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## DYSPLASIA EPIPHYSEALIS CAPITIS FEMORIS

### *A Clinical-radiological Syndrome and its Relationship to Legg-Calvé-Perthes Disease*

*By*

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At the Coastal Hospital, Refsnæs, and at the Finsen Institute, Copenhagen, Denmark, about 300 cases of Legg-Calvé-Perthes disease have been treated so far.

Detailed analysis has shown that these cases do not make up a uniform, homogenous group.

In addition to the typical syndrome there is at least one well-defined group of cases differing from the typical one, radiologically as well as clinically—not least in respect to prognosis.

Let me first specify what I consider characteristic of the typical Legg-Calvé-Perthes syndrome. This brief description will be based mainly on the radiological appearances. Of course, the clinical symptoms are of great importance, but so uncharacteristic as to be of limited interest in this context.

*Legg-Calvé-Perthes disease develops in an epiphysis which previously has appeared completely normal—in shape as well as size.*

On this item there is by now general agreement, but this has not always been so.

In the nineteen twenties and thirties a group of orthopaedic surgeons, headed by *Calot*, held that the disease always arose in a hip joint which was already the seat of slight congenital subluxation. *Calot* made the sharp statement that: "L'ostéochondrite" ou "maladie de Legg" ou de Perthes, *n'existe pas*. Il s'agissait de *subluxations meconnues*".

These views were so severely criticized (*Calvé*, *Perthes*, *Sorrel*, *Waldenström* and others) that the orthopaedic surgeons of those times were gradually left with the impression that *Calot's* views were erroneous. Since then, there has been practically no discussion about this

question, and to-day it is considered an established fact that the Legg-Calvé-Perthes disease develops in a hip joint which has previously been completely normal, clinically as well as radiologically.

This related to possible *premorbid* changes in Legg-Calvé-Perthes disease.

As to the development of the disease itself it may be briefly stated that:

The first radiological sign of Legg-Calvé-Perthes disease is—in addition to slight widening of the joint space—*massive uniform condensation* of the bone tissue in the epiphysis which is otherwise normal in shape and size. However, soon the epiphyseal contours become a bit angular, and at the same time the epiphysis shrinks a little—but the diffuse, uniform condensation remains.

This is the stage of condensation (Fig. 1a).

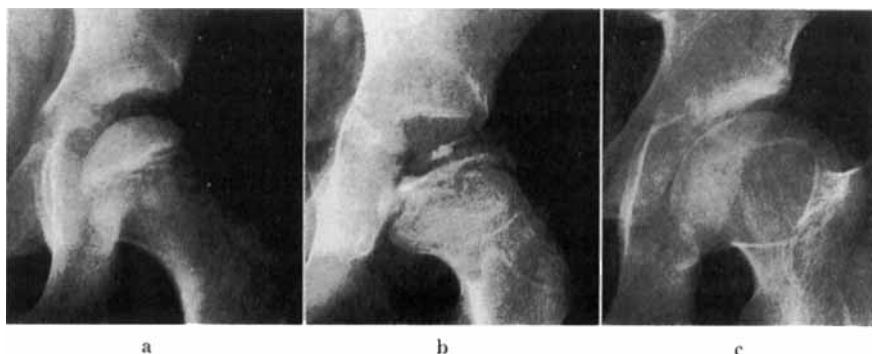
It is replaced by what is called the stage of fragmentation, the condensed epiphysis being split up into a number of *angular or streaked*, but *still condensed fragments*, frequently separated by *considerable defects*. At the same time there will often be *flattening* of the epiphysis and *broadening of the neck*, but these phenomena depend essentially upon the treatment applied (efficacy of non-weight bearing) (Fig. 1b).

This entire development—from massive condensation in an epiphysis of normal shape and size to a completely split-up and deformed epiphysis may rightly be designated as constant radiological “*deterioration*”.

This period of “*deterioration*” lasts for about 18 months, and not until then is a quite delicate, granular new-formation of bone visible in the epiphyseal defects. From this time on, the healing continues, new-formation of bone increases, but the bone structure in the epiphysis does not return to normal until at the end of 5–7 years.

The disease usually leaves a major or minor *flattening* of the femoral head and *broadening of the neck*—depending upon the efficacy of the treatment. These changes in shape do not immediately give rise to clinical symptoms, but may involve osteoarthritis of the hip joint at a later date (Fig. 1c).

This *typical radiological appearance and course of Legg-Calvé-Perthes disease* is observed mainly at an age over 6–8 years. In younger patients the appearances are not quite so typical, and among the youngest group, under 4–5 years, there is a *group of patients showing an entirely different picture—radiologically as well as clinically*. In our series they make up 10 per cent of all patients and are chiefly boys,



*Fig. 1.*

*Typical Legg-Calvé-Perthes disease.*

a. Stage of condensation. b. Stage of fragmentation. c. Healing.

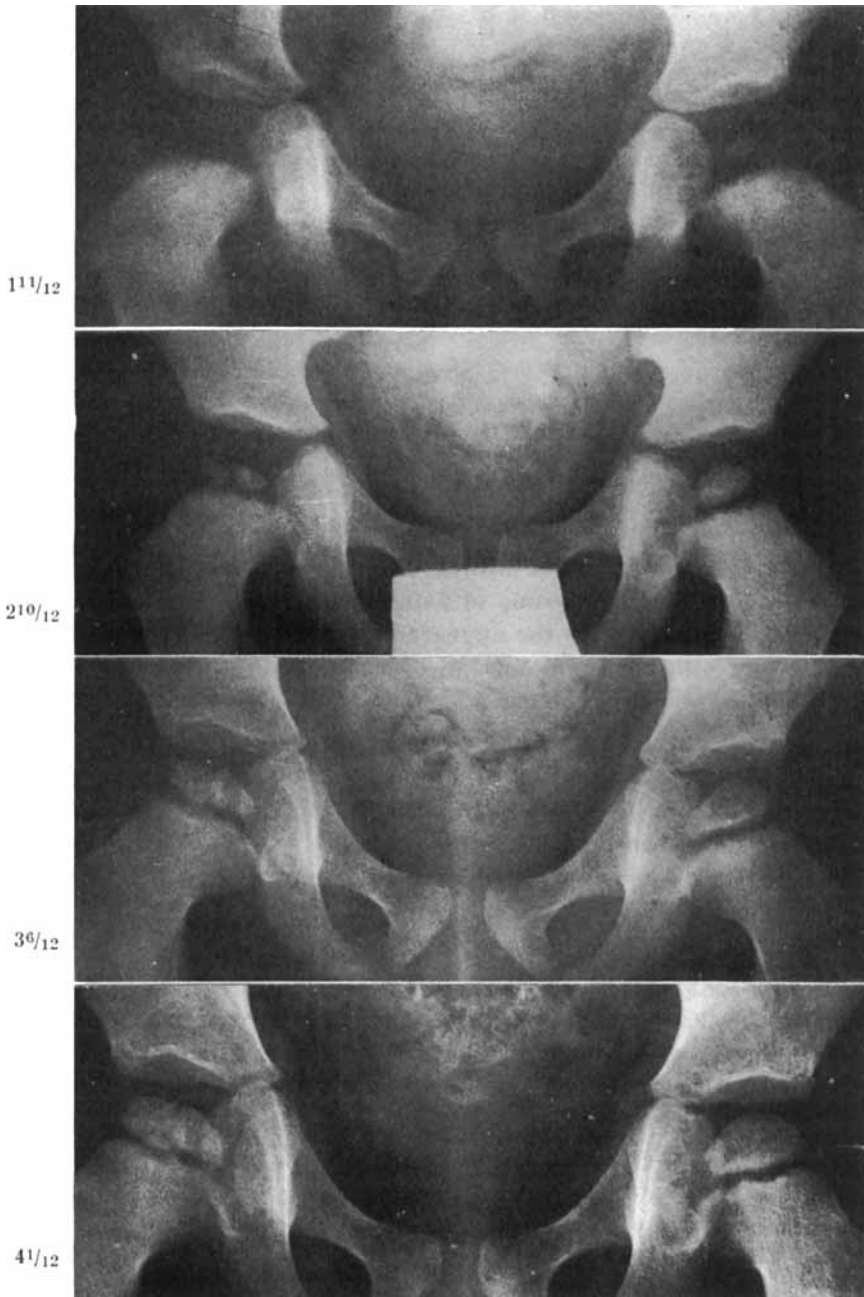
just as in all other groups of the material. Incidentally, there are in this age group too cases of entirely typical Legg-Calvé-Perthes disease.

When in this special group of patients we look at films of the children's hip joints *prior* to the appearance of typical radiologically changes (Fig. 2, 1<sup>11/12</sup>) they do not—just as in Legg-Calvé-Perthes disease—show a completely normal hip. At this early date the hip joint (or joints) will show *a marked delay in the development of the epiphyseal nucleus*. The ossification usually does not set in until about 2 years of age, and when at last it starts *the small epiphyseal nucleus will be of a pathological appearance*.

At the time when the patients start having constant symptoms—incidentally the same as those of typical Legg-Calvé-Perthes disease—the radiological appearances are as follows (Figs. 2, 3, 4, 5):

*A diffuse, granular structure* in the small, delayed epiphyseal nucleus. The size of the granules ranges from millet-seed to a grain of rice. The epiphysis may bear a certain resemblance to a blackberry. Another characteristic is that *the condensation is very inconspicuous* in these granular epiphyses. It may be entirely absent, but when it is present it is situated in the basal part of the epiphysis, and its shape is determined by the granules which are themselves or in their periphery condensed, especially down towards the epiphyseal line. This gives a patchy or rather netlike, basal condensation. *The uniform, massive diffuse condensation which is so characteristic of the typical cases of Legg-Calvé-Perthes disease is never seen.*

The picture which has just been described is that encountered when



*Fig. 2.*

*Dysplasia epiphysealis capitis femoris dx.*

First picture prior to appearance of right epiphyseal nucleus. Left: Delayed epiphyseal nucleus. The figures indicate the patient's age. ♂.

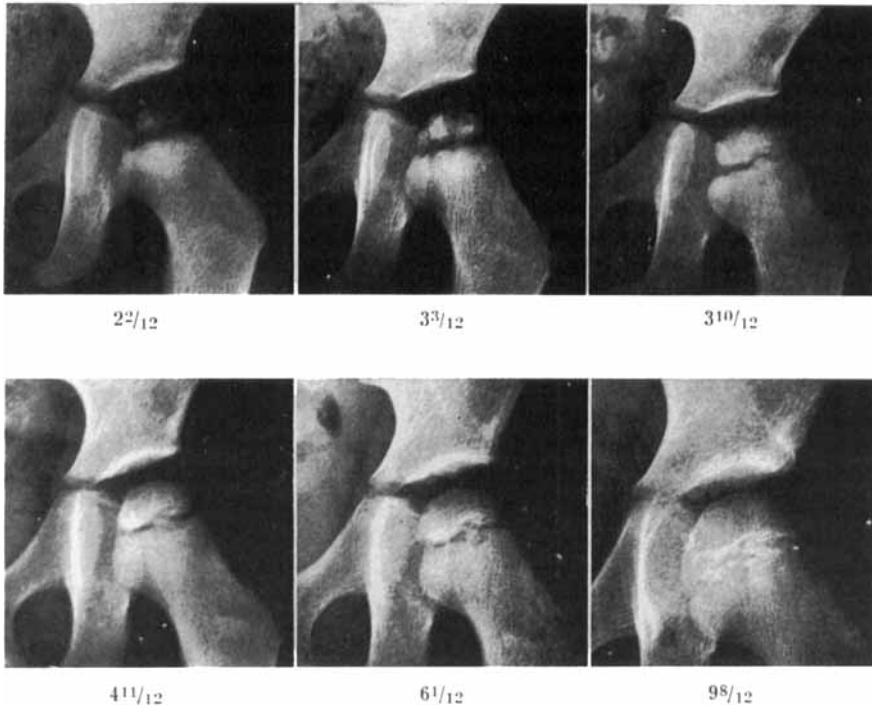


Fig. 3.

*Dysplasia epiphysealis capitis femoris.*

Course until end result. The figures indicate patient's age. ♂.

the patients are admitted for treatment, *i.e.* it represents an early stage of the disease.

A study of the *further course* (Figs. 2, 3, 4) reveals that this too differs perceptibly from that generally seen in typical Legg-Calvé-Perthes disease.

In the typical cases of Legg-Calvé-Perthes disease it is characteristic that during the first 18 months or so, *i.e.* through the fragmentation stage, X-rays show a striking deterioration. In contrast, the small group described above is characterized by a *constant improvement in the X-ray appearances after the first film*. There will be a steady homogenization of the epiphysis with slow disappearance of the granular structure. This development, until a completely normal structure is re-established takes only about *3 years*, while in typical Legg-Calvé-Perthes disease it takes 5-7 years. To this is added an extremely important factor, *viz.*

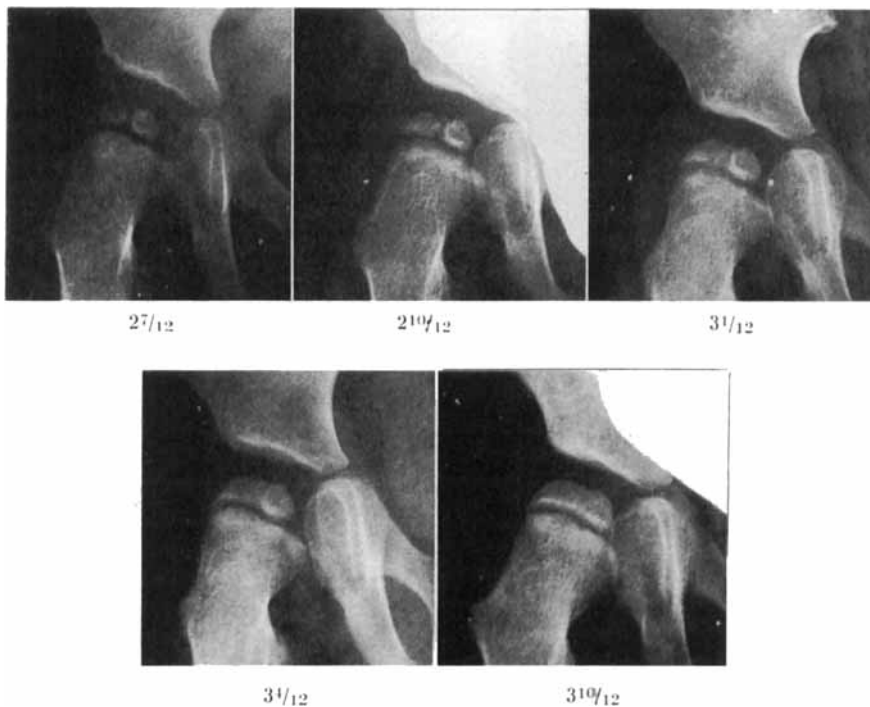


Fig. 4.

*Dysplasia epiphysealis capitis femoris.*

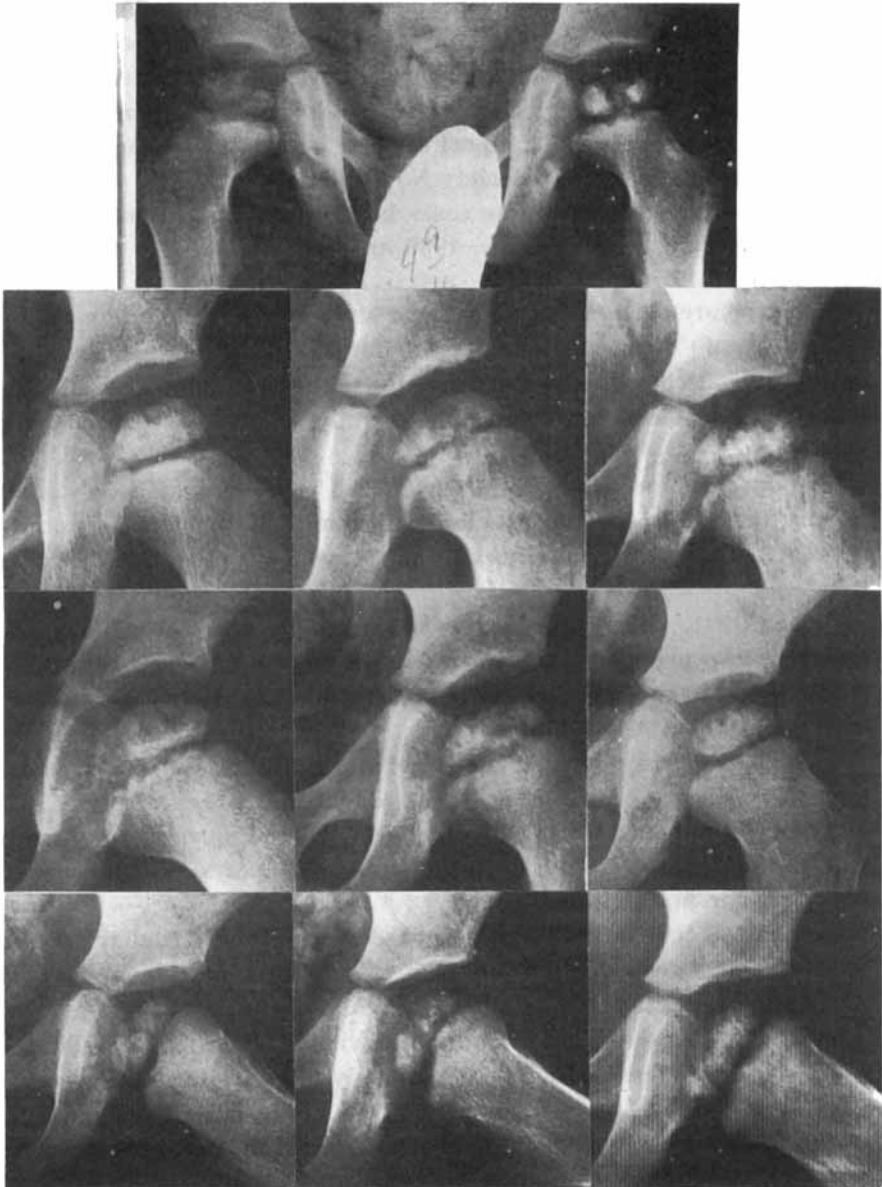
Typical course. The figures indicate the patient's age. ♀.

that during this course the epiphysis + neck *undergo very little deformation*: There will be no essential flattening of the epiphysis or broadening of the neck. In brief: In about 3 years a large number of cases show the head and neck of the femur to be of *completely normal shape and structure*.

Yet another two properties characterize this special type of case.

It is more often bilateral than typical Legg-Calvé-Perthes disease (in our series in 42 per cent as compared with 7 per cent).

It is more often *familial* in the sense that there is a history in the immediate family of not only similar cases, but also of other diseases of the hip (typical Legg-Calvé-Perthes, congenital dislocation, etc.—*vide infra*). In our series the family history was positive for hip joint disease in 17 per cent of the special cases described above as compared with 8 per cent of the typical cases of Legg-Calvé-Perthes disease.



*Fig. 5.*

*Dysplasia epiphysealis capitis femoris.*

10 different examples, all representing a single stage of a typical course.

All ♂. Age 2 years 9 months—6 years 9 months.

In order to render the following discussion of this special syndrome in young children more perspicuous, it had to be given a short and convenient name. For descriptive reasons we have chosen the neutral term: *Dysplasia epiphysealis capitis femoris* or for short: *Capital dysplasia*. This was the term used in a report of these cases by *E. Karup-Pedersen* in the Danish Orthopaedic Society Nov. 1959.

Scattered reports on similar cases have appeared in German orthopaedic literature (*Hilgenreiner*, 1936 and *Gickler*, 1937).

As is apparent from what has been stated above, then, our approx. 300 cases of "Legg-Calvé-Perthes syndrome" included at least two groups of patients having fundamentally different clinical and radiological appearances: capital dysplasia and typical Legg-Calvé-Perthes syndrome.

*The dysplasia* occurs in children under 5 years of age, in very small, greatly delayed epiphyseal nuclei, and manifests itself in a granular bone structure of the epiphyseal nucleus without essential condensation.

By contrast, Legg-Calvé-Perthes disease was encountered in older children, appearing in quite normally developed epiphysis and manifesting itself in the early stages as diffuse, massive condensation of the entire capital epiphysis.

This alone clearly characterizes the two syndromes, but *more important still* is maybe the difference in *course* and *prognosis*:

Dysplasia is headed, from the very outset, for *improvement*: A normal bone structure is re-established in about 3 years with no or *very slight change* in epiphyseal shape.

In contrast, the course of Legg-Calvé-Perthes disease is characterized by *radiological deterioration* with increasing fragmentation and further flattening during the first 18 months. Not until this stage does incipient, scattered newformation of bone become visible, leading slowly, in 5–7 years, to normal bone structure in a usually rather *deformed epiphysis*.

The question, then, is: Do these two characteristic syndromes represent *two entirely different diseases* or are they *merely two different manifestations of the same disease*?

There is obviously *no* question of different diseases.

Despite the fact that both syndromes are extremely rare—Legg-Calvé-Perthes disease developing in 1 out of 1000 births and dysplasia being even less common—they are relatively often *combined in the same*

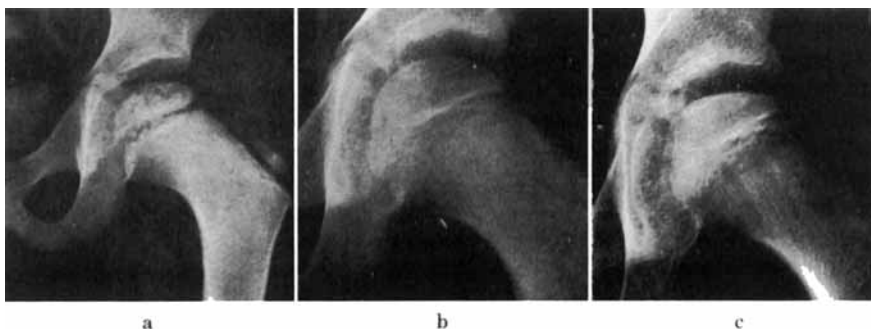


Fig. 6.

*Dysplasia epiphysealis capitis femoris with secondary Legg-Calvé-Perthes disease in the same hip joint.*

- a. 6 years 0 months: dysplasia epiphysealis capitis femoris.
- b. 13 years 9 months: the dysplasia has disappeared; capital epiphysis normal.
- c. 14 years 1 month: Legg-Calvé-Perthes disease: stage of condensation.

*patient. Among our 30 cases of dysplasia 6 were combined with typical Legg-Calvé-Perthes disease.*

In such *combined cases* we have the clearest and most characteristic pictures of the two types, when *dysplasia occurs first and Legg-Calvé-Perthes disease at a later date* (incidentally, we have never met with the reverse).

Two of our six combined cases were of this type (Figs. 6 and 7). Considering the rare occurrence of both types, it is very unlikely that we should be dealing with change coincidence in these patients.

The case depicted in Fig. 6 is of particular interest: A 6-year-old boy was treated for typical dysplasia which healed without any sequelae. After an accident at the age of 13, he developed typical Legg-Calvé-Perthes disease of the *same* hip. Fig. 7 illustrates another case: A 3-year-old boy was treated for typical dysplasia. After it had healed, the boy was allowed out of bed, but supported only on the apparently normal contralateral hip which rapidly developed severe, typical Legg-Calvé-Perthes disease.

Case histories like these two must leave a definite impression that the preceding dysplasia has predisposed to the subsequent Legg-Calvé-Perthes disease, or that there has been a common *state of preparedness* for the two conditions. It must be mentioned that the two secondary cases of Legg-Calvé-Perthes syndrome occurred following adequate traumas.

On the other hand, if two types of hip disease *are present at the same*

2<sup>10</sup>/<sub>12</sub>



3<sup>7</sup>/<sub>12</sub>



4<sup>1</sup>/<sub>12</sub>



4<sup>1</sup>/<sub>12</sub>



*time in the same patient*, the radiological appearances of the dysplasia and of the Legg-Calvé-Perthes disease often turn out to be not quite so typical. Frequently, both have to be characterized as *transitional cases*: One predominantly bearing the marks of dysplasia and the other of Legg-Calvé-Perthes disease.

Lastly, one may not infrequently come across what might be called *pure transitional cases*, *i.e.* cases which cannot be classified at all as being predominantly dysplastic or predominantly Legg-Calvé-Perthes: The condensation of the capital epiphysis is considerably more marked than in dysplasia—but yet far from the massive, diffuse condensation in Legg-Calvé-Perthes disease. The granular structure is *so loose* that it starts looking like fragmentation, without by any means attaining the irregularly streaky and angular fragmentation of Legg-Calvé-Perthes disease. The course too occupies an intermediate position, particularly in regard to the duration.

Such “pure” transitional cases may occur in one hip only or in both hips and are *most often encountered at the age of 4–6*.

It is not, however, only in these *combined cases* and *transitional cases* that dysplasia and Legg-Calvé-Perthes disease are associated, but they also show *familial association*. In 5 of our 30 patients with dysplasia (17 per cent) there was a history in the *immediate* family of several cases of Legg-Calvé-Perthes disease and dysplasia: In these 5 families there was a total of not less than 11 cases of dysplasia, 9 of Legg-Calvé-Perthes disease, and 2 of congenital dislocation. (The familial cases are *minimum values*, as the families were not studied systematically.)

Such an accumulation of dysplasia and Legg-Calvé-Perthes disease in a fairly small group of persons as well as the relatively common coincidence of the two conditions in the same person can certainly not be explained as a *change coincidence* of two different diseases. Indubitably, they must represent *two different manifestations of the same disease*.

But which disease? And why does it have *different radiological-clinical manifestations in the different age groups*?

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← Fig. 7.  
*Dysplasia epiphysealis capitis femoris sin.* with *secondary Legg-Calvé-Perthes disease* in the *contra-lateral* hip joint (which had previously been slightly dysplastic). The figures indicate the patient's age. ♂.

Early X-ray films and biopsy from capital epiphyses (*Jonsäter*) show without doubt that the initial stage of Legg-Calvé-Perthes disease must represent a rather rapidly developing ischaemic *capital necrosis*, *i.e.* a serious circulatory disturbance. The later stages of Legg-Calvé-Perthes disease, then, merely represent the breakdown of the bone necrosis and re-formation of the capital epiphysis by live bony tissue. The X-ray appearance of *capital dysplasia*, on the other hand, cannot by *any* means be interpreted as capital necrosis. If anything, it seems tempting to interpret it as the appearance of an *ossification disturbance*, *i.e.* delayed, irregular ossification. Can this picture, then, also represent a circulatory disturbance at the site of the capital epiphysis?

By injection experiments *Trueta* has demonstrated that the blood supply to the capital epiphysis is *ample in children up to the age of 4*, from metaphyseal as well as from lateral and medial epiphyseal vessels. *From the age of 4–8* the blood supply is far more sparse and only from one source: The lateral epiphyseal vessels. *From the age of 8 and until puberty* the blood supply slowly increases, the vessels in the ligamentum teres starting their development. However, it is not until about puberty that a real improvement in vascularization takes place, the epiphysis becoming again connected with the metaphyseal vessels.

According to these findings it is not difficult to understand that although circulatory disturbances at an age of up to 4 years may induce disturbances of growth in the rapidly growing epiphysis, it can probably not cause a complete cessation of the blood supply and subsequent capital necrosis. On the other hand, this must be a likely possibility in children over 4 years of age in whom the epiphysis receives its blood supply from only *one* source: The lateral epiphyseal vessels. In children approaching puberty the blood supply is again more ample and the growth rate slower, so that the possibility of capital necrosis decreases anew.

In brief: According to *Trueta's* findings, the capital dysplasia *as well as* the Legg-Calvé-Perthes disease may be explained as sequelae of circulatory disturbances in the capital epiphysis: According to these investigations, it is not surprising that circulatory disturbances at a late stage of childhood induce capital necrosis, *i.e.* Legg-Calvé-Perthes disease, while at an early age they cause only ossification disturbances at the site of the capital epiphysis.

However, this is not saying *for certain* that the two syndromes have really been induced by circulatory disturbances at the site of the capital epiphysis, but such a genesis would be *in keeping with* the view at

which we had arrived merely by clinical-radiological studies: That Legg-Calvé-Perthes disease and capital dysplasia are merely two different clinical-radiological manifestations due to the same cause.

But if both syndromes are caused by circulatory disturbances in the capital epiphysis, it is but natural that they should not be *absolutely* sharply distinguished or *completely* well-defined: In addition to the *typical* cases there is a certain number of *transitional* ones.

Of course, these transitional varieties may give rise to difficulties in diagnosis and classification.

However, it pays to try to group these cases between the pure dysplasias at one extreme and typical Legg-Calvé-Perthes syndrome at the other.

As a matter of fact, there is a great difference between the course and prognosis of the extreme cases, and by attempting to group the transitional cases in relation to the extremes we can *determine the prognosis with a far greater degree of certainty*: The more pronounced the dysplastic character of a case, the more it is predominated by ossification disturbances and the less by actual necrosis of bone, the better is the prognosis, in respect to duration as well as end result.

On the other hand, if necrotic changes predominate, the course will be considerably longer, the end result usually poorer, and the treatment far more trying, to the patient as well as to the doctor.

As far as I can see, therefore, a knowledge of the two syndromes described above affords a more rational basis for *individualized treatment* of the large group of patients who have so far been referred to us as cases of Legg-Calvé-Perthes disease.

Radiological changes, exactly identical with the capital dysplasia described in this paper, is present in the capital epiphysis of most children with *congenital dislocation of the hip*.

In children *with* as well as *without* dislocation the capital dysplasia may be followed by capital necrosis—which in children *without* dislocation is called Legg-Calvé-Perthes disease.

Orally as well as in writing, it has been maintained by several German orthopaedic surgeons, but most energetically by the French author *Calot* that *Legg-Calvé-Perthes disease* and *congenital dislocation of the hip* are merely *two different manifestations due to the same cause*—two aspects of the same disease.

The *common occurrence* of the dysplasia in cases of congenital dis-

location of the hip and its *close relationship* to Legg-Calvé-Perthes disease perhaps indicate that *maybe Calot was right!*

#### S U M M A R Y

Among the 300 cases admitted to the Coastal Hospital, Refsnæs, and the Finsen Institute, Copenhagen, as Legg-Calvé-Perthes disease there is, apart from typical cases, in the younger age classes a group of cases which differ clearly from typical Legg-Calvé-Perthes disease, clinically as well as radiologically.

This special syndrome is described and designated *dysplasia epiphysealis capitis femoris*.

The relation of capital dysplasia to typical Legg-Calvé-Perthes disease is described.

The pathogenesis of the two syndromes is discussed on the basis of *Trueta's* studies.

Attention is called to a possible pathogenetic relationship between congenital dislocation of the hip and Legg-Calvé-Perthes disease.

#### R E S U M E

Parmi 300 cas admis à l'Hôpital de la côte de Refsnæs et à l'Institut Finsen de Copenhague souffrant de la maladie Legg-Calvé-Perthe, on a constaté en dehors des cas typiques dans les classes jeunes un groupe de cas qui diffèrent nettement de la maladie typique de Legg-Calvé-Perthe, aussi bien cliniquement que radiologiquement.

Ce syndrome spécial est décrit et désigné *dysplasia epiphysealis capitis femoris*.

La relation entre la dysplasia de la tête fémorale et la maladie typique Legg-Calvé-Perthe est décrite.

Il est discuté de la pathogénèse de ces deux syndromes sur la base études de *Trueta*.

L'attention est attirée sur une parenté pathogénétique éventuelle entre la dislocation congénitale de la hanche et la maladie de Legg-Calvé-Perthe.

#### Z U S A M M E N F A S S U N G

Unter den 300 Fällen die unter der Diagnose Legg-Calvé-Perthes Erkrankung am Küstenhospital Refsnæs und am Finsen Institut, Kopenhagen, aufgenommen wurden, findet man, abgesehen von den typischen

Fällen, bei der jüngeren Altersklasse eine Gruppe von Fällen, die sich scharf von der typischen Legg-Calvé-Perthes Erkrankung sowohl klinisch als auch röntgenologisch unterscheiden.

Dieses besondere Syndrom wird beschrieben und als *dysplasia epiphysealis capitis femoris* bezeichnet.

Die Beziehung von Kopfdysplasie zur typischen Legg-Calvé-Perthes Erkrankung wird beschrieben.

Die Pathogenese beider Syndrome wird unter zugrundelegung von *Trueta's* Untersuchungen besprochen.

Die Aufmerksamkeit wird auf einen möglichen pathogenetischen Zusammenhang zwischen angeborener Hüftverrenkung und der Legg-Calvé-Perthes Erkrankung hingelenkt.

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