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Diagnosis of Legg-Calvé-Perthes disease

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SUMMARY

Despite the use in recent years of sophisticated imaging methods, the diagnosis of Legg-Calvé-Perthes disease is predominantly based on clinical examination, standard x-rays in two planes, and ultrasonography. What is essential is early detection of the collapse of the femoral head, which can be very rapid, especially in older age groups. Delayed diagnosis in these cases, usually associated with developed extrusion of the femoral head, prevents the application of principles of conservative treatment, and palliative femoral extension-abduction osteotomy is necessary.

BACKGROUND

Legg-Calvé-Perthes disease (LCPD) was described independently by three observers in 1910-Perthes [1], Calvé [2], Legg [3]. Naturally it was due to the recent introduction of the x-ray imaging. LCPD occurs most commonly in children between 4 and 8 years [4]. However cases beyond this age period are not rare. Meanwhile in children below 4 years the course of the disease is usually benign not needing any form of treatment at all, in children over 8 years the collapse of the femoral head is very dramatic issuing in the rapid development of the femoral head deformity [5]. It seems to us, that recently the number of new cases approaching the age limit of 15 years had increased. Then the differential diagnosis from the idiopathic avascular necrosis of the femoral head in adults concerning the prognosis is needful. Boys are affected more frequently than girls (ratio 5 : 1). It is known, that especially very active boys with short stature are at higher risk for development of LCPD. Retardation of bone age is a very common finding [6].

MEDICAL HISTORY

Affected children complain of the hip or knee pain. Parents observe the limping which can be intermittent. The pain is usually mild and located in the groin. Symptoms can be mild issuing in the delay of the first examination at doctor. The pain is originated from the inflammation of the synovialis of the hip joint.

Physical Examination

Range of motion (ROM) of the affected hip is limited. Predominantly is limited abduction and internal rotation. It seems, that the degree of limited motion corresponds with the involvement of the hip joint by synovialitis. The typical finding is a mild tenderness in front of the hip joint in the groin. Later however the limitation of the hip joint is determined more by the secondary changes in the hip joint. When the femoral head is round or oval the centre of the hip rotation is inside of the socket („Ball and Socket type” of joint). In the cases with rapid development of collapse of the femoral head, the center of hip rotation moves out from the hip joint. It develops articulation between the deformed and extruded femoral head (saddle deformity) and the acetabular edge („Roller bearing shape” of the hip joint) [7]. In clinical examination the extended affected extremity assumes the position in adduction and in flexion in abduction (Sign „Abduction with flexion”).

To maintain free motion of the hip is a demanding task for physiotherapists as well as for parents. Natural development of the affected hip joint under different forms of LCPD (sex, age, degree of involvement, compliance of patient and his family with the treatment) seems to be however more decisive for the prognosis. Until now nobody proved, that the intensive physiotherapy can change the natural development of LCPD. On the other side, under the condition of surgical treatment, deep involvement of patient in physiotherapy is very worthy as the surgical treatment can limitation of ROM make worse.

X-ray diagnosis

The natural history of LCPD is generally thought to go through 4 stages: 1. Synovitis, 2. Fragmentation, 3. Reossification, 4. Late deformity. On the basis of AP and frog x-rays Catterall classification was developed involving four groups concerning the degree of involvement. Moreover the concept of „Head at risk” was defined [8]. Simpler classification was given by Salter and Thompson differentiating groups A and B [9]. Recent classification by Herring [10], based on the lateral pillar concept is at present days mostly used and it seems to be decisively predictive for the outcome of the LCPD. Nevertheless Wiig et al. [11] proved that all three classifications are reliable if the categorisation is done by experienced examiner. Contrast arthrography of the hip joint is a very sensitive method how to disclose the beginning femoral head deformity and lateral extrusion [12]. At the stage of full ossification of the extruded femoral head the possibilities of surgical treatment are limited. Although the epiphyseal extrusion is measurable by US-scanning [13], the anatomical landmarks are much more clear on contrast arthrography.

Bone scan

Although bone scan is a very sensitive method for early depiction of LCPD, associated however with many false-positive findings, its role to differentiate hips with benign course and those with poor prognosis following former Conway [14] interpretation is not highly specific (recanalisation versus revascularisation) [15,16].

MRI imaging

Its value is based on the possibility to make an early diagnosis before x-ray. Moreover MRI can show early the extent of the lesion in the femoral head. Recently introduced dynamic gadolinium enhanced subtraction technique [17] allows early identification of ischemia and the pattern of revascularisation.

Hip arthroscopy

Endoscopic examination is more useful for diagnosis and treatment of sequelae of LCPD [18].

Differential diagnosis of LCPD

Children with LCPD may complain of knee pain. In the case of absence of local knee finding the physician must carefully examine the hip and obtain AP and frog lateral x-rays of the hips. The differential diagnosis from other reasons is given by a characteristic clinical and laboratory data [19]. Some attention necessitates the x-ray differential diagnosis. It must be differentiated Meyer's dysplasia for which is

pathognomical the absence of progress of fragmentation, on the other side the bone growth of the ossified particles of the affected head is apparent [20,21]. Systemic disorders and skeletal dysplasias have characteristic clinical and x-ray findings. „Seven x-ray approach” should be undertaken:

1. Lateral skull
2. AP chest including shoulders
3. Lateral spina
4. AP hands and wrists
5. AP knees
6. AP elbows
7. AP pelvis and hips.

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