PERTHES' DISEASE OF THE HIP

E. W. SOMERVILLE, OXFORD, ENGLAND

From the Nuffield Orthopaedic Centre, Oxford

It is now sixty years since Legg (1910), Calvé (1910) and Perthes (1910) described the condition of coxa plana, or Perthes' disease as it is more often called today, thus allowing its differentiation from the much more serious tuberculosis of the hip with which it had formerly been confused. It is ironical to find all these years later that a diagnosis of tuberculosis nowadays entails no more than a few months in hospital whereas a diagnosis of Perthes' disease sometimes leads to a period of rest in bed with traction, splintage or plaster

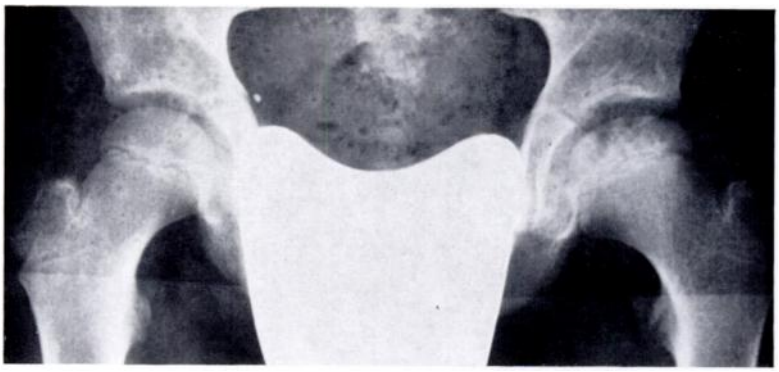

FIG. 1

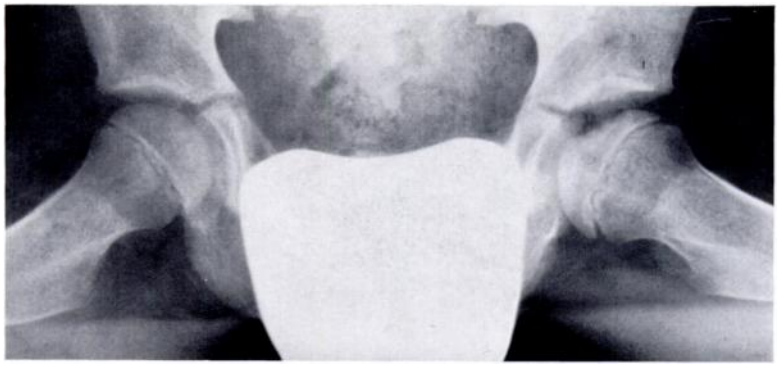

FIG. 2

Figure 1—The ischaemic changes in the left hip suggest that the whole of the femoral head is involved. Figure 2—A lateral view of the same hip shows that only half the head is involved.

immobilisation for one to three years, with no certainty of getting anything better than a moderate result at the end of it. In fact during these sixty years we have advanced very little in our knowledge or treatment of this condition.

PATHOLOGY

There is little doubt that the underlying lesion in Perthes' disease is ischaemia of the ossific nucleus of the head of the femur. Classically the disease has been divided into three phases: first the phase of ischaemia when the nucleus becomes dense, rather irregular in shape

and relatively smaller; then the second phase of so-called fragmentation; and finally the third phase of reossification. These three phases require further elaboration.

The ischaemic phase—The ossific nucleus of the head of the femur normally develops as a single entity but it has in fact two main blood supplies with very little connection between them. The distribution of this blood supply has been demonstrated by the injection experiments of Trueta and Harrison (1957), and their findings confirm what can be deduced from a radiological study of the developing femoral head in both health and disease. There is an antero-lateral leash of vessels from which the antero-lateral part of the nucleus develops, and there is a postero-medial leash from which the postero-medial part arises. The antero-lateral segment is usually the larger. The small and inconsistent blood vessels in the ligament of the femoral head play a very small and variable part in the vascular supply.

FIG. 3

FIG. 4

Figure 3—Typical early ischaemic changes seen in the left hip of a child aged 6 years. Figure 4—The same hip nine months later. Without passing through any of the usual stages the femoral head is restored to normal.

The ischaemia varies both in extent and in degree, depending on whether it affects one or both leashes and how many vessels in each are involved. In most cases only one part of the epiphysis is affected, usually the antero-lateral part, supplied by the antero-lateral vessels. In an antero-posterior radiograph this is not always obvious (Fig. 1) but in an oblique view it is clear (Fig. 2). Less often the whole of the nucleus is involved. The changes seen in the former type are consistent with damage to the antero-lateral leash, which because of its exposed position is particularly vulnerable to trauma. But local trauma could hardly be the cause of damage to both leashes, which must be damaged if the whole nucleus is involved. In this case some more generalised lesion must be present—possibly a transient synovitis producing thrombosis in the veins or a tense effusion damaging all the blood vessels.

The degree of ischaemia may also vary widely. In some the effect is only transitory, the typical changes of early Perthes' disease being resolved in a few months (Figs. 3 and 4). In such cases the nucleus is restored to normal without passing through either of the later phases. Presumably there has been a transient ischaemia with only partial or temporary obstruction of blood vessels and consequent rapid recovery.

At the other extreme there may be complete or almost complete involvement of the whole nucleus. In the early stages it is impossible to differentiate between the different types, and only the subsequent progress of the disease makes it clear how extensive the vascular damage has been. There is no evidence in the radiograph in Figure 5 to suggest that the course will

FIG. 5

FIG. 6

FIG. 7

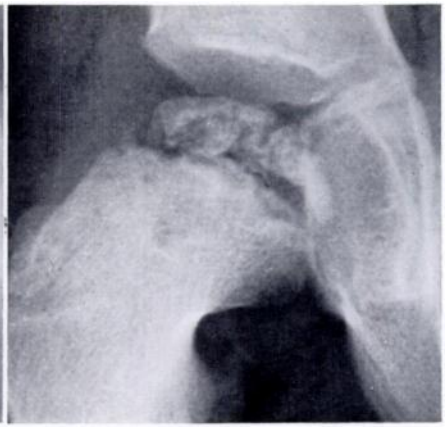

FIG. 8

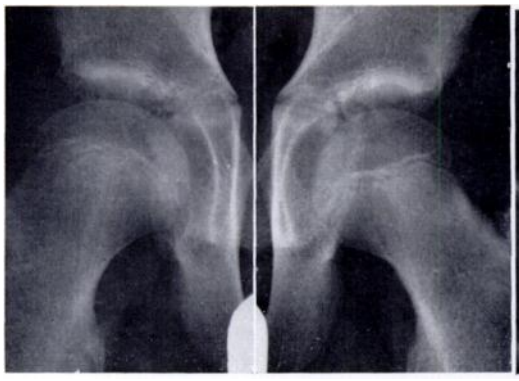

FIG. 9

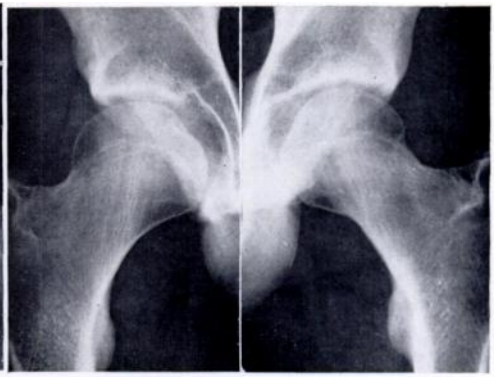

FIG. 10

Figure 5—Early ischaemic changes in the head of the right femur of a child aged 6 years. Figure 6—The same hip eighteen months later. It is clear that the whole nucleus was involved and all but a small fragment has been absorbed. Figure 7—The same hip three months later. Multiple small nuclei have developed in the shape of the nucleus. Figure 8—Nine months later the small nuclei are beginning to coalesce. This is most noticeable at the periphery. Figure 9—Six years later the nucleus is completely consolidated but it is broad and shallow. Figure 10—A further four years later the nucleus has returned to normal.

be different from that shown in Figure 3, but a radiograph eighteen months later shows that the ischaemia must have been complete. The whole of the ossific nucleus was affected and has been gradually absorbed (Fig. 6) in spite of continuous treatment, first on a frame, then in broomstick plasters.

Phase of fragmentation—The phase of fragmentation would be better termed the phase of absorption. The affected part of the nucleus is progressively eroded and removed (Fig. 6). It can be seen clearly in this case, and in all others that are followed carefully, that there is no evidence of new bone formation until the phase of absorption is complete, whether the whole or part of the nucleus is involved.

Phase of reossification—Reossification takes place not by replacement going on side by side with absorption, but as a distinct process beginning after the phase of absorption is completed. The radiograph shown in Figure 7 was taken three months after that shown in Figure 6. The appearance suggests that multiple small nuclei have developed, scattered throughout the substance of the part of the head of the femur that would have been the ossific nucleus if ischaemia had not developed. A small part of the old ossific nucleus can be seen deeply placed among these new nuclei. In spite of its appearance this is not fragmentation but just the opposite. Nine months later (Fig. 8) the small nuclei are beginning to coalesce, mostly at the periphery. The shape of the nucleus has been maintained and it has continued to grow as if the ischaemia had never occurred. Six and a half years later, at the age of fifteen (Fig. 9) the head of the femur is completely consolidated; it is round, but is still a little flattened and the depth of the epiphysis is less than that on the normal side.

Phase of remodelling—A further phase should be added to those usually described and called the phase of remodelling. Almost all hips which have suffered from Perthes' disease present some degree of deformity, but for many years afterwards remodelling of the nucleus continues. Four years later at the age of nineteen the shape and size of the head have very considerably improved (Fig. 10). There is now very little difference between the two sides. This progressive remodelling makes it difficult to assess the final results for at least ten or twelve years after treatment has been completed.

TREATMENT

In the past, treatment has always been directed towards the avoidance of pressure on a presumably softened femoral head with the object of preventing deformity. Such treatment has often involved a prolonged period of rest and protection, usually in recumbency, with or without traction or splintage, though sometimes treatment has been carried out with appliances designed to allow walking without weight-bearing. Pressure obviously plays a part in the development of deformity but the radiological appearance has probably overemphasised this, because the area of the head that would be likely to take most pressure is also the part affected by the ischaemia in most cases, and it is easy to mistake absorption of the ossific nucleus for collapse and distortion of the femoral head itself.

The degree of pressure on the femoral head will vary according to its position in the acetabulum. The weight-bearing area is decreased slightly in lateral rotation and increased in medial rotation when it is better covered by the roof of the acetabulum. The larger the area of contact the lower will be the load per unit area. Should there be an element of subluxation the loading per unit area will be still further increased. Kemp and Boldero (1966) have shown that an element of subluxation may be present at a very early stage in the disease.

Radiographs in early Perthes' disease often show a well-marked element of subluxation (Fig. 11). This amount of subluxation in an otherwise normal hip would inevitably lead to marked deformity if it were allowed to persist. In the presence of bony softening this deformity will be considerably increased. The subluxation can be reduced either by medial rotation alone or by medial rotation combined with abduction (Fig. 12). This position can easily be maintained by the use of external splintage, but this involves a prolonged period of immobilisation which has many physical and psychological disadvantages.

Correction can be maintained equally well by operation, either subtrochanteric osteotomy as demonstrated by Axer (1965), or pelvic osteotomy (Salter 1966, Salter and Bell 1968). How the subluxation is corrected is relatively unimportant, provided it is corrected, but it seems wise to use the simplest technique and the one least prone to complications. During the past thirteen years twenty-seven hips affected by Perthes' disease have been treated in this centre by subtrochanteric osteotomy.

Technique—Skin traction is applied for one or two weeks to relieve any spasm or pain. The hip is then abducted about 45 degrees and as much medial rotation as possible is obtained by using serial plasters strictly without using force. To do this properly may take three or four

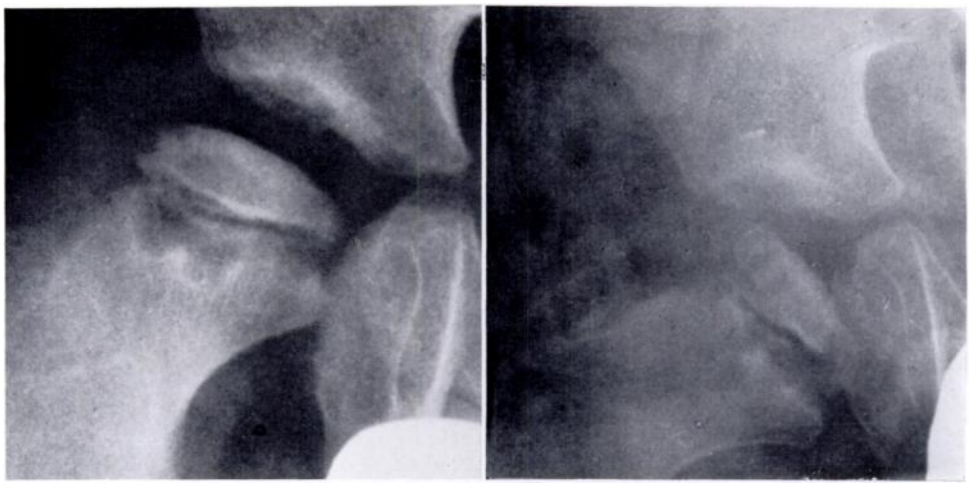

FIG. 11 FIG. 12

Figure 11—Early Perthes' disease of the right hip with subluxation. Figure 12—The subluxation has been corrected by medial rotation and abduction.

weeks. It can then be determined by radiography whether adequate reduction of the head of the femur will be obtained by lateral rotation osteotomy alone or whether some adduction will also be required. This determines the type of osteotomy. Since there is no question of correcting anteversion, rotation at the osteotomy should not exceed 20 or 25 degrees and the varus 10 or 15 degrees; if possible, rotation alone is carried out. The fragments unite in six weeks and the child is then allowed free. As soon as the hips and knees have regained mobility the patient is allowed to walk and to resume full normal activities without any restrictions and whatever the appearance of the capital epiphysis.

PROGRESS

Because a period of ten to twelve years after treatment is required to assess final results it is not yet possible to carry out a comprehensive statistical survey, but it is possible to review the cases that have been treated for an adequate time. The greatest value of such a review is to observe the redevelopment of the head in serial radiographs. In this series twenty-seven hips have been treated; a further three hips in which there have been changes of Perthes' disease but no evidence of subluxation or loss of medial rotation were kept under observation but not subjected to any restraint.

The treated hips have been divided into three groups according to the stage reached at the time the hip was first seen: Group 1 in which absorption had not yet started or was incomplete; Group 2 in which reossification had begun; and Group 3 in which consolidation was already well advanced or complete.

FIG. 13

FIG. 14

FIG. 15

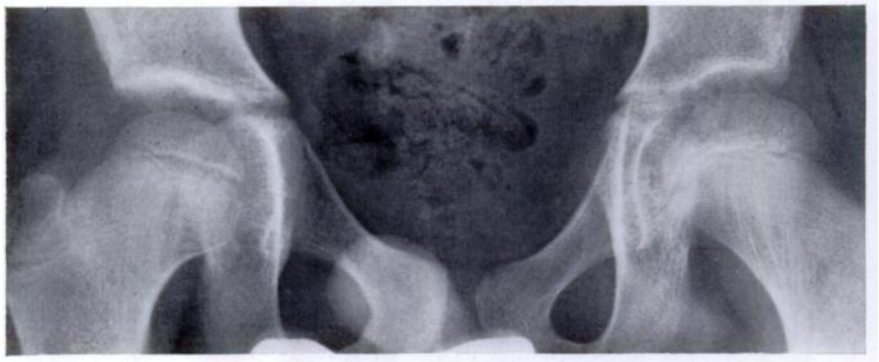

FIG. 16

Figure 13—Early Perthes' changes in the left hip of a boy aged 6. Some subluxation is present. Figure 14—One year after osteotomy a large part of the nucleus has been absorbed. Figure 15—A further year later there is considerable reossification. A gap is still visible in the centre of the nucleus but several small nuclei can be seen developing in it. Figure 16—Six years after operation the head is completely consolidated but it is broad and the epiphysis is shallow.

Group 1—There were nineteen hips. The ages at diagnosis were between four (one patient) and eleven (one patient); in three the age was eight. The follow-up period was from eighteen months to six years. The four patients who were under observation for less than four years have been omitted but these four have been following the same pattern as those followed for longer.

Fifteen hips have been followed up with regular radiographs for from four to seven years. In three the head of the femur is restored to a normal shape without any loss in depth of the epiphysis or broadening of the head and neck. Nine hips have run a very similar course

FIG. 17

FIG. 18

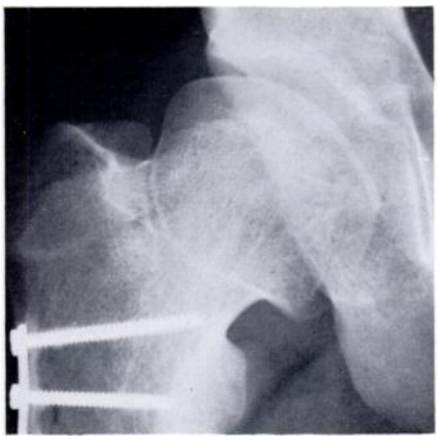

FIG. 19

Figure 17—Perthes' changes involving the outer segment of the head of the right femur in a boy aged 11 years. Figure 18—Arthrograph shows that there is real flattening of the femoral head. Figure 19—Six years after osteotomy the nucleus is consolidated. The head is round and reasonably congruous but broad and the neck is short. The acetabulum is a good shape.

(Figs. 13 to 16). In the case shown in Figure 13 there is very little deformity of the nucleus but it is dense and reduced in size and some subluxation is present. The subsequent radiographs show that absorption occurred in the usual way and was followed by reossification, which eventually became complete. The epiphysis is still broad and shallow, but at eleven years of age the stage of remodelling has still some way to go. If the pattern of the previous example is followed there is still plenty of time for complete remodelling to take place.

In the child aged eleven at the time of diagnosis the changes were already well advanced but absorption was not complete (Fig. 17) and an arthrograph showed that there was actual

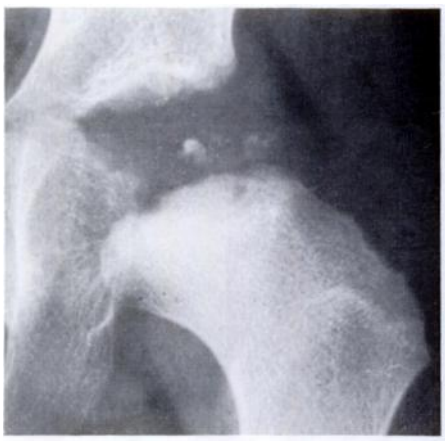

FIG. 20

FIG. 21

Figure 20—Perthes' changes in the hip of a boy aged 4½ years. Absorption is complete and several new ossific nuclei have just appeared. The hip is subluxated. Figure 21—Four years after osteotomy the head is almost consolidated.

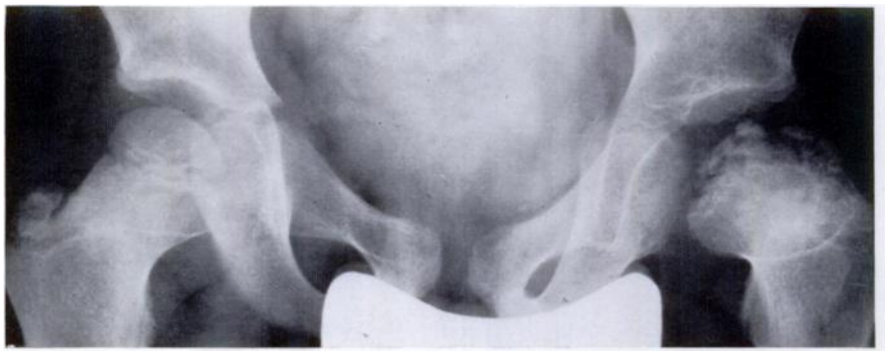

FIG. 22

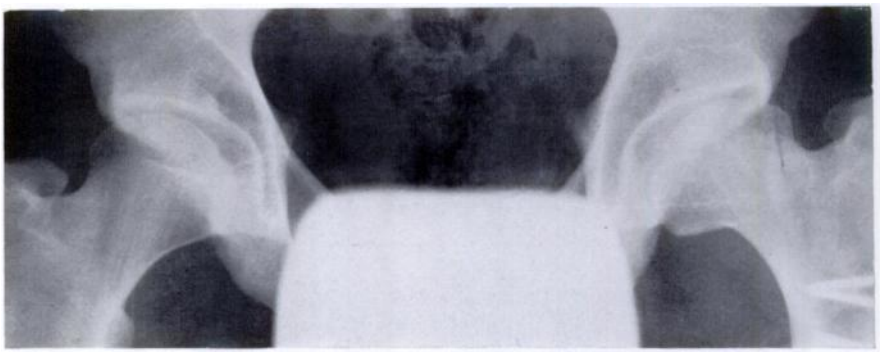

FIG. 23

Figure 22—Perthes' changes in the left hip of a boy aged 8 years. Ossification is well advanced but the hip is subluxated. Figure 23—The same hip thirteen years after osteotomy. The head is congruous but broad and shallow. The acetabulum is a little shallow but it is interesting that the other acetabulum is similar.

deformity of the head (Fig. 18). Six years later the head of the femur was round though rather large and the acetabulum was a reasonable shape (Fig. 19). There was shortening of the neck from disturbance of growth.

In two hips the result was disappointing and the deformity such that some permanent deformity must be inevitable.

Group 2—There were five hips in which the stage of absorption was past by the time the diagnosis was made. The progress of the hip of the youngest patient is shown in Figures 20 and 21. That of the hip of the oldest is shown in Figures 22 and 23. It is interesting that on the opposite side the shape resembles that on the affected side, although there was never anything to suggest Perthes' changes on that side and this hip was never immobilised. Of the other three, one is now normal but two still show some stigmata.

Group 3—There were three hips. Consolidation had already taken place at the time of diagnosis. No appreciable improvement has yet been noticed and if it is to take place it may take many years. It seems likely that treatment in this stage has little or nothing to offer.

FIG. 24

FIG. 25

Figure 24—Perthes' changes in the right hip of a boy aged 6 years. There is no clinical or radiological evidence of subluxation. Figure 25—The same hip three and a half years later. No treatment was given and the boy has led a normal active life. The condition has healed and the head is almost normal.

If the correction of subluxation is of therapeutic value, those hips in which there is no radiological evidence of subluxation and no clinical loss of medial rotation should not require any treatment at all. Three hips have been seen which in the first place fell into this category. Figure 24 shows such a hip in a boy aged six. There is no displacement but there are well-marked changes in the femoral head. The boy was kept under close observation but no treatment was given and no restriction was put on his activities. Three and a half years later the head is restored almost to normal. One hip in this small group was found to be developing increasing displacement and deformity and has recently been subjected to operation, but the third hip has shown no deterioration in two years and appears to be developing normally.

DISCUSSION

If the results described here were no better than those obtained by means of prolonged splintage they still would have the advantage that they were obtained by a method of treatment lasting little more than two months, after which the children were allowed to lead normal lives with no physical restraint. The radiographic follow-up of these hips suggests 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 wholly ischaemic. It may also be that the stimulus of normal active use promotes rather than retards the process of redevelopment. There does not appear to be any evidence here to suggest that the osteotomy itself speeds the process of revascularisation or of reossification. It appears rather that reossification will take place whatever form of treatment is carried out, and whether treatment is started early or late: we probably have no control over it whatever. But we can control the shape of the mould in which the reossification will take place, and it is towards this end that treatment should be directed. In studies of the progressive absorption and regeneration of the ossific nuclei in cases of Group 1 certain points of interest have been seen. It appears that reossification does not begin to be shown radiologically until absorption of the ischaemic bone is complete. Reossification takes place by the appearance of multiple separate ossific nuclei which ultimately fuse as consolidation progressively takes place, though this process may take several years to complete. This fusion of individual nuclei starts at the periphery and spreads towards the centre, where a defect may persist for some time until it finally fills. This is only what would be expected, because the blood vessels enter at the periphery.

But there are still some awkward questions to answer. Absorption of the ischaemic bone must indicate that blood vessels are invading it. Why is new bone not laid down until absorption is either complete or very nearly complete? When the bone has been absorbed what takes its place?

Jonsäter (1953) compared the radiological with the histological changes obtained from punch biopsies of the ossific nucleus at various stages during the disease. This showed initial ischaemia with continued growth of the epiphysial cartilage around it. Salter (1966) described the "head within a head" in which the normal growing ossification continued outside the shell of the old ossific nucleus, which itself never increased in size. It seems probable that the deeper layers of the epiphysial cartilage surrounding the ischaemic bony nucleus continue to grow outwards, enlarging the femoral head in the normal way; it will ultimately be in this newly formed cartilage that the multiple ossific nuclei will develop, and it is not until sufficient new cartilage has grown that ossification can occur. The fibrotic remains of the old nuclei are absorbed over many years and gradually replaced by bone.

It has for long been assumed that the basic lesion in Perthes' disease is ischaemia of the ossific nucleus of the femoral head, but it is certainly a very different condition from avascular necrosis. In avascular necrosis reossification does not take place; a sequestrum forms which only gradually absorbs, with extensive destruction of the femoral head and disorganisation of the joint; the normal anatomy is never restored. On the other hand in Perthes' disease reossification always occurs, and if the shape of the head has been maintained the newly formed ossific nucleus ultimately develops into a normal femoral head. It is possible that the difference is simply a matter of degree. In Perthes' disease, although the lumen of blood vessels may become obliterated by one means or another the vessels are probably never divided and remain in continuity so that recanalisation can readily occur, whereas in avascular necrosis the result of severe trauma such as a fracture or slipping of an epiphysis, or of sepsis, the blood vessels are completely divided, the continuity is lost and there is no chance of revascularisation. In fact revascularisation can only take place when the epiphysial line has closed. In this case the deeply placed cartilage cells of the epiphysis will have lost their nutrition and become necrotic, and growth of the femoral head would then not be possible. But it may be that the ischaemia in Perthes' disease is of an entirely different nature. Instead of the arteries being occluded it may be that there is a venous thrombosis, and the ischaemia results from acute congestion rather than from arterial occlusion. It is much easier to envisage venous thrombosis resulting from mild trauma, often causing no symptoms at all, which could be the precursor of Perthes' disease. It could also explain the great difficulty in producing truly typical Perthes' disease experimentally.

SUMMARY

1. Perthes' disease is an ischaemic lesion of the ossific nucleus of the head of the femur which may vary both in extent and degree. It is probably never quite complete.
2. When part of the ossific nucleus only is affected, as is usually the case, it is almost invariably the antero-lateral part.
3. The process of absorption of the damaged bone is complete radiologically before there is radiological evidence of reossification.
4. Reossification always occurs in Perthes' disease.
5. The aim of treatment must be to see that the mould in which the head is shaped is the right shape when ossification occurs.
6. The deformity of the head of the femur does not occur from pressure alone, but from pressure combined with subluxation. Full unrestricted weight-bearing can be allowed with safety on a femoral head in which there are ischaemic changes provided the femoral head is well contained.
7. The time of treatment can be very greatly reduced by using operation to correct the subluxation instead of relying on external splintage. This can be achieved by subtrochanteric osteotomy with rotation, or rotation combined with varus angulation.
8. Perthes' disease and avascular necrosis of the head of the femur are different conditions with different characteristics.
9. Suggestions are made as to the nature of the disease in relation to absorption, continued growth and reossification.

I am indebted to those of my colleagues who have referred their cases to me for treatment, to Mr R. L. Emanuel for the reproduction of radiographs and to my secretary, Mrs J. H. Anderson, for typing the manuscript.

REFERENCES

AXER, A. (1965): Subtrochanteric Osteotomy in the Treatment of Perthes' Disease. *Journal of Bone and Joint Surgery*, **47–B,** 489.

CALVÉ, J. (1910): Sur une forme particulière de pseudo-coxalgie greffée sur des déformations caractéristiques de l'extrémité supérieure du fémur. *Revue de chirurgie*, **42,** 54.

JONSÄTER, S. (1953): Coxa Plana. *Acta Orthopaedica Scandinavica*, Supplementum 12.

KEMP, H. S., and BOLDERO, J. L. (1966): Radiological Changes in Perthes' Disease. *British Journal of Radiology*, **39,** 744.

LEGG, A. T. (1910): An Obscure Affection of the Hip-Joint. *Boston Medical and Surgical Journal*, **162,** 202.

PERTHES, G. C. (1910): Über arthritis deformans juvenilis. *Deutsche Zeitschrift für Chirurgie*, **107,** 111.

SALTER, R. B. (1966): Experimental and Clinical Aspects of Perthes' Disease. *Journal of Bone and Joint Surgery*, **48–B,** 393.

SALTER, R. B., and BELL, M. (1968): The Pathogenesis of Deformity in Legg-Perthes' Disease. *Journal of Bone and Joint Surgery*, **50–B,** 436.

TRUETA, J. (1957): The Normal Vascular Anatomy of the Human Femoral Head During Growth. *Journal of Bone and Joint Surgery*, **39–B,** 358.

TRUETA, J., and HARRISON, M. H. M. (1953): The Normal Vascular Anatomy of the Femoral Head in Adult Man. *Journal of Bone and Joint Surgery*, **35–B,** 442.