascia shown (1—rectus femoris; 2—vastus intermedius; 3—vastus lateralis).](image)

**Fig. 7**

Incision over medial border of rectus femoris. The vastus intermedius displayed and the line of division of its fascia shown (1—rectus femoris; 2—vastus intermedius; 3—vastus lateralis).

![Gaping of fascial incision on bending of the knee.](image)

**Fig. 8**

Gaping of fascial incision on bending of the knee.

At operation the subcutaneous connective tissue overlying the extensor apparatus of the knee was always found to be penetrated by rather rigid fibres running vertically from the skin to the fascia, while the subcutaneous layer of fat was always rather thin. There were no very obvious changes in the muscles except perhaps a smaller volume and a higher rigidity on passive stretching. The muscles were stimulated by galvanic and faradic current; the contractions were small, slow, or completely absent (Cases 2, 3, 5, 7).

In our first cases we used the operation described by Bennett (1922), sometimes with the modification introduced by Ramadier and Lacheretz (1953). It was however seen that elongation of the contracted fascia and of the vastus intermedius was quite sufficient to permit full
flexion. In later cases therefore we limited the intervention to this procedure (Figs. 7 and 8). In some cases where the patella was markedly higher than normal the ligamentum patellae was plicated.

HISTOLOGICAL FINDINGS

Histological examination of pieces of muscle was carried out by Dr V. J. Fischer of the Institute of Pathological Anatomy (Director Professor V. Jedlička). He reports: "The diameter of muscle fibres is considerably narrowed and in many places they completely disappear.

The striping is slightly effaced. On transverse sections it can be seen that all muscle fibres show a decrease of their width and that a considerable part of all fibres of the quadriceps muscle, especially the rectus femoris and vastus intermedius, are replaced by fat. Subsarcolemmal nuclei are, in some places, activated, and endomysial nuclei increased (Figs. 5 and 6). In some places there are clusters of myophages. There is a slight increase of interstitial fat. Muscle bundles are moderately innervated, the termination of the nerve fibres giving the
impression that in some places they end blindly. This blind ending is not observed in all cases. From all investigations it is evident that there is a primary muscular disturbance. The changes in the nerves are not marked and seem to be secondary. The blind termination of nerve fibres found on histology is not constant."

The histological picture is reminiscent of that in arthromyodysplasia (amynoplasie congenitale, arthrogryposis) as reported by Poli (1929), Rocher (1933), Potel (Wiemuth 1901), Sheldon (1932), Middleton (De Palma 1954) and others.

AFTER-TREATMENT AND RESULTS

The knees were immobilised in a position of 90 degrees of flexion. Full flexion was not used because of the possibility that this might cause difficulty in re-educating the quadriceps. In all children the range of flexion of 90 degrees was maintained, though there was never even a slight improvement later (Figs. 9 to 11). The range of active flexion was nearly always the same as that of passive flexion. Knee movement up to 90 degrees is sufficient for all important movements and eventually mobility can be still more improved by a further operation in later years.

DISCUSSION

The clinical picture described resembles superficially an incomplete syndrome of arthromyodysplasia (arthrogryposis). In this condition only one part of an extremity may be affected (Cuilleret 1937), but it persists without any change from birth, and may be sometimes improved by treatment (Rocher 1933). In our cases, however, the condition first appeared in children at the age of one or more years and steadily led to increasing restriction of movement in spite of conservative treatment.

Gradual restriction of mobility due to a fibrous degeneration of certain muscles in childhood is known to occur in several syndromes: 1) Some cases of so-called congenital dislocation of the patella becoming manifest in later years only (Payr 1934, Cuilleret 1937, Leveuf and Pais 1946). Three such cases have been studied in this clinic. 2) Camptodactylia, occurring especially in girls, is often of familial incidence and is usually manifested after the tenth year of life and during puberty (Rocher and Rouill 1934, Aurousseau 1937). 3) The so-called Pouce bot flexus. 4) Some cases of digitus quintus supraductus. 5) Some cases of pectus excavatum, manifested as late as at puberty (Hněvkovský). The muscular origin of this deformity was recognised by Lester (1950).

From these analogies we may presume that in our cases there was a muscular dysplasia of congenital origin of the vastus intermedius and rectus femoris muscles, manifesting itself in later life only, progressive and not responding to conservative methods of treatment. The progressive nature of the condition is also shown by histological findings such as activation of subsarcolemmal nuclei and the presence of myophages. The congenital nature of the condition is suggested by the radiological findings of deviations and hypoplasia of the patella. The changes in the subcutaneous tissue found at operation are analogous with findings in congenital anomalies of similar character (Rocher 1933).

Fibrosis of a part of the quadriceps muscle may be the cause of different clinical conditions, according to the part of the affected muscle. If the vastus lateralis muscle is involved a lateral luxation of the patella occurs; if the middle part of the quadriceps muscle is involved it results in a “pseudoluxation”—the condition described here.

As far as I am aware, no case of deviation of the patella in a medial direction has, as yet, been described. Perhaps involvement of the ilio-tibial tract is necessary for the production of deviation, and clearly this tract cannot exert any influence on the medial side of the thigh.

As far as I am acquainted with the literature no similar case of progressive limitation of flexion of the knee caused by congenital myodysplasia has been described. Domanský (1932) described the case of a man aged forty-nine who, from childhood, had had restriction of
movement of one knee. The radiograph showed a well preserved joint space, the patella being smaller and situated rather higher than that of the other knee. Because of the limited mobility of the knee this case cannot be classified as a simple patella alta (Jansen 1929). Another similar case of congenital ankylosis with patella alta in a child of nine was published by Lombard (1934): because the operative lengthening of the quadriceps tendon was successful the author accepts the opinion of Potel (1897) that the ankylosis is of muscular origin. It is possible that more similar cases could be found but under some other diagnosis.

As to etiology, the author's view is that late disturbance of ontogenesis of the affected muscles—a dynamically active tissue—may be presumed. A congenital dysplasia of any tissue can be of varying degree and may be clinically manifested in the growth of the embryo, the foetus or the child.

SUMMARY
1. Twelve children are described in whom gradual limitation of knee flexion developed, without previous injury or inflammation.
2. Clinical and histological investigation showed progressive fibrous degeneration of the vastus intermedius and rectus femoris.
3. As the patella showed signs of retarded development a particular form of myodysplasia, probably congenital, may be supposed.
4. Conservative treatment always failed to stop the progress of this disease.
5. Surgical treatment (Bennett's method or an elongation of the ligamental apparatus of the intermedius muscle) was carried out in ten cases and gave good permanent results. Flexion remained possible only to the extent obtained at operation; rehabilitation gave no further improvement.
6. In the opinion of the author, these cases occur more often than is generally believed, but they are recorded under a different diagnosis and consequently are not submitted to rational treatment.

REFERENCES


