Legg-Calvé-Perthes disease
(Hip osteonecrosis)

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ABSTRACT

Legg-Calvé-Perthes disease is a form of idiopathic avascular necrosis of the femoral head, evolving to classical “coxa plana”, which will lead to secondary hip arthritis. Legg-Calvé-Perthes syndrome occurs most commonly in boys than in girls by a ratio of 4 to 1, commonly in the age range of 4 to 8 years. Etiological factors are represented by vascular, traumatic, constitutional, endocrine, genetical, racial and socioeconomical factors.

Three phases have been described: avascular necrosis, fragmentation and healed phase. Pathogenesis is represented by two concomitant processes, located in femoral ossific nucleus: resorbtion of necrotic bone and new bone genesis. Two forms are described: Potential form – no fracture occurs in subcondral level; Real form – after the fracture occurs. Clinical signs are represented by: pain – of mild nature, usually activity-related; lameness walking, and limited hip motion.

Radiological findings: three stages are described: Initial stage – characterized by a pathognomical sign “nail scratch”; Second stage – The bony epiphysis begins to fragment, presenting areas of increased radioluency and radiodensity; Third stage – normal bone density returns. Alterations in the shape of the femoral head occurs. Other exams – IRM, arthrography, scintigraphy.

Classifications: Catteral – associating “head at risk” signs, groupe 1 or 2 and no “head at risk sign” having good prognosis, 3 or 4 plus “head at risk sign” having poor prognosis; Salter and Thompson; Herring.

Evolution of long term, leading to epiphyseal deformities in severe forms.

Treatment: has been based on the containment principle, being represented by orthopedic treatment or surgical treatment.

Key words: femoral head; idiopatic avascular necrosis; epiphyseal deformities; radiological findings; classifications; orthopedic and surgical treatment.

Hip osteonecrosis represents a clinical and radiological entity, characterized by avascular necrosis of proximal femoral ossification center, evolving to classical “coxa plana”. This illness may spontaneously heal, but important modifications, which will lead to secondary hip arthritis (1).
**EPIDEMIOLOGY AND ETIOLOGY**

Legg-Calvé-Perthes syndrome occurs most commonly in boys than in girls by a ratio of 4 to 1. It occurs most commonly in the age range of 4 to 8 years, but cases have been reported in children from 3 to 11 years of age. Usually affects one hip, the incidence of bilaterality being reported as 10% (1,2,3).

**ETIOLOGICAL FACTORS**

- **Vascular factors** – arterial obstruction, especially in posterior circumflex artery; intraosseous venous hypertension; coagulation disorders (1,2,4,5);
- **Traumatic factors** – especially repetitive microtraumatism, seems to have an important etiological role. That is reflected by an increased frequency in hyperactive children involved in sports activities, explaining male predominance.
- **Constitutional factors** – causing congenital epiphyseal weakness (Lehmann) the involved individuals having skeletal maturation delay. There is considerable evidence of anthropometric abnormalities in children with Legg-Calvé-Perthes syndrome, affected children being smaller in all dimensions, comparative with unaffected children (6,7).
- **Endocrine factors** – affected children presents abnormalities of growth hormones (IGF, STH) (8,9)
- **Genetic factors** – pose an inheritance risk of 10% (10)
- **Racial and socioeconomical factors** – there is an increased frequency of Legg-Calvé-Perthes syndrome in Japanese people, other Asians, Eskimos, and central Europeans, and a decreased frequency in native Australians, Americans Indians, Polynesians, and blacks. Another factor is represented by malnutrition, which is related to the reportedly increased incidence of Perthes disease in low-income families (3).

**PATHOLOGICAL ANATOMY**

The main lesion is avascular necrosis of ossific nucleus histologically counting three phases.

1. **Avascular necrosis phase** – characterized by osteocytes degenerescence and destruction. Bone marrow degenerative changes are early. In the same time we observe adjacent soft tissue modifications – thicken synovial and articular capsule caused by congestion and edema. Blood vessels are dilated, surrounded by lymphocytic infiltrate (2,11).

2. **Fragmentation phase** – characterized by necrosis process in ossification center and adjacent cartilaginous tissue, followed by invasion of conjunctivovascular buds. There are areas of osteolytic, necrotic bone, alternating with areas of unaffected bony trabeculae. Between bony trabeculae, is an amorphous mass of extravasate eritrocites, degenerated fat cells and bone marrow. The new formed granulation tissue and capillars from metaphyseal periosteum will absorb dead bone tissue by osteoclastic process, causing trabecular fragmentation (12).

3. **Healing phase** – characterized by partial or total substitution of necrotic bone, by newly formed bone tissue. If the dead bone was partially replaced, necrotic bone will persist leading to a specific aspect: osteochondritis dissecans. In case of non weight bearing during all evolution period, normal shape of femoral head can be restored (12).

**PATHOGENESIS**

Initially an ischemic episode occurs, leading to avascular necrosis of epiphyseal ossific nucleus. As result of this process enchondral ossification of growth cartilage is ceased, while the articular cartilage, being nourished by synovial liquid, keeps growing (2,13). In this stage, at the affected hip, radiologically, it can be observed a widening of the articular space and a smaller ossific nucleus. From periphery to center ischemic areas who kept normal structure are revascularized and the vascular buds are occupying vascular chanelles. In this way the osteocytes and osteoclasts intake will lead to new bone formation at the necrosed trabeculae, causing their thickening. This can be observed radiologically, as an increased radiodensity, in early stages.

Two concomitant processes are located in femoral ossific nucleus: resorption of necrotic bone and new bone genesis. In subchondral level resorption process is predominant causing local fracture. There are two forms described:
potential form, no fracture occurs and real form, after the fracture occurs.

**Potential form** – ossification process is uninterrupted, leading to normal growth and development. Epiphysial deformation is not present and hip motions are in normal ranges. Radiologically, it can be observed “head in a head” image, which represents the contour of growth stope line, which limits the ossific nucleus a the initial ischemic episode (7).

**Real form** – subcondral fracture starts at the anterolateral area of the epiphysis adjacent to growth cartilage (maximum stress area during weight bearing, extending superior and posterior. A second ischemic process, of mechanic etiology, due to collapsing trabeculae and neocapillaries obstruction (12). Afterwards, a new vascularization process occurs, starts from the periphery. The necrotic bone is replaced by a vascularized fibrous tissue, which later can ossify. This process is called “creeping substitution”, when the femoral head is remodeled by the applied stress.

The deformities degree depends on the stage of necrosis, head-acetabulum interactions. Reconstruction stage may lead to a spherical unmodified femoral head, or to lost of sphericality, causing a flat head – coxa plana. Secondarily, acetabulum changes occurs, as a reaction of head shape and volume changes. This is reflected in an increased volume of acetabulum, characterized by a high opening angle, and by increased acetabulum roof. In severe cases, this acetabulum modifications, are insufficient, to prevent a deformed and uncontainable femoral head.

The physeal plate in Legg-Calvé-Perthes syndrome shows evidence of cleft formation with amorphous debris and extravasation of blood. In the metaphyseal region, endochondral ossification is normal in some areas, but in others the proliferating cells are separated by a fibrillated cartilaginous matrix that does not calcify. The cells in these areas do not degenerate but continue to proliferate without endochondral ossification, leading to tongues of cartilage extending into the metaphysis as bone growth proceeds in adjoining areas. As a specific element, there are no bone necrosis areas in metaphysis.

Another lesion is represented by extrusion of growth cartilage plate, in cases of severe deformities of femoral head. The alterations of growth cartilage and metaphysis causes a defected longitudinal growth of proximal femur, leading to a shorter and thicker femoral eck, associated with a widened femoral head – “coxa magna” (Figure 1) (maximum level in fragmentation phase) (7). At the same time, the unaffected great trochanter, is developing normally, over the femoral head level. All this modifications, are leading to functional coxa vara, with abductor muscles insufficiency (13).

Due to shortening of femoral neck, clinical changes occurs, consisting in a 1-2 cm shorter inferior limb.

**CLINICAL PRESENTATION – STARTS INSIDIOUSLY**

**Pain** – of mild nature, usually activity-related. The pain the patients experience generally is localized, to the groin, or referred to the anteromedial thigh, following obturator nerve’s innervations area (15).

**Walking** – with a history of the insidious onset of a limp. Trendelenburg sign may be positive due to abductor muscles insufficiency. It can be observed a moderate thigh muscles atrophy, and a 1-2 cm inferior limp shortening (1,15).

**Hip motion** – limited, particularly abduction and medial rotation (15).

**RADIOLOGICAL FINDINGS**

Radiological examination plays an important part in establishing diagnosis, evolution and prognosis (16).

There are three phases described:

**Initial phase** – characterized by a pathognomonic sign – “nail scratch” represented by a fine radiolucent line parallel to the subchondral surface of femoral head (Figure 2).

In this stage, one of the first signs of this condition is failure of the femoral ossific nucleus

**FIGURE 1.**
to increase in size because of a lack of blood supply. The affected femoral head appears smaller than the opposite, unaffected ossific nucleus.

The femoral ossific nucleus appears radiodense. This relative increased radiodensity may be caused by osteopenia of the surrounding bone.

Growth cartilage plate becomes thin and irregular. Widening of metaphyseal area may be early, always being observed during the evolution of the disease (16).

Widening of the medial joint space is another early radiographic finding, being measured by distance between femoral head contour and the radiological U.

**Second phase** — characterized by opacity, fragmentation and flattening of the ossific nucleus. The increased opacity of ossific nucleus in this stage is real reflecting it’s collapse and overlapping of trabecular fragments. Also the opacity can be explained by formation of new bone tissue at the unresorbed trabeculae (17).

**Third phase (healing)** — characterized by the presence of newly formed bone tissue. The outer contour of the epiphyseal nucleus becomes irregular. The femoral head is shorter and wider, and the femoral head may be modified becoming wide and flat. Secondarily flattening of acetabulum can be observed (16).

**MRI** — represents a frequently used investigation method, having a high sensitivity in detection of ischemic areas. Therefore in stage 1 the extension of necrotic process is difficult to evaluate without MRI investigation (18).

**Artography** — plays an important part in establishing the possibility of centering the femoral head and the position in which this can be performed. Also it can reveal incipient flattening of femoral head and the abduction hinge phenomenon.

**Scintigraphy** — has a defining role in diagnosis and prognosis facilitating the determination of the epiphyseal revascularization.

**Classifications** — Several classification methods have been developed to help establish therapeutical strategies and also playing a part in prognosis.

**Catteral** classification represents the most important one and is based on femoral head radiological aspect (13). It has four groups:

- **Group 1** — limited involvement of anterior part of the femoral head. Profile radiography shows subchondral fracture which is not visible in AP incidence because it doesn’t extend in the superior region. Good prognosis.
- **Group 2** — lateral epiphyseal column is maintained intact. Subchondral fracture is visible in AP incidence also because it extends posterior over the superior part of epiphysis. Good prognosis.
- **Group 3** — lateral epiphyseal column is involved. AP incidence shows subchondral fracture extending from the lateral part towards the medial part of the epiphysis, involving a major part of it. The posterior part is partially uninvolved. Reserved prognosis.
- **Group 4** — the major radiological feature is the involvement of the entire epiphysis. In AP and profile incidences the fracture extends on the entire subchondral area. Poor prognosis.

**Salter and Thompson** described a simplified two-group classification based on prognosis and determined by the extent of the subchondral fracture line, which appears early in the course of the disease: in group A, less than half of the head is involved (Catterall groups 1 and 2) (Figure 3), and in group B, more than half of the head is involved (Catterall groups 3 and 4) (Figure 4). The major determining factor between groups A and B is the presence or absence of a viable lateral column of the epiphysis. This intact lateral column (i.e., Catterall group 2, Salter-Thompson type A) may shield the epiphysis from collapse and subsequent deformity.

For patients included in groups 2, 3, 4 Catteral have been described certain radiological signs called “head at risk signs”. These are: 1. Lateral
extrusion of femoral head; 2. Pericapital ossifications; 3. Metaphyseal cysts; 4. Horizontal growth cartilage plate; 5. Gage sign, V-shaped radiolucent defect localized laterally at both epiphysis and metaphysis (Figure 5).

Herring and colleagues proposed a radiographic classification based on the radiolucency of the lateral pillar of the femoral head on anteroposterior films during the fragmentation phase of the disease (Figure 6). The lateral pillar occupies the lateral 15 to 30% of the femoral
head width on an anteroposterior radiograph. The central pillar occupies approximately 50% of the head width, and the medial pillar occupies 20 to 35% of the medial aspect of the head width on anteroposterior radiography.

*Herring A* – the lateral column has normal height. Excellent prognosis.

*Herring B* – the lateral column is collapsed up to 50% of normal height, +/− central column involvement. Therapeutical results are good in patients up to 8 years.

*Herring C* – lateral column is collapsed with over 50% of normal height. Reserved prognosis.

**EVOLUTION AND PROGNOSTIC – PROGNOSTIC FACTORS**

**Good prognosis** – mobile hip; sufficient external column (Catteral 1,2); no head at risk signs; good vascularization, scintigraphically determined; no extrusion of femoral head (12).

**Reserved prognosis** – extrusion of femoral head; advanced external column collapse (Catteral 3,4); presence of head at risk signs; articular incongruency (19).

**Evolution** – long term, 3-4 years. In severe forms, Catteral 3-4, in which treatment methods failed to ensure a good centering of the femoral head, will lead to severe deformations. This is represented by femoral head flattening and slightly external subluxation. Articular incongruence will lead to secondary arthritis, which in some cases may appear early in teen subjects. In order to evaluate these modifications secondary to remodeling process and establishing a prognosis, several evaluations methods have been developed.

*Moses method* – it evaluates femoral head sphericity using concentrating circles, 2mm one from each other. Using this method there are three types of femoral head shapes: 1. normal, femoral head inside the same circle; 2. regular, femoral head is in between two circles; 3. irregular (18).

*Stulberg method* – based on femoral head sphericity and head-acetabulum congruency. There are 5 groups: 1. normal hip; 2. coxa magna type modifications whith spherical femoral head; 3. irregular but congruent femoral head; 4. flattened but congruent femoral head; 5. irregular and incongruent femoral head (18).

The extension of femoral head involvement is directly linked duration of evolution. Generally, the larger epiphyseal surface involvement, the longer evolution of the condition and the final outcome is poor.

The starting age has an important role in prognosis. Therefore, the younger the patient the more favorable the prognosis. The younger patients possess a high capacity of epiphyseal and acetabulum remodeling, and in the same time head a risk signs are rare, especially in patients under 5 years of age. In a reduced number of cases (Catteral 1,2) it may be obtained full anatomical and functional recovery, with the femoral head regaining spherical form.

**TREATMENT**

The main goal is the relief of weigh bearing on femoral head and preventing its extrusion, the final goal consisting in a spherical femoral head at the time of healing. The basic principle is that of containment.

The essence of containment is that, to prevent deformities of the diseased epiphysis, the femoral head must be contained within the depths of the acetabulum to equalize the pressure on the head and subject it to the molding action of the acetabulum. Containment is an attempt to reduce the forces through the hip joint by actual or relative abduction and moderate internal rotation positioning.

Treatment must be early started, in necrosis or fragmentation phases, and maintained during the whole evolution period.

No form of treatment is indicated if the child demonstrates none of the clinical or radiographic at-risk signs; if he or she has Catterall group 1 or 2, or if the disease is already in the reossification stage.

A child who demonstrates clinical or radiographic at-risk signs, regardless of the extent of epiphyseal involvement, should receive treatment. Even patients with Catterall group 2 disease (or lateral pillar type B disease) who are at risk may have poor results without treatment.
Demonstration of the hinge abduction phenomenon, or the inability to contain the hip, is a contraindication to any type of containment treatment. Serious damage to the femoral head and acetabulum may result from trying to contain a noncontainable head.

**Orthopedic treatment** – It ensures stress relief on the hip achieving immobilization weight bearing relief and a good containment of the femoral head avoiding its collapse (4).

Methods: Petri broomstick abduction long leg plasters. The disadvantage of this method is head-acetabulum contact in the same area impairing articular cartilage’s nourishment. Also it limits the knee and ankle range of motion.

Continuous skeletal traction with the leg in abduction and moderate internal rotation is one of the most efficient treatments. The major disadvantage is the long immobilization period ranging from 10 to 12 months. The next 6 months walking is allowed with discharging braces.

Much more useful are the discharging orthosis with ischiatic support of Tachdjian type or ankle suspension bandages. This kind of orthosis allows walking with weight bearing on healthy leg.

Another type of discharging orthosis are Atlanta and Chicago types. The most widely used abduction orthosis is the Atlanta Scottish Rite orthosis or a modification thereof. These devices were thought to provide for containment solely by abduction without fixed internal rotation. These orthotic devices allow free motion of the knee and ankle. Containment is provided by the abduction of the brace and the hip flexion required walking with the legs in abduction.

**Surgical treatment**

Surgical methods of providing or maintaining containment are advocated by many investigators. Surgical containment methods offer the advantage of early mobilization and the avoidance of prolonged bracing or cast treatment. In addition, no end point for discontinuing treatment is required, and any improved containment is permanent. Surgical containment may be approached from the femoral side, the acetabular side, or both sides of the hip joint.

**Femoral Osteotomy** – with or without associated derotation, offers the theoretical advantage of deep seating of the femoral head and positioning of the vulnerable anterolateral portion of the head away from the deforming influences of the acetabular edge. The varus position reduces the joint forces on the femoral head. This procedure also relieves the intraosseous venous hypertension and improves the disturbed intraosseous venous drainage reported in Legg-Calvé-Perthes syndrome, thus speeding the healing process. This belief, however, has been disapproved.

Prerequisites for varus derotation osteotomy include a full range of motion, congruency between the femoral head and the acetabulum, and the ability to contain the femoral head in the acetabulum in abduction and internal rotation. As with nonoperative treatment, the procedure must be performed early in the initial or fragmentation stage of the disease to have any effect on head deformity (11).

The negative aspects of this treatment modality must be considered. Varus osteotomy, with or without derotation, usually requires the use of internal fixation and external mobilization in plaster for 6 weeks. The patient must incur the inherent risks and costs associated with at least one surgical procedure and most likely a second surgical procedure for hardware removal. The limb is temporarily shortened by the procedure. The varus angle must not exceed a neck–shaft angle of less than 110 degrees. The varus angle generally decreases with growth, however, if there has been physeal plate damage secondary to the disease, this remodeling potential may be lost, and the patient may have permanent shortening and temporary or permanent weakness of the hip abductors. The proponents of varus osteotomy, with or without derotation, report 70 to 90% satisfactory anatomic results using this method (18,20).

**Coxal osteotomy** – The most efficient method is the Salter osteotomy which provides for containment by redirection of the acetabulum, providing better coverage for the anterolateral portion of the femoral head. The head is placed in relative flexion, abduction, and internal rotation with respect to the acetabulum in the weight-bearing position. Any shortening caused by the disease can be corrected, and the need for bracing is eliminated. Prerequisites for innominate osteotomy include restoration of a full range of motion, a round or almost round femoral head, and joint congruency demonstrated arthrographically. Treatment must be performed early in the course of the disease, and the head must be well seated in flexion, abduction, and internal rotation (21).
Salvage procedures are used for uncontrollable hips, especially the ones with abduction hinge. These procedures are Chiari osteotomy, lateral shelf arthroplasty, cheilectomy, and abduction extension osteotomy. These procedures are palliative having as a goal improvement of articular range of motion reducing abductor muscle insufficiency, reducing pain and correcting the shortening of the leg (19,20).

Therapeutical indications

Necrosis phase – just a simple supervision of patients is indicated in good prognosis forms: partial forms, and in under 4 years of age patients (Figure 7). Orthopedic treatment is recommended in uncertain prognosis forms: extended forms, in patients under 4-years old, and in all necrosis beside the partial ones after 6 years of age. The most used procedure is continuously skeletal traction, followed by limitation of physical activities, wheelchairs, discharging orthosis and restarting traction.

Fragmentation phase – if the prognosis is apparently good: mobile hip, sufficient lateral column, no head at risk signs, good vascularization, no extrusion, treatment can be reduced to limitation of physical activity, under clinical, radiological and MRI, monitorization until reconstruction phase. If the prognosis is poor, with extrusion advanced collapse of external column, the option are orthopedic and surgical treatment, the latter being prefered due to apparently superior result.

Reconstruction and remodeling phase – in case of a flattened, extruded femoral head, or reticular incongruency, the surgical treatment can be taken in consideration.

FIGURE 7

REFERENCES


