Legg-Calvé-Perthes disease: classifications and prognostic factors

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Summary

***Introduction*. Legg-Calvé-Perthes Disease (LCPD) represents idiopathic avascular necrosis of femoral head in pediatric population. Indications for treatment depend mostly on prognosis about femoral head sphericity and hip congruence at the end of growth. The aim of this review is to highline prognostic factors of LCPD.**
***Methods*. Bibliographic search in PubMed allowed selection of 33 articles concerning prognostic factors and/or classification of LCPD.**
***Conclusion*. Clinical factors of poor prognosis are overweight, female sex, age exceeding 6 years old, and lack of hip abduction. Radiologically, Herring's classification is consensual because of its high prognostic value and very good reproducibility. The other signs of femoral head "at-risk" and the assessment of the reduction in abduction of the femoral head in the acetabulum are also prognostic of late evolution. MRI seems to be a future tool in assessing the fate of hips in LCPD. It is likely that a better understanding of LCPD etiology would precise the prognosis of this disease.**

KEY WORDS: Legg-Calvé-Perthes disease; prognostic factors.

Introduction

Legg-Calvé-Perthes Disease (LCPD) represents idiopathic avascular femoral head necrosis in pediatric population, affecting children aging 2-10 years old. The exact etiology of this disease is not well known. The affected femoral epiphysis undergoes varying degrees of self-limited necrosis, leading to a broad spectrum of pathology and evolution, from mild forms without sequelae to severe deformities with limited range of motion and early osteoarthritis of the hip joint (1-4).
Clinically, LCPD usually presents with limping and localized pain in the thigh and/or knee, occurring after sustained physical activity in a healthy child, and/or deficits in abduction and internal rotation, and Trendelenburg gait in advanced stages.
Plain X-ray is the most current additional diagnostic procedure in order to assess the stage of the disease, the containment of the femoral head in the acetabulum and the extent of capital involvement. The methods of treatment evolved during numerous decades, since the abduction casts and tractions until the advanced methods of surgical containment in order to obtain a spherical congruity of the hip joint, to restore range of motion and to prevent osteoarthritic deformity of the femoral head.
Indications for invasive treatment depend mostly on prognosis, which can be assessed during childhood or adulthood. Numerous factors have been studied in order to determine their involvement in the prognosis of LCPD
The aim of this review is to highline prognostic factors of LCPD.

Methods

Computerized bibliographic search in PubMed was performed using keywords as follows: "osteochondritis of the hip" or "perthes" or "coxa plana" and "prognosis" or "prognostic factor" or "classification". Articles published after 2000 concerning prognostic factors and/or classification of LCPD were selected. On 485 studies, we selected 21 for systematic review. We manually added 12 historical major articles.

Main criteria of judgment

The overall prognosis depends on hip congruence at the end of growth, which directly influences degenerative changes and osteoarthritis in adulthood (1). Therefore, all possible prognostic factors are assessed during childhood on their influence on femoral head shape and hip congruence at the end of the growth, which is usually classed according with Stulberg (1) (Figure 1) (Table 1), in 5 stages. This classification is strongly prognostic of the long-term evolution of the hip. Unfortunately, it does not help to the choice of treatment.
Mose (2) also proposed a classification of the femoral head at the age of 16 years, prognostic of the long-term evolution of the hip. It is based on the shape of this head.

- Spherical head: good prognosis (Figures 2-4)
- Spherical but crescent-shaped head: fair prognosis
- Non-spherical head: poor prognosis (Figures 5-9).

We hereby list prognostic factors during childhood.

Table 1 - Stulberg classification.

Class	Description	Radiologic aspect	Prognosis
I	Spherical congruency	Normal	Good
II	Spherical congruency; Loss of head shape <2mm	Spherical head with one or more of the following findings: coxa magna, short femoral neck, upper located great trochanter, obliquus acetabulum	Good
III	Aspherical congruency; Loss of head shape >2mm	Non-spherical head but not flat	Mild-to-moderate arthritis
IV	Aspherical congruency	Flat head and acetabulum	Poor: moderate arthritis
V	Aspherical incongruency	Flat head, normal neck and acetabulum	Bad: severe early arthritis

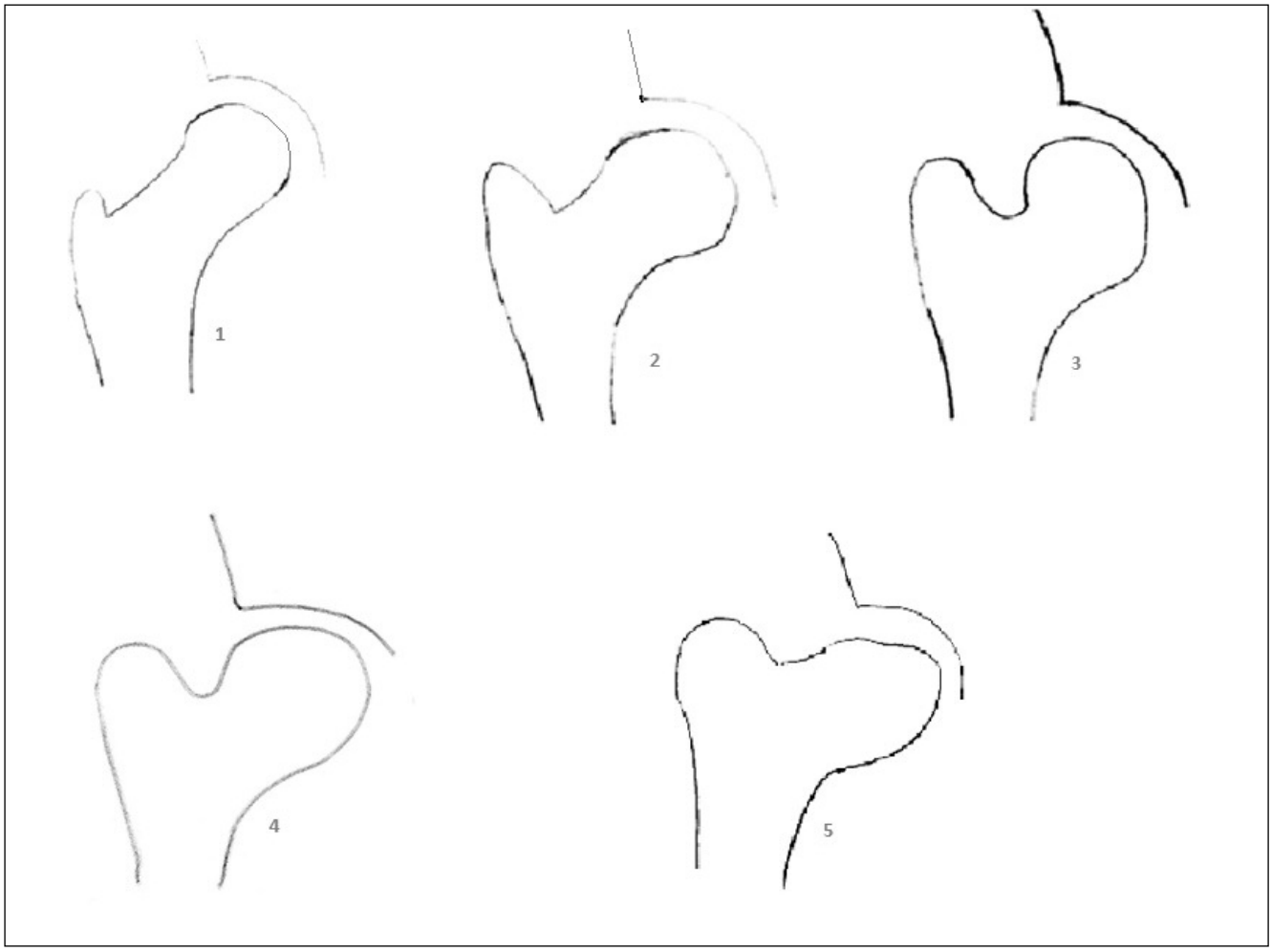

Figure 1 - Stulberg classification.

I. Clinical prognostic factors

A. Age

This is the main prognostic clinical factor. In the literature, the border age between good and poor prognosis oscillates between 6 and 9 years (3-8). It actually concerns more the skeletal age than the chronological age. For Herring in 2004, the limit beyond which the prognosis is worse is a chronological age of 8 years or bone age of 6 years.

B. Gender

Female sex is a non-consensual risk factor for poor progno-

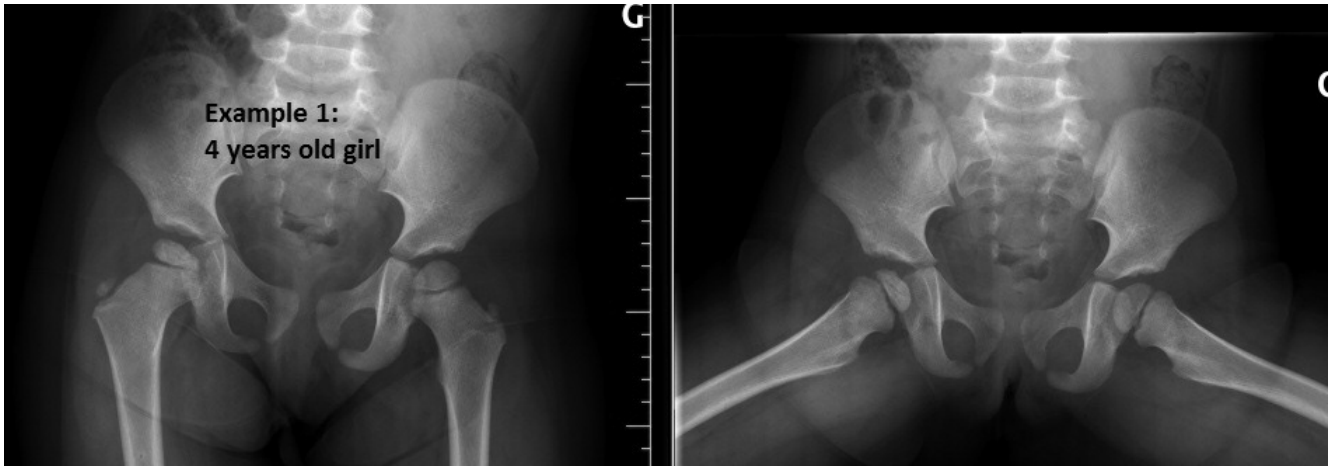

Figure 2 - 4 years old girl, pelvic X-Ray showing osteochondritis, Catterall 1, Herring A.

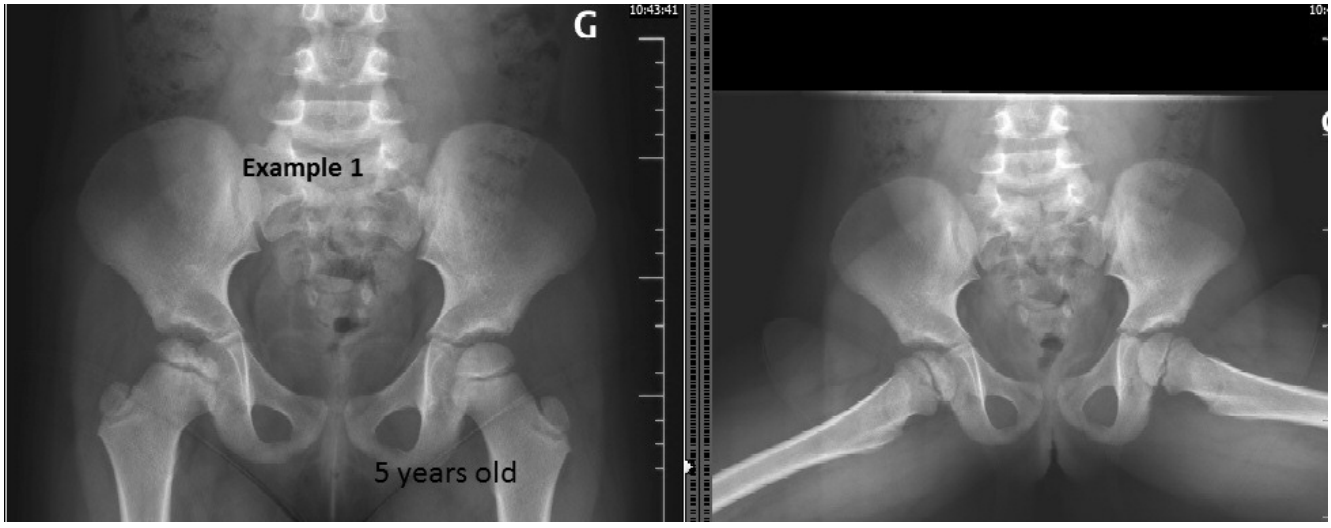

Figure 3 - The same patient, 1 year later.

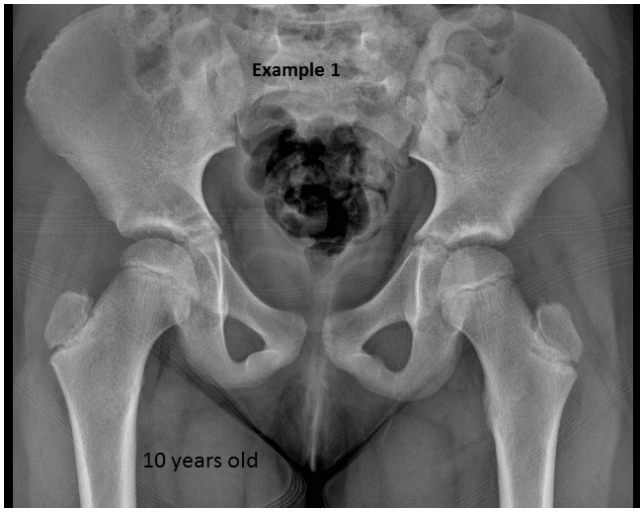

Figure 4 - The same patient at last follow-up, restitutio ad integrum, Stulberg I.

sis (4-6). There is a bias associated with the advancement of bone maturation in girls compared to boys of the same age, reducing the duration of remodeling by equal age (4, 5).

C. Overweight
It is a risk factor for poor prognosis (9, 10).

D. Clinical examination
Hip stiffness and especially contraction in adduction or lack of abduction are pejorative factors (9, 10).

II. Radiological prognostic factors

A. The extent of necrosis
This is the main radiologic prognostic factor of final Stulberg grading (11). It is also a prognostic factor of length discrepancy of the lower limbs (12). It is evaluated according to different classifications as follows.
- Salter and Thomson classification (13) (Figure 10) (Table 2)
This classification is based on the extension of the chondral fracture line. It must be assessed after 4 months of evolution

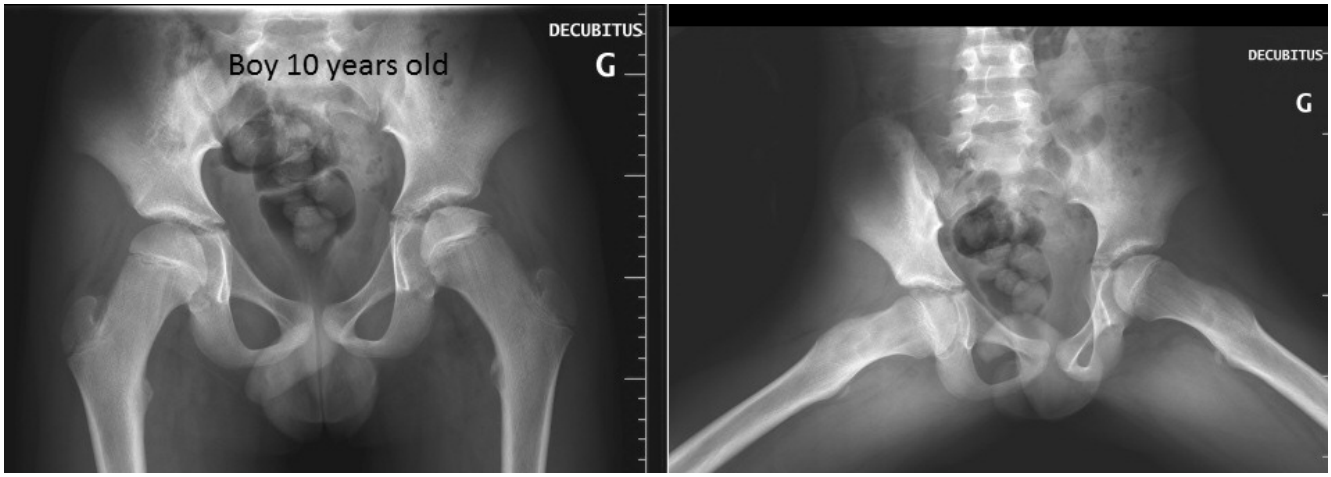

Figure 5 - 10 years old girl, X-Ray showing osteochondritis, Catterall 1, Herring B.

Figure 6 - The same patient, MRI showing osteochondritis.

Table 2 - Salter and Thomson Classification.

	Extent of subchondral fracture line	
A	A1	Medial half on AP view
	A2	Medial half on lateral view
B	B1	Lateral half on AP view
	B2	Whole head

of the disease, on hip antero-posterior and lateral X-ray. This accuracy of the date of realization makes it difficult to be applied in common practice. In addition, the chondral fracture line is often demanding to assess.

- Catterall classification (3, 9) (Figure 11) (Table 3).

It is the oldest classification (1971) and encompasses 4 stages, each of which is associated to a prognostic value (9). In 2008, a modified Catterall classification was proposed, due to poor reproducibility of the original one (6). It joins stage 1

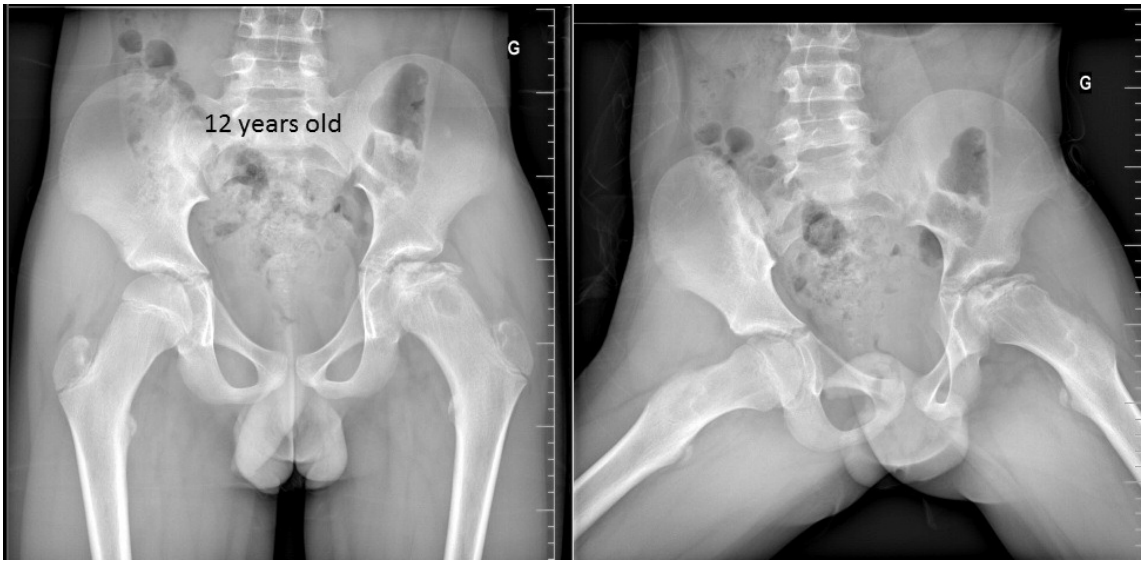

Figure 7 - The same patient 1 year later, Catterall 3-4, Herring.

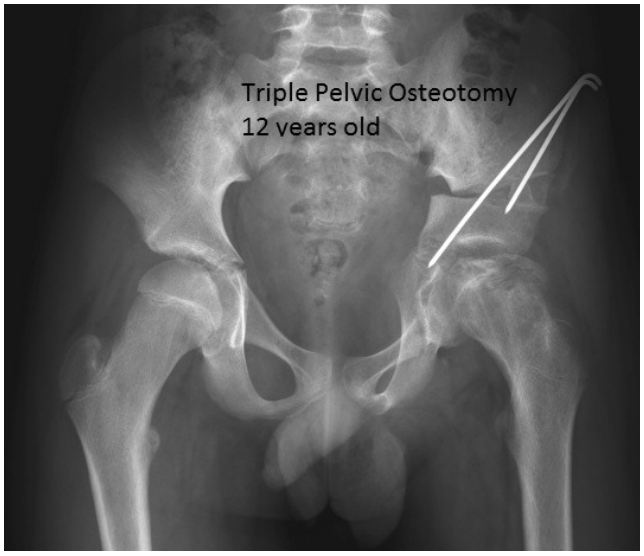

Figure 8 - The same patient after triple pelvic osteotomy.

height (> 50%), but thin (2 to 3 mm), or with decreased density, or lower than the central pillar.

In 2009, another modification of this classification (7) was published, dividing Stages C into Stage C1, involving <75% of the lateral pillar, and C2, involving > 75%. The prognostic value of this classification has been confirmed since (5, 6, 8, 15, 16). It is also the classification with the best intra- and inter-observer reproducibility (17).

- The Growth Plate Involvement, GPI (18)

The total width of the epiphysis (entire W) and the width of the affected epiphysis (affected W) are measured on the frontal radiogram and on the Lauentstein view.

The GPI is then calculated as follows:

GPI = (affected W / entire W) on AP x (affected W / entire W) on Lauenstein.

GPI <0.25 (type I) is associated with a Herring group A or B.

GPI ≥0.25 (type II) is associated with a Herring B/C or C group.

The inter and intra-observer reproducibility of this index is very high (17).

B. Other signs of hip "at-risk". They are typically 5 as follows (9):

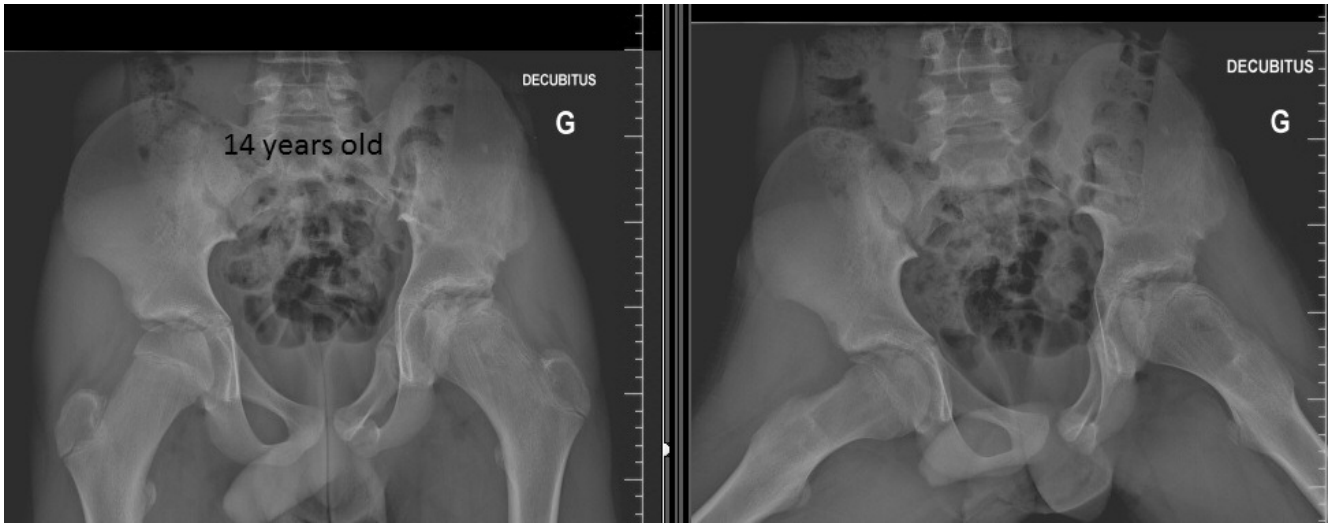

Figure 9 - The same patient at last follow-up, Stulberg V.

with 2 (> 50% necrosis) and stages 3 with 4 (<50% necrosis). The Author of this classification reveals a highly significant influence to the final result according to Stulberg. However, in the same year, another Author (14) demonstrates the importance of differentiating stages III and IV of Catterall classification, since prognosis is significantly different.

- Herring classification (4) (Figure 12) (Table 4)

It was published in 1992, because of the poor reproducibility of that of Catterall. It is also known as the "lateral pillar classification".

For its Author the femoral head is divided into 3 pillars:

- The lateral pillar represents the 15 to 30% of the lateral part of the head
- The central pillar, the central 50%
- The medial pillar, the medial 20-35%.

This classification distinguishes 3 groups according to the extent of the involvement. Herring further describes a B/C border stage, characterized by a lateral pillar of conserved

- The presence of a clear lateral image (sign of "V de Gage"),
- The eccentricity of the head or lateral subluxation
- The horizontal position of the conjugal plate
- The presence of a metaphyseal reaction
- The presence of lateral calcifications.

The lateral subluxation can be evaluated by the acetabular head index described by Heyman (19) with a threshold value between 86% (20) and 90% (21). This AHI is calculated by the A/B ratio (Figure 4).

C. Reduction in abduction

It is appreciated by X-ray in abduction position, in order to search for a conflict and to check the reducibility of the head in the acetabulum. It is a good prognostic factor. The criteria for reduction were published in 1991: the widening of the median space by at least 2 mm, and the reduction of the super-lateral space (22).

Table 3 - Catterall classification.

Grade	Description	Prognosis
Grade I	Very anterior involvement of epiphysis No metaphyseal involvement	Excellent prognosis without treatment at any age
Grade II	Anterior involvement < 50% Possible metaphyseal involvement	< 4 y.o.: good prognosis > 4 y.o.: 50% of good prognosis without treatment
Grade III	Anterior involvement > 50% Frequent metaphyseal involvement	Poor prognosis
Grade IV	Total epiphyseal involvement Metaphyseal involvement	Poor or bad prognosis

III. MRI

Publications about its value in the assessment of LCP disease are multiplying. However, it appears that its interest is still limited to clinical research protocols (23). A publication confirms the feasibility of MRI with Gadolinium injection in this indication (24).

MRI with Gadolinium makes it possible to evaluate the severity of the epiphyseal lesion very early, before the fragmentation phase. There is a correlation between the percentage of residual perfusion of the epiphysis and the level of the infusion index at this early stage and the evolution towards the Herring classification (which occurs later). A low residual percentage and a low perfusion index correlated with a greater deformation of the femoral head at 2 years of mean recoil (25, 26).

Table 4 - Herring classification.

Group	Description	Prognosis		
Group A	No involvement of lateral pillar	Stulberg 1 et 2 in 100% of group A patients		Excellent prognosis
Group B	> 50% of lateral pillar height maintained	<9 years	Stulberg 1 and 2: 92%	Fair prognosis
		> 9 years	Stulberg 2: 30% Stulberg 3: 50% Stulberg 4: 20%	Uncertain / poor prognosis
Group C	Involvement of lateral pillar height > 50%	Stulberg 2: 13% Stulberg 3: 52% Stulberg 4 or 5: 35%		Poor prognosis

Table 5 - Arthrographical classification according with Laredo.

	Relation between acetabulum and femoral head
I	Normal
II	Spherical Coxa magna. Subluxation in neutral position, which reduces at 30° of abduction
III	Egg-shape Coxa magna. Persistent subluxation at 30° of abduction
IV	Flat Coxa magna; subluxation at 30° of abduction; impingement on abduction
V	Deformed Coxa magna; subluxation at 30° of abduction

An increase in Apparent Diffusion Coefficient (ADC) and Diffusion Weighted Imaging of the femoral metaphysis relative to the contralateral (healthy) hip appears to be related to a more severe Herring stage (27). On the other hand, there is no link between these parameters (ADC and DWI) and the Herring class.
Finally, the “acetabular head index” described on the radiograph by Heyman can be transposed to MRI images, with a threshold value of 77% indicating a shedding of the femoral head (20). The benchmarks for calculating AHI are then cartilaginous (Figure 5).
However, there is no evidence that visible subluxation earlier on MRI than on X-rays is a significant factor in poor progno-

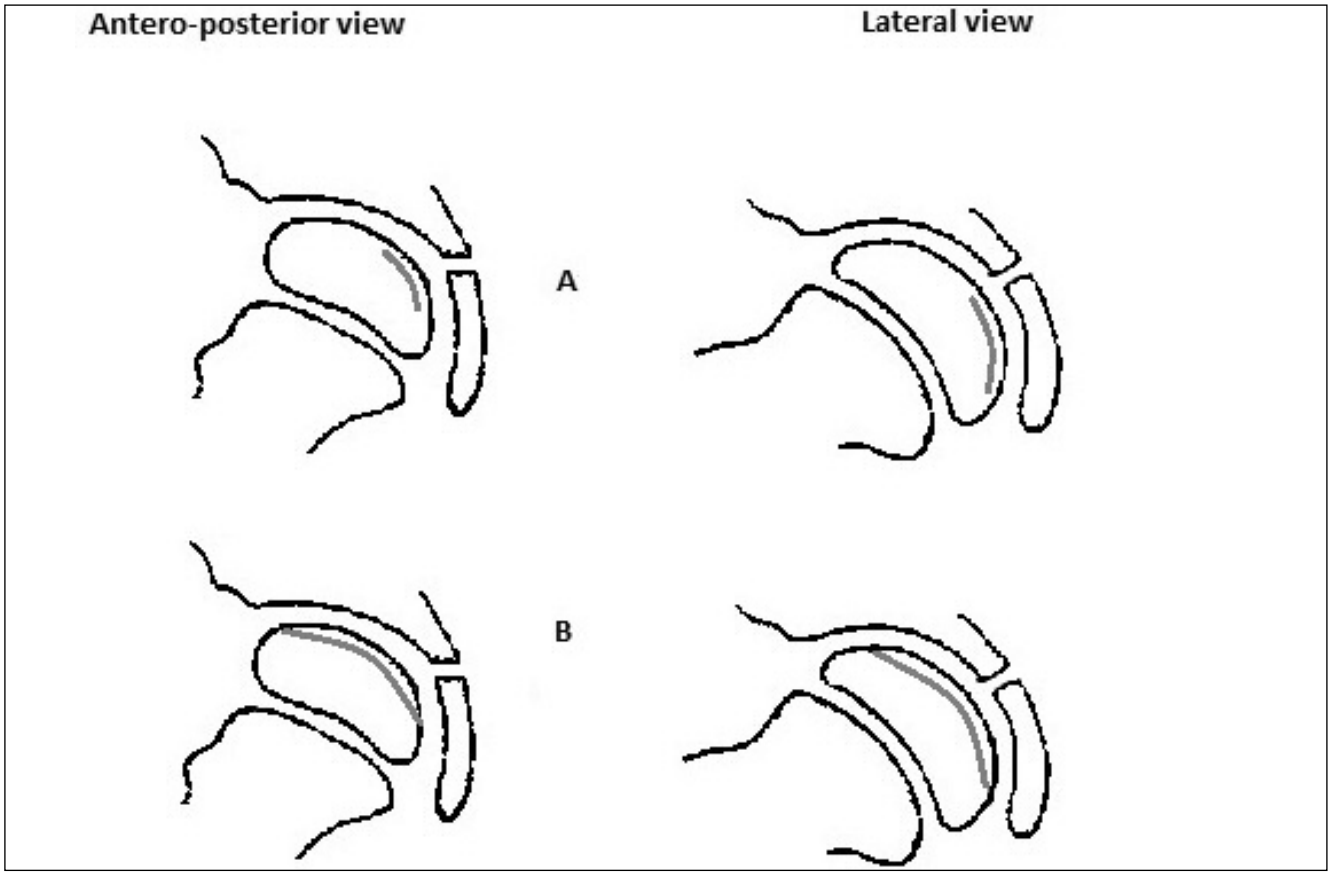

Figure 10 - Salter and Thompson classification.

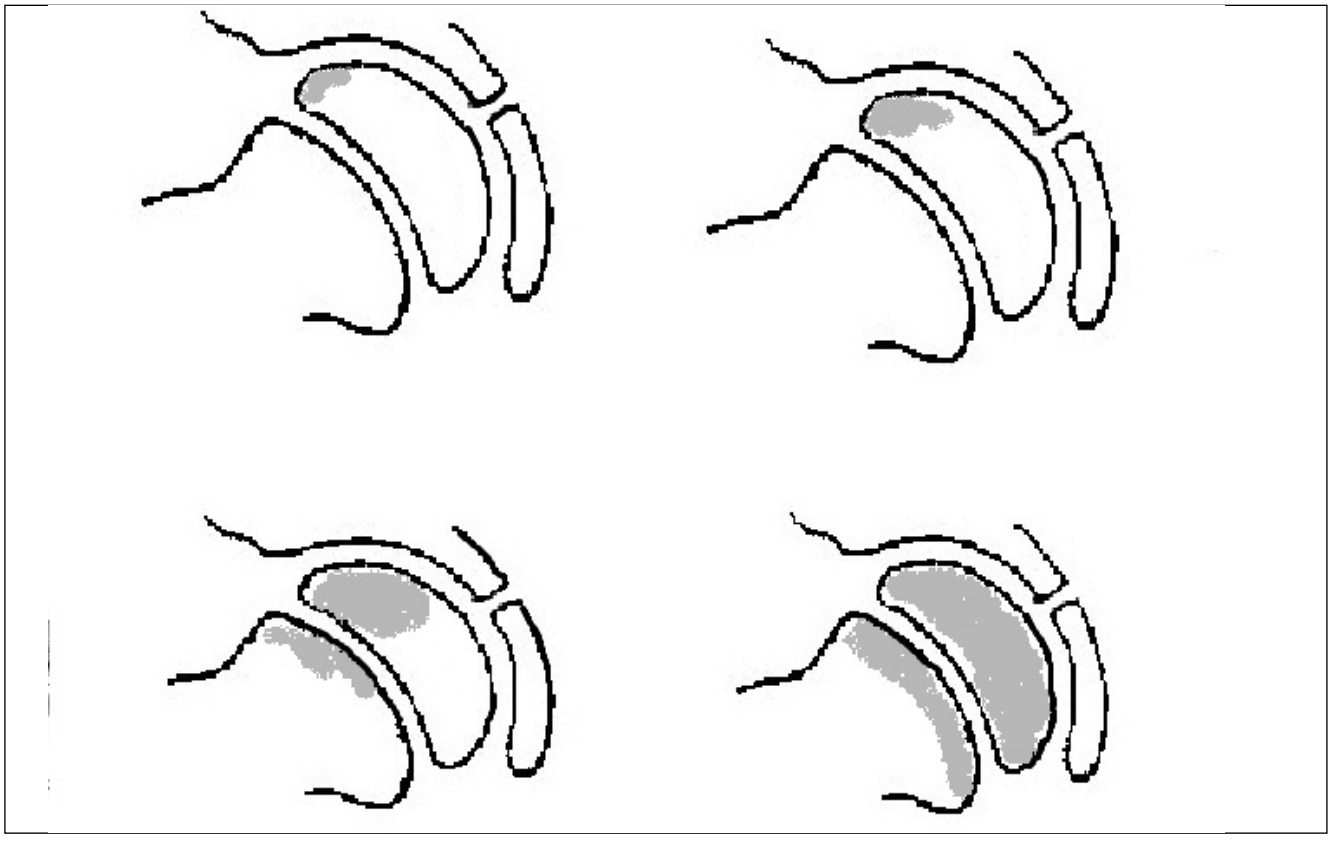

Figure 11 - Catterall classification.

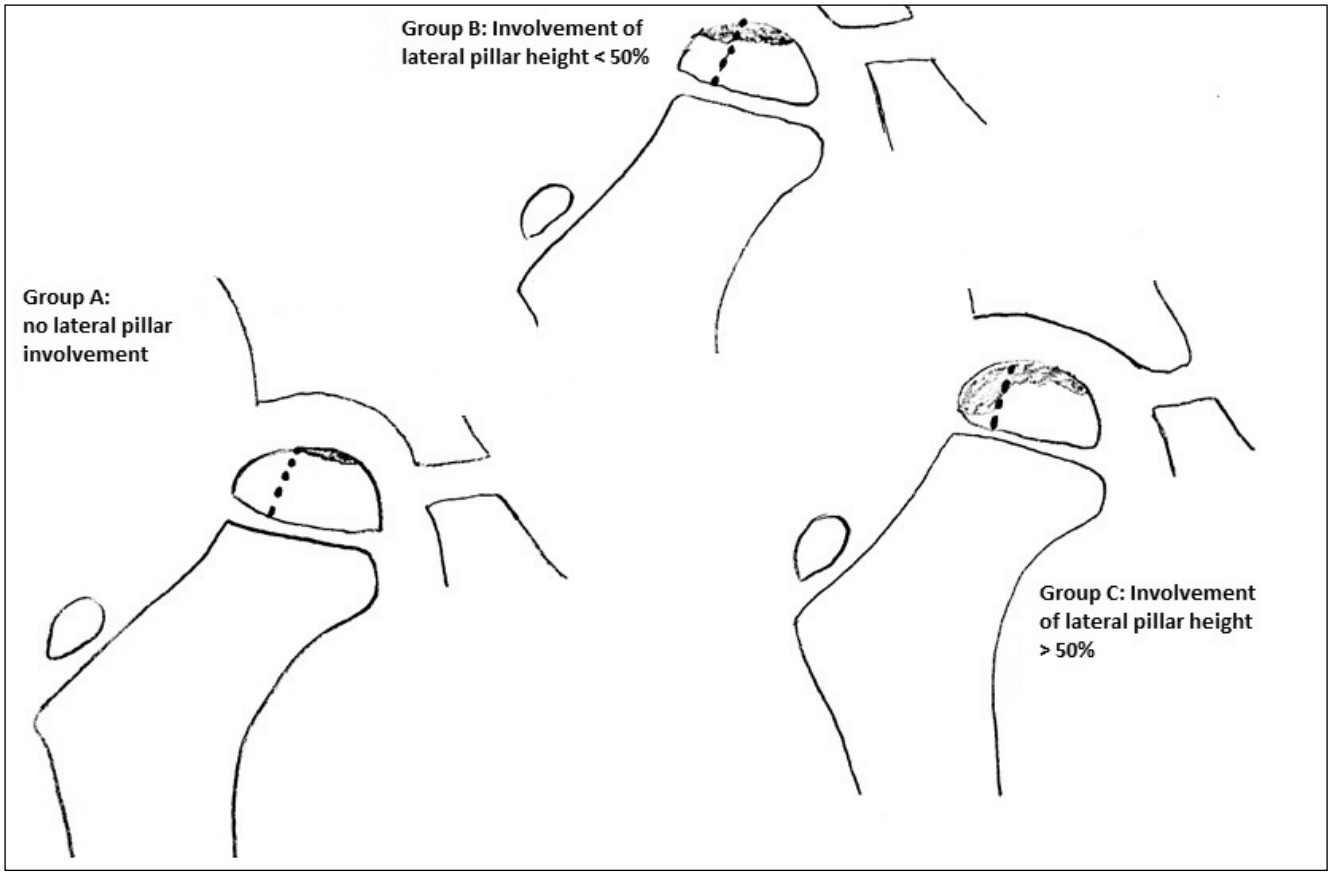

Figure 12 - Herring classification.

sis, as this subluxation is related in part to the thickening of the medial articular cartilage. This thickening, as the importance of intra-articular synovitis increases and then decreases as the disease progresses, thus does not represent a significant prognostic factor (28, 29) but simply a marker of LCPD progression.

IV. Arthrography

The classification of Laredo (30) (Table 5) has a high prognostic value (31). Nevertheless, its difficulties of realization make it little usable in clinical routine.

V. Scintigraphy

It has no prognostic value. It therefore has no indication here (32).

Conclusion

From the above, one can summarize that LCPD presenting with stiffness in an overweight girl, over the age of 6 years old is likely to have a poor prognosis at the end of growth and in adulthood.

Radiologically, Herring's classification is consensual because of its high prognostic value and very good reproducibility. The signs of "at-risk" femoral head and the evaluation of the reducibility in abduction of the femoral head in the acetabulum must be considered.

MRI seems to be a future tool in assessing the fate of these hips, studying more precisely the extent of necrosis. However, no specific prognostic elements have yet been described. Our understanding of the natural history and causes of LCP disease is still limited, and it is likely that a better understanding of its causes will precise the prognosis of this disease (33).

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