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Prognostic Evaluation of Legg–Calvé–Perthes Disease by MRI Part II: Pathomorphogenesis and New Classification

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Summary: The most widely used system of grading Legg–Calvé–Perthes disease (LCPD) is still the radiographic grouping by Catterall, although it lacks interobserver reliability, especially in the early stages. The predictive value of Salter–Thompson classification and Herring's "lateral pillar" classification are still being studied. In Part 1, we demonstrated the good reliability and the predictive value of four magnetic resonance imaging (MRI) indices (extension of necrosis, lateral extrusion, physeal involvement, metaphyseal changes) through their correlation to clinical and radiographic conditions of the hips at follow-up. The same good results were obtained by submitting to statistical analysis a second group of 31 patients (French series). On the basis of these statistical studies, a new classification has been proposed. It takes into account the extent of necrosis and two MRI risk signs: lateral extrusion and physeal involvement. The extent of necrosis up to or more than 50% separates two main groups, A and B. The associated MRI risk factors distinguished six classes with different prognoses. Appropriated treatment also is suggested for each class. Our experience on MRI in LCPD led us to draw a pathomorphogenetic model called "packed capsule." According to this biomechanical model, the femoral head is considered a segment of a sphere made of viscoelastic material and hermetically sealed. The deformation of the head depends on the behavior of the necrotic fluid collected inside the capsule under the weight-bearing forces. Finally, our suggestion in the treatment of Perthes disease is to relieve weight bearing up to the fragmentation stage, whether the diagnosis has been made by the use of MRI or without it. During the fragmentation stage, MRI is extremely useful in performing prognosis; at this time our classification can be applied, and the corresponding treatment can be followed. **Key Words:** MRI and Perthes disease—Perthes disease—Prognostic classification.

Most of the studies (3,4,12) on the natural history of Legg–Calvé–Perthes disease (LCPD) proves that the great majority of the cases (60–70%) heal spontaneously without functional impairment at maturity. There also is evidence (10) that a considerable number of functioning hips at the fifth decade become painful and require arthroplasty during the sixth decade. It is reasonable that these late-degraded results concern those hips that were considered well healed at a follow-up of 10–30 years for their good clinical condition, although an imperfect radiographic outcome was present. Consequently, at the very long term follow-up (40–50 years), the percentage of good results is much lower than observed at the maturity follow-up. Prognosis in LCPD is therefore of great importance in selecting patients to be treated and in choosing the type of treatment.

Since the publication by Anthony Catterall in 1971 (3), orthopedists have been prognosticating LCPD on the basis of his radiographic classification and head-at-risk signs, although successive studies (5) have demonstrated the poor intra- and interobserver reliability and the late appearance of the characteristic fragmentation. Decision making about LCPD also is conditioned by its long duration and by the concern about the consequences that a prolonged orthopedic treatment might have on a child's psychology. Moreover, Herring (6) in 1994 demonstrated that even the literature of the last three decades concerning the treatment of LCPD is inconclusive, because it lacks scientific rigor. He underlined that there is a bias in the selection of the patients who are included in the study and that this selection was based on Catterall classification, which itself lacks interobserver reliability.

Other radiographic classifications have been proposed in the last years to simplify the interpretation of the radiographic pictures and reduce the interobserver error. Unfortunately even these classifications have some drawbacks that make them unreliable. The subchondral fracture, on which the Salter–Thompson classification is based (13), is rarely encountered in the very early and in the quite late stages of the disease. The evaluation of the lateral pillar height by Herring et al. (7) is difficult in very young patients and impossible in bilateral cases.

Arthrography, taking into account the flattening and

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the extrusion of the cartilaginous epiphysis, may give very useful prognostic information even in the early stages of LCPD, but it is an invasive and unrepeatable procedure.

Bone scan also is useful in the early stages the better to quantify the extent of epiphyseal ischemia and to predict the severity of necrosis, but it does not allow appreciation of the shape of the femoral head and its relationship with the acetabular fibrocartilage. Moreover, it is not routinely used because of the radiation risk.

The most common tools do not allow investigation of the pathologic alterations of the various parts of the femoral head and the changes in physical status (mechanical resistance, consistency, etc.) The role of the ischemic process is universally accepted as a pathogenetic factor, but the load also has been considered of relevant influence (2). Therefore we think that a prognostic classification should take into account both the biologic effects of the ischemia in a certain phase of the disease and the mechanical consequences of the weight borne until the time of the clinical onset.

The various radiographic aspects of Catterall classifications, actually defining the groups of gravity and the risk signs, are described as pathologic transformations of the bony epiphysis being part of the natural history of the disease. In this sense they do not consider the influence of the load as an extrinsic factor. Moreover, Catterall groups and risk signs tell us very little about physical changing and about the pathomorphologic alterations of the femoral head. Since the end of the 1980s, MRI has been used in LCPD, and many authors have demonstrated its usefulness in detecting the epiphyseal necrosis in the early stages and in showing details of different hip joint structures.

THE "PACKED CAPSULE" MODEL

Some of the mentioned morphologic details have contributed to formulate a new interpretative model about the general process that leads to the onset of the deformity. This model considers the interaction of the two mentioned factors: the intrinsic factor (ischemia–necrosis) and the extrinsic one (the load).

From the literature we know that

- even in the severe and advanced forms of LCPD, the epiphyseal cartilage undergoes very few alterations consisting of a slight thickening;
- according to Mitchell's MRI studies (11), a cavity with a low-intensity signal in T_1 (Fig. 1A) and high-intensity signal in T_2 (Fig. 1B) contains a watery fluid;
- the growth cartilage is thinner than the epiphyseal cartilage: at MRI, growth cartilage needs high-resolution techniques to be distinguished from the very low intensity signal bands (in T_1) related to the subchondral bone epiphysis and metaphysis (Fig. 2);
- we have never observed ruptures of epiphyseal cartilage but only distortion (sometimes severe) in its shape. Conversely, the growth cartilage often had lesions of variable extension; and
- at MRI, metaphyseal cysts are nearly always associated with growth cartilage lesions.

Some years ago, Mercer-Rang (14) used a model called "ice-cream scoop" to explain the progressive femoral head deformation in LCPD, according to which this is crushed between the acetabulum and the neck, extruding laterally like an ice-cream ball between the scoop and the cone. This model is based on the principle that the epiphyseal necrosis transforms the solid mass of the femoral head into a soft and pasty mass whose physical behavior is that of a nonnewtonian high-viscosity liquid.

Our new interpretative model of the biomechanical pathogenesis, called packed capsule, is substantially different from that of Mercer-Rang and is explained as follows (Fig. 3).

In a child the femoral head is physically comparable to a segment of a sphere of composite material (cartilage, bone trabecular lamellae made of organic mineralized matrix, marrow cells, intra- and extravascular fluids) with a smaller segment of a solid sphere above transmitting the load (acetabulum). The chondroepiphysis is considered as a shell in the form of 4/7 segment of a sphere, made of viscoelastic material with a thicker convex surface above (epiphyseal cartilage) and a thinner plane or

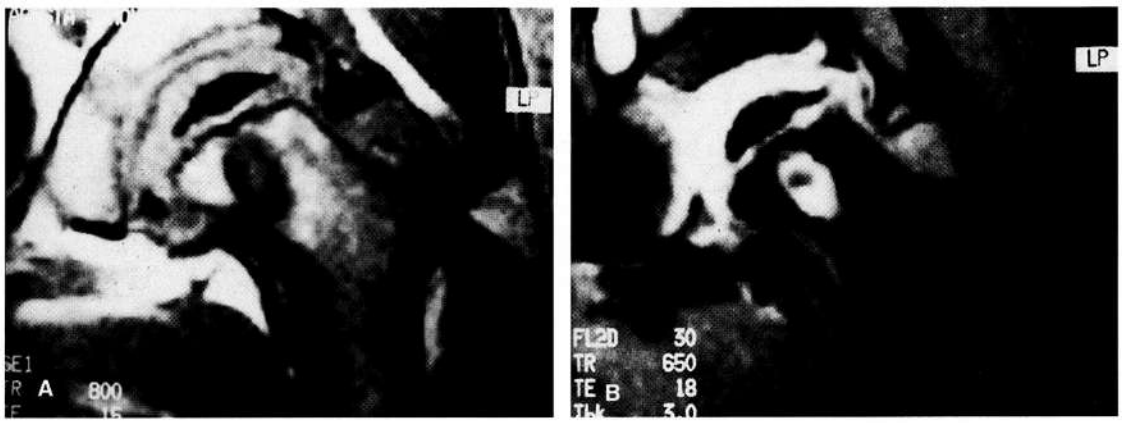

FIG. 1. A: Large metaphyseal cyst in a 5-year-old boy with LCPD: T_1-weighted coronal view. **B:** The same view in T_2-weighted scan: high-intensity signal indicating a watery fluid.

FIG. 2. High-resolution T_2 gradient-echo images of normal hip in a 4-year-old child: the cartilaginous layer of the physis is thinner than the epiphyseal one.

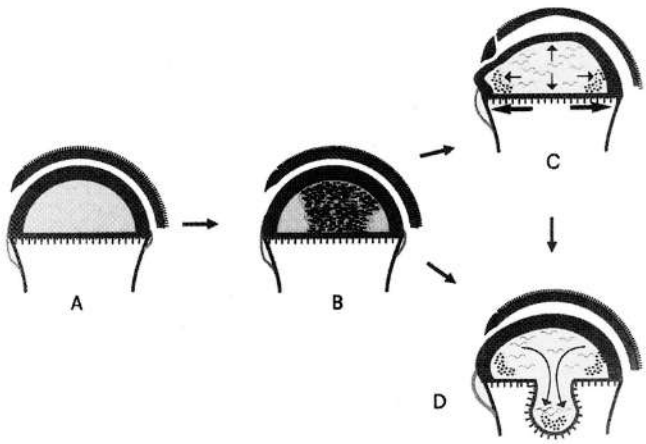

FIG. 3. The "packed capsule" model. The chondroepiphysis is considered as a shell made of viscoelastic material (cartilage) whose spherical form is supported by the trabecular bony frame **(A)**. The shell remains hermetically sealed and impermeable, even in pathological conditions and under load pressure. When the necrosis is >50%, the necrotic fluid deriving from the liquefaction of fatty marrow will be collected inside the capsule and increases its pressure as a consequence of load and for the lack for trabecular neutralization **(B)**. The increased pressure inside the capsule causes the deformation of the sphere and the extrusion of the epiphysis anteriorly and laterally; the increase of the growth plate diameter with distention of the perichondral ring (arrow) **(C)**; and the stressing of the thinner portion of the capsule, the growth cartilage, with possible rupture into the metaphysis, transphyseal connection with fluid transfer from high-pressure compartment to low-pressure compartment, with possible excavation in cysts **(D)**.

slightly curved surface below (the physis). In normal and pathologic conditions, the shell integrity is preserved, remaining hermetically sealed and practically impermeable even under high and brief pressures (for example, the standing phase). The sphericity in the superior convex part as well as the dimensions of the diameter of the inferior part (growth plate) are guaranteed by the trabecular frame of the epiphysis. Repeated epiphyseal ischemic episodes cause osteocyte and bone marrow cell necrosis. When it is applied to a restricted portion ($\leq$50%), the residual trabecular frame will be resistant to load, and the viscoelastic shell will not undergo any deformations. When the necrosis is more extended, the necrotic fluid, produced by the liquefaction of the fatty marrow, will be collected inside the sealed capsule. This fluid raises its pressure as an effect of the load and for the lack of trabecular neutralization.

The consequences of the increased pressure of the necrotic fluid inside the cartilaginous capsule are according to a newtonian fluid behavior:

- the deformation into a segment of an ellipse of the segment of a sphere. This deformation is asymmetric because the expansion is toward the uncovered portions (anterolaterally);
- the increase of the mean diameter of the growth plate due to rolling of the external part of the shell on to the metaphysis (like a track of a tracked vehicle);
- the distention of the perichondral ring (groove of Ranvier);
- the stressing of the thinner portion of the shell—the growth cartilage—with possible rupture into the metaphysis. The lesion may vary from piercing to the total derangement of the tissue architecture. Transphyseal connection causes the fluid transfer from the high-pressure compartment (epiphysis) to the low-pressure compartment (metaphysis). This may explain the cystic excavation within the metaphysis and transphyseal bony bridging. The hydraulic model of the packed capsule allows us to explain some relevant clinical data:
- the younger the child, the better the prognosis: the growth plate is thicker in a younger child, and the amount of the necrotic fluid is relatively less;
- the statistical correlation, in our study, between the extent of necrosis and the severity of physeal involvement; and
- the incontrovertible effectiveness of weight-bearing relief realized since the early stages of the disease by reliable methods such as bed rest and plaster cast (2).

The concept of packed capsule is valid in the early stages of LCPD when trabecular derangement and marrow necrosis are prevalent. In a successive phase, with the onset of the regeneration process and of the formation of fibrocartilaginous tissue, the involved part of the femoral head will be deformed by the load, as a semisolid and plastic material. The weight-bearing pressure

TABLE 1. *Correlation to Stulberg class and total score for the 31 hips*

	Stulberg class		Total score	
MRI signs	S	*p*	S	*p*
EXT	0.70	<0.001	0.65	<0.001
LAT	0.74	<0.001	0.84	<0.001
PHY	0.86	<0.001	0.88	<0.001
MET	0.66	<0.001	0.71	<0.001

S, Spearman coefficient; *p*, statistical significance.

TABLE 2. *Prognostic classification of LCPD by MRI*

Group	Class	*n* (%)	MRI risk factors	Risk	Prognosis	Treatment
A: necrosis ≤50%	A0	9 (16)	PHY.0–LAT.0	No risk	Very good	No treatment
	A1	7 (12)	PHY.1–LAT.0 or PHY.0–LAT.1	Slight decreasing of epiphyseal index	Good	No treatment. Limitation in sport and physical activities
B: necrosis >50%	B0	1 (2)	PHY.0–LAT.0	Transient class: evolution to a more severe B class in a successive MRI	One of the new B class	Unloading until sucessive MRI
	B1	11 (18)	PHY.1–LAT.1 or PHY.0–LAT.1 or PHY.1–LAT.0	Coxa magna, short neck, decreased epiphyseal height	Fairly poor	Unloading (bed rest, plaster cast)
	B2	14 (24)	PHY.1–LAT.2 or PHY.2–LAT.1	Coxa plana, incongruity	Poor	Weight-bearing relief in containment position. Containment surgery after age 7
	B3	17 (29)	PHY.2–LAT.2	Hinge abduction, subluxation	Very poor	Early containment surgery

PHY.0–LAT.0, no MRI risk signs; PHY.1–LAT.1, minor MRI risk signs; PHY.2–LAT.2, major MRI risk signs.

forces, according to the fluids laws, will be concentrated in the restricted loading zone. This is typical of the wedge action of the acetabular edge on the lateral portion of the femoral head—more or less extruded—that is able to inhibit the regeneration and the enchondral ossification of the epiphysis, to damage the physis beneath and to generate the epiphysiodesis and then the asymmetric growth of the neck. The eccentric site of epiphysiodesis also determines an unbalanced growth of the epiphysis that develops aspherically (1). It is well known that a

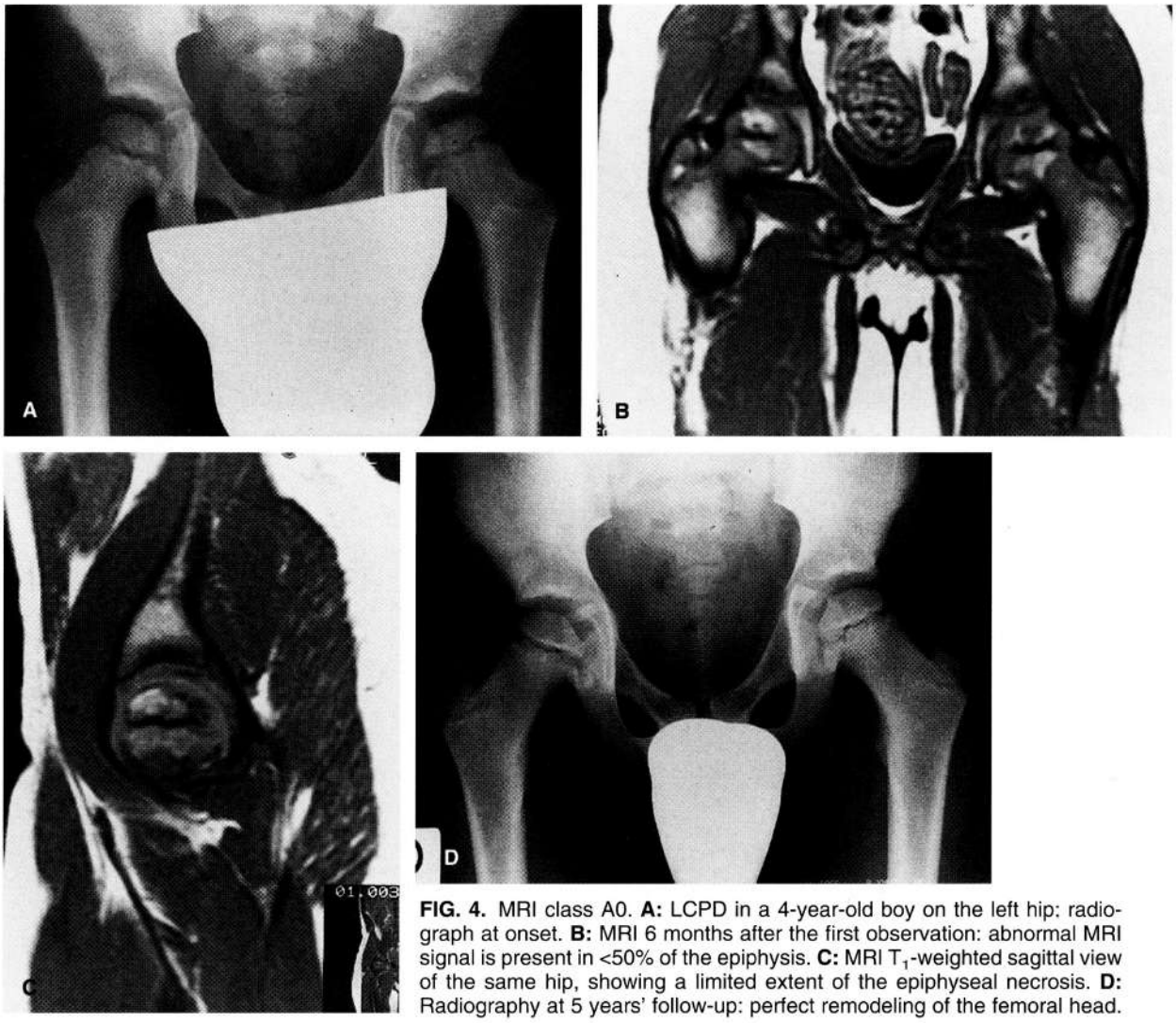

FIG. 4. MRI class A0. **A:** LCPD in a 4-year-old boy on the left hip: radiograph at onset. **B:** MRI 6 months after the first observation: abnormal MRI signal is present in <50% of the epiphysis. **C:** MRI T_1-weighted sagittal view of the same hip, showing a limited extent of the epiphyseal necrosis. **D:** Radiography at 5 years' follow-up: perfect remodeling of the femoral head.

subsequent increasing radial growth. If the head is maintained contained, with a fair range of motion and weight-bearing free, the severe flattening can be prevented.

In class B3, vertical growth potential of epiphysis and neck is inhibited by the large lesion of the physis (Fig. 9). Unbalanced growth between the covered (and crushed) portion of the head and the extruded portion, with residual or normal growth potential, leads to hinge abduction. Early containment surgery must be performed to avoid this catastrophic evolution.

CONCLUSION

Our experience with MRI in LCPD is based on its use in all observed patients for the last 10 years. It has been strengthened by a statistical correlation study between MRI risk signs and clinical–radiographic condition at follow-up on our 28 hips and on 31 hips offered by University of Montpellier. This experience induced us to draw a pathomorphogenetic model (packed capsule) that agrees with modern scientific knowledge. This model cannot answer all the points questioned by the physicians about LCPD, but it is a way to explaining the majority of clinical and radiologic findings.

Our prognostic classification proceeds both from a statistical correlation study and from the concepts held in the biomechanical model we proposed.

Although a primary ischemic injury of the physis cannot be *a priori* excluded, this possibility still remains unascertained, whereas the packed capsule model explains the secondary lesion due to the load. Physeal lesion and lateral extrusion are therefore caused by the combined action of necrosis and load, and they themselves are the intermediate factors of the ultimate damage to femoral head.

There are many controversies about the appropriate treatment of LCPD depending on the difficulties encountered in evaluating the results of a given type of treatment (6). The purpose of this classification is to provide guidelines based on the prevention of the worsening in physeal involvement and in lateral extrusion by means of weight-bearing relief (class B1) and on the prevention of

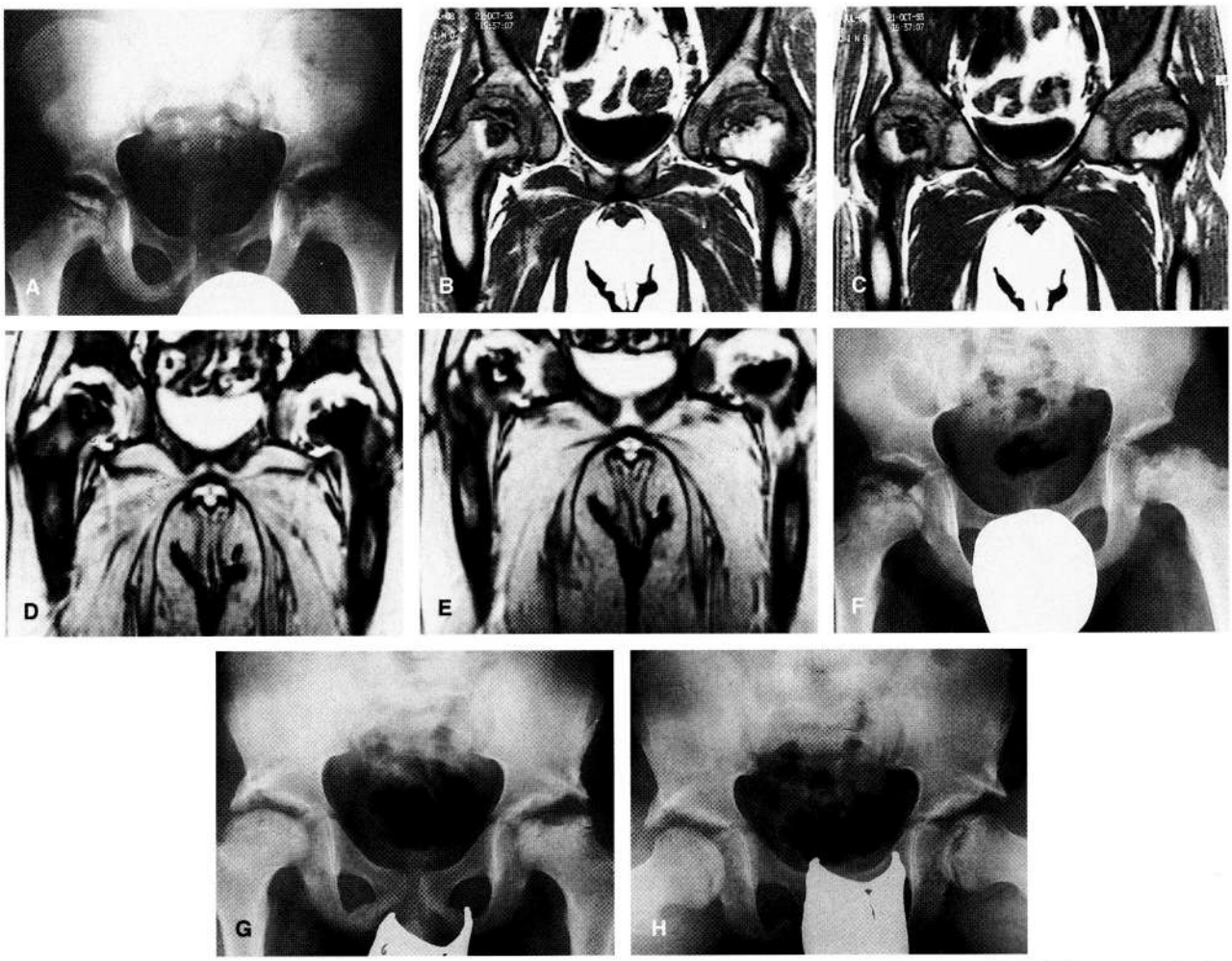

FIG. 9. MRI class B3. **A:** Radiographic picture at onset of bilateral Perthes disease in 5-year-old patient. **B:** MRI T_1 scan: in both hips, there is a severe flattening with lateral extrusion of the epiphysis. On the right hip, a wide metaphyseal cyst is evident, whereas on the left hip, an irregular undulation of the physeal band appears. **C:** Sequential MRI T_1 scan confirming the previous observation. **D:** MRI T_2 scan shows the right hip intrusion of the epiphysis into the metaphysis and a watery cyst in a wide metaphyseal excavation; on the left hip, a large epiphysiometaphyseal fusion is evident. **D:** AP radiograph 1 year later. **E:** Radiographic aspect at follow-up after bilateral shelf procedure. Centrolateral epiphysiodesis and large epiphyseal defect are evident on left hip in the AP and **(F)** lateral views.