THE ETIOLOGY AND TREATMENT OF CONGENITAL DISLOCATION OF THE KNEE*

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Congenital dislocation of the knee was described by Chatelaine in 1822 (quoted by Shattock 1891) and then by Bord in 1834 (quoted by Forgon and Szentpétery 1961). It was at first thought to be more common in girls, but in a review of a larger series of cases the sex distribution was found to be equal (Provenzano 1947). Kopits (1925) reviewed 2,393 patients with congenital anomalies and found eleven cases of congenital dislocation of the knee as compared with 923 cases of congenital dislocation of the hip, or one dislocated knee for every eighty-four dislocated hips. Sixty per cent of the patients with congenital dislocation of the knee have other congenital anomalies, congenital dislocation of the hip and foot deformities being the most common. Cleft palate, harelip, chest cage deformities, hypoplasia of the fibula and dislocation of the elbow are some of the other deformities seen with congenital dislocation of the knee (Forgon and Szentpétery 1961).

In some of the families studied genetic transference of the condition has been evident. In Provenzano's (1947) review of 200 cases, seven patients had a family history of congenital dislocation of the knee. McFarlane (1947) described a family in which a mother and her three children by three different fathers all had congenital dislocation of the knee.

**Etiology**—Both intrinsic and extrinsic causes have been suggested. The intrinsic causes are genetic abnormalities; the extrinsic causes are mechanical factors. Duraiswami (1955) pointed out that it is often difficult to distinguish between genetically inherited anomalies and those produced by adverse factors occurring during the development of the embryo. Some of the extrinsic causes suggested have been abnormal foetal position, primary contracture of the quadriceps muscles, and traumatic dislocation during parturition.

Shattock (1891) and Drehmann (1900) postulated an abnormal foetal position during gestation as the cause of congenital dislocation of the knee. In cases that they reported the patients had breech deliveries with the knees in a hyperextended position and the feet locked under the chin. This position produced a hyperextension force at the knee, causing it to dislocate. This theory failed to explain the etiology in those cases—the majority—in which there is not a breech delivery, although the percentage of breech deliveries with congenital dislocation of the knee is higher than the expected 3 to 4 per cent of breech deliveries in the general population.

Middleton (1935) believed that the deformity was the result of a primary contracture of the quadriceps muscle. He came to this conclusion after finding these muscles to be partly replaced by fibro-fatty tissue. This has not been accepted because the author failed to prove that the muscle changes were really primary and not a secondary effect of the dislocation.

Trauma to the knee during birth was implicated as a cause of the dislocated knee by Mauclaire (quoted by Forgon and Szentpétery 1961). This is no longer an accepted theory because it has been shown in dead infants that when an attempt is made to produce anterior dislocation of the knee, the femoral epiphysis will displace before the knee will dislocate.

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Most reported patients treated early have been managed successfully by non-operative means. Mayer (1913) reviewed sixty-eight patients and found that a cure had been achieved in 81 per cent of those who were treated before three months of age; a cure rate of only 33 per cent was achieved in those treated after three months of age. The general plan of non-operative treatment was gradually to flex the knee to a reduced position using serial plasters, traction or strapping. Most patients were kept in some form of external fixation to maintain the reduced position until six months of age; follow-up examinations three to four years later revealed knees that were completely normal. In some of the patients treated after six months of age it is often necessary to lengthen the anterior structures of the knee to achieve a reduced position. Niebauer and King (1960) found it necessary in the older infants to lengthen the quadriceps and patellar tendons, and to release the contracted anterior capsule in order to reduce the knee in a flexed position.

Mayer (1913) also reviewed thirty necropsies and surgical reports of patients with congenital dislocation of the knee; these reports revealed similar findings. There was often anterior dislocation of the tibia on the femur and the quadriceps was contracted. The anterior cruciate ligament was found to be attenuated and elongated. The posterior margin of the tibial plateau was rounded, and cartilage was found covering the upper end of the posterior surface of the tibia. McFarland (1929) and O'Dell and Holt (1954) also found the anterior cruciate ligament to be elongated and poorly developed. Less frequently the anterior cruciate has been noted to be hypertrophied; in one patient the posterior cruciate ligament was completely absent (Niebauer and King 1960).

**Classification**—Many terms have been used to describe various hyperextension deformities of the knee. Finder (1964) classified congenital hyperextension deformities of the knee into five distinct clinical entities. The category to be considered here is congenital dislocation of the knee. In this form of hyperextension of the knee the tibia dislocates in front of the femoral condyles. The posterior surfaces of the tibial condyles lie adjacent to the anterior surfaces of the femoral condyles (Fig. 1).

**Prenatal development of the knee joint**—Henke and Regher (1874) believed that the bony configuration of the knee joint and the intra-articular structures were the result of an adaptation by the foetus to movements (Magnus 1905). Fell and Canti (1934) disproved this when they demonstrated experimentally that a "normal knee joint" continued to develop in the avian foetus after the mesenchymal knee joint had been transplanted to another site. It has been shown that the cruciate ligaments and menisci develop within the knee and are not the result of an ingrowth of tissue. The sequence of differentiation is first the menisci, then the capsule and cruciate ligaments (Bardeen 1910). Chondrification of the femoral blastema is complete in the embryo by six weeks; at seven weeks the cartilaginous form of the tibia is complete. During the eighth week the anterior and posterior cruciate ligaments are completely differentiated, and the foetal knee joint is a miniature replica of the adult knee joint (Gray and Gardner 1950).
CLINICAL MATERIAL

There were five cases of congenital anterior dislocation of the knee in three children—two boys and one girl. Their ages at the time of operation were respectively four, four and nine years.

There was no history of breech delivery in any case. The only instance of maternal illness during pregnancy was that of recurrent pyelonephritis in one mother. This was treated with antibiotics. In no case was there a family history of dislocating hips or knees. All three patients had normal mentality.

Physical examination—All the patients walked with their knees dislocated and flexed 15 to 20 degrees. The femoral condyles were easily palpable in the popliteal fossa. The tibia dislocated anteriorly on the femur when brought from 90 to 20 degrees of flexion. Flexion of the knee was limited to 90 degrees and all the knees could be hyperextended by at least 20 degrees. The range of movement was completely painless.

All the children had a definite anterior “drawer” sign and all demonstrated lateral instability in the dislocated position. Two of the knees had a positive posterior drawer sign.

Two patients had signs of dislocated or subluxated hips. All three patients had signs of hypoplasia of the fibula. There was lateral subluxation of the ankle joint in three of the extremities that had an absent or hypoplastic fibula. All of the patients had subluxation of the subtalar joint (Fig. 4).

There were no signs of generalised ligamentous laxity in any of the patients. One patient had a very short neck, thoracic scoliosis and pectus carinatum.

Radiographic findings—At zero degrees of flexion the five knees showed an anterior dislocation of the tibia on the femur (Fig. 1). In the lateral view the tibial plateau was severely inclined posteriorly.

The intercondylar view of the dislocating femur showed the height of the intercondylar notch to be markedly reduced as compared with that in a child of a similar age (Figs. 2 and 3). In all the patients subluxation of the subtalar joint was demonstrated by cineradiography (Fig. 4). One of the patients with bilateral dislocation of the knees had also bilateral congenital dislocation of the hips (Fig. 5); another patient had a unilateral subluxation of the hip. The third patient had normal hips. Hypoplasia of the fibula was present in three of the five knees. The patient with scoliosis had multiple hemivertebrae in the lower cervical and upper thoracic region.

Findings at operation—Four of the knees showed complete absence of the anterior cruciate ligament; in the fifth knee this ligament was thin and elongated. Two of the knees showed complete absence of the posterior cruciate ligament; in one knee this ligament was attenuated.

The tibial plateau sloped posteriorly 30 to 45 degrees in all five knees. All the knees showed a shallow intercondylar notch and poorly developed tibial spine.

One knee had a bucket-handle tear of the medial meniscus; in three knees the menisci were unattached to the tibia in the anterior half of the joint, so that the menisci were abnormally mobile.

All the knees could be reduced without lengthening the anterior structures, although flexion was limited to 90 degrees.

Treatment and results—Unfortunately, no patient was brought for treatment before the age of three years. The knees could not therefore be held reduced and functional in a brace or plaster. The aim of our treatment was to reduce the dislocation and to allow weight bearing. After reduction we allowed function; in doing this we thought that the joint musculature, the collateral ligaments and bony configuration would develop normally to give the patient a stable knee.

In all cases the cruciate ligaments were found to be either absent or hypoplastic. To provide the knee with extra stability so that it would remain reduced in a brace an anterior cruciate ligament was reconstructed. For this purpose we used with slight modification the
technique described by Jones (1963) for repair of the ruptured anterior cruciate ligament (Fig. 6). The anterior cruciate ligament was fashioned from the retinaculum and tendon medial to the patella. A hole was drilled through the lateral femoral condyle so directed as not to pass across the epiphysial plate. The reconstructed ligament was pulled through this hole so as to be taut enough to prevent anterior dislocation at zero degrees of flexion. After

Fig. 2
Figure 2—Dislocating knee: intercondylar view shows the reduced height of the intercondylar notch, and the poor development of the intercondylar eminence. Figure 3—Normal knee: intercondylar view of the knee joint of a child of 9.

Fig. 4
Subluxation of the subtalar joint, shown in all five limbs affected by congenital dislocation of the knee.

operation the limb was put into a long plaster with 10 to 15 degrees of flexion of the knee. The plaster was retained for six weeks, after which a long leg brace allowing 20 to 90 degrees of flexion was fitted. The patients were allowed up walking in their braces and were given isometric exercises to strengthen the quadriceps. The braces were worn day and night.
Results—Four of the five knees treated have remained reduced. One patient was able to walk without his brace ten weeks after operation without any signs of subluxation or dislocation; the other knees are still being protected in the braces. The knee that redislocated had a very small area of horizontal tibial plateau for articulation with the femur. An osteotomy of the tibia with angulation was done to bring the tibial condyles parallel to the femoral condyles.

![Image of a knee with bilateral congenital dislocation of the hip](Fig. 5)

**Fig. 5**
Bilateral congenital dislocation of the hip was present in one patient.

![Diagram of a knee with anterior cruciate ligament](Fig. 6)

**Fig. 6**
Technique for reconstructing an anterior cruciate ligament (Jones 1963). Reproduced by kind permission of the Editor and Dr K. G. Jones.

THEORY OF ETIOLOGY

In our opinion the primary cause of congenital dislocation of the knee is absence or hypoplasia of the cruciate ligaments. The dysplasia may be genetically inherited or may be induced in the developing embryo before nine weeks of age.

In the adult knee the femoral condyles are elliptically shaped; their horizontal diameter is greater than their vertical diameter (Figs. 7 and 8). In the young foetus the femoral condyles

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Figure 7—Foetal femur (twenty-two weeks): the femoral condyles are relatively round compared with those of the adult. Figure 8—Adult femur: the femoral condyles are elliptically shaped.

Figure 9—Foetal tibia (twenty-two weeks): the tibial plateau slopes posteriorly 35 degrees in the young foetus. Figure 10—Adult tibia: the posterior slope of the tibial plateau is approximately 10 to 15 degrees.
are not fully developed; they are round as compared to the elliptically shaped adult condyles (Figs. 7 and 8). The foetal tibial plateau slopes posteriorly 35 degrees whereas in the adult it slopes 10 degrees (Figs. 9 and 10).

As the adult knee extends the tibial condyles follow an elliptical path around the femoral condyles. As the distal attachments of the collateral ligaments move in this elliptical path they are stretched to their limit at zero degrees of flexion and further forward gliding of the tibia on femur is prevented. When the foetal knee is extended the collateral ligaments move along an arc of a circle. This allows the collateral ligaments to remain relatively lax in extension.

As the adult knee extends the posterior half of the tibial plateau abuts against the femoral condyles, blocking further extension. As the foetal knee extends the posterior slope of the tibial plateau allows the tibia to glide around the femoral condyle (Figs. 11 and 12). The structure shown spanning the posterior aspect of the knee joint in Figures 11 and 12 represents the posterior joint capsule, the hamstrings and the gastrocnemius. As the adult knee extends these posterior structures abut against the projecting femoral condyles at zero degrees of flexion and in this way assist in preventing hyperextension of the knee. In the foetus the posterior condylar projections are not developed and a greater range of hyperextension can occur before the posterior structures abut against the femoral condyle.

If the cruciate ligaments are absent or hypoplastic in a foetus a dislocating force is necessary to produce the clinical deformity. In a random radiological study of 3,875 pregnancies it was found that 25 per cent of the foetuses were in a breech position at some time before the thirty-fourth week of gestation. After the thirty-fourth week most underwent spontaneous version. Of those foetuses that were in a breech position 75 per cent were noted to have their knees in an extended or hyperextended position (Vartan 1945). There is therefore a force tending to produce an anterior dislocation of the knee in approximately 18 per cent of all pregnancies. If there is an absence or hypoplasia of the cruciate ligaments the end result will be a congenital dislocation of the knee.

Giorgi (1956) described a patient in whom routine radiographs of the knee showed that the intercondylar eminence was absent. Giorgi further noted that this patient had a positive anterior "drawer" sign, but no signs of dislocation or subluxation of the knee. This may be an example of knee with congenital absence of the anterior cruciate ligament that did not have a hyperextension force applied to it during intra-uterine life. In this event the para-articular structures and the bony configuration of the knee developed normally so as to give the knee sufficient stability to allow normal activities.

**Experimental dissections**—To test our hypothesis further the knees of thirty aborted foetuses of between fourteen and thirty-two weeks of development were dissected. The cruciate ligaments, the collateral ligaments and the posterior joint structures (hamstring tendons, gastrocnemius origins and posterior capsule) were observed while an anterior dislocating force was applied to the knee. It was found that the cruciate ligaments were the only structures that in themselves could prevent anterior dislocation; when only the cruciate ligaments were
cut an anterior dislocation could easily be produced. These findings applied only to foetuses that were less than twenty-eight weeks old. By the twenty-eighth week the femoral and tibial condyles had approached the configuration of the adult knee joint.

In dissections of foetuses older than twenty-eight weeks and also in adult knees we found structures other than the cruciate ligaments, that prevented anterior dislocation. The posterior joint capsule, the collateral ligaments the hamstring tendons and the gastrocnemius were all found to be deterrents to anterior dislocation. These findings in the older foetuses and in the adult seem to be corroborated by Kennedy’s (1963) study of traumatic anterior dislocation of the knee. In most of his cases the dislocating force ruptured the cruciate ligaments, the posterior joint capsule, and the tibial collateral ligament; in one case the biceps femoris was ruptured and in another the origin of the gastrocnemius was avulsed from the femur.

ASSOCIATED ANOMALIES

In the last thirty-eight years at the seventeen Shriners Hospitals, 155 children with congenital dislocation of the knee have been treated. There were ninety-nine girls and fifty-six boys. Eighty-two per cent of these patients had associated anomalies of the musculoskeletal system. The most commonly occurring anomaly was congenital dislocation of the hip: this was present in 45 per cent of the patients. Thirty-one per cent had congenital deformities of the feet, the most common deformity being talipes equinovarus. Ten per cent of the children had congenital dislocation of the elbow. The dislocations of the hip, the elbow and subtalar joint that occur in association with congenital dislocation of the knee may all be due to a common defect. In the dislocating knees we found the intra-articular ligaments to be absent or hypoplastic. It is possible that this condition exists in the other dislocating joints, the defective intra-articular ligaments being respectively the ligamentum teres of the femoral head, the annular ligament and the interosseous talo-calcaneal ligament.

SUMMARY

Five knees with congenital dislocation were explored. The cruciate ligaments were found to be absent or hypoplastic. The results obtained by reconstructing an anterior cruciate ligament appear to be good. On the basis of dissections of foetal knees we believe that the cruciate ligaments are the main structures preventing an anterior dislocation of the knee in early foetal life; this deviation from the adult pattern is due to the bony configuration of the foetal knee. We therefore postulate that the basic defect in congenital dislocation of the knee is an absence or hypoplasia of the cruciate ligaments.

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REFERENCES

HENKE and REGHER, (1874): Quoted by Magnus (1905).