

# Perspectives on Legg-Calvé-Perthes disease

Colin Moseley MD, CM, FRCS

Clinical Professor of Orthopaedics, University of California at Los Angeles  
Emeritus Chief of Staff, Shriners Hospitals for Children, Los Angeles

## Correspondence:

Prof C Moseley  
4865 Glencairn Rd  
Los Angeles  
USA 90027

Email: colinmoseley@mac.com

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## Introduction

The world of Legg-Calvé-Perthes disease (LCP) has been full of activity but there has been little progress made. We are as unsure of the indications for treatment now as we were half a century ago and, in fact, the evidence that any treatment is effective is weak. Our understanding of the disease is incomplete; nevertheless each of us, as a surgeon, must develop a working approach to the problem in order to deal with the patients who present with this condition. This article, therefore, will not attempt to provide recommendations for management, but will present ideas and a framework that might help orthopaedic surgeons to gather their thoughts about this condition, and to develop their own working approach.

**Key words:** paediatric, hip, femoral head, Legg, Perthes

## Evidence

It is worth considering why we do not have stronger evidence about a disease that is fairly common, and has been the subject of countless papers, very few of which have resulted in strong evidence or useful guidelines. Herring conducted a review of the literature and concluded that the literature is not very helpful.<sup>1</sup> He initiated a rigorous and well-managed multi-centred study in 1984 to examine several surgical and non-surgical treatments.<sup>2</sup> After 30 years of collecting data, and with a 20-year follow-up the difference in the results of these several treatment modalities were not obvious. He found that young patients did not all do well, and that patients over 8 years of age with moderate femoral head deformity might benefit from surgery.

Why has strong evidence been so hard to come by? One of the difficulties stems from the fact that we do not have an objective and reliable classification to use in grading the severity of this condition. The Salter-Thompson classification is reputed to give an early indication of the severity in terms of the extent of epiphyseal involvement, but the sub-chondral fracture they describe is only visible in the early stages, and in less than half of those cases.<sup>3</sup>

Catterall's classification with the 'head at risk' signs is, unfortunately, a staging classification since the classification can change during the course of the disease as the apparent extent of involvement of the epiphysis increases.<sup>4</sup> Herring's 'lateral pillar' classification,<sup>5</sup> which has become widely accepted, is likewise a staging, not a grading, classification. The categories in the original classification were difficult to assign, requiring the addition of the 'B/C' class to accommodate those cases that could not be clearly assigned to either the B or C categories.

At the other end of the disease we have a similar problem with respect to defining outcomes. Outcomes are played out over the lives of our young patients, but no prospective long-term studies have been reported. Retrospective studies of late outcomes are difficult to perform, and Weinstein's study<sup>6</sup> is one of the few good ones available, showing that about 50% of all LCP patients go on to need total hip arthroplasty.

*We do not have an objective and reliable classification to use in grading the severity of this condition*

Because of the difficulty in following patients over the long term we have resorted to intermediate, or proxy, outcomes that we can determine at skeletal maturity. The Stulberg classification<sup>7</sup> is used most commonly but is not completely adequate because it uses some imprecise terminology and is difficult to apply. In addition, it is not completely clear how the Stulberg classes relate to the development of osteoarthritis and later difficulties.

It seems clear, then, that as long as we have difficulty classifying the patients as they enter our studies, and have difficulty assessing their outcomes, we will have difficulty gathering evidence and reaching strong conclusions about the management of this condition.

## Two diseases

I have found it helpful in thinking and teaching about this condition to consider it as two diseases that are marching hand-in-hand. The first is a biologic disease that is described in biologic terms as shown in *Figure 1*. The consequence of the biologic disease is that the femoral epiphysis goes through a period of softness. The second is a mechanical disease that is described using the mechanical terms in the same figure. The mechanical disease takes advantage of the softness with the consequence that the femoral epiphysis loses its sphericity. As orthopaedic surgeons we are limited to altering structure and mechanics in the hope of ameliorating the mechanical disease, but have been powerless in affecting the biologic disease. A possible exception to this is the claim by Joseph<sup>8</sup> that early surgery can speed up the natural history and perhaps skip stages.

Until now all of our treatments of LCP have been based on the principle of containment. It is important to stress that 'containment' is a word used only in LCP, and is different from 'coverage' which is measured by the centre-edge angle. Containment reflects the proportion of the articular surface of the femoral epiphysis that is apposed to the acetabular surface. It makes sense to think that increased containment minimises the loss of sphericity that results from variable pressure on the soft femoral epiphysis. The analogy of a scoop of ice cream on a cone may be illustrative.

Biological Perthes	Mechanical Perthes
<ul style="list-style-type: none"> <li>• Vascular compromise</li> <li>• Cell death</li> <li>• Bone resorption</li> <li>• Vascular ingrowth</li> <li>• Bone formation</li> </ul>	<ul style="list-style-type: none"> <li>• Subchondral fracture</li> <li>• Softening</li> <li>• Collapse</li> <li>• Extrusion</li> <li>• Loss of sphericity</li> <li>• Lateral hinging</li> <li>• Containment</li> </ul>

*Figure 1. LCP can be thought of as two diseases marching hand-in-hand: a biological disease, and a mechanical disease, each with its own terminology*

## Ice cream

A scoop of ice cream that is placed on a cone is perfectly round because it has been formed by the round scoop. It is rigid enough to maintain its round shape against gravity, but if you push on it with your thumb you will form a depression and the ice cream will extrude in other areas of lesser pressure with a consequent loss of sphericity. Similarly, if you push on it with the round scoop it will extrude in areas outside the margin of the scoop even though the part within the scoop will remain perfectly round. If, however, the entire scoop of ice cream is 'contained' within the scoop, it will maintain its sphericity no matter how soft it is and no matter the pressure because there is no route for it to extrude. It is, in other words, perfectly contained.

## Containment

Perfect containment of the proximal femoral epiphysis cannot be achieved. The fact that the hip can move means that even if perfect containment can be achieved in one position it will be compromised when the hip moves to a different position. It is reasonable, however, to adopt a surgical goal of maximally containing the epiphysis in the weight-bearing standing position when maximum joint reaction forces occur.

*Perfect containment of the proximal femoral epiphysis cannot be achieved*

The articular surface of the femoral head is part of a spherical surface that includes not only the epiphysis, but also the physis and part of the metaphysis. The articular surface of the head is larger than the acetabulum with the result that part of the head is always outside the acetabular articular surface. In the context of LCP however, we are concerned with the epiphysis, not the head. The epiphysis occupies about 50% of the head and, since the normal acetabulum is about half of a sphere, it corresponds well to the epiphysis. There is, therefore, some position of the hip in which the epiphysis is fully contained within the acetabulum. The goal of containment treatment is to ensure that this position corresponds to the standing weight-bearing position of the hip.

In the standing position the lateral part of the epiphysis is usually not contained and achieving containment involves changing the relationship of the femoral head to the acetabulum. It is worth noting that, in the normal hip, it is the lateral part of the epiphysis that is uncovered, and that this is the part of the head most consistently involved in LCP.<sup>6</sup> Containment of this part of the epiphysis can, theoretically at least, be accomplished by changing the attitude of the acetabulum (by redirectional pelvic osteotomy), the shape of the proximal femur (by proximal femoral varus osteotomy), or the position of the femur (by casting or bracing).

## Containment and the lateral pillar

Herring,<sup>5</sup> in coining the term and stressing the importance of the 'lateral pillar', is usually given credit for bringing the importance of the lateral part of the epiphysis in LCP to our attention but Somerville expressed this concept many years before.<sup>9</sup> In his important article in 1971 he made the point that 'when part of the ossific nucleus only is affected it is almost invariably the antero-lateral part', and went on to state that:

'...provided the head of the femur is well contained in an undeformed acetabulum it will develop normally even though the ossific nucleus may be in part or in whole ischaemic. ...The aim of treatment must be to see that the mould in which the head is shaped is the right shape when ossification occurs.'

He used the term 'subluxation' to mean much the same as we would use the term 'uncontained' today. We can give Herring credit for pulling these concepts together in the concept of the lateral pillar.<sup>5</sup>

## The future

We have good evidence that surgical treatment can be of benefit in certain patients. We cannot help but suspect that the benefit to other groups is masked by the difficulty in assessing their disease. We must hope that new techniques to evaluate the circulation in the femoral head will improve our classification of this disease in its early stages and facilitate meaningful studies and strong evidence for indications for treatment.

Multi-centre international registries that follow patients into their adult lives will provide functional and radiologic information by which to assess outcomes. We must do what we can to encourage the development of such registries.

Attention is being devoted to the possibility of modifying the biological disease with medications such as the bisphosphonates. It is hopeful that such treatment will help to maintain the strength of the bone and minimise extrusion and deformity.

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