Legg-Perthes-Calvé Disease: A Case Report with Review of the Literature

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INTRODUCTION

Legg-Perthes-Calvé disease (LPC) or primary hip osteochondritis (PHO) is an ischemic necrosis of the upper femoral epiphysis. It was described in 1910 simultaneously by Legg in the USA, Calvé in France, Perthes in Germany and Waldenstroem in Sweden. It is a serious and disabling disease. It attacks the upper extremity of the growing femur, and evolves in four phases: condensation, fragmentation, repair, and deformation. The frequency of this disease is not well known and varies according to the authors between 1 in 2000 in Europe and 1 in 5000 in black Africa. Legg-Perthes-Calvé disease is a hip disorder of children between three and ten years of age, with the majority of cases occurring in children under 14 years of age. The sex ratio is 4 boys to 1 girl and the mean age of diagnosis is 6.5 years. We report the observation of a 13 year old female patient who presented for 4 years after a road accident with mechanical pain involving the hips.

Keywords: Legg-Perthes-Calvé disease, female, over 10 years.

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OBSERVATION OF THE PATIENT

Anamnese

13 year old Senegalese patient, black, student (on leave this year) who has presented for 4 years, after a public road accident (hit by a motorbike), oligoarthralgia of mechanical appearance involving the hips and the right knee. The pain was triggered by walking and calmed at rest with nocturnal positional awakenings and a morning wake-up call lasting about 10 minutes. There is also a dodging limp on the right side. Given this symptomatology, the family consulted the regional hospital in Thiès, then referred to Dakar in Orthopaedics at the Aristide LeDantec Hospital (HALD) from where she was referred to the rheumatology department (HALD).

Clinical examination

The general examination reveals a good general condition, a weight of 49 kg, a height of 1.49 m and a body mass index (BMI) of 22.07 kg/m2. On inspection, there was a right-sided dodging limp, on clinical examination there was flessum of both knees, a valgus knee on the left, and inequality of the lower limbs with the distance between the right umbilicus and medial malleolus at 88 cm and the left at 90 cm. Pain is triggered only by mobilisation of the right knee and both hips. Examination of the knee showed no patellar instability (no patellar shock), nor any positive drawer sign. The right hip showed slight joint laxity but this
could not explain the limp. A second, more careful, examination revealed pain on palpation with digitative pressure on the diaphysis of the right femur. Examination of the spine revealed an inversion of the dorsal kyphosis with a thoracic ampliation of +5 cm, a lumbar hyperlordosis, the hand-ground distance was 8 cm. There is no scoliosis (absence of gibbosity in anteflexion). The hypotheses evoked, given the age of our patient (13 years), her road accident, the symptoms and her clinical examination, are a subluxation of the right hip, coxo-femoral dysplasia or primary osteochondritis of the hip (Legg-Perthes-Calvé disease).

Paraclinical examination

The blood count was normal and the haemoglobin electrophoresis was also normal (AA). Immunology : HLA-B27 and antinuclear antibodies were negative. On imaging, the CT scan of the pelvis showed epiphyses with decreased height, flattened heads, slightly short and stubby necks and slightly protruding greater trochanters. Focal upper polar pinching of the right coxofemoral joint and bilateral osteophytes. These features are consistent with sequelae of osteochondritis or Legg-Perthes-Calvé disease. CT scan of the knees was normal.

Diagnosis

In a growing 13 year old girl, a right lower limb dodging limp can have several origins. The most common is a dislocation of the patella. The other main causes of lameness are a rupture of the anterior cruciate ligament, panosteitis or a fracture. The clinical examination ruled out most of these hypotheses. Imaging was very helpful and provided evidence of osteochondritis or Legg-Perthes-Calvé disease.

Treatment and evolution

An analgesic treatment based on ibuprofen (1400 mg every 6 hours) was started at 30 mg/kg and Paracetamol 1000mg 3 times a day at a dosage of 20 mg per kg every 8 hours. This reduced the pain, but in no way prevented its progression. A follow-up telephone call was made. Six months later, no new clinical signs have been noted. We act in concert with the orthopaedic surgeons and have recommended close radiological follow-up for our patient, so as not to miss the onset of excentration of the femoral head, which is an indication for surgery. Subsequently, regular follow-up will take place until bone maturation.

REVIEW OF THE LITERATURE

From an epidemiological point of view

The figures in the literature on the incidence of the disease vary between 5.1 and 29 per 100,000 children under 14 years of age (Table 1) [5].

<table>
<thead>
<tr>
<th>Region</th>
<th>Annual incidence per 100,000 children</th>
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<tbody>
<tr>
<td>British Columbia, Canada (Gray)</td>
<td>5.1</td>
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<tr>
<td>Wessex, Great Britain (Barker)</td>
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<tr>
<td>Massachusetts, USA (Molloy)</td>
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<tr>
<td>Yorkshire, Great Britain (Hall 1989)</td>
<td>6.1</td>
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<tr>
<td>Trent, Great Britain (Barker)</td>
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<td>Zealand, Denmark (Moberg)</td>
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<td>Uppsala, Sweden (Moberg)</td>
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<td>South Jutland, Denmark (Moberg)</td>
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<td>Eastern Cape White, South Africa (Purry)</td>
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<tr>
<td>Mersey, Great Britain (Barker)</td>
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<td>Northern Ireland (Kealey)</td>
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<td>South Scotland (Pillai)</td>
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<tr>
<td>Liverpool, Great Britain (Hall 1983)</td>
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<td>Faroe Islands (Niclasen)</td>
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The older the age, as in our patient’s case, the worse the prognosis. Beyond the age of 9 years, the results of treatment are poor in almost three quarters of cases. Diarra Kadja Founè [11], Marcel Fèvre et al [6], d’Allaine François, and Fotturuso O Ritter [13] made the same observation. B.S. Soumana et al [10], and J Kany et al [14] found lower age ranges (4-8 years).

The clear male predominance is admitted by all. For some authors, it is 80% for boys and 20% for girls [8]. For Troueta [14], this is in favour of the traumatic origin of LPC (same distribution of leg fractures), whereas for Chung the explanation lies in a particular vascular disposition.

The black race would be relatively spared from LPC in Africa as well as elsewhere. According to Fisher, Wynne Devis [15], and Lozar, the frequency of LPC in the black race is 1/5000. Trueta found 10 cases in blacks out of a total of 286 cases [14]. No explanation has been found for this phenomenon. Goff...
also found that the disease was 16 times less frequent in the black race [7].

**Diagnostically**

Gait disorders were the most frequent cause of consultation in several studies. This is due to the fact that muscle contractures constantly accompany the disease on the one hand and the shortening of the affected limb on the other. The same observation was made by Diarra Kadja Founè [11] who found 61.36% of walking disorders as a reason for consultation.

For many authors, sickle cell disease is an aetiological factor in LPC, especially in the black race [9, 12]. The mechanism is believed to be vascular obstruction by sickle cells. The polymerisation of Hb S favours the occurrence of morphological and functional changes in red blood cells: dehydration, membrane rigidity, increased adhesion to the vascular endothelium. Vascular obstruction, responsible for tissue anoxia, leads to bone infarction [9]. Hemoglobin electrophoresis in our patient was normal (AA).

The disease is usually unilateral but may involve both hips in 15% of cases [16]. The diagnosis is often made late, after a few months of evolution [16].

The onset of symptoms is insidious, and in 85% of cases the pain is unilateral and localised to the hip, but the young child often points to the knee as the site of pain [16]. The pain increases with physical effort, with a particularly high point at the end of the day with a dodging limp. Painful limitations are in internal rotation and abduction movements.

Clinical examination shows limitation of internal rotation of the hip (patient lying prone, allowing testing of this rotation on both hips and detecting asymmetry [16]. The Thomas sign is positive (patient supine, healthy hip flexed so that the lumbar spine is flat on the table. The diseased hip is flexed by 15°, indicating hip stiffness). In some cases a hydarthrosis (visible on ultrasound) can be observed.

Standard X-rays are essential with two mandatory views, the face and the Lauenstein view. At the beginning, they are normal. If the clinical symptoms persist, these X-rays must be repeated. Then "small signs" appear: the decrease in height of the cephalic nucleus and the relative widening of the joint space, an irregularity of the nucleus, sometimes, more visible on the Lauenstein view, the clear subchondral border indicating a subchondral fracture.

The technetium-99m bone scan shows a fixation hole in the epiphysis in the ischaemic stage. This is a sign of ischaemia which in most cases will develop into Legg-Perthes-Calvé disease. This examination is of particular interest at the infraradiological stage because the presence of hypoxia has a predictive value of 92% of ischaemia but, crucially, a normal scan eliminates the diagnosis of Legg-Perthes-Calvé disease with a predictive value of 99% [18]. According to Kohler [19], it appears that bone scans, although they do have a diagnostic role in ischaemia which does not necessarily lead to Legg-Perthes-Calvé disease, do not have any prognostic value and have only a limited role in this disease.

Magnetic resonance imaging can be used to identify hip damage more accurately than conventional radiology by visualising the epiphyseal cartilage and also the labrum. The protocol must be specified in order to use the most informative sequences and sections.

The protocol generally used includes coronal sections acquired in T2 STIR, then in T1 SPIN, and sagittal sections in T2 STIR and/or T1 SPIN. The need for gadolinium injection is not clear.

The study of the MRI images should make it possible to specify four parameters [20]:
- The extent of epiphyseal necrosis (more or less than 50%)
- The extent of lateral excentration
- The involvement of the conjugation cartilage
- The extent of metaphyseal involvement

On T2, the inflammation appears as a hypersignal (white) as well as the liquid effusion, the necrosis in white or black.

In T1, the necrosis is visualized in black, the cartilaginous structures are very well visible during this acquisition. It should be noted that normal physeal tissue appears as a wavy band with no signal in T1 and hyperintense in T2, whereas in the case of abnormalities, the cartilage band is enlarged or deformed with "W" or "M" shaped undulations. At the maximum, an epiphyseal-metaphyseal cystic image bridging the cartilage or an epiphyseal-metaphyseal fusion may be seen, indicating ephysisodesis. Metaphyseal signs are cystic images with T1 hypersignal.

MRI with gadolinium injection is at least as effective and early as scintigraphy in making the diagnosis before the appearance of radiological signs [17]. It should therefore be preferred to scintigraphy unless sedation or anaesthesia is required for its performance in small children.

**Therapeutic aspect**

The aim is to achieve optimum remodelling of the epiphysis by revascularisation and reconstruction through fitting the femoral head into the acetabular mould. To achieve this, numerous orthopaedic treatments have been proposed, such as simple unloading with a cane or wheelchair, abduction devices allowing walking or casts [21]. The minimum duration
of one year of treatment raises doubts about the strict adherence to these rules in children. Long-term bed traction, associated with daily mobilisation, imposed, at the time when it was still proposed [22], hospitalisation in a rehabilitation centre, with the inevitable socio-psychological consequences in children who were generally indolent. These treatments have never proved to be effective. Short-term traction can only be justified in the event of an acute painful episode or significant stiffness. In the latter case, it must provide quantified radiological proof of improvement in the abduction angle within a few days, without which it can be quickly discontinued [23]. Surgical treatments are also aimed at improving the fit of the epiphysis into the acetabulum, allowing for better reciprocal remodelling during the revascularisation and reconstruction phases. To achieve this, several techniques are used:

- femoral derotation varisation osteotomies [24].
- acetabular reorientation osteotomies (triple osteotomy, innominate osteotomy [25]).
- Enlargement acetabuloplasty (osteoplastic stop [26], Chiari osteotomy [27, 28]). The indications therefore seem to be as follows.
- unloading under English canes in the necrosis phase with a volume greater than or equal to 50% of the total epiphyseal volume and maintenance of mobility (abduction and internal rotation) by physiotherapy. Support may be restored in the revascularisation phase.
- bed traction for a few days in case of acute pain or frank stiffness, with radiological control of the mobility gain obtained.
- surgical treatment in the early phase of revascularisation [29], in children with a bone age of at least six years (although this is not universally accepted [30] in the case of progressive excetration of the hip, in Herring B or B/C. Monitoring should be done clinically and radiologically every two to three months. Heavy and punishing orthopaedic treatments have been progressively abandoned. It is currently recognised that 60% of untreated hips evolve favourably without treatment. In the remaining 40%, with poor prognosis signs, treatment is recommended because it shortens and positively modifies the natural history of the disease. Finally, Herring C appears to have a poor prognosis in the majority of cases, with or without treatment.

Evolution

In the acute phase, the radiological evolution has been described in three radiological phases by Waldenström. The first is a hyperdense core, which he describes as the necrosis stage, the second is the fragmentation stage and the third is the reconstruction stage with the appearance of neoformed bone. The correspondence of the radiological images with the histological stage that he proposes can no longer be accepted today. Indeed, stage 1 of hyperdensity corresponds to the beginning of revascularisation and not to avascular necrosis. Reconstruction begins at this stage, continues with bone resorption at stage 2 of fragmentation and intensifies at stage 3. The interest of this classification is above all to appreciate the stage of fragmentation at which excetration occurs, which must be quantified early on, and the duration of the disease. The longer the disease lasts, the worse the prognosis. More interesting seems to be Joseph's [37] modification of Canale's [31] Elizabethville classification into 7 evolutionary stages. Stages Ia to Ib last 3 to 4 months, stages IIa and IIb 9 to 16 months. The transition from stage Ib to stage IIIa is tricky, with possible signs of severity (lateral calcifications, metaphyseal and acetabular signs). The deformity appears from stage IIIa onwards. In 30% of cases, stage IIIb includes lateral calcification.

At the end of the evolution, the restoration of the sphericity of the femoral head is variable, ranging from perfect sphericity with some irregularity to significant deformities with a "square head". It is here that the deformities of the head with flattening (coxa plana) or coxa magna are appreciated, associated with deformities of the acetabulum, of the neck which is short and causes a shortening leading to an inequality of length of the lower limbs, as a consequence of the remodelling of the growth plate. Stulberg's classification [42] allows results to be assessed according to Mose's criteria of sphericity and congruence of the femoral head and acetabulum. It is based on a study that compared the radiological images of the femoral heads at bone maturity and in the long term (30 and 40 years). There is no change in class between these two dates. Class I corresponds to a completely normal hip, class II to a spherical and congruent femoral head but with abnormality (ies) in the head, neck or acetabulum, class III where the head is non-spherical but not flattened, still concentric, with or without the same associated signs as before, class IV where the head is flattened but congruent with associated signs, class V with a flattened and incongruent head without associated signs. Stulberg found that heads classified as I and II had no signs of long-term OA, classes III and IV had mild to moderate OA and class V had severe OA at less than 50 years.

Catterall, in 1971 [32], described 4 radiological signs of severity, which were increased to 5 in 1981 [33]:

- the modified Gage sign, which corresponds to a small osteoporotic segment forming a clear V-shaped image on the lateral edge of the epiphysis,
- lateral epiphyseal calcification,
- Lateral eccentricity according to Green [35] corresponding to the percentage of epiphyseal uncovering in relation to the width of the physeal on the healthy side E=(AB/CD) x 100. If this percentage exceeds 20%, it is a serious factor.
- horizontalization of the physeal,
- diffuse metaphyseal damage.
These are the so-called "head at risk" signs.

Lateral excentration or subluxation is the major sign of severe forms. It must be carefully looked for. Heyman [36] has described an index (acetabulum head index AHI) measuring the relationship between the head and the acetabulum on frontal radiographs. Sales de Gauzy and Cahuzac [41] calculated this index on MRI and showed that it was more reliable because MRI takes into account the cartilage structures of the acetabulum and femoral head. Worsening of this index is diagnosed earlier on MRI, which is the reference examination to assess the evolution.

When the clinical examination suspects a hinge abduction, it was recommended that a so-called “refocusing” abduction film be taken to assess the reducibility of the eccentricity and the reintegration of the external calcification into the acetabulum. This image is certainly useful before making a therapeutic decision. Quain and Catterall [40] performed arthrography, which has now been superseded by MRI. Kruse gave precise radiographic criteria on the abduction view: widening of the medial joint space by more than 2 mm and reduction of the superior-lateral joint space [38]. Revascularisation is assessed by scintigraphy according to Conway's classification [34] into two groups; group A with early and rapid revascularisation of the lateral column with a good prognosis, group B with central hyperactivity or hypofixation persisting for more than 5 months which has a poor prognosis. Lamer [39] has shown that MRI with gadolinium injection allows early diagnosis of revascularisation and better assessment of its extent, which confirms the limited use of scintigraphy at this stage.

CONCLUSION

Legg-Perthes-Calvé disease (LPC) or primary hip osteochondritis (PHO) is an ischaemic necrosis of the upper femoral epiphysis occurring in children aged 4 to 8 years with a male predominance (20% in girls and 80% in boys). It is a condition of unknown cause, but there seems to be a consensus that it is of ischaemic origin in the territory of the posterior circumflex artery. However, in the multifactorial origin of LPC disease, sickle cell disease in the black race is mentioned. The evolution is progressive over several years and always ends in recovery with or without sequelae. After an asymptomatic period of three to four months (initial ischaemia stage), there follows an evolutionary period of 18 months to 3 years (successive stages of necrosis, fragmentation and reconstitution), then a phase of remodelling of the femoral head lasting several years until the end of growth. During this period, the head recovers a satisfactory shape and congruence (adaptation of the acetabulum) or, on the contrary, presents a sequential state which weighs on the future: in severe cases, the subject will be exposed in adulthood to the risk of premature osteoarthritis of the hip.

REFERENCES


