Opinions on and Comparative Observations about the Etiology of Legg-Calvé-Perthes Disease

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Summary. The article describes the various opinions on the etiology of Legg-Calvé-Perthes disease:
1. The theory of double infarction.
2. The theory of venous stasis.
3. The latent prolonged ischemic phase.

Due to the varying development of the vasculature of the proximal end of the femur, with a diminishing number of arteries ensuring blood supply and the subsequent reduction in the speed of flow in the myeloic sinuses, we describe here the origin of the latent ischemic phase of the proximal epiphysis of the femur, which constitutes the predisposition for the development of Perthes' disease. In the discussion about the diverse alterations connected with Legg-Calvé-Perthes disease, like the retardation of skeletal maturity, the dysplasia capitis or the necrosis following the treatment of congenital hip luxation, the different interpretations about the etiology of Perthes' disease are compared.


The term Legg-Calvé-Perthes disease implies the aseptic necrosis of the proximal epiphysis of the femur and the secondary centre of ossification which occurs between 2 and 12 years of age, reaching its peak during the years from 5–8. To-date, many different opinions have been put forward as to the origin of the aseptic necrosis of the proximal epiphysis of the femur, involving the adjoining metaphyseal parts. Arthur Legg in his publication in June 1909 already mentioned the possibility of a traumatic, avascular necrosis of the proximal epiphysis of the femur as the reason for changes in the epiphyseal nucleus of ossification among children. Further publications followed: that by J. Calvé in July of 1910 and that of G. Perthes in October of 1910. Other possible causes of this illness were suspected, such: inflammatory diseases, transitory synovitis, coxitis, osteomyelitis as well as minor or major traumas, Jacobs (1960), for example, described that 12 per cent of his patients suffering from the Legg-Calvé-Perthes disease had previously gone through transitory synovitis. Most authors, however, agree that the cause lies in the occlusion of blood levels which diminishes, or totally interrupts, the area's alimentation and supply of oxygen.

Up to now innumerable authors have tried to explain the reasons for this reduced, insufficient or pathological blood supply of the proximal epiphysis of the femur. According to the views expressed, the results of these trials may be summarized by means of three different theories:
1. The theory of the double or repeated infarct;
2. The theory of a venous stasis;
3. The theory of a latent ischemic phase in the development of the proximal epiphysis of the femur, as a result of variable development, with a diminished number of blood vessels as a consequence of retarded development.

The Theory of the Double Infarct

On the basis of their experiments with dogs, Zahir and Freeman (1972) and Sanchis, Zahir and Freeman (1973) became
convinced that Perthes disease was the result of repeated infarcts within the region of the epiphysis of the femur. Inoue, Freeman et al. (1976) compared the histological findings of animal experiments with the histological findings among 57 hips of 56 Japanese children suffering from Perthes disease. In 51% of these histological investigations they discovered changes characteristic of repeated infarcts within the region of the proximal epiphysis of the femur, namely the presence of atrophied, reparative granular tissue. These authors are of the opinion, that the second infarct occurred shortly after the appearance of the first symptoms, that is to say shortly after the first infarct. In 34% of the cases under investigation, they found criteria pointing to a double infarct which, roentgenologically, were still in the initial stage according to Jonsäter (1953). The authors believe that these findings make a prognosis of Perthes disease more difficult in the early stage, since it cannot be determined whether a second infarct has already occurred or is yet to come.

In the view of the authors, repeated infarcts may explain not only the aseptic necrosis but also the chronicity of the changes. In their opinion, the first infarct leads primarily to the atrophy of the entire nucleus of ossification, whereby the nucleus becomes diminished. In contrast to the changes during the first infarct, which are of a general nature, pathological changes which occur during the second infarct are localized and affect above all the reparative, granular tissue. Findings by McKibbin and Rališ (1974) also provide criteria supporting this opinion.

The Theory of Venous Stasis

Suramo, Puranen, Vuorinen and Heikinen (1974) advocate the concept of a venostasis in the extra-capsular region as the cause of pathological changes in terms of Perthes disease. The authors studied the venous outflow within the area of the neck of the femur among 28 femurs affected by Perthes disease and 20 normal femurs. Among the 28 cases suffering from Perthes disease, nine were found to be in stage I according to Jonsäter (1953), nine in stage II and ten in stage III. In all 18 cases in stage I and II, the authors determined that the venous outflow from the proximal end of the femur took place by means of the sinuses of the stem of the femur rather than through the gluteal veins. The intra-articular veins, above all the vena circumflexa femoris lateralis and the vena circumflexa femoris medialis were filled with contrast media. The gluteal veins were presented with contrast media in only one of the 18 cases in stages I and II. In the cases classified under stage III as well as in the 20 normal femurs, venous outflow proceeded readily through the gluteal veins. This led the authors to assume that they might be confronted with an extracapsularly limited venostasis. On the basis of this assumption, the authors recommended inter-trochanteric osteotomy for treatment of Perthes disease, which has a positive effect on venostasis, or respectively on the elevated intra-osseus pressure within the region of the proximal epiphysis of the femur. The intrinsic origin of the venostasis, however, remained obscure.

Another group of authors sees the cause of venostasis in the elevated intra-articular pressure in cases of synovitis or coxitis, which might lead to the compression of intra-articular veins or which might constitute the basis for repeated infarcts (Barta, Szepesi, Molnar 1978/1981). These authors regard the typical Perthes disease as an illness which occurs in swoops and which is explicitly recurrent in character. They have attempted to furnish proof of their opinion by means of animal experiments, in terms of 300 operated rabbits, and are engaged in enlarged their material. These authors were successful in all their cases of Perthes disease in furnishing the anamnestical proof of one or more instances of synovitis, and they also believe to be able to furnish histological proof for inflammatory processes in operated cases. Children suffering from Perthes disease, who were operated by them, are said to have shown as a rule strongly pronounced exudates under pressure.

Our Conception of the Latent Ischemic Phase

Anatomy

Three groups of blood vessels are responsible for the blood supply of the proximal epiphysis of the femur from the prenatal phase of development up to maturity. These are:

1. Branches of the lateral circumflex artery;
2. branches of the medial circumflex artery;
3. branches of the artery of the acetabulum.

Branches of the medial circumflex artery of the femur which, according to investigations by Lanz-Wachsmuth are missing in 0.33% of cases, are:

1. The dorso-medial retinacular arteries;
2. the dorso-lateral retinacular arteries.

The branches of the lateral circumflex artery of the femur are:

1. The ventral retinacular arteries.

Branches of the artery of the acetabulum are:

1. Arteries of the ligament teres.

According to investigations by Tucker (1949), Trueta (1957), Chung (1976) as well as our own, the arteries of the ligamentum teres may be found in only 40–50% of human beings.

As far as dorso-lateral arteries are concerned, most authors list numbers which vary from 2–6. The diameter of these arteries increases by 100% during the period from childhood to maturity.

As for dorso-medial arteries, authors agree that their number varies (1–3) and is reduced with advancing age. The diameter of these arteries is also reduced during human growth.

As for ventral retinacular arteries, most authors agree that they end in the proximal metaphysis of the femur.

Anatomical Observations

Among 59 sections at an age-level of 5 weeks of pregnancy up to 12 and a half years, I have observed a hypoplasia of only two arteries among the dorso-lateral arteries in five out of fiftyseven cases. In one case I observed seven dorsolateral arteries.