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Congenital dislocation of the knee

Congenital dislocation of the knee is a very rare condition, the incidence in Scandinavia hitherto not being known with certainty. Thirteen patients treated during the years 1960 to 1983 with 19 affected knees were reviewed and followed up. Findings of muscular imbalances in nine of these cases and of spinal abnormalities in four cases strongly indicate neuromuscular imbalance as an aetiological factor.
The minimum incidence was estimated to be 0.017 per mille or approximately 1 per cent of the incidence of congenital dislocation of the hip.
Recommendations for treatment are that manipulation should be carried out gently, and if not successful within 2–3 months and in all cases of Grade III, an operation is indicated. Too many newborn children in this series had manipulation fractures or slipped epiphyses. However, signs of fracture had disappeared at follow-up in most cases.

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Congenital dislocation of the knee (CDK), although very rare, has drawn the attention of orthopaedic surgeons during the last century and a half. A great number of cases has been reported and the contribution of a few more will be of interest therefore only if they can throw some light on the aetiology of this condition.

We have focused our interest on the association of CDK with certain syndromes and with neuromuscular imbalance, and on the relative merits of closed versus open treatment.

Definition of CDK

In agreement with Laurence (1967) and Curtis & Fisher (1969), all kinds of hyperextended knees present at birth were included in the term CDK, classified in three grades (Figure 1). *Grade I* represents hyperextension of the knee joint at birth without displacement of the joint surfaces of the femur in relation to the tibia (the axes of the long bones pointing at each other at the joint line). *Grade II* represents a subluxation, the tibial epiphysis sliding onto the anterior part of the femoral condylar articular surface. *Grade III* represents total dislocation of the tibial epiphysis in front of the femoral condyles.

Knees not spontaneously extended beyond the neutral position at birth but only presenting resistance to flexion at some point were not included.

Patients and methods

Thirteen patients with 19 congenitally dislocated knees were treated at the Orthopaedic Hospital of Copenhagen and the orthopaedic department for children, Rigshospitalet, in the period 1960 to 1983; two patients were primarily seen and treated in other hospitals. All charts and radiographs were reviewed, and all patients except one (dead) were followed up clinically and radiographically.

All the patients had been followed closely since the day of delivery; the diagnosis had been made immediately and an orthopaedic surgeon had seen them within a few days. The possibility of the condition having been caused by quadriceps fibrosis associated with injections could thus be ruled out.

All cases were initially treated by manipulations and plaster casts; resistant cases were later operated, the principle being elongation of the quadriceps tendon by a reverse V-Y-plasty in the tendon and by necessary additional transections or plastic operations on the extensor apparatus.

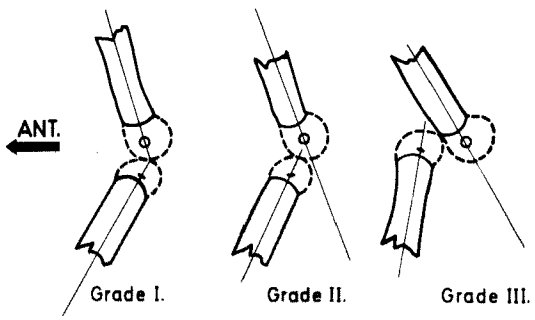


Figure 1. Grading of congenital dislocation of the knee using lateral radiographs.

Evaluation of muscle power in early postnatal life is difficult beyond observations such as "good power of quadriceps, 0 power of hamstrings". Later in life, muscle power was routinely graded 0–5.

At follow-up a semiquantitative scoring scale was used to characterize the overall physical function of the patient in daily life. The Kettelkamp knee scoring scale II (Kettelkamp & Thompson 1975) was used with modification: the section on flexion contracture in the Kettelkamp scale was substituted by a section on flexion deficiency with the same range of scores (0 to 8 points). The best possible result in both scoring scales is a total of 97 points.

The clinical signs as well as the activities of daily life were reflected in the knee scores. A high score meant a high activity level, no pain in the knee joint, ability to walk up and down stairs and on rough ground, no synovial thickening, little or no flexion defect, and no instability.

Patients with CDK not part of a syndrome generally had high knee scores, normal radiographs and normal range of motion at follow-up.

Attention was paid to certain syndromes: arthrogryposis, Down's syndrome, Larsen's syndrome, myelomeningocele (MMC), and cerebral palsy.

Table 1. Congenital dislocation of the knee; coincident syndromes and musculoskeletal disorders

Case	Sex	Side	Grade	Heredity	Breech presentation	Syndrome
1	M	R L	II I	Hip dislocation 3 generations on maternal side		Down's
2	F	R L	III III	Hip dislocation 3 generations on maternal side		Down's
3	F	L	I	None		
4	F	R L	III III	Mother hypertelorism + slightly hyperextended knee	+	Larsen's
5	F	R L	I I	None	+	
6	F	R	I	None		
7	F	R L	I I	None		MMC (big. lumb.)
8	M	R	II	None		
9	F	L	I	Grandmother on paternal side hyperextended knees		
10	M	R	III	None	+	
11	F	R L	II II	None	+	
12	F	L	III	None		
13	F	L	III	None, twin-sister is normal		Arthrogryposis multiplex

Observations

The ratio females/males was 10/3 and the condition was found equally often at the right and the left knee. Six of the patients had bilateral CDK, generally those with the highest degree of CDK at birth, totalling 19 knees in 13 patients. Breech presentation had occurred in four cases (Table 1).

In the strict sense of the word, only Case 13 could be classified as *arthrogryposis;* she had at the same time rigid, prone superior extremities, club feet, dislocated hip and knee on the left side, degeneration of muscles and subcutaneous fibrotic tissue to a great extent. Two siblings, Cases 1 and 2 had *Down's syndrome;* one had MMC (Case 7); none had cerebral palsy. Our case of *Larsen's syndrome* (Case 4, Figure 2) was classified in agreement with Silvermann's criteria (1972): CDK plus dislocation of the hips, positional deformities of the feet, coupled with a distinctive facies (hypertelorism, prominent frontal bone).

In all 11 operated knees, the cruciate liga-

Coincident disloc. hip	Coincident club foot	Spinal deformity	Muscle imbalance	
			At birth	At follow-up (0–5)
None	None	Coalescent vertebrae scoliosis	Totally dominant quadriceps – no spontaneous flexor activity	Quadriceps (5) still stronger than hamstrings (4)
R L	None	Butterfly vertebrae and coalescent vertebrae	Not recorded	Dead, 13 years
None	None		Not recorded	Quadriceps (5), hamstrings (5)
R L	R L	Bifid spine L_V/S_I broadened lumbar vertebrae	Stronger quadriceps than hamstrings	Atrophy of m. rectus femoris, mm vasti (5), hamstr. (5)
L	R L		Stronger quadriceps than hamstrings	Quadriceps (5), hamstrings (5)
None	None		Stronger quadriceps than hamstrings	Quadriceps (5), hamstrings (5)
R L	R L	Bifid spine L_{III-IV}	Quadriceps with good force bilat.; hamstrings & lower leg 0	Quadriceps (5), hamstrings and crural mm. (0)
R	None		Slight hypotonia of right hamstrings	Quadriceps (5), hamstrings (5)
None	None		Not recorded	Quadriceps (5), hamstrings (5)
R	None		Isolated diminished power of right hamstrings (2)	Quadriceps (5), hamstrings (4)
R L	R L		Diminished hamstring power bilat.	Still in treatment
None	None		Isolated diminished power (1) of left hamstrings	Still in treatment
L	R L		Marked, widely scattered muscle inactivity	Still in treatment

ments (Table 2) were of normal thickness, but stretched to some degree. At follow-up, slight to marked anterior drawer sign was found in seven knees, being absent in the very slight cases and in the patient with Larsen's syndrome, who had a very limited range of motion because of the deformity of the femoral condyles (squared off).

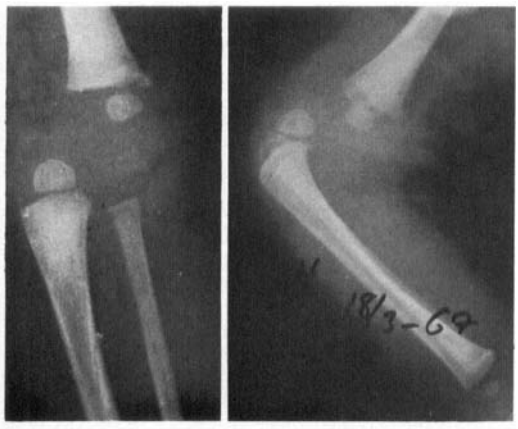
We noted a high (6/19) incidence of real fractures through the metaphyseal bone or even the femoral or tibial shaft recognized during manipulation therapy and radiographically verified (Figure 3); in addition, four knees had small infractions close to the epiphyseal line which were considered secondary to the manipulative treatment. The tibial head in some cases had an extremely retroposed position.

A fibrosis, as described in cases with a history of intramuscular injections was not found in any of our cases.

Table 2. Congenital dislocation of the knee; treatment and results

Case	Manipulation fractures	Findings in operated cases[a]	Age at follow-up	Findings at follow-up	
				Score[b]	Radiography
1	Small infraction of proximal tibial shaft; only R	–	22	88	Slightly squared lat. fem. cond.; R + L
2	Fracture of distal part of femoral diaphysis; both R & L (Fig. 3)	–	(+13)	–[c]	–
3	Small infraction of epiphyseal line of L tibia	–	16	97	Normal
4	Fracture and major epiphysiolysis of tibial head; R + L (Fig. 2)	Quadriceps very tense and hamstrings dislocated ant.; R + L	15	71	All fem. and tib. cond. squared off; R + L
5	None	Quadriceps, especially rectus femoris, very tense and hamstrings dislocated ant.; R + L	14	72	Normal
6	None (Fig. 4)	–	12	95	Normal
7	None	Strong quadriceps not fibrotic; R + L	10	61	Slightly squared off fem. condyles; R + L
8	Early fracture of R tibial shaft. Later small infractions at both tibial and femoral epiphysis; R	–	6	84	Normal
9	None	–	4	95	Normal
10	None	Distal part of vastus lat. m. fibrotic. Hamstrings dislocated ant.; R	2	84	Normal
11	Small infractions of tibial metaphysis; R + L	Only suprapatellar pouch fibrozed	1	–[d]	–
12	Tibial mid-shaft fracture L, small lysis of tibial growth plate L	Rectus muscle contracture	1	–[d]	–
13	None	Quadriceps normal, hamstrings 0	1/2	–[d]	–

[a] In all operated knees the cruciate ligaments were of normal thickness, but stretched to some degree.
[b] Modified Kettelkamp knee score.
[c] Dead before follow-up.
[d] Still in treatment.



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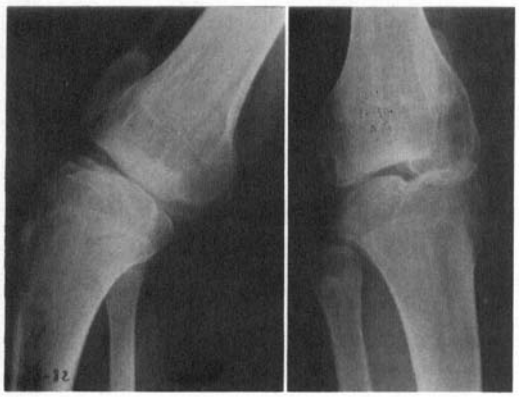

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Figure 2. Epiphysiolysis following manipulation of the right knee in Case 4 who had CDK of both knees and Larsen's syndrome. Same events and nearly identical radiographs of the left knee.

A. Patient 15 days old. Grade III CDK, untreated. The ossification centres at their normal positions in the tibial head and distal femoral epiphysis – side by side.
B. Same knee after forced manipulation. Knee flexed, tibial epiphysis slipped. Age 22 days.
C. Same knee at age 15 years. Squared off condyles.

At follow-up most of the patients had radiographically normal condyles (Figure 4), the retroposition of the tibial head had disappeared, and only two knees had slightly squared off femoral condyles (one with MMC and one with Down's syndrome), while one had markedly squared condyles (Case 4, with Larsen's syndrome). This patient had had severe manipulation fractures in early life but angulations disappeared during the observation period and squared off condyles developed after a V-Y-plasty operation with sectioning of the collateral ligaments (the only case in which this was considered necessary). At follow-up she could

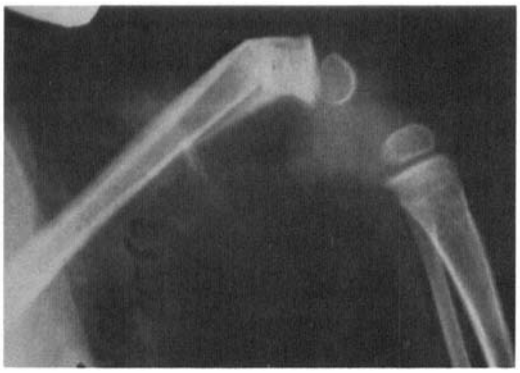


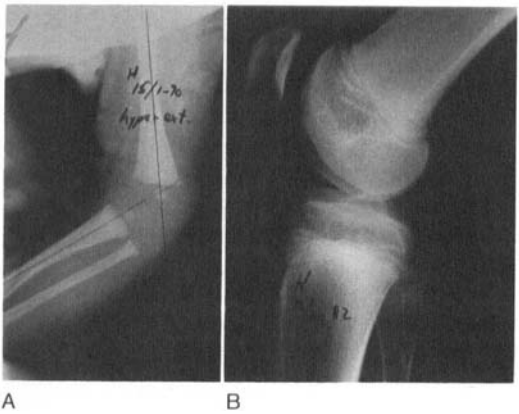
Figure 3. Manipulation fracture of distal part of femur. Case 2, 5 months old.

only bend her knees from 0 to 20 degrees. It was impossible to determine whether her deformity was due to damage to the young epiphysis, later postoperative looseness, or the syndrome itself.

Based on 11 children with CDK born in the islands east of Store Bælt from 1963 to 1982 and the 653 521 live-born children in the same area, the minimum incidence was 0.017 per mille.

Discussion

The first reports on CDK date back to Chaussier in 1812 (Rechmann 1914) and Chatelain in



A B

Figure 4.

A. Case 6. Hyperextended right knee at the age of 7 days. CDK Grade I, treatment only by manipulation and casts. No fracture or epiphysiolysis.
B. Follow-up at the age of 12½ years. Normal condyles.

1822 (Drehmann 1900), and by 1947, 213 cases had been reported (Provenzano 1947). Since then we have found 134 *new* cases described in the literature (Larsen et al. 1950, Niebauer & King 1960, Forgon & Szentpetery 1961, Karlén 1964, Charif & Reichelderfer 1965, Katz et al. 1967, Laurence 1967, Curtis & Fisher 1969 and 1970, Silvermann 1972, Bose & Chong 1976, Ahmadi et al. 1979, Austwick & Dandy 1983, and Nogi & McEwen 1983).

Larsen et al. (1950) credited the first exhaustive description of a case of his syndrome to Kaijser (1935) in Stockholm. This type of CDK is extremely rare as we had only one in our series.

The incidence of CDK is not known, but was estimated by Steindler (1924) and Kopits (1925) to be about one for every 90 cases of congenital dislocation of the hip (CDH), which in Scandinavian populations is about 1.7 per mille (von Rosen 1962). Our minimum incidence of CDK of 0.017 per mille would thus correspond to a CDK/CDH ratio in Scandinavia of 1/100, i.e. in good agreement with the previous estimates.

Charif & Reichelderfer (1965) found an incidence of CDK of 0.7 per mille in a black community in Washington D.C., most of the cases being slight.

As to the aetiology, breech presentation may play a role (Niebauer & King 1969), but as we have only four cases in our material, there must be other aetiological factors. Also, it must be borne in mind that the condition itself may predispose to breech presentation (Vartan 1940).

We could not find any support in our material for the suggestion of Katz et al. (1967) that lack of the anterior cruciate ligament would be of importance for developing CDK. We rather feel that increasing CDK causes the cruciates to lengthen gradually.

Neither do we have any certain evidence of fibrosis in the quadriceps muscle as an aetiological factor in our cases of CDK as suggested by Karlén (1964) and Hněvkovsky (1961); quadriceps myofibrosis caused by intramuscular injections (Hagen 1968, Alvarez et al. 1980) was ruled out in our material. In our operated cases we always found fibrosis of the suprapatellar pouch and often a thick and tense lateral knee capsule and iliotibial tract, with the vastus lateralis inserting more proximally than normal. This may be an aetiological factor or a secondary phenomenon. What we consider of greater importance were the findings of muscular imbalance in nine cases as well as findings of spinal abnormalities in four of these cases, which strongly indicates neuromuscular disturbance as an aetiological factor.

Our material had a remarkable number of fractures following manipulation of the newborn children; we do not believe this to be unique for our hospitals but rather that it was not noted earlier; in the literature we have found only five single cases. We found that the bones remodelled totally during growth so that no radiographic signs of earlier fracture were found at follow-up from about 6 years of age. Kaijser (1935) found some changes in the epiphyses at the knee in some young children, but he did not ascribe them to manipulation; he thought they were probably hereditary deformities.

Two of our patients had slightly squared off condyles at follow-up and had marked anteroposterior looseness (the patients with MMC and Down's syndrome). One had marked squared condyles (the patient with Larsen's syndrome).

To conclude, we recommend careful, non-violent manipulation and early operation, i.e. at about 3 months of age, if manipulation is without effect. Especially CDK Grades II and III should be operated early; until 1981 all our cases fractured at manipulation. Because of the small number of patients, treatment should be centralized in very few hands.

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