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COXA VARA INFANTUM

I. Clinical Appearance and Aetiological Problems

By

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The normal development of the skeletal and muscular systems depends on normal use of muscles and joints. Even when our small patients do not always experience any subjective discomfort of importance, they instinctively perceive the failing stability of the affected hip, and they will from the outset try to spare the diseased limb with the result that the whole of this region lags behind in its normal development.

We may differentiate between five groups of non-inflammatory conditions in the caput-collum region, all presenting the above-mentioned features, though not always easily distinguishable. These groups are as follows:

(1) Congenital defect of the upper end of the femur.
(2) Coxa vara infantum.
(3) Coxa plana (Legg–Calvé–Perthes).
(4) Coxa vara adolescentium (epiphyseolysis colli femoris).
(5) Coxa vara, either primary or secondary, associated with various other conditions.

The synonyms of coxa vara infantum are:
Coxa vara congenita.
Developmental coxa vara.
Coxa vara.

In 1905 *Hoffa* gave the first detailed account of this condition. Since then descriptions of coxa vara infantum have been published at regular intervals.

AETIOLOGY

There are still differences of opinion over the aetiology of coxa vara infantum.

Reiner and Drehmann regarded this condition as a transition to a partial defect of the femur.

Nilsonne holds disturbances of ossification responsible, possibly due to a flaw in the vascular supply of the embryo. He assumes that coxa vara infantum belongs to the group of congenital defects in the upper end of the femur.

Camitz presumes that coxa vara infantum is due to disturbances of ossification, as in coxa plana, not to disturbances in the vascular system. He points out that both he and *Hoffa* before him have demonstrated areas of increased vascularization. He locates the most severe histological changes in the epiphysis of the caput, and he maintains that the development of the disease is post-foetal.

The material presented by *Camitz* does not include radiograms, and the material he has examined with reference to the metaphysis seems, as far as we can ascertain, somewhat scanty.

Brailsford regards coxa vara infantum merely as a dysostosis–as a developmental defect in the zone of ossification.

Gütig and Herzog regard this condition as an aseptic collum necrosis acquired at an early age, not as a congenital lesion.

Walter assumes that coxa vara depends on a congenital faulty anlage of the median area of the collum.

Schinz assumes that coxa vara congenita is merely an epiphyseolysis capitis femoris, manifest already at birth or directly afterwards.

Nille claims that transitions between coxa vara infantum and coxa vara adolescentium are to be found, and he has demonstrated a case of the latter condition, in which changes in the collum and the acetabulum were also observed.

Lange maintains that coxa vara is a traumatic–mechanical disease, and no congenital lesion. He believes in the existence of a rarefied zone along lines of abnormal stress and strain. He dismisses the theory of a congenital factor when there is bony healing or ossification of the involved zone within a few months following an operation.

Tucker has studied the blood supply of the caput in children and has found three main sources: one deep via the femur, a second via the ligamentum teres, and a third via a retinacular group of vessels. The retinacular group in its turn is composed of three vascular areas with wide variations. He regards a concentration of the blood supply in a single retinacular group of vessels as incurring danger of avascular

necrosis, since the group constitutes the main source of supply to the metaphysis and the head of the femur. *Tucker* maintains that the importance of the blood supply via the ligamentum teres increases with age.

Merenschow has tried to induce fracture of the collum femoris post mortem, and he has found that the line of the fracture invariably passes through the collum distal to the epiphyseal line, never through the cartilage. Hence his conclusion that epiphyseolysis must be regarded as an intrinsic problem, not traumatic.

Höxter says that it is almost impossible to produce experimental epiphyseolysis. Instead the bone is fractured. He claims that epiphyseolysis is due partly to trauma, perhaps by accumulation of repeated minor injuries, partly to endocrine factors (adiposogenital).

Harris has found that the administration of a growth hormone to rats reduces the resistance of cartilage to shearing-strength, whereas a sex hormone increases it.

Waldenström holds endogenous factors responsible for coxa plana.

Sundt regarded coxa plana as a general systemic disease elicited by a secondary factor.

THE INCIDENCE OF COXA VARA INFANTUM

Information with regard to the incidence of coxa vara infantum is scanty. Reports have appeared from time to time on the numerical relationship of patients treated for congenital dislocation of the hip to patients treated for coxa vara infantum. *Mesurier* gives this ratio as 14:1.

In an attempt to calculate the incidence of coxa vara infantum in Norway we have arrived at the conclusion, based on our material although with certain corrections, that it is at least 1 per 25,000 live births.

THE SERIES

The material dealt with in the present study consists of 42 patients, 22 males and 20 females, a total of 55 hips (see Table I).

This material comes from the University Orthopaedic Hospital (Sophies Minde), Oslo, Norway, and covers the past 20 years. Four patients come from Surgical Department A[1] of the University Hospital (Rikshospitalet), Oslo, over the same period. An attempt has been made to limit this material very strictly by exclusion of doubtful cases, particularly in the older age groups. We further increased this limitation by requiring an adequate case history, and that at least traces of the caput nucleus must be demonstrable in its proper place in the acetabulum.

[1] Chief: Professor Johan Holst, M.D.

The material is classified in three age groups (Table I, page 282):

(I) Patients (total 16) under 10 years old.
(II) Patients (total 11) between 10–15 years.
(III) Patients (total 15) over 15 years old.

The patients were grouped as above in accordance with the date of the first osteotomy. The non-operated patients were classified according to their age when they first came under our observation.

As shown in the second half of Table I the material comprises 13 patients with bilateral coxa vara and 29 with unilateral.

Bilateral disease was found in four males and nine females. Among the unilateral cases 11 were affected on the right side and 14 on the left side.

The two first columns of the second half of Table I show the operative hips and the two last columns show those not operated on or still under treatment.

PERSONAL INVESTIGATIONS

A. The Clinical Picture.

The clinical picture of coxa vara infantum in our material is rather oligosymptomatic. Most of the children are well developed at birth. They show signs of tiring readily when beginning to walk.

Soon they start limping, and in bilateral cases they develop a characteristic, waddling gait reminiscent of dislocation of the hip. In some cases the Trendelenburg test is positive. At first there is no upward displacement of the trochanter, and the head of the femur is palpated in its proper place. Pain is not constant.

Quite exceptionally the symptoms may decrease; as a rule they increase considerably with advancing years. About puberty, instability of the hips is found to be considerable and upward displacement of the trochanter is measurable. Gluteal deficiency, tilting of the pelvis in unilateral cases, and limitation of mobility are demonstrable. Increased abdominal lordosis and growing discomfort form part of the clinical picture.

The upward displacement of the trochanter may cause a rather considerable shortening of the limb, up to 9–10 cm. Quite exceptionally a shortening of the limb may be considered due to hypoplasia of the upper end of the femur as shown in Fig. 7.

Secondary symptoms in the back are common, particularly when the affected limb is much shortened.

In higher age groups the symptoms increase rapidly, finally rendering the patient completely invalid at a comparatively early age. As one of our patients remarked: "I could not even walk as far as the tram stop".

A definite large trauma is said to have started the first symptoms in four patients only.

B. The Radiological Picture.

The diagnosis of coxa vara infantum depends on the changes seen in the radiogram, the characteristic varus deformity of the collum-caput axis, a short and poorly developed collum, and widening of the angle formed by the epiphyseal line in relation to the horizontal.

Radiographic findings vary greatly according to the stage of development of the disease at the time when the patient comes for examination.

(a) Age Group I.

In our youngest patients we find the epiphyseal line in the collum femoris to be more or less steep; in the frontal plane it is often seen as a vertical, rarefied gap, wide and poorly delimited (see Fig. 1, Case 1).

On closer scrutiny an irregular deposit of lime may be seen in the zone of rarefaction. The lime may appear more linear, thereby indicating the boundary between epiphysis and metaphysis.

In this group we find the following:—

Eleven cases in which the epiphyseal line runs vertically (90°). One case in which the angle formed with the horizontal line is 20° and in another case 50°.

Six cases in which the angle is about 75°.

One case in which the angle is 100° (Case 14).

The median collum lip, which normally passes under the caput epiphysis, carrying this, does not develop, but retains its cartilaginous character or shows irregular deposits of lime, usually tapering into a narrow, labiate bony process. In many respects this bony process resembles periosteal callus or the changes to be found adjoining pseudarthroses, changes which reflect the reactive attempts of the organism to prevent a gliding tendency in the pseudarthrosis area (see Fig. 1 a–e and Fig. 2 a–c).

When we follow these cases radiologically, we find that in most of them this supporting structure will fracture sooner or later (see Fig. 1 e).

In other cases we find in the collum a triangular area of bone with

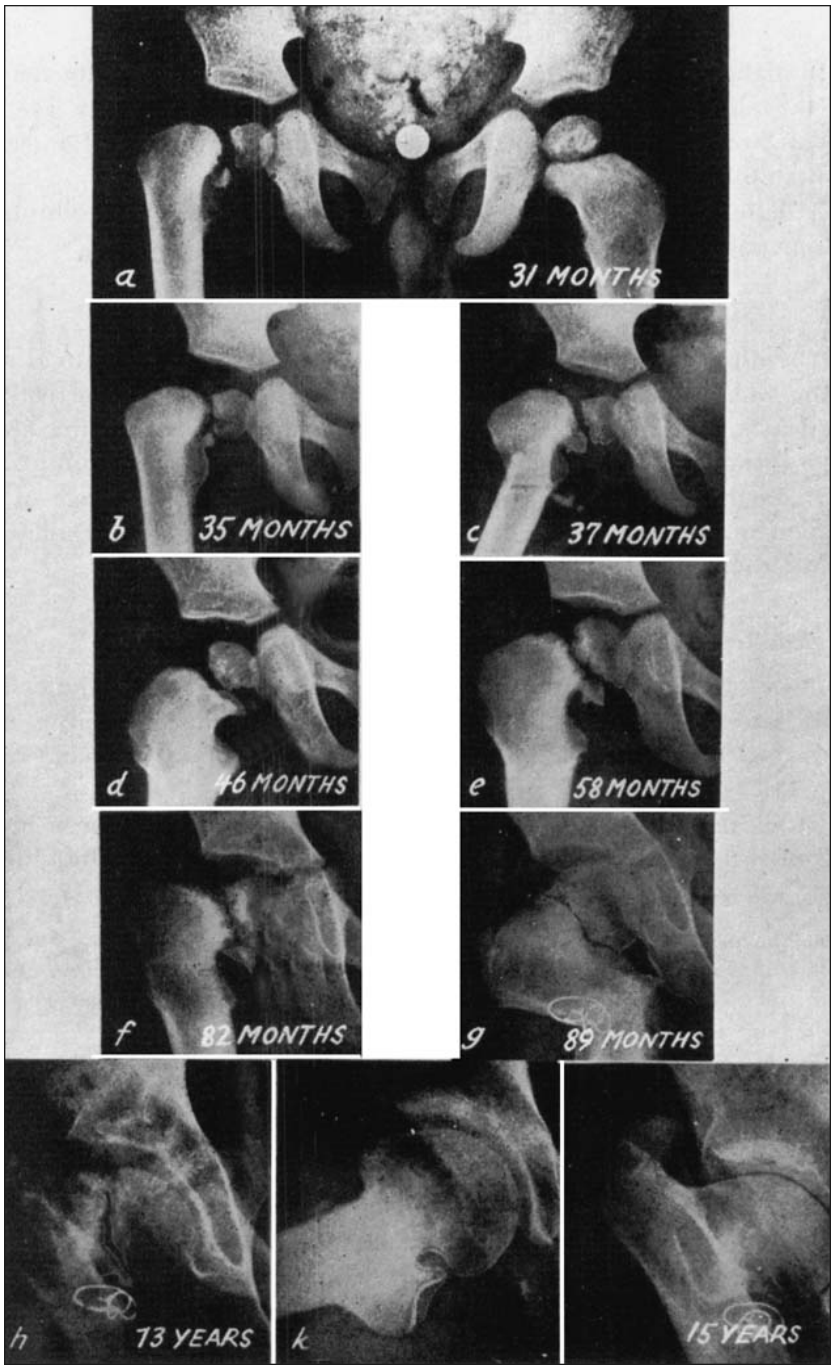


Fig. 1, Case 1. N. A. Age: 31 months-15 years.
Right-sided coxa vara infantum. Subtrochanteric osteotomy when 37 and 85 months old. On each occasion the pseudarthrosis recurred, but ossification occurred spontaneously at puberty. Note how the metal thread has wandered from the outer to the inner side of the femur because of the post-operative reconstruction of the trochanter mass. Post-operative observation period eight years.

its base below, apparently limited by a λ-shaped epiphyseal line. It has been maintained that this triangle is an artefact due to radiographic illusion. We have been unable to find evidence in support of this point of view, and the course of events in our Case 4 seems definitely to negate such an explanation (see Fig. 2, left side).

In this way the part of the collum which supports the caput nucleus becomes very poor and greatly varying in shape. It may become long and narrow, triangular or less regular.

In our material there are great variations in the development of the collum. The wider the rarefied zone in the metaphysis and the more deficient the calcification, the poorer is the development of the collum; it may also fail to develop at all. In most cases the trochanteric mass lags in development in comparison with the healthy side, but this state of affairs varies greatly in different cases. The tip of the trochanter reaches higher than normal, approaching the iliac bone in several cases.

Arthrography with Perabrodil shows a spacious joint capsule whose attachment to the bone is immediately lateral to the translucent belt in the metaphysis. At the same time the joint capsule shows an annular constriction corresponding to the circular ligamentous reinforcement normally found in the capsule of the hip-joint. Both acetabulum and caput are well developed, their outlines being even and sharply defined.

In two cases the epiphyseal angle diminished spontaneously while under observation (see Fig. 2, Case 4). Even when the development of the collum and the ossification of its metaphysis are poor, it is characteristic of this group that good contact is maintained between the caput epiphysis and the residual collum. We still find only slight signs of epiphyseal slipping.

Only in one case, in which the coxa vara was bilateral, did we find at as early an age as 2 years that considerable slipping had taken place in the metaphysis, with poor contact (see Fig. 3).

In *Case 4* (Fig. 2) the radiograms were taken at 5-year intervals, a and b with an interval of one year before the operation, c four years after subtrochanteric, bilateral osteotomy.

It is easy to see how on the right side an attempt has been made to build out the metaphysis of the collum from below with a view to forming a support for the epiphyseal line and the caput nucleus. The support, however, is very weak, and in Fig. 2 b it seems to have broken down.

The development of the metaphysis on the left side is much better. The epiphyseal line does not run so steeply, forming an angle of only 30°. But the lower, median corner has broken loose in the form of a triangular area of bone whose base faces downwards.

A year later (see Fig. 2 b) matters can be seen to have improved. There is no

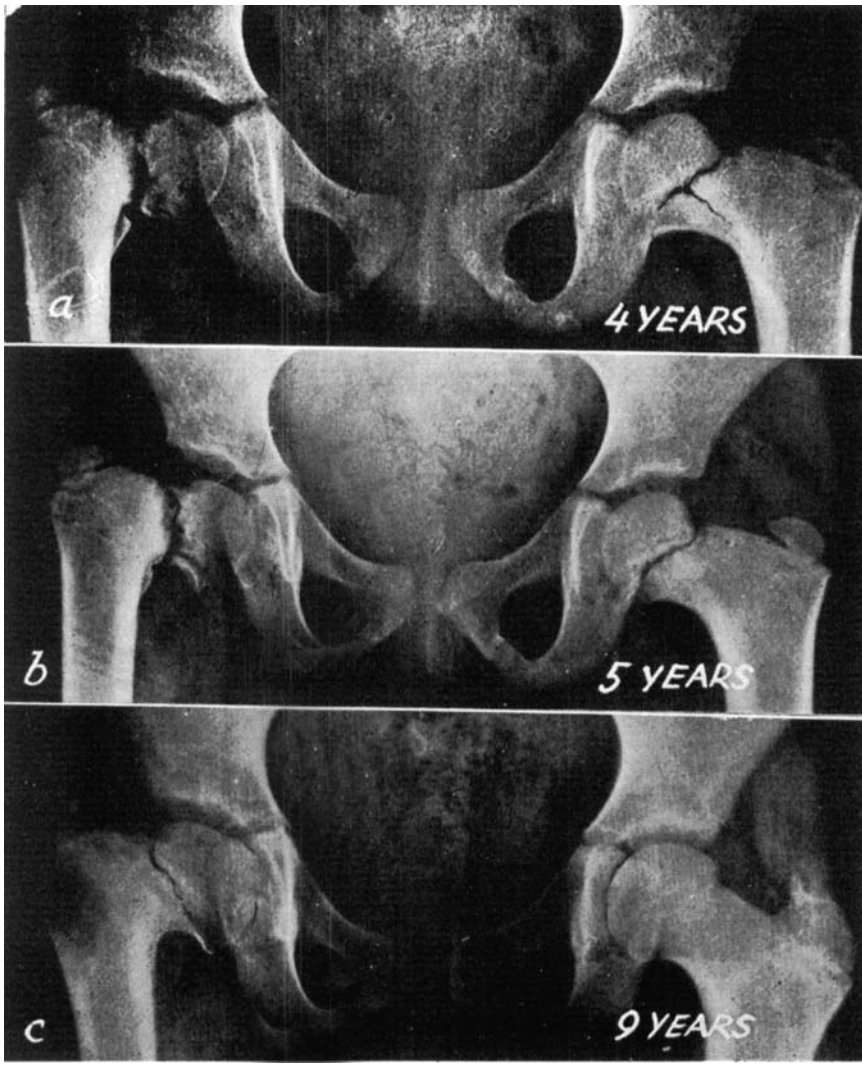


Fig. 2, Case 4. K. T. Age: 4-9 years.
Bilateral coxa vara infantum. Note the left side. In the area marked *a* a triangle of bone is apparently detached from the lower part of the collum, and *b* shows that this condition has subsided spontaneously. The right side of *b* shows a lip-shaped, lime-saturated shadow under the caput, separated from the collum by a zone of rarefaction. Treatment consisted of bilateral subtrochanteric osteotomy.

longer any sign of this "fracture line", which we moreover take to be evidence of an osteitis. The support of the metaphysis is well built out, and the angle with the epiphysis continues to be only 30°.

With such radiological findings on the left side, it would be quite justifiable to keep this hip under observation in the future.

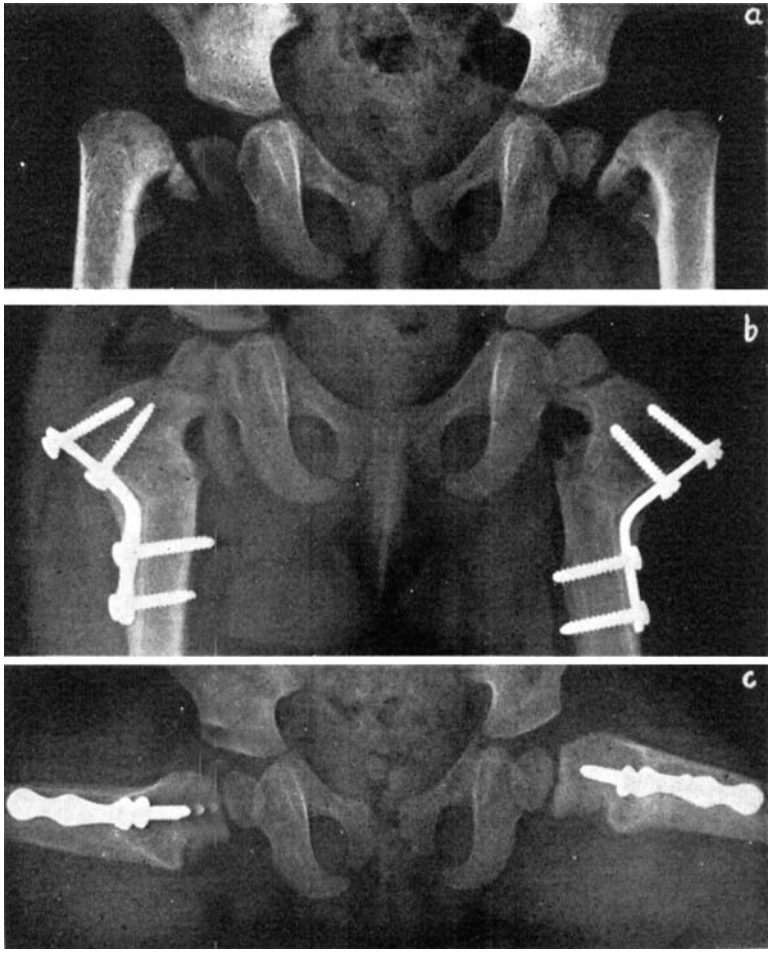


Fig. 3, Case 15. F. Age: 2 years.
Bilateral coxa vara infantum, our youngest patient operated on when only 2 years old. *b* and *c* are taken four months after subtrochanteric osteotomy. Note the reconstruction of the collum metaphysis.

Four years after bilateral subtrochanteric osteotomy it will be seen that the result on the left side is excellent. On the right side, no powerful metaphysis which can give adequate support for the caput nucleus has developed in spite of osteotomy (see Fig. 2 c).

(b) Age Group II.

The most conspicuous feature of the radiograms taken of our patients between the ages of 10 and 15 years is the slipping of the caput which has become more marked.

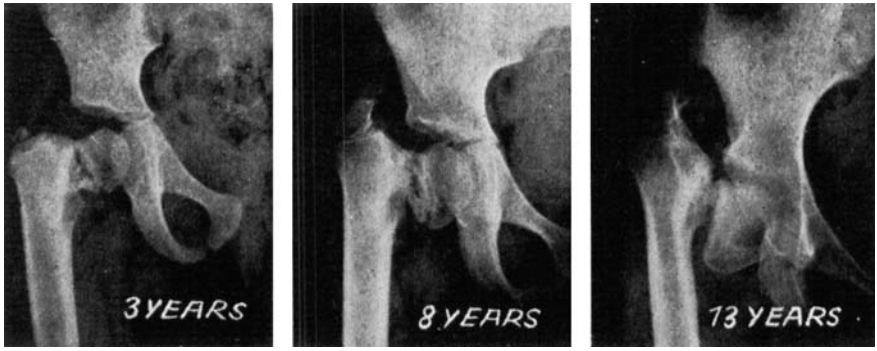


Fig. 4, Case 25. F. Age: 3-8-13 years.
Typical development of coxa vara infantum during the course of ten years. Untreated. Note in the last picture, how the caput has already partly passed the trochanter minor.

The rarefied belt in the metaphysis usually becomes narrower in patients in Group II and even when normal bone structure cannot be found the density of the lime increases. In the cases in which the caput nucleus has not succeeded in finding support against the collum or the lesser trochanter, it may be found far down along the shaft of the femur, without any appreciable osseous union with the collum.

In this group we find only two hips which show no sign of slipping of the caput nucleus.

TABLE I

A comprehensive survey of the whole material classified in age groups.

	I. Number of patients			II. Number of hips treated				
				Hips operated on in cases of		Hips not operated on or under treatment		Total
	Men	Women	Total	unilateral c. v.	bilateral c. v.	unilateral c. v.	bilateral c. v.	
Group I under 10 years cases 1-16	8	8	16	9	10	1	2	22
Group II between 10-15 years cases 17-27	5	6	11	6	6	2	—	14
Group III over 15 years cases 28-42	9	6	15	8	2	3	6	19
Total	22	20	42	23	18	6	8	55

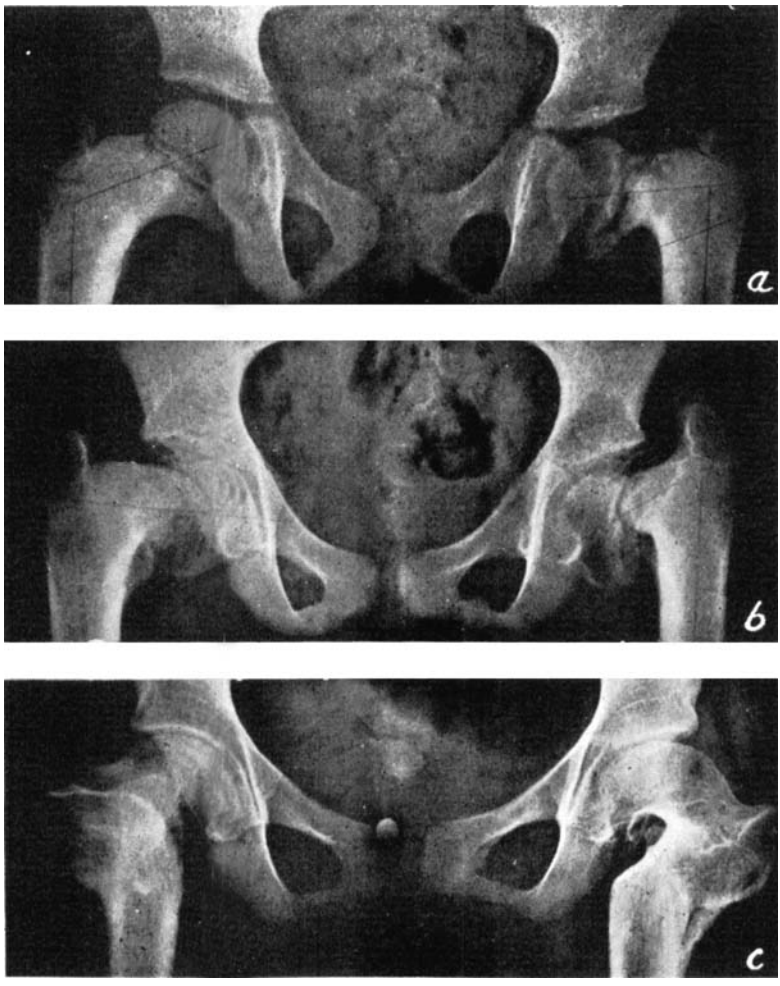


Fig. 5, Case 21. F. Age: 7-12-17 years.
Bilateral coxa vara infantum. Note on the right side how the radiogram shows aggravation of the disease from *a* to *b*, and how the radiological changes resemble those of epiphyseolysis adolescentium. Treatment consisted of bilateral subtrochanteric osteotomy. Good readjustment of the trochanter region.

Five hips show considerable slipping of the caput, and five other hips show such advanced slipping that more than half of the caput epiphysis lies below the inferior edge of the collum. In one case the whole of the caput nucleus lies below the collum femoris.

Fig. 4 is a good example of the rate at which this process develops; there is an interval of ten years between the first and last radiogram.

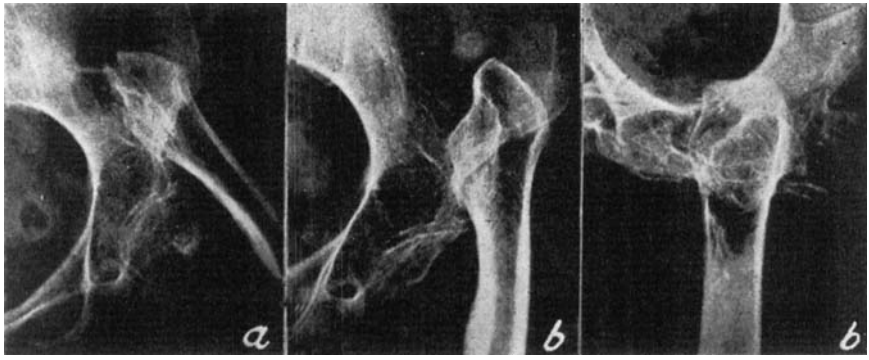


Fig. 6, Case 36. F. Age: 47 and 52 years.
Coxa vara infantum with a complete pseudarthrosis between the remnants of the caput and the trochanter mass. Treated by removal of the caput alone. Freedom from pain and good mobility, though the hip is unsteady.

During the observation period we found in one case development of coxa vara on the previously apparently healthy side (see Fig. 5, Case 21).

(c) Age Group III.

All these patients, who began treatment after the age of 15, show marked signs of slipping of the caput femoris. In half these cases either the connection between the caput epiphysis and the collum appears completely interrupted in the radiograms, or a definite pseudarthrosis has developed at the site of contact (see Figs. 5 and 6).

C. Histological Findings.

We have had occasion in only a few cases to make a histological examination of sections taken from our patients. These sections show the characteristic changes: A cartilaginous zone, wider than normal, with scattered islands of bone against the metaphysis. Areas showing a normal transition from cartilage to bone are hardly to be seen, and we find instead areas with deposits of lime and signs of ossification and residual cartilage alternately. We have found that radiological and histological changes correspond quite well, but unfortunately our histological material is not sufficiently large for a complete comparison between pathological-anatomical, radiological and histological findings. Here reference may be made to the histological studies of *Schwarz, Hoffa, Camitz, Ludloff* and *Helbing.*

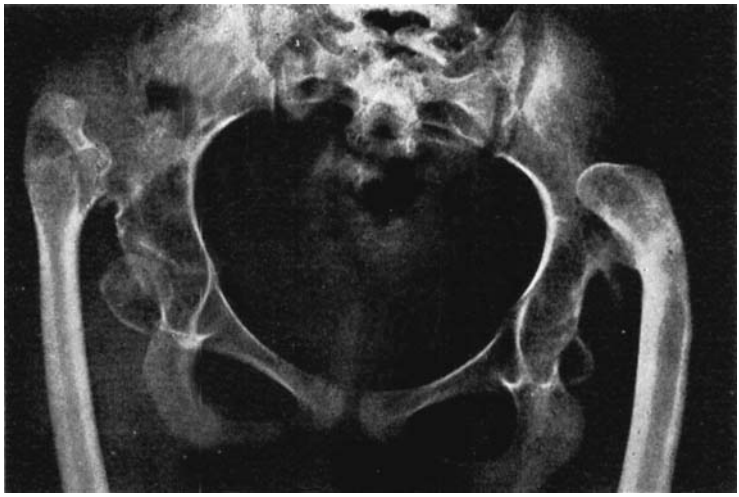


Fig. 7, Case 37. F. Age: 15 years.
Bilateral coxa vara infantum approaching hypoplasia of the upper end of the femur. Note the remnants of the caput in the right acetabulum and a corresponding bony shadow on the left side.

D. Complicating Pathological Processes in the Collum Region.
1. Legg–Calvé–Perthes' Disease.

In our material, this disease was found in two patients in Group I (Cases 9 and 10), in two patients in Group II (Cases 17 and 20) and in one patient in Group III (Case 42). The disease was found in a total of seven hips (see Table II).

It will be noted that the lesion occurs in patients of the typical age groups. In five cases the disease was observed before treatment was instituted, in three of them on the same side as the coxa vara.

Only in two cases is the development of this disease postoperative, in one of them on the coxa vara side, and in the other one on the non-operative "healthy" side.

Fig. 8, Case 9.

The patient came for treatment when 7 years old, and presents typical coxa vara infantum on *the right side*. The collum forms an angle of 80° with the shaft of the femur. The epiphyseal line runs vertically and in a fairly irregular way, not strictly sagittal either. The caput epiphysis is well developed, without signs of rarefaction.

The left side exhibits coxa vara, but also marked fragmentation and deformity of the epiphyseal nucleus. The epiphyseal line does not run vertically, but follows a more arched course. As there was no sign of slipping apart from the varus position of the collum, there was considered to be no indication for operative intervention on this side at the time.

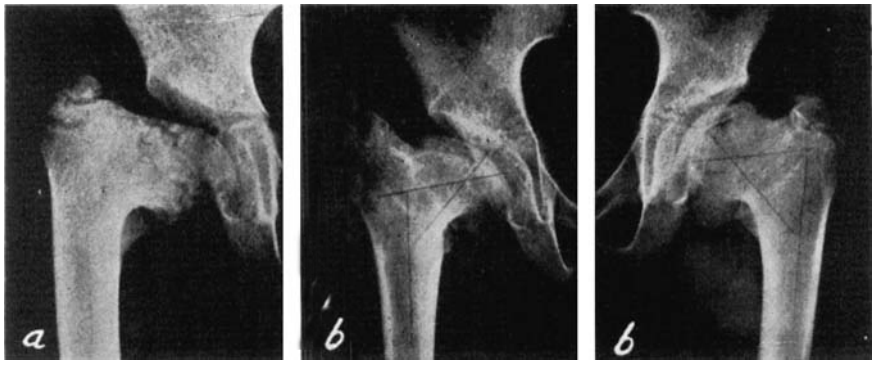


Fig. 8, Case 9. F. Age: 7 and 11 years.
Coxa vara infantum on the right side with secondary development of Legg-Calvé-Perthes on both sides. *b:* After subtrochanteric osteotomy on the right side.

We may assume that the left side presents Legg-Calvé-Perthes' disease in a pure form. The disturbances in the ossification of the metaphysis are insignificant and do not present the radiological picture seen in typical cases of coxa vara infantum.

Case 10.

This patient who began to receive treatment when 7 years old, presents marked Legg-Calvé-Perthes' disease on the left side plus coxa vara with a steep epiphyseal line.

Subtrochanteric osteotomy was performed, and excellent support was provided for the epiphyseal line and caput. However, the deforming process in the caput nucleus has not subsided, and as the period of observation has lasted only nine months, it is difficult to say what may be the final result of the operation. In this case the changes in the caput epiphysis dominate the radiological picture. The case might just as well be registered under the heading: Legg-Calvé-Perthes' disease. We have included the patient in this group, however, because the varus position of the collum is so marked. The case undoubtedly represents borderline features and shows that there may be ill-defined, transitional forms between Legg-Calvé-Perthes' disease and coxa vara infantum.

2. *Epiphyseolysis Capitis Femoris Adolescentium.*

Three of our patients present a radiological picture which may be regarded as a transition to the epiphyseolysis of puberty or a combination of this disease and coxa vara. (Fig. 5, Case 21, right side, Case 13 and Case 27, left side). (Table II).

The radiological evidence in these cases suggests that the flexion of the collum takes place in a more backward direction, at the same time as in a frontal view the caput nucleus is seen with the epiphyseal side more forwards as in typical epiphyseolysis. Thus the rather sharply defined rarefaction in the sagittal plane, which is characteristic of coxa vara infantum, is lacking.

Case 21, Fig. 5, right side.

At the age of 7 years a radiogram shows a slight indication of coxa vara infantum on the right side and a fully developed coxa vara infantum on the left side. At the age of 12 years both sides show poor calcification and poor development of the collum metaphysis. Owing to the position of the caput and the backward slipping, the radiogram is suggestive of the epiphyseolysis of puberty on the right side.

Case 13.

At the age of 3 years a characteristic and typical coxa vara infantum is seen on both sides. At the age of 10 years the radiogram is suggestive of an epiphyseolysis just before puberty. The calcification in the rarefied zone of the metaphysis is relatively ample and fairly dense. The lime deposits, however, are irregular and the collum is short. The epiphyseal line is quite narrow, and the caput nucleus seems displaced downwards and backwards. For anyone familiar with the evolution of such cases it is evident that this is a case of coxa vara in the process of healing and not an apparently incipient epiphyseolysis adolescentium.

Case 27.

A mongoloid, 12 year old boy of the adiposo-genital type, presents features similar to those of Case 21, but with considerably greater slipping of the caput calotte. Also here, the belt of rarefaction in the metaphysis is less sagittally, but more frontally placed, and the caput epiphysis seems to slip further backwards than usual in typical coxa vara infantum.

3. Other Skeletal Anomalies.

A. Hypoplasia of the Trochanteric Mass.

The most common skeletal anomaly is a hypoplasia simultaneously affecting the whole trochanteric mass. As a rule, this hypoplasia is proportional to the changes found in the collum metaphysis. As soon as the pathological process subsides or is arrested, the trochanteric mass will develop to an almost normal degree. This hypoplasia, therefore, appears to be a secondary process.

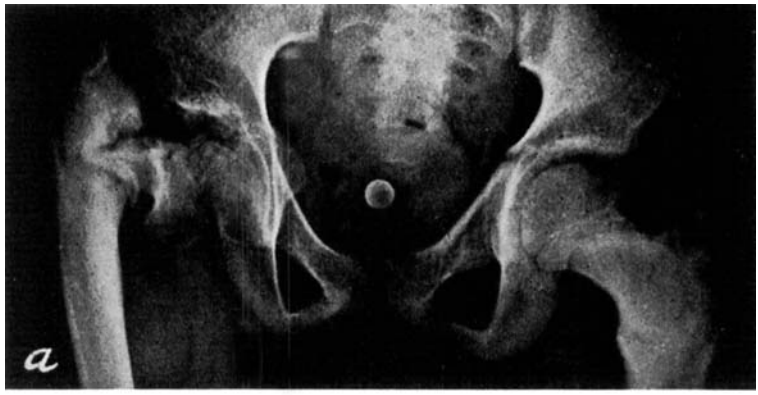
B. Hypoplasia of the Upper End of the Femur.

Conditions which we could characterize as genuine hypoplasia of the upper end of the femur occur with considerably less frequency than those belonging to type A. We have observed two such cases. A third case represents a transitional form of both types of hypoplasia.

As pointed out above, cases of hypoplasia showing no remnants of the caput nucleus, have not been included in our material.

Case 34, Fig. 7.

This patient came to the hospital when she was 15 years old. Unfortunately her previous radiograms have been destroyed. On the right side a distinct, though poorly developed caput nucleus is found in place in the acetabulum. The mass of



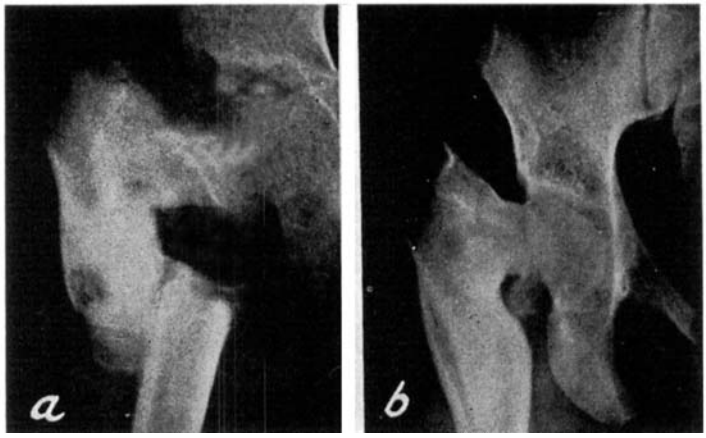


Fig. 9, Case 19. M. Age: 10 and 25 years.
Coxa vara infantum on the right side, with osteitis (dysostosis?) on both sides. Subtrochanteric osteotomy was performed on the right side. Note the post-operative reconstruction of the bone.

the collum and trochanter is markedly hypoplastic, and connection with the caput nucleus has been lost. The trochanter is situated at a high level outside the pelvis.

The changes appear even more marked on the left side, and only a trace of the caput nucleus can be seen in the acetabular region. There is no distinct articular space as seen on the other side. The mass of the collum and trochanter is even less developed than on the right side.

Multiple skeletal anomalies are found in the lower limbs. The fibula is absent on both sides, the astragalus and os calcis show block formation, and some of the radiating osseous arches of the foot are missing. These conditions cannot be traced to the hypoplasia of inactivity, but must be included in the group of genuine hypoplasia.

Case 16.

In this case a typical coxa vara is found on the left side, whereas the right side presents total aplasia of the entire upper end of the femur. Only a small bone nucleus corresponding to the caput epiphysis appears in the acetabular region.

Case 15, Fig. 3.

Our youngest patient underwent subtrochanteric osteotomy when only 2 years old. Already at this age there was a marked slipping of the caput and the epiphyseal line with a triangular area of the metaphysis, coincident with a very marked hypoplasia of the trochanter region. As a whole this case may be interpreted as transitional to genuine hypoplasia of the upper end of the femur. Nevertheless after osteotomy a surprising reconstruction of the collum-trochanter region occurred.

4. Traumatism.

Two patients in Group I give a history of definite and considerable traumatism, Case 3, Fig. 10 and Case 6. Three patients in Group II give a doubtful history of trauma, Cases 20, 24 and 25 (Fig. 4).

Two patients in Group III report a doubtful history of trauma (Cases 31 and 36), and two patients report a history of definite traumatism at the age of 4 years, Case 36, (Fig. 8) and Case 28, (see Table II).

Both patients in Group I exhibited similar radiological findings originally. In Case 3 there can be no doubt as to the traumatic origin of the disease, and even in Case 6 we regard such a genesis as quite likely.

Judging by available radiograms, however, we cannot definitely exclude the possibility that the metaphysis showed a slight disturbance of ossification even before the trauma. In all the other cases there is insufficient radiogical evidence before and after the trauma on which to form a definite opinion with regard to the influence of the alleged trauma on the development of coxa vara.

Case 3, Fig. 10.

When 3 years old the patient suffered from an intertrochanteric fracture on the left side after a fall from a barn bridge. The position of the fracture was good, and treatment consisted of rest in bed.

Fig. 10 a shows a roentgenogram taken two months after the accident. A faint irregular fracture line is seen running right through the trochanteric mass. The collum shows marked atrophy with increased density of bone. It will be seen that the lower median corner of the metaphysis is sclerotic and drawn out into a labiate promontory. The epiphyseal line forms an angle of 30° with the horizontal. The patient was allowed to get up five months after the accident. She did not at first complain of her hip, but she gradually experienced increasing discomfort.

A radiological examination one year after the accident (Fig. 10 b) showed complete detachment of the caput epiphysis from the remnant of the collum. The

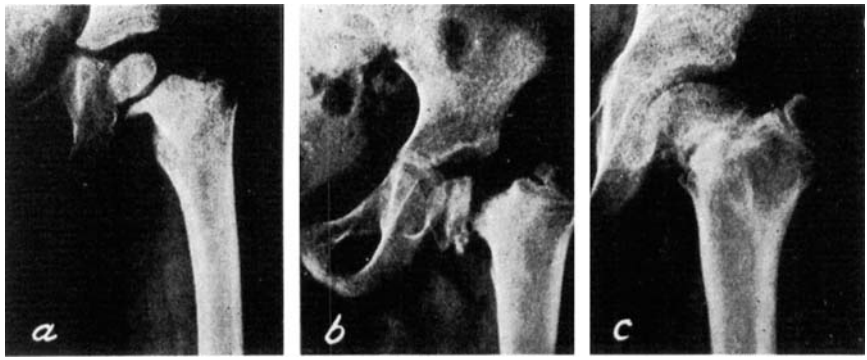


Fig. 10, Case 3. F. Age: 3-4-8 years.
Post-traumatic coxa vara infantum.
(a) Sclerosis of the fractured collum area.
(b) Slipping a year later with marked resorption.
(c) Four years after subtrochanteric osteotomy.

line of this detachment does not follow the previous fracture line, but runs about 1-2 cm. medially to this. It will also be seen that the caput nucleus is in place in the acetabulum together with the epiphyseal line and the adjoining border of the metaphysis. The metaphyseal rarefaction shows an irregular outline and, particularly in the lower field, irregular deposits of lime. This picture is very reminiscent of the changes found in coxa vara infantum. The final result of the subtrochanteric osteotomy (Fig. 10 c) is very satisfactory. The patient should continue under observation, however.

5. Osteitis.

Our material contains several cases which we could describe as examples of localized trauma of osseous tissue with resorption of bony tissue depending on weight-bearing.

The changes appear partly in a triangular area in the lower part of the collum becoming separated by a poorly calcified belt from the rest of the collum (Fig. 2 a), partly in the labiate bony process, which seeks to pass under the caput, being delimited from the metaphysis by a similar rarefied zone running vertically through the bone substance (Fig. 2 b). Both conditions have been described previously.

We cannot, however, dismiss the possibility that the changes in some cases may be due to a fracture caused by a minor trauma, with secondary resorption.

Case 19, Fig. 9.

This case is in a class by itself. Here we see how the whole of the collum has been broken out of the shaft of the femur as a triangular wedge, with its base facing the caput epiphysis.

TABLE II

Pathological processes in the collum region complicating our cases of coxa vara infantum.

	Total number of hips	Group I		Group II		Group III	
		with c.v.	without c.v.	with c.v.	without c.v.	with c.v.	without c.v.
Legg-Calvé-Perthes' disease	7	2	1	2	1	—	1
Epiphyseolysis capitis femoris adolescentium ...	4	2	—	2	—	—	—
Hypoplasia of the upper end of the femur	5	2	1	—	—	2	—
Trauma	4	2	—	(3?)	—	2(+2?)	—
Osteitis	1	—	—	1	—	—	—
	21	8	2	5	1	4	1

Signs of a similar process are found in the left trochanter minor, in which a distinct linear rarefaction is plainly visible. The collum on this side also shows a horizontal streak with greater lime density than in the adjoining bone. This must be interpreted as the outcome of a process similar to the one on the right side. In this case it is probably not so much the weight-bearing conditions as the disturbances of ossification which are responsible for the patient's discomforts. The case should therefore be included in the dysostosis group.

6. Rickets.

Only in one case (Case 23) do we find an arched flattening of the collum-shaft-angle reminiscent of the statically determined changes in rickets. In this case the lesion is unilateral, however, and the epiphyseal line is typical in shape. The subsequent behaviour of the case is characteristic of coxa vara infantum and the changes found can hardly be regarded as due to rickety changes in the bones. Thus, not a single case in our material presents clinical or radiological evidence of rickets.

E. Discussion.

On scrutinizing this material, we very soon encounter difficulties in distinguishing between the four groups of diseases mentioned in the introduction to this work. Not all cases can be placed definitely in one group. In many cases it is rather a matter of both/and than of either/or.

This seems to be one of the reasons why there are such great differences of opinion over the aetiology of coxa vara infantum.

Legg–Calvé–Perthes' Disease.

Many authors have found that the changes characteristic of Legg-Calvé-Perthes' disease are not limited to the caput epiphysis, but also affect the metaphysis and the acetabulum. These changes in the metaphysis are localized mainly in the superior part, i.e. in the upper limits of the collum when seen in a frontal exposure.

Waldenström, however, has published in the Scandinavian textbook of surgery (Nordisk Lærebok i Kirurgi) a reproduction of a macroscopic specimen of coxa plana in which the changes are situated in the base of the metaphysis, i.e. in the lower border of the collum when seen in the frontal plane.

Camitz insists that there is no difference in the histological aspects of Legg–Calvé–Perthes' disease and coxa vara infantum. His investigations are thorough and of great interest. Coxa vara infantum is still first and foremost a radiological diagnosis, however, and as *Camitz* does not submit any radiogram, it seems to us that the last link in his chain of evidence is incomplete.

Our material includes a total of seven hips representing Legg-Calvé-Perthes' disease which appeared either on the "healthy" side or on the coxa vara side, in 5 cases before the patients came under treatment. In only two cases was the development of the disease postoperative.

It will thus be seen that Legg-Calvé-Perthes' disease occurred in our material in five out of 42 patients, among whom it was therefore more frequent than among the general population. The disease developed more frequently as a spontaneous process than in connection with operative treatment. The coxa vara limb does not seem to be much more predisposed to this disease than the "healthy" limb on the opposite side, the relative incidence of the disease on the two sides being as 4:3.

Thus, it is evident that the development of coxa plana is independent of our treatment. The increased frequency with which it occurs, in some cases on the coxa vara side, in other cases on the other side, may suggest a certain aetiological relationship. But we have been unable to find evidence indicating that coxa vara infantum, as defined by us, is always associated with considerable caput necrosis.

Coxa Vara Adolescentium.

In several cases we have encountered difficulties with regard to the differential diagnosis of coxa vara infantum and epiphyseolysis

adolescentium. There are, however, certain special features which may be useful in distinguishing between the two.

Epiphyseolysis occurs just before puberty and attacks an anatomically well developed collum. Its lime content is good, and also its architectural structure. The epiphyseal line runs horizontally. The osseous border between the epiphyseal line and the metaphysis, as a rule, is well defined and marked in such cases, and the caput nucleus is seen to slip backwards and downwards, a displacement which comes out most clearly when a radiogram is taken in Lauenstein's position. In coxa vara infantum, on the other hand, the collum is hypoplastic, its lime content being poor or irregular, and the metaphysis often tapering. The transition between the metaphysis and the epiphyseal line is apt to be blurred and frayed.

In five cases in our material we have found signs of slipping of the epiphysis in conjunction with marked hypoplasia of the remnants of the collum. We have taken these findings to indicate the concurrence of coxa vara infantum and epiphyseolysis capitis femoris. In one case in particular (Fig. 5, Case 21) the above mentioned condition is well illustrated.

Our material gives support to the opinion that epiphyseolysis capitis and coxa vara infantum are two essentially different conditions. On the other hand, coxa vara infantum, particularly of the hypoplastic type of the disease, predisposes to slipping of the epiphysis just before puberty. We must consider that on account of the disabling strain to which patients suffering from coxa vara infantum are exposed, even a slight disturbance in the hormonal balance may cause increasing failure of the epiphyseal line.

From the aetiological point of view, these two diseases probably have nothing in common, as it may now be considered as proved that epiphyseolysis adolescentium depends on hormonal action.

Congenital Defects in the Upper End of the Femur.

As a rule it is easy to distinguish between coxa vara and congenital defects in the upper end of the femur. Yet, there are transitional forms in which the anatomical structure of the upper end of the femur is fairly normal, but in which its degree of development lags considerably behind that of the healthy limb.

In a few cases the changes are so marked that they must be taken to indicate primary faulty development. We have placed one of our cases (Case 37, Fig. 7) in this group. The patient did not come for treatment until a late stage. The remnant of the epiphysis remains

in the acetabulum and its connection with the trochanter has been broken. We do not know whether this happened before or after birth. This case supports the opinion that coxa vara belongs to the group of congenital defects of the femur. On the other hand, our material gave no data on which to base the opinion that every case of coxa vara infantum belongs to this group.

Heredity.

In several cases we have found a familial predisposition to coxa vara, and two of our patients are father and daughter, the father suffering from unilateral and the daughter from bilateral disease.

However, we have not been able to produce radiological proof of all the assumed cases of familial occurrence of coxa vara in our material. So many familial cases have been reported from other quarters, however, that we have to reckon with a certain familial predisposition, perhaps an endogenous factor.

Traumatism.

Only in two cases in our material did trauma play an important part (Case 3, Fig. 10).

The final course of the disease in one of these cases was exactly the same as that observed in the majority of our patients. The patient developed necrosis of the cranial collum metaphysis directly after the accident. After the resorption of the necrotic area, the radiological picture was typical of that of coxa vara infantum.

The child reacted to subtrochanteric osteotomy in the same way as cases of coxa vara infantum; the whole of the trochanter area developed to a surprising degree and with amazing speed after the operation.

The factors throwing light on the aetiology of coxa vara infantum in our material may be summarized thus:

1. Bone necrosis in the caput epiphysis occurs more frequently on both the coxa vara side and on the "healthy" side than in the normal population.
2. Several cases show a concurrence of coxa vara infantum and coxa vara adolescentium.
3. Transitional forms occur towards the genuine hypoplasia in the upper end of the femur.
4. Heredity probably played a part in the case of the patients who were father and daughter.

5. Severe trauma was undoubtedly responsible for one of our cases.
6. The process in the metaphysis usually ends in recovery, provided that the normal static-functional conditions of stress and strain are restored in the collum femoris.

The collum and the trochanteric region develop much more strongly than before (see the results of treatment to be published in a future issue of Acta Orthop. Scand.). We would like to emphasize specially this last point as it seems to give us the key to the aetiological problems. We believe that coxa vara infantum is due to disturbances of nutrition which may be caused by several factors, some congenital, others acquired, usually a combination of both. The intersecting static-dynamic forces in a region of great importance to upright gait render the pathological process worse because of the varus position of the collum-caput axis, that is, until the static-dynamic balance is successfully restored by operation.

The congenital factors may be classified partly in the dysostosis group, partly in the group referred to by *Tucker* when he speaks of a "concentration of the blood supply to a retinacular group of vessels constituting a potential risk of avascular necrosis".

Partly with, partly without these conditions, trauma seems to play a very important part in the development of coxa vara infantum. Trauma need not necessarily result from a single outstanding event, but may be the outcome of abnormal stress and strain, pressure and the like of a more or less fortuitous character, interfering with the nutrition of the parts involved, or it may consist of a summation of "unterschwellige" irritants conditional on the daily wear and tear on the hip.

In this way a local disturbance of nutrition leads to a breakdown of ossification with a secondary rarefaction along abnormal zones of strain, an increase of decalcification and reactive changes.

It is in this way that floating transitions are found both in hypoplasia of the upper end of the femur and avascular caput necrosis. We consider the concurrence of this condition with epiphyseolysis capitis adolescentium as a fortuitous circumstance. It might perhaps be more correct to say that coxa vara infantum creates conditions which favour the development of an epiphyseolysis rather than a collum whose anatomical structure is apparently normal.

SUMMARY

The author has examined a series of 42 patients with a total of 55 hips, suffering from coxa vara infantum.

The series is classified in three age groups:

(I) Patients under 10 years.

(II) Patients between 10 and 15 years.

(III) Patients over 15 years.

The sex distribution of the disease was found to be approximately even, and its incidence in Norway is calculated as being at least 1:25,000 live births.

The disease was bilateral in 13 cases. Spontaneous recovery is very rare, and as a rule matters become steadily worse from early childhood to the completion of puberty, by which time a complete pseudarthrosis has developed between caput and collum. Legg–Calvé–Perthes' disease was found in seven hips, in four cases in combination with coxa vara.

Epiphyseolysis capitis femoris was found in four cases, always on the coxa vara side.

The changes were so far-reaching in four hips that the condition was transitional to hypoplasia of the upper end of the femur. One patient presented coxa vara on one side and marked hypoplasia of the upper end of the femur on the other side.

In four cases there was a history of severe trauma to the hip, but in only one of these cases was a single severe trauma quite obviously responsible for the genesis of the disease. The author emphasizes the importance of trauma as a contributory aetiological factor. When there is a moderate degree of irregularity in the processes of ossification in the collum metaphysis, the strain entailed by the daily use of the hip may start a vicious circle and prevent the normal development of the collum support for the caput epiphysis. In the opinion of the author, the good results achieved by the operative readjustment of the epiphyseal line give support to the theory that traumatic factors are of importance to the development of the disease and its subsequent course.

The transition to genuine hypoplasia of the upper end of the femur is floating. The comparative frequency with which Legg–Calvé–Perthes' disease and coxa vara infantum occur in one and the same patient suggests the possibility of a genetic factor common to both.

Heredity and concurrence with several developmental anomalies have been found. The author concludes that aetiologically we have to

regard the interplay of several factors as being responsible for the genesis of coxa vara infantum.

ZUSAMMENFASSUNG

Der Verfasser hat eine Serie von 42 Patienten mit insgesammt 55 Hüften, die an coxa vara infantum litten, untersucht.

Die Fälle wurden in drei Altersgruppen aufgeteilt:

(I) Patienten unter 10 Jahren.

(II) Patienten zwischen 10 und 15 Jahren.

(III) Patienten über 15 Jahre.

Die Erkrankung war auf beide Geschlechter ungefähr gleichmässig verteilt und ihr Vorkommen in Norwegen wird als zumindest 1:25,000 Lebendgeburten berechnet.

Die Erkrankung war bilateral in 13 Fällen. Spontanheilung kommt sehr selten vor. In der Regel sieht man eine ständige Verschlechterung vom Kleinkindalter bis zur Vollendung der Pubertät, in welcher Zeit sich eine komplette Pseudarthrose zwischen Caput und Collum entwickelt.

Legg–Calvé–Perthes' Erkrankung wurde in 7 Hüften gefunden, und zwar immer auf der Coxa vara-Seite.

In vier Hüften waren die Veränderungen so weitgehend, dass man sie als ein Übergangsstadium zur Hypoplasie des oberen Femurendes ansehen konnte.

Ein Patient zeigte coxa vara auf der einen und ausgesprochene Hypoplasie auf der anderen Seite.

In vier Fällen wurde über ein schweres Trauma der Hüfte berichtet, aber nur in einem der Fälle konnte ein einziges schweres Trauma mit Sicherheit mit der Entstehung der Krankheit in Zusammenhang gebracht werden. Der Verfasser hebt die Wichtigkeit des Traumas als einen mitwirkenden ätiologischen Faktor hervor.

Wenn ein mässiger Grad von Unregelmässigkeit im Verknöcherungsprozess der Collum-Metaphyse vorhanden ist, kann die Beanspruchung, die durch den täglichen Gebrauch der Hüfte bedingt ist, einen circulus vitiosus hervorrufen und hierdurch die normale Entwicklung der Stütze des Schenkelhalses für die Kopfepiphyse verhindern.

Nach der Meinung des Verfassers machen die guten Ergebnisse, die bei der operativen Wiederanpassung der Epiphysen-linie erzielt werden, es wahrscheinlich, dass traumatische Faktoren in der Entwicklung der Krankheit und in ihrem weiteren Verlaufe von Wichtigkeit sind.

Der Übergang zur genuinen Hypoplasie des oberen Femurendes ist unscharf. Die relative Häufigkeit, mit welcher Calvé–Legg–Perthes' Erkrankung und coxa vara infantum bei ein und demselben Patienten vorkommen, macht einen gemeinsammen genetischen Faktor wahrscheinlich.

Erblichkeit und Zusammenauftreten mit mehreren Entwicklungsanomalien sind gefunden worden. Der Verfasser zieht den Schluss, dass das Zusammenspiel mehrerer Faktoren für die Entstehung der coxa vara infantum verantwortlich ist.

RESUME

L'auteur a examiné une série de 42 malades avec un total de 55 hanches souffrant de coxa vara infantum.

Cette série a été classée en trois groupes d'âge:

1) malades au-dessous de 10 ans.
2) malades entre 10 et 15 ans.
3) malades âgés de plus de 15 ans.

La distribution par sexe de la maladie a été trouvée à peu près égale, et son incidence en Norvège est calculée comme étant au moins de 1 sur 25.000 naissances.

La maladie était bilatérale dans 13 cas. La guérison spontanée est très rare, et d'une manière générale les cas s'aggravent progressivement de la tendre enfance jusqu'au moment de la puberté, époque à laquelle il s'est développé une pseudarthrose entre la tête et le col du fémur.

La maladie de Legg–Calvé–Perthes a été observée dans sept hanches, dans 4 cas simultanément avec le coxa vara.

Dans 4 cas, on a trouvé une épiphyseolyse de la tête du fémur, toujours du côté du coxa vara.

Les modifications étaient si profondes dans 4 hanches qu'il s'agissait d'un état transitoire d'hydroplasie de la partie supérieure du fémur.

Un malade avait le coxa vara d'un côté et une hydroplasie marquée de la partie supérieure du fémur de l'autre côté.

Dans 4 cas, il y avait eu un grave trauma de la hanche, mais dans un seul cas le trauma était apparemment responsable de la genèse de la maladie. L'auteur souligne l'importance du trauma comme facteur étiologique contributoire. Lorsqu'il y a une légère irrégularité dans le processus d'ossification du col, l'effort imposé par l'usage constant de la hanche peut créer un cercle vicieux et empêcher le développement normal du col supportant l'épiphyse de la tête. Dans l'idée de l'auteur,

les bons résultats obtenus par le réajustement opératoire de la ligne épiphyséale viennent appuyer la théorie que les facteurs traumatiques présentent de l'importance pour le développement de la maladie, et son cours subséquent.

La transition à l'état d'hypoplasie de la partie supérieure du fémur est flottante. La fréquence relative de l'apparition simultanée de la maladie de Legg–Calvé–Perthes et du coxa vara infantum chez un même malade permet de supposer l'existence d'un facteur génétique commun aux deux.

On a découvert des facteurs héréditaires et des rapports avec plusieurs anomalies de développement. L'auteur conclut qu'au point de vue étiologique il faut considérer que plusieurs facteurs sont responsables de la genèse du coxa vara infantum.