

Editorial

LEGG-CALVÉ-PERTHES DISEASE: 100 YEARS

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Legg-Calvé-Perthes disease (LCPD) started to be studied 100 years ago (1910), thanks to the advent of the possibility of conducting clinical studies by means of radiographic images (1895). However, many questions remain open, both in relation to the etiology of this orthopedic condition and in relation to which therapeutic measures are valid. Thus, great controversy has been generated.

In 1910, three studies were published, respectively by A. Legg, J. Calvé and G. Perthes. These studies described a new condition affecting children's hips that differed from joint tuberculosis, which was the commonest disease found at that time. Calvé (1875-1954) worked at the "Berck Sur Mer" hospital, which was the biggest specialized tuberculosis service in France. In 1898, he acquired an X-ray machine and, using this, he identified 10 patients with the new disease, which he named pseudo-coxalgia. In turn, working in Leipzig, Germany, Perthes (1869-1927) was one of the first surgeons to use radiographs in his country, and he defined six clinical cases of a condition that he named "juvenile deforming arthritis", which differed from tuberculosis of the hip. In the same year, in Boston, United States, the Harvard graduate Legg (1874-1939) described a new condition in a study on "an obscure hip condition".

Since then, over all the intervening years, around 2000 studies on this topic have been published. Some techniques and concepts have been described both for conservative treatment and for surgical treatment.

The incidence of LCPD is around 15 cases per 100,000 individuals, and it is more common in boys than in girls, with a ratio of 5:1. The disease is very rare among Afrodescendent and Chinese individuals. It is interesting to note that its incidence is greater among individuals of poorer social class: 26 per 100,000 in class 5 versus 4 per 100,000 in class 1.

What is known in relation to its etiology? The etiology continues to be obscure, with occurrences of disorders of the arterial circulation or the venous drainage of the growing femoral head, or both of these. Trueta (1949) attributed the cause of the necrosis to closure of the epiphyseal vessels located in the posterosuperior region of the hip, thus demonstrating that in children aged four to seven years, the only source of blood supply to the proximal femoral epiphysis is the lateral epiphyseal vessels. Trauma or inflammatory processes affecting these vessels, with consequent obstruction, may lead to LCPD. Camargo et al (1984) and Godoy Jr. (1988) demonstrated through selective angiographic studies on deep arteries that patients with LCPD present partial or total occlusion of the medial circumflex artery of the affected hips, which may make the proximal femoral epiphysis susceptible to ischemia within the age group from four to eight years.

Dr. Robert Salter made an especially large contribution towards the anatomopathology of this condition through an experimental model in pigs. However, there is no reason to question the hypothesis that LCPD is caused by ischemia of the epiphysis. A deficit in the venous drainage of the hip may also occur. Hence, the etiopathology of this disease may be explained thus: 1) at birth, the child carries vascular abnormalities that are probably of genetic origin; 2) with the passage of time, the child develops abnormalities of skeletal growth and bone maturation; and 3) within the age group from four to eight years, when the epiphysis is most susceptible to ischemia, because of the vascular pattern in this age group, the process could be triggered by a factor capable of breaking down the precarious circulatory equilibrium that had been maintained until that

time. This triggering factor could be: trauma to the joint, thereby causing arterial and/or venous obstruction; an episode of traumatic or inflammatory synovitis (due to a distant infectious focus), thus producing arterial and/or venous plugging and obstruction; or fractures due to stress or pathological conditions in the epiphysis, thereby interrupting the blood irrigation to a greater or lesser degree. The vascular abnormalities seen in LCPD consist of diminished vascularization on the affected side or absence of the medial circumflex artery, or alternatively, presence of an atrophied medial circumflex artery or one with obstructions in its distal branches. Dimeglio (1995) advocated these ideas in relation to delayed development of the hip joint, occurrences of local microtrauma and vascular abnormalities.

Research and papers on this topic in Brazil go back more than 50 years. In 1957, Prof. Flavio Pires de Camargo published the results from an original study on so-called “bone grafting through inversion” which produced satisfactory surgical treatment for LCPD. His study showed the possibility for “biological” treatment, in which vascularization of the proximal femoral epiphysis would be stimulated based on theories put forward by Trueta, who had studied and investigated bone tissue circulation. Thus, Camargo proposed that revascularization of the femoral neck could be achieved using an inverted bone graft and through stimulating the formation of collateral vessels that would undergo anastomosis with the epiphyseal vessels.

What is known about the treatment? The aim in treating LCPD is always to improve hip mobility and the anatomical relationship between the femoral head and the acetabulum, in an attempt to diminish the deleterious effect of the disease on the joint. Precise indication of the best type of treatment is sometimes difficult. The treatment ranges from clinical and radiographic follow-up of the child, to conservative treatment consisting of the use of traction, orthopedic braces or plaster casts for hip abduction; and to operative treatment when the criteria that have been defined allow such indications. Operative treatment is indicated primarily in the initial phases of LCPD and is based on two distinct recommended approaches: proximal femoral osteotomy for varization and derotation; or osteotomy on the iliac bone, in the innominate region of the bone. Both of these osteotomy procedures have the aim of improving the cover of the compromised femoral head. Several studies have compared the femoral and iliac osteotomy techniques, and it has been demonstrated that the results regarding the shape and containment of the femoral head are similar, thereby increasing the polemic about this issue.

Cordeiro (1972, 1980) analyzed the role of intertrochanteric femoral osteotomy as a “mechanical and biological” operative treatment method for LCPD and showed that around 60% of the results were satisfactory. Guarniero et al (1995, 1997) published new analyses on the results from femoral osteotomy procedures and showed that 65% of the results were satisfactory. Rossi (1995) suggested an easier technique for performing the osteotomy.

Telöken (1992) followed up 31 patients who had been treated by means of Salter acetabular osteotomy and concluded that the normal duration of the fragmentation phase of the disease became shortened, which could be explained by the increased blood flow to the femoral head that the surgery promoted, through an increase in the collateral vascular network going to the epiphysis. In a study conducted in 1993, Kuwajima found that the time taken for evolution from the necrosis stage to fragmentation decreased when Salter osteotomy was performed, with an increase in the vascularization of the epiphysis.

What would the role of arthrodiastasis using an external fixator consist of in cases of LCPD? According to the studies and investigations conducted in our orthopedics service since 1991, we can affirm that arthrodiastasis using a monolateral external fixator applied to the hip, with or without articulation, promotes acceleration of the reossification of the femoral head and induces improvement of the degree of joint mobility. This procedure is indicated in the necrosis and fragmentation phases of LCPD.

The prognostic factors involved in this disease include: the patient’s age at the time of disease onset; the extent to which the femoral epiphysis is compromised; presence of two or more so-called “radiographic signs of a head at risk” (as described by Catterall); the height of the lateral pillar (Herring classification); and premature closure of the epiphyseal growth plate. It can be concluded from reviewing the recently published studies that the treatment used for LCPD is based much more on the orthopedist’s personal experience than on truly scientific foundations. Additional prospective studies are needed in order to deepen our knowledge about different results from orthopedic treatment. There is also a need for guidelines and protocols, in order to ensure uniformity between the different types of therapeutic possibility.