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Early operation of the dislocated knee in Larsen´s syndrome

A report of 2 cases

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Two brothers with Larsen´s syndrome were operated on for bilateral anterior dislocations of the tibia at the knee already in their infancy. The children were followed for 12 years and had better results than is commonly seen in Larsen´s syndrome knee dislocations operated on at a later stage.

Larsen´s syndrome was first described by Larsen et al. (1950). The condition is congenital and characterized by dislocation of the knees and by facial deformities with prominent forehead, wide-spaced eyes, and a depressed nasal bridge. Dislocation of other joints and deformities of the feet and the spine are also seen. Operative treatment of the knee dislocations has often been delayed until late infancy. In 2 cases presented here, open knee reduction was performed early in life, and the patients were examined regularly until the age of 12.

Case 1

A boy was born after an uncomplicated pregnancy . There was no family history of congenital deformity. The child had facial abnormalities, bilateral hip dislocations (Figure 1), and bilateral anterior dislocations of the proximal tibiae. Immediate closed reduction of the hips and knees was possible, but could not be retained. At 3 months of age, a bilateral open knee reduction was performed. The distal part of the quadriceps muscle was short and was replaced by fibrous tissue. Bilateral V-formed quadriceps lengthening was performed according to the technique of Curtis and Fisher (1969). Closed reduction of the hips was done. The position of the hips and knees was maintained in a Lorenz frog-leg plaster for 3 months with the knees in 70° of flexion.

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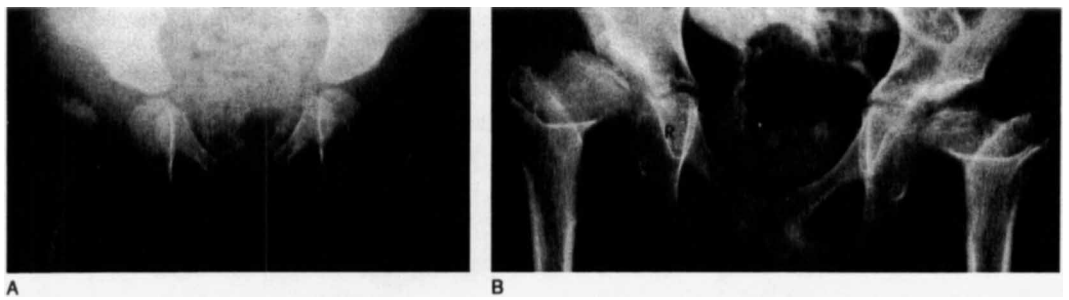
After plaster removal the hips and knees remained reduced.

At the age of 3 years, radiographic examination of the hips showed widening of the femoral metaphysis and absence of the ossification center in the femoral head. At aged 5 years, both capital epiphyses were flattened and irregular. At aged 12 years, both femoral heads were deformed. In the knees, a patella baja was observed (Figure 1). At aged 12 years, the child was walking with pelvic rotation, but without support and without a limp. Hip flexion was 15°–120° and 20°–90°, respectively. The knees were stable, with a range of flexion of 15°–130° and 25°–90°. respectively. The child was mentally retarded.

Case 2

A brother to Case 1 was also born after an uncomplicated pregnancy. He had facial abnormalities and dislocation of the knees like his brother (Figure 2), but no hip dislocation. Closed reduction of the knee dislocations failed. One week after birth the child had open reduction of the knees, which were held in plaster in 30° of flexion for 3 months.

The knees remained reduced, and the child could walk withut support at 3 years of age. Hip mobility was normal. Appearances of ossification centers in the femoral heads were delayed, and after 3.5 years, both capital epiphyses were irregular. At aged 5 years, both femoral capital epiphyses were normal. At aged 11 years, limited motion of the right hip was observed. Radiographic examination showed flattening of the right capital epiphysis with a lateral defect. The boy was limping and the mobility of the right hip was limited. The knees were stable, with a range of flexion of



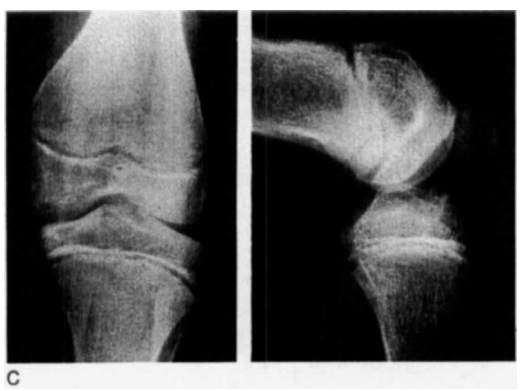


Figure 1. Case 1. A boy with Larsen's syndrome.
A. At aged 3 months. Bilateral hip dislocation. The metaphyses are broad.
B. At aged 12 years. Both femoral heads are severely deformed. The acetabuli are shallow and steep.
C. At aged 12 years. Left knee. Reduction has been maintained. Both the femoral and the tibial epiphyses are slightly dysplastic. The patella is low-riding.

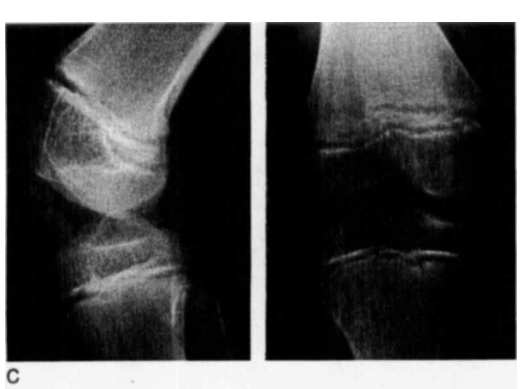


Figure 2. Case 2. Larsen's syndrome in a boy, brother to Case 1.
A. Left leg at birth. Anterior dislocation of the tibia on the femur. Arrow is pointing forward.
B. At aged 11 years. The right epiphyseal head is collapsed, sclerosed, and fragmented. The hips were not dislocated at birth.
C. At aged 11 years. Right knee. The reduction has been maintained. Hypoplastic patella baja.

0°–120° and 10°–120°, respectively. Radiographic examination showed a bilateral patella baja (Figure 2). The child was mentally retarded like his brother.

Discussion

According to Silverman (1972) the combination of anterior knee dislocation and facial deformities are diagnostic for Larsen's syndrome. The typical finding in

congenital knee dislocation is quadriceps fibrosis (Curtis and Fisher 1969, Johnson et al. 1987). Most children with congenital knee dislocation not associated with Larsen´s syndrome may be successfully treated without operation (Nogi and MacEvan 1982). However, in Larsen´s syndrome, conservative treatment often fails (Table 1), and better results are obtained after open reduction. Larsen et al. (1950) found in one case of open knee reduction in a 19-month-old girl marked elongation of the anterior cruciate ligament and secondary changes in the bony architecture. The patellar tendon was short and therefore lengthened before reduction. In the reports of Latta et al. (1971), Triqueros et al. (1978) and Samuel and Davies (1981) a quadriceps lengthening procedure was performed before reduction. In our 2 patients reported here, quadriceps lengthening was performed in early infancy. At follow-up, the radiographic examination showed patella baja in both children, but the knees were free of pain and stable with a fair range of motion. Development of secondary ligamentous instability and secondary changes of the bony architecture were probably prevented by the early reduction.

Table 1. Results of conservative and operative treatment for knee dislocations in 9 patients with Larsen's syndrome

Author	Knee	A	B	C
Larsen et al. 1950	R	19	O	R, L
	L	19	O	R, I
	R+L	33	O	R
	R+L	13	C	
	R	16	O	R, I
	L	25	O	R, I
Latta et al. 1971	R+L	0–8	C, T	D
		8	O	R, I
Steel and Kohl 1972	R+L	21	C	D
Oki et al. 1975	R+L	1	C	D
		3–4	T, C	R, I
Triqueros et al. 1978	R+L	12	O	R, L
Samuel and Davies 1981	R	1w	C	R
	L	5	O	D
		12	T	D

A Age of the child when treatment was started (in months), w week.
B Treatment: C Closed reduction, T Traction, O Open reduction.
C Results: D Remaining dislocation, R Maintained reduction, I Instability, L Limitation in mobility.

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