Perthes' disease or late avascular necrosis after developmental dislocation of the hip?

10 children followed for 6–35 years

Pawel Koczewski and Marek Napiontek

Department of Pediatric Orthopaedics, Karol Marcinkowski University of Medical Sciences, Poznan, Poland. Correspondence: Dr. Marek Napiontek, Department of Pediatric Orthopaedics, Samodzielny Publiczny Szpital Kliniczny nr 4 im. W. Degi, Ul. 28 Czerwca 1956 nr 135/147, PL-61-545 Poznan, Poland. E-mail: ortop@webmedia. pl
Submitted 00-07-27. Accepted 01-01-07

ABSTRACT – We studied 10 patients treated because of late avascular necrosis (AVN) mimicking Legg-Calvé-Perthes' disease (LCPD) after developmental dislocation of the hip (DDH). DDH was recognized late at an average age of 5.4 months and in all children it was treated without surgery. In 4 children, the treatment was complicated by mild AVN of the femoral head, which had disappeared before 3 years of age. The first clinical signs of LCPD were noted at a mean age of 5.8 years. They all had Catterall's type III or IV of LCPD. The course was typical of LCPD. 8 children were operated on at mean age of 7.4 (5–12) years. In 7 of them, subtrochanteric derotation-varisation osteotomy of the femur with shortening combined mainly with Dega's pelvic osteotomy was done. The operative treatment resulted in a concentric position of the femoral head and good coverage of the acetabulum. Follow-ups were done at 10 (6–35) years. Shortened femoral neck and trochanteric overgrowth occurred in 4 operated hips. According to the Stulberg classification, 2 hips were classified as type I, 1 as I/II, 5 as II, 1 as II/III and 1 as IV. We conclude that LCPD mimicking late AVN can occur in hips treated because of developmental dislocation.

■

The development of Legg-Calvé-Perthes' disease (LCPD) in the same joint in which developmental dislocation of the hip (DDH) was previously diagnosed and treated is extremely rare. Lindholm et al. (1978) reported 3 cases, and Williams et al. (1982) 1 case. The results in patients with both diseases are not known.

Patients and methods

In about 4,700 patients with DDH and 370 with LCPD treated in our institution since 1966, 15 patients were suspected of having both diseases. We found complete records in 10 (5 girls) of them (Table). They had been followed for 10 (6–35) years. Follow-up examinations were done at a mean age of 18 (11–43) years.

DDH had been diagnosed late at an average age of 5.4 (1–18) months. Bilateral DDH was present in 8 and unilateral in 2 patients. 8 children were treated nonoperatively with a Frejka pillow in the first year of life. 1 child was treated with the Lorenz method (closed reduction and stabilization in a plaster cast in frog position) and 1 had no treatment, but spontaneous reduction occurred.

In 9 children, nonoperative treatment resulted in concentric reduction. In the oldest child, in whom the diagnosis of bilateral DDH was made at age 18 months, nonoperative treatment (closed reduction and casting) did not result in concentric reduction, because of coxa valga antetorta. Residual dysplasia was seen in 8 children. In 4 of them, the nonoperative treatment of DDH was complicated by mild avascular necrosis of the femoral head (AVN) which, however, disappeared before 3 years of age.

Clinical signs of LCPD such as pain and limping were noted at a mean age of 5.8 (4.5–7.5) years. Nonoperative treatment mainly consisted of the non-weight bearing started mean 3 months later, and was applied for 2–3 years.

Patients with both DDH and late AVN/LCPD

Pts	Sex	DDH side	DDH Age at diagnosis (months)	DDH treatment (months)	DDH Results	LCPD side	LCPD Age at diagnosis (years)	LCPD Surgical treatment (years)	LCPD Follow-up (years)	CE angle (°)	LCPD Results (Stulberg classific.)
1	F	L/R	3	Frejka pillow (12)	Reduction, spherical femoral head	R	5.4	1. DVO, Dega's osteotomy (6.0) 2. Transfer of the greater trochanter (12.0)	10.6	27	III/II
2	M	L/R	2	Frejka pillow (12) DVO, Dega's osteotomy (77)	Residual dysplasia	L	5.0	Dega's osteotomy (6.5)	6.0	25	I
3	M	L/R	2	Frejka pillow (?)	L/R: reduction, spherical femoral head	R	5.0	VO, Dega's osteotomy (5.7)	6.0	23	I
4	F	L/R	9	Frejka pillow (15)	L/R: residual dysplasia, AVN	L	5.8	DVO, Dega's osteotomy (6.9)	6.2	23	II
5	F	L/R	2	Frejka pillow (8)	L/R: megacoxa, trochanter altus	R	6.8	Shelf acetabuloplasty (9.5)	11.3	36	II
6	F	R	1	Frejka pillow (12)	L/R: residual dysplasia, AVN	R	4.6	DVO, Salter osteotomy (6.7)	12.4	38	I/II
7	M	R	3	Frejka pillow (12)	L/R: residual dysplasia, megacoxa	R	7.2		11.8	27	II
8	M	L/R	–	Spontaneous reduction DVO, Dega's, osteotomy (77)	L/R: reduction spherical femoral head	L	4.5	DO, Dega's osteotomy (6.3)	7.5	24	II
9	M	L/R	18	Closed reduction (24)	L/R: residual dysplasia	L	7.5	DO, anteflexion, shelf (12.3)	35	17	IV
10	F	L/R	9	Frejka pillow (12), Ortolani splint	R: residual dysplasia	L	6.5		6.0	18	II

DO subtrochanteric derotation osteotomy, DVO subtrochanteric derotation-varization osteotomy

All children had Catterall's type III or IV LCPD. The course of LCPD and its phases were typical, even in 8 patients with a persistent dysplastic hip joint.

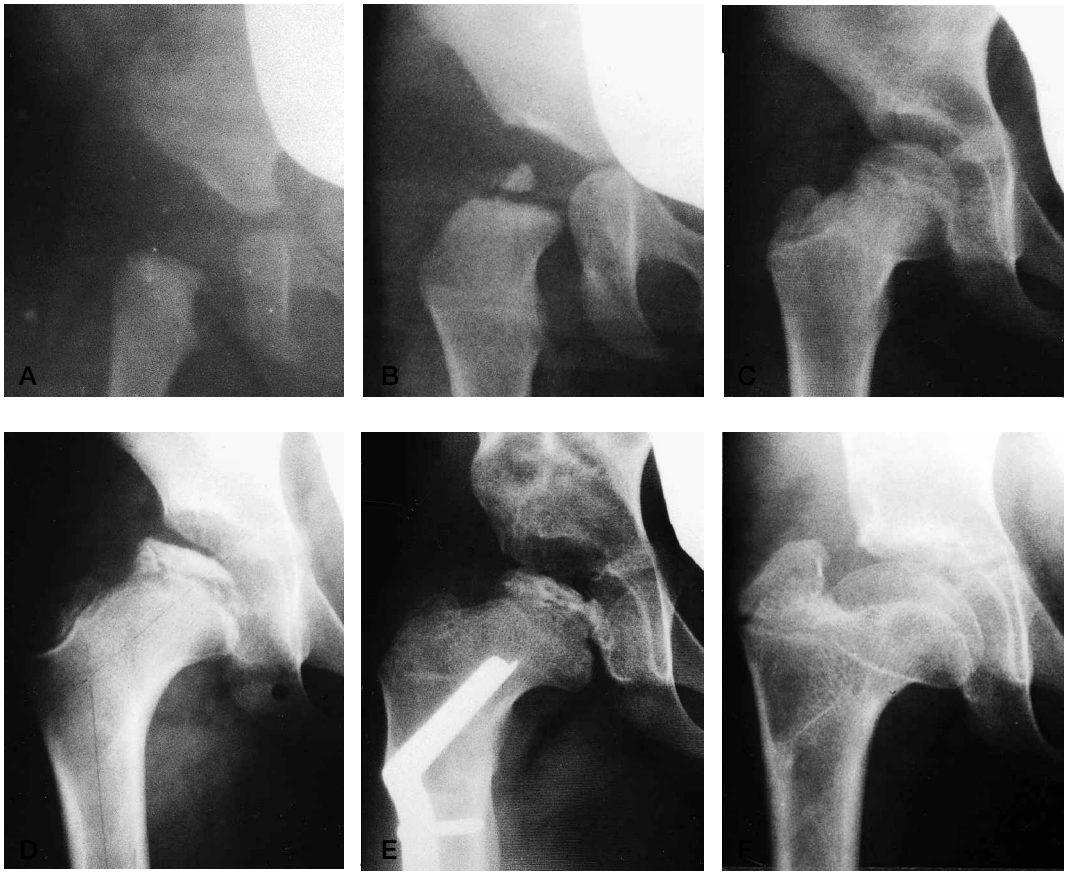
8 children were operated on because of LCPD at the age of 7.4 (5.7–12) years. In 8, subtrochanteric osteotomy, with derotation (from 5° to 35°), varisation (from 10° to 15°) and with anteflexion (15°–20°) was done. In 6 of them, it was combined with Dega's pelvic osteotomy, in 1 with Salter osteotomy. 1 patient had a subtrochanteric osteotomy supplemented with shelf acetabuloplasty, in another, only a shelf plasty was done. During subtrochanteric osteotomy, the distal femur was routinely shortened mean 1.2 cm.

In all of the children, operative treatment resulted in both a concentric position of the femoral head in the hip joint and its good coverage by the acetabulum. Shortened femoral neck and greater trochanteric overgrowth occurred in 4 operated hips (in 1 of them, distal and lateral transfers of the greater trochanter had been done). 1 of our patients is presented in the Figure.

Results

According to McKay's clinical classification (1974) modified by Barrett et al. (1986), 7 hips were classified as excellent, 1 as good, and 2 as fair. The latter 2 cases were classified as fair because of limping caused by trochanteric overgrowth.

According to Severin's classification of radiographs (1941), 5 hips were classified as Ia, 4 as IIa, and 1 as IIb. According to Stulberg et al.'s classification (1981), 2 hips were classified as type I, 1 as I/II, 5 as II, 1 as II/III and 1 as IV. The mean CE angle of Wiberg during follow-up was 26° (17°–38°).



A. DDH on the right side in a 3-month-old girl at the time of diagnosis.

B. At the age of 7 months. Mild AVN after conservative treatment.

C. At the age of 5 years 4 months. Complete remodeling of the femoral head.

D. At the age of 5 years 11 months. Condensation phase of LCPD.

E. At the age of 6 years 4 months after surgical treatment (subtrochanteric derotation-varisation osteotomy of the femur with shortening combined with Dega's pelvic osteotomy).

F. At the age of 11 years 3 months with short femoral neck and greater trochanteric overgrowth after LCPD.

G. Final result at the age of 15 years (3 years after distal transfer of the greater trochanter).

Discussion

The main diagnostic problem in patients with both DDH and LCPD is to distinguish between late AVN after DDH and LCPD. In all patients analyzed, complete remodeling of the femoral head had occurred over a period of 3–6 days before LCPD had been detected. Restoration of the femoral head after AVN was well documented by radiographs, leaving no doubt about destinguishing between AVN after DDH and LCPD. Similar observations were made by Williams et al. (1982) and Lindholm et al. (1978). Kruczynski (1987) indicated that the condensation/fragmentation sequence typical of Perthes' disease is rare in AVN after DDH.

The incidence of LCPD reported by Tachdjian (1972) is 1 in 750 for boys and 1 in 3700 for girls. The incidence of DDH is 1–1.5 in 1,000 (Weinstein 1996), so it is obvious, that both of these conditions can occur at the same time. The question arises: is this only a simple coincidence, or is there any etiopathological relation between them?

Wynne-Davies and Gormley (1978) reported that patients with LCPD had a higher (11%) incidence of abnormal birth presentation than that seen in the normal population (2%–4%). The same risk factor exists regarding DDH, so the incidence of LCPD in patients with DDH can be expected to be higher than in a normal population. Coincidence of two such serious pathological conditions as DDH and LCPD should increase the risk of hip destruction. Such a view was confirmed by Lindholm et al. (1978), who described the course of LCPD after DDH as severe and having a poor outcome. In our study, we noted that the course of LCPD after DDH was typical, no more severe than usual, and the outcome (especially clinical) was mostly excellent. Trying to find the reasons for this controversy, we noted few differences between Lindholm et al.'s (1978) and our patients. In all 3 cases which they reported, DDH was diagnosed very late (18–24 months of life) and treated nonoperatively for a long time with closed reduction, plaster casting, and a Denis-Brown splint. In our patients, DDH was diagnosed earlier and treated more conservatively. The surgical treatment was also different. Lindholm et al. (1978) used only a varus intertrochanteric osteotomy. In 2 cases after 2 and 4 years, he performed a shelf acetabuloplasty. Their results were better with this procedure when it was done earlier, and they concluded that "early Salter operations might have been more effective".

Most of our operated patients had a double-level operation, subtrochanteric derotation-varisation osteotomy with shortening combined with a pelvic osteotomy or shelf acetabuloplasty. Those who underwent the latter procedure had a worse outcome with persistent discongruency of the hip, because they were not suitable candidates for this technique.

No benefits in any form have been received or will be received from a commercial party related directly or indirectly to the subject of this article.

Barrett W P, Staheli L T, Chew D E. The effectiveness of the Salter innominate osteotomy in the treatment of congenital dislocation of the hip. J Bone Joint Surg (Am) 1986; 68 (1): 79-87.

Kruczynski J. Rebuilding of the hip with avascular necrosis after nonoperative treatment of congenital hip dislocation. Thesis, Karol Marcinkowski University of Medical Sciences, Poznan, Poland 1987.

Lindholm T S, Laurent L E, Osterman K, Snellman O. Perthes' disease of a severe type developing after satisfactory closed reduction of congenital dislocation of the hip. A report of three cases. J Bone Joint Surg (Br) 1978; 60 (1): 15-7.

McKay D W. A comparison of the innominate and the pericapsular osteotomy in the treatment of congenital dislocation of the hip. Clin Orthop 1974; 98: 124-32.

Severin E. Contribution to knowledge of congenital dislocation of hip joint: late results of closed reduction and arthrographic studies of recent cases. Acta Orthop Scand (Suppl 63) 1941; 84: 1-141.

Stulberg D, Cooperman D R, Wallenstein R. The natural history of Legg-Calvé-Perthes' disease. J Bone Joint Surg (Am) 1981; 63: 1095-108.

Tachdjian M O. Pediatric Orthopedics. W. B. Saunders Company, Philadelphia 1972: 385-406.

Weinstein S L. In: Lovell and Winter's Pediatric Orthopaedics (Eds. Morrissy R T, Weinstein S L). Lippincott-Raven Publishers, Philadelphia, New York 1996: 903-50.

Williams L, Wientroub S, Canario A T, Fixsen J A. Severe Perthes' disease noted 5 years after successful conservative treatment of congenital dislocation of the hip. J Pediatr Orthop 1982; 2 (4): 424-6.

Wynne-Davies R, Gormley J. The aetiology of Perthes' disease. J Bone Joint Surg (Br) 1978; 60 (1): 6-14.