ACTA ORTHOPAEDICA SCANDINAVICA · SUPPLEMENTUM XII

From the Orthopaedic Clinic of Karolinska Institutet, Stockholm.
Chief: Professor S. Friberg

COXA PLANA

A HISTO-PATHOLOGIC AND ARTHROGRAFIC STUDY

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STOCKHOLM 1953

ESSELTE AKTIEBOLAG
STOCKHOLM 1953
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Preface

During discussion of roentgen pictures of coxa plana, Professor Folke Knutsson, at that time chief of the Roentgen Department of the Orthopedic Clinic of the Karolinska Institutet, pointed out that we lack knowledge of the relation between the roentgen pictures and the histologic changes in this disease and thus gave me the impulse to start this investigation.

My chief, Professor Sten Friberg kindly gave his consent to the investigation and later took great interest in my work. He let me benefit by his great clinic and scientific experience and with his friendly advice helped me to get over many difficulties on my path.

Professor Olof Reuterwall has been amiable enough to allow me to perform the histological work at the Radiopathologic Institution of Konung Gustaf V:s Jubileumsklinik. Miss Eva Schultz made the microscopic slides.

Professor Åke Wilton, chief of the Pathologic Department of the Karolinska Institutet gave me very valuable assistance at the final interpretation of the histologic material going through the manuscript and putting at my disposal his great experience as bonepathologist.

During the work I got valuable advice from Docent Bengt Engfeldt in evaluating the histologic pictures.

Even after leaving our clinic Professor Knutsson has given me opportunities to discuss the arthrographic pictures, which has been of great importance for my work.

The translation was carried out by Docent Max Gorosch, Ph. D. and Captain Jack O'Brien-Hitching, M. C.

To all those who have helped me i want to express my sincere thanks.

Stockholm in March 1953

Stig Jonsäter

CHAPTER 1

Historical Review

In the year 1909, H. WALDENSTRÖM described a diseased condition of the hip-joint in children which he called "the upper tuberculous focus in the collum". On the basis of ten cases, all of them under the age of nine years, he gave an account of the roentgenological changes consisting of a focus beneath the epiphysial cartilage that was often connected with a focus in the epiphysis. The complaint manifested itself in a series of insignificant pains, intermittent limping and restricted mobility in the hip-joint more especially in regard to inward rotation and abduction. Waldenström pointed out that the process never passed over to the joint but was prevented by the joint cartilage which formed an impenetrable cover. In a couple of cases he was able to observe a healing accompanied by a complete disappearance of the focus, but with a flattening of the epiphysis as a final result.

All cases reacted positively to tuberculine; in one or two cases he obtained focal reactions to tuberculin tests, and in a couple more there was a manifestation of enlarged glands in the groin, all of which made Waldenström presume that the disease was of a tuberculous nature.

The following year (1910), WALDENSTRÖM gave a more detailed report of the disease in his dissertation, "Die Tuberkulose des Collum Femoris im Kindesalter und ihre Beziehungen zur Hüftgelenkentzündung". The material was then enlarged by two more cases and the disease was still considered as being a tuberculous manifestation.

The same year, and independent of one another, three authors reported a hip disease in children with a symptomatic picture resembling that described by Waldenström.

CALVÉ (1910) had observed 10 cases during a period of 3 years, about which he gave a detailed report as to the clinical and roentgenological findings. Clinically the disease was characterized by pains, limping, restricted movement in the hip-joint especially in regard to abduction, and a palpable enlargement of the caput femoris. The roentgen pictures showed the coxa-vara position, hypertrophy of the head and collum together with atrophy and plate-shaped deformation of the epiphysis and in certain cases insular bone formations of the epiphysis, which later on became confluent. Calvé emphasized strongly that the surface of the cartilage was intact. He described

the disease as being arthritic of a chronic or sub-acute nature, and after discussing several alternatives, came to the conclusion that the cause should be looked for in an abnormal or delayed osteogenesis. Calvé's roentgen pictures are hazy in reproduction and give very little information, but he has excellent sketches of the pictures with the details showing up clearly.

LEGG (1910) reported on 5 cases between the ages of 5 and 8 years. One of these cases had been operated on by Goldthwait by curetting a focus in the collum immediately below the epiphysial cartilage. Cultures showed a growth of staphylococci. Legg was most inclined to believe that the disease was caused by trauma because he had found such in all of his cases. Trauma would have caused a decrease of the blood supply to the epiphysis accompanied by a subsequent deformation of the bone nucleus. The focus in the collum was, in his opinion, due to hyperaemia caused by trauma.

PERTHES (1910) had collected 6 of his own cases, one of which was bilateral. In his first work he calls the disease "Arthritis deformans juvenilis" and describes it in approximately the same way as the previously mentioned authors did. He thought that the most typical thing about the roentgen picture was the decrease in the height of the epiphysis. He compared the head to a frustum of a cone or to a cylinder, and considered this deformation as being typical for the disease in its beginning. He also showed the lighter areas in the epiphysis and considered them as being resorption foci, which when shrinking caused the deformation of the bone nucleus. In one or two cases Perthes showed secondary changes in the socket that adapted itself to the form of the head. The most important clinical symptoms were the restricted movement and faulty position of the hip, of which the restricted movement was caused by the deformation of the joint head alone, while reflex muscular spasms and joint excitation did not play any part at all. Perthes was of the opinion that the deformation assumed the appearance described by him because the hip mainly performed flexion and extension movements during the time when the epiphysis was soft and shrunken. With regard to the etiology, Perthes denied the importance of trauma because he had not found any such in his cases. He inclined more to the opinion that the disease was the result of an inflammatory process in the joint that had occurred during the years of infancy.

The works of these four authors are the first reliable reports of the disease that is now usually known as coxa-plana, Calvé-Legg-Perthes-Waldenström's disease. As will be seen the description of the disease that has been given by all seems to agree fully in all essentials, but they have each different conceptions of the nature of the affliction.

During the following years the disease, which I shall call coxa-plana in in this work, aroused a lively interest, and numerous works were published about it both in Europe and in America. Most of them, however, were short

reports about a few cases and are no longer of any great interest. It is my intention in this study to engage myself only with the works that will throw new light on the nature of the disease and especially those that deal with its pathological anatomy.

In 1913, PERTHES published a new work entitled "Ueber Osteochondritis deformans juvenilis" in which he characterizes the disease as "ein durch subchondrale Destructionsherde bedingter, im Laufe von Jahren sich vollziehender eigenartiger Schwund der oberen Femurepifüse". In this work he describes the histological picture for the first time. Perthes had obtained the preparation by excising a piece of the head from a 9 year old boy who had suffered from the disease for 2 years. He stated that the joint cartilage had a thickness of 3 to 4 mm throughout the section. He found numerous cartilaginous islands in the bone under the cartilage, some of which were connected to the joint cartilage by "strings". The bone was of the same hard consistency as that of healthy bone, and histologically, the joint cartilage had the appearance of hyalin cartilage with good, stainable nuclei, while the cartilage in the inner part of the epiphysis was of a more thready structure. There were strikingly wide marrow-spaces with fat marrow. There were no signs of inflammatory processes to be seen anywhere. Perthes considered that the cartilaginous islands in the bone were due to a new cartilaginous formation for which reason he called the process osteochondritis and thereby considered its nature as being non-tuberculous, but, at the same time, emphasized the agreement with the clinical and roentgenological picture described by Waldenström.

The following year (1914), a new work on coxa-plana came from Perthes' clinic written, this time, by his assistant, SCHWARZ, who also made a report on the histological findings in a case, a 7 year old boy who had been afflicted for 4 years. The histological picture was described in a similar manner to Perthes' case, but Schwartz drew attention to the fact that, in some parts of the epiphysis there were trabeculae that did not form a network but lay like short, spinous formations in the preparation.

In 1918, EDBERG had an opportunity of examining histologically a preparation which he had obtained by excising a specimen from a 7 year old boy who had had hip trouble for 2 years. Edberg found the bone rich in blood in both the collum and the head. The epiphysial cartilage was normal and approximately 3 mm in thickness. On the surface of the specimen he observed bone islands that were embedded in the cartilage proximate to its surface. No signs of inflammatory changes were manifest anywhere. The joint cartilage was normal in appearance and the bone was rich in red bone marrow and fat marrow.

In 1920 WALDENSTRÖM published an article dealing mainly with the denomination of the disease. Till then there had been great confusion in this respect,

and the disease was known by different names in different countries, often after one of the authors who had described it. This latter system seemed unpractical to Waldenström. The name suggested by Perthes, "osteochondritis deformans coxae", which was often used in the German literature, he thought was faulty because the histological examinations that had been made up to then showed that the disease was not of an inflammatory nature. Instead Waldenström suggested the name "coxa-plana" owing to the deformation of the hip that takes place during the more advanced stages of the disease, and in analogy with other deformations of the joint, coxa-vara and coxa-valga. The suggestion did not gain immediate favour, but, by degrees, the name forced its way into the literature and is now used by most authors either alone or in combination with some personal name.

The same year, 1920, the first real monograph about coxa-plana was published by the Norwegian author SUNDT. In his historical survey, he gave an account of most of the works on the subject that had been published up to then, and suggested that the disease should be called "Malum coxae Calvé-Legg-Perthes". Sundt's material comprised 66 cases that had been examined from many different points of view, among which that of age, pains, restriction of movement, limping, social conditions, locality (town or countryside) and so on. On the whole his observations coincided with those made by previous investigators. He referred to the histological investigations made by Perthes and Edberg, and gave as the essential feature in the pathological anatomy of the disease, the cartilaginous islands that had been observed in the epiphysis, and expounded several theories as to the origin of these formations. As far as the nature of the disease was concerned, he was more inclined to think that it was an "osteodystrophy" due to a "dysendocrinia" of hereditary disposition. If a person so disposed were to be exposed to an injury to the hip (trauma, infection, etc), Sundt is of the opinion that the disease would be manifest and give rise to the characteristic changes.

The first one who seems to have realized that coxa-plana was necrosis of the bone, or at least who has stated it clearly, was PHEMISTER who, in 1921, gave a report of a case that had been operated on after having been subject to trouble in one hip for 8 months. Phemister exposed the joint and made an opening in the anterior part of the head through which the epiphysis was curetted. To a large extent the epiphysis was damaged by necrosis and compensated by granulation tissue. Here and there could be seen remnants of old living bone as well as, in places, newly formed bone. In addition there were giant cells of osteoclastic type. Phemister interpreted the changes as being of an inflammatory and infectious nature, even if cultures and guinea-pig tests of the curetted material gave a negative result.

In 1922 WALDENSTRÖM had had occasion to follow the course of 22 cases right from the beginning of the disease until the growth was completed.

The same year he made a report of his experiences in an article. The definite form of the head had been reached before the growth had been completed. The disease ran its course in a typical manner no matter how it was treated. Waldenström classified it into 4 periods mainly from the roentgenological point of view.

I. The period of development comprised 3—4 years, and was sub-divided into 2 stages:

A. The Initial stage (½—1 year), where the epiphysis is dense, flattened and uneven at the edge. The collum often manifests decalcifications in the region proximate to the epiphysial line. The joint cartilage is of ordinary height.

B. The Fragmentation stage (2—3 years), where the epiphysis is markedly flattened and divided into many pieces. Atrophy of the bone.

II. The Healing period comprising 1—2 years. The epiphysis becomes homogeneous and the borders visible.

III. The Growing period, which continues until the normal growth has been completed. During this period the head attains its final shape.

IV. The Definite period.

This classification seems to be the most logical one, and has been used by numerous authors when it was a matter of stating the different phases of the disease.

After Phemister had pointed out necrosis as being the essential factor in the patho-anatomic picture, it was not long before several authors published works which confirmed the observations, among whom are Riedel and Lang (1922) and Axhausen and Heitzmann and Engel (1923). Riedel's paper is worthy of interest in so far as that it gives a description not only of the bone necroses but also of the changes occurring in the cartilage. The latter consisted for the most part of unchanged hyalin cartilage, but here and there one found a cell proliferation with oedematous swelling and a thready transformation of the hyalin basic substance, as well as, in places, a high degree atrophy of the cell nuclei. The normal fat marrow had been turned into a fibrous marrow which, in places, was poor in cells, and in others it was the seat of an infiltration of inflammatory cells and, here and there, passing over to a granulation tissue where polynuclear giant cells were arranged in heaps in the depressions in the remaining trabeculae, as a sign of a lacunar resorption. In other places there was a profusion of osteoblasts denoting a new formation of bone. But Riedel was unable to find any bacteriae in the sections; to judge from the histological picture, however, he thought, like Phemister, that the disease was of an inflammatory nature.

Axhausen described the histological picture in approximately the same way as Riedel did. On the question of etiology he offered opposition to the

authors who were of the opinion that the disease was of a traumatic nature. On test animals he had produced wedge-shaped fractures on the epiphysis. All the cases healed without any manifestation of bone necrosis, which caused him to draw the conclusion that trauma played no part in the origin of the necrosis. Instead he propounded the theory that the necrosis was due to a circulatory disturbance caused by a bacillary embolism, where the infection had either not got manifest, or had been so weak that it healed quickly.

During the years immediately following it seems that the question of the etiology of coxa-plana has dominated the discussions. In 1925, MURK JANSEN was of the opinion that the cause was a congenital flattening of the joint socket, which caused an incongruence to arise between the head and the socket. The uneven loading of the head brought about in this way caused vascular damage with consequent necrosis of the bone.

BENTZON (1926) emphasized the importance of the cartilaginous insulae in the epiphysis whose existence preceding authors had shown, and put forward the theory that the disease could be due to a hyperaemia caused by small traumata which had not blocked the vessels but had paralyzed the vasoconstrictors.

ROCKEMER (1927) followed the same lines of thought. He found necrosis in one of his own cases and referred to previous examinations with the same findings, but did not consider that the necrosis was ischaemic but was rather haemorrhagic due to inhibited flow of venous blood from the epiphysis.

CALOT (1929) allied himself more closely to Murk Jansen's theory and thought that he could conclude that all the cases described up till then were merely overlooked congenital subluxations and founded this idea on the statement that one could not show one single case where the roentgen picture proved that the hip had originally been normal.

HEILGENREINER (1923) would like to attribute the cause of the disease to an inhibitive malformation of the same nature as the congenital dislocation and subluxation of the hip but of a lower degree, and considered that the later appearance of the changes in coxa-plana was due to the fact that these hips had not been exposed to the trauma which the reposition entails.

In 1934, WALDENSTRÖM, in his paper "The First Stages Of Coxa-Plana", described 10 early cases, of whom none had been afflicted for a longer period than 5 months. He thought it probable that the necrosis gives symptoms right from the beginning, and that the roentgen picture in this stage can still be normal. The increased density of the epiphysis in the initial stage might be due to the bone beginning to shrink. The lighter areas in the fragmentation stage might be caused by the resorption of the bone. Waldenström also described 2 early symptoms, the subchondral thinning and the increased

head-socket distance, the former due to resorption of necrotic bone and the latter due to the fact that the head decreases in volume, whereby the intra-articular pressure decreased with an increase of the blood supply to the soft tissues in the fossa acetabuli as a consequence, and these swollen soft tissues pressed the head laterally.

FERGUSEN and HOWORTH (1934) had operated on 21 hips with coxa-plana by drilling up into the epiphysis. They had exposed the hip-joint and found in the active stage the synovial membrane thickened, soft and vascularized besides being often irregular with villus formations. The periosteum was thickened and oedematous, the cartilage and synovia being, on the contrary, normal. Microscopically the synovial membrane, the capsule and the periosteum showed signs of chronic inflammation. In later stages the soft tissues were taut, thin and sclerozed. Microscopically there was a manifestation of fibrosis with obliterated vessels. No report was made in this work about any histological examinations of the bony tissue, but the bone resistance in drilling was said to be normal. Also the majority of the cases in the later stages manifested a normal hardness of the bone, but in some cases there were regions of a softer nature, and these regions were considered as being representative of the more advanced stages of the disease. Ferguson and Howorth, on the basis of their investigations, considered that most cases of coxa-plana were due to a slightly infectious arthritis that caused a sclerozing of the capsule and the synovial membrane accompanied by obliteration of the vessel and ischaemia of the bone as a consequence and this all the more so as 75 % of their cases had had an infection not long before the appearance of the symptom.

LERICHE (1934) after having carried out a series of tests on animals arrived at approximately the same result as Bentzon in regard to the etiology. In a group of test animals he had severed the supplying arteries without getting any roentgenological or histological changes in the epiphysis. In another group he injected sodium salicylate around the collum in order to bring about a blocking of the vessels, but even this did not give him any epiphysial changes. In the third experimental group he crushed the ligamentum teres and obtained in the roentgen picture a flattening of the epiphysis, but no changes in the density of the bone. During the histological examinations the vessels in the ligamentum teres were obliterated, the joint cartilage was changed round the attachment of the ligament, but there was no sign of bone necrosis in the epiphysis. By extreme rotation of the hip in the fourth group he brought about a stretching of the ligamentum teres. The roentgen picture showed no changes, but in the histological examination fewer and more thinly scattered trabeculae were observed than on the control side in addition to a marked hyperaemia. And in the fifth group, too, in which he had injected alcohol of 95 % strength around the

collum he obtained, in the histological examination, a pronounced hyperaemia together with narrower and fewer trabeculae. The roentgen picture manifested thinnings in the bone. In the sixth group, finally, he injected ammonia in the same manner in order to obtain a strong toxic effect. The roentgen picture showed necrosis around the place of injection but otherwise nothing of note. The histological examination showed the epiphysis to be richer in marrow than usual besides being rich in blood.

From these experiments, then, Leriche came to the conclusion that the changes in coxa-plana should be due to hyperaemia, in all probability caused by trauma that was either one great trauma or a repetition of lesser traumas.

Axhausen's theory about mycotic emboli in the capsular vessels being the cause of coxa-plana was also embraced by KONJETZNY (1934) who discussed the question on the basis of 2 cases which he had examined histologically. He showed the presence of bone necrosis and giant cells as well as the same changes in the joint capsule as those found by Ferguson and Howorth.

In 1938, WALDENSTRÖM, after examining his bi-lateral cases, was able to conclude that coxa-plana arises in a primarily normal hip and so refute the statement that these hips are the seat of congenital changes that produce or favour the origin of coxa-plana.

GALL and BENNETT (1942) became the advocates of the theory of ischaemia being the cause of coxa-plana, and in support of this opinion they pointed out that when carrying out their histological examinations they had discovered pronounced necrosis in the epiphysis, whereas there were no changes in the joint cartilage. This circumstance should be due to the fact that, to a large extent at least, the joint cartilage got its nutrition from the synovia.

Quite early the opinion was suggested that metabolic disturbances should be the basis for the occurrence of coxa-plana, and hypothyreoidism came to take a dominating position in the discussion, especially due to a work written by CAVANAUGH, SHELTON and SUTHERLAND (1936). They had examined 5 cases, all of whom manifested signs of hypothyreoidism. This was opposed by GILL (1940 and 1943) who, in a series of 20 children with coxa-plana, had carried out all the examinations that usually form the basis for diagnosing hypothyreoidism. He was unable to find any sure signs of lack of function in the thyroid gland. Gill became an adherent of the ischaemic theory but considered that the primary localization of the changes was the metaphysis and the epiphysial cartilage, and that the necrosis in the epiphysis was secondary to these changes.

In 1948, HOWORTH published a new work on the examinations carried out by him on 50 cases that had been operated on by drilling. The histological report is approximately the same as that in his earlier work in collaboration with Ferguson.

HAYTHORN (1949) had operated on 33 cases by exposing the hip-joint

and curetting the epiphysis. The curetted material is described as being soft, necrotic and containing bony spicules as well as bits of cartilage. He classified the cases in accordance with the predominant changes in the following manner:

I. Degenerative changes: Necrosis or necrobiosis was manifest in all of the cases. In some of them there were no signs of reparation, but in others there was a fibrous tissue infiltrated with a few lymphocytes and mononuclear cells. There were so few segmented leucocytes that infection could not be suspected.

II. Crushing of the necrotic areas: In 6 cases there was a marked crushing and conglomerating of the degenerative elements. There was no pressing together of the head in these cases which, in Haythorn's opinion, made the changes difficult to explain.

III. Reparative processes: Side by side with the degenerative changes there were regions that had been regenerated by means of a fibrous replacement of marrow, and, in places, bands of compact fibrous tissue.

IV. Giant cell and cystic reaction: In 12 cases there was a manifestation of giant cells of the foreign-body type.

V. Cartilaginous changes: The cartilage cells had partly lost their polarity and normal arrangement. In some places the cartilage was saturated with arterioli. Such changes were manifest in both the epiphysial and joint cartilage.

VI. Vascular changes: As a rule no vessels were found in the degenerated regions, but capillaries and thick-walled arterioli were found in such regions where there was regeneration. No signs of embolism or thrombosis were found in the vessels.

VII. Similarity with scurvy and rachitis: Some pictures with incomplete ossification as in rachitis were described.

Haythorn gave no descriptions of the roentgen pictures. He finished his article with a statement that there was no relation between the age of the patient, the manifestation of the symptom and the nature of the changes.

CHAPTER 2

The Aim of the Examinations

Despite the large number of investigations and the profuse flora of literature, our knowledge of coxa-plana is still in many ways defective. This applies especially to the etiology about which opinions are still in opposition to each other, and where no really definite proofs have been established for one or other of the many theories that have been propounded.

The fundamental principles of the pathological anatomy are relatively well known, and at present the majority of pathologists and orthopedic surgeons will be unanimous in their opinion that the essential part of the pathologic histology of the disease is the necrosis of the bone and that other manifested changes are secondary to this necrosis. All investigators, however, when describing the pathologic changes, have treated the disease as a unity and, as far as I know from the literature, no one has systematically classified the changes in accordance with the different roentgenological stages which, nevertheless, are well known and generally characteristic.

The shape of the head has become an object of numerous studies. With the exception of a few resectional and post-mortem preparations, all of which represented relatively advanced stages, it is only the bone head itself, that is to say, that part of the caput femoris that is roentgen-opaque, that has been judged. No series dealing with the conditions prevailing in the joint cartilage and the "cartilaginous head" have been encountered in the literature.

The present investigation will treat two main problems:

1. *Which histological changes correspond to the different roentgen stages?*
2. *What appearance has the "cartilaginous head" in the same stages?*

When evaluating the roentgenological stages I have followed Waldenström's classification of the initial stage, the fragmentation stage, the reparative stage, the growing stage and the definite stage.

All cases where the roentgen picture shows an increase in the head-socket distance, a subchondral thinning as well as a dense and at times shrunken epiphysis, but where the epiphysis is still homogeneous and where one cannot observe any thinning zones, have been placed in the initial stage.

The fragmentation stage comprises all cases where the epiphysis presents alternately dense and thin areas but where normal bony structure has not yet begun to return.

The reparative stage includes cases where the roentgen picture shows a reappearance of normal bony structure together with a disappearance of the sclerotic areas.

For the sake of simplicity Waldenström's two last stages have been made into one and called the definite stage. I consider this justifiable as Waldenström himself has pointed out that the head in this stage assumes its definite form despite the fact that the growth has not been completed. Cases belonging to this category are characterized by the fact that, in all places, the bone epiphysis has an even, normal bony structure and an even line of demarcation towards the joint cartilage.

It must be pointed out here that these stages are not sharply separated from each other but that the roentgen picture successively assumes the appearance that characterizes the new stage, and that the demarcation between the stages becomes more clear the longer the interval is between two roentgen examinations. Where there has been any doubt as to the evaluation, the case has been placed in the stage where the characteristic has, to the largest extent, been included in the picture.

PART I

HISTOLOGICAL EXAMINATIONS

CHAPTER 3

Material

The histological examinations have been carried out on preparations obtained from biopsies on patients that were admitted to the Orthopedic Clinic of the Karolinska Institutet for treatment. The material comprised 34 patients of whom 26 were boys and 8 were girls. At the time of the examination the youngest patient was 3 years and 2 months of age and the oldest was 11 years and 3 months. Seven of them manifested changes in both hips, while 27 had unilateral changes, and in all of them the diagnosis coxa-plana had been roentgenologically confirmed. The material does not include any case where changes of any other kind (dislocation, sub-luxation, coxa-vara, etc.) were suspected or confirmed in the hips.

In some of the cases biopsy was carried out on the same side during different stages of the disease, and some of the bilateral cases were examined on both sides. Table I shows a more detailed classification of the biopsies carried out on the different patients.

Table I. Number of biopsies in the investigated material

	Number of patients		Number of biopsies
	♂	♀	
Bilateral cases with 1 biopsy	1	—	1
Bilateral cases with 2 biopsies on the same side	—	1	2
Bilateral cases with 1 biopsy on each side	3	—	6
Bilateral cases with 2 biopsies on one side and 1 biopsy on the other	2	—	6
Unilateral cases with 1 biopsy	18	7	25
Unilateral cases with 2 biopsies	2	—	4
Total number	26	8	44

The cases have not been selected in any other way than that approximately the same number of biopsies have been carried out during the different roentgenological stages. The material has been collected during the years 1946 to 1948, but does not include all the patients that were treated during this period as, in many cases, the parents objected to the examinations being made. For the same reason I have not been successful in getting any patients in whom the disease had reached the definite stage.

All in all, 44 biopsies were carried out.

As a normal material, preparations have been taken from children of the same ages, in which coxa-plana usually appears, and who had died from other diseases, or as the result of accidents. In order to preclude the possibility that there was some hip disease, the hips that had been removed were roentgenographed before the preparations were taken out. And also from these normal hips a preparation was taken with the same technique as was used for coxa-plana hips, in order to obtain fully analogous conditions.

CHAPTER 4

Technique

The biopsies were carried out under general anaesthesia and undertaken with a hollow needle, the inner diameter of which was 2 mm. In the beginning a needle with a cutting edge was used, but it was incapable of penetrating the bone in the epiphysis which was often hard, for which reason it had to be changed for a needle with a sawing edge. The needle is fitted with a mandrin which completely fills its lumen and the cutting point of which reaches to approximately 2 mm beyond the edge of the needle.

An incision of about 5 mm in length is made with a narrow lancet in the skin immediately above the trochanter major into which opening the needle with its mandrin is inserted in the estimated direction towards the epiphysis until one feels the elastic resistance offered by the joint capsule. After roentgen exposures a number of corrections have had to be made as to the position and direction of the needle, before one has succeeded in getting the needle to point to the often very low epiphysis above the epiphysial line. After this the needle has been bored through the joint capsule and into the joint cartilage.

Following a further roentgenological control, the mandrin is withdrawn and a drilling chisel, similar to that used by carpenters and which transmits a backwards and forwards movement in a rotary movement, is attached to the needle. In this manner the needle is then bored into the bone epiphysis for about 1 cm, thus cutting out of the bone a cylinder of the same length. Before the needle with the preparation is withdrawn, a roentgen picture is taken in order to show the position of the needle as well as that of the preparation in the epiphysis. (Fig. 1).

As a rule the preparation has consisted of a connected cylinder of bone with, in most cases, a piece of the joint cartilage in the one end. The preparation thus obtained is fixed in a solution composed as follows:

Sublimat	235 g
Water	3,760 g
Formalin	1,000 g
Concentrated Acetic Acid	200 g

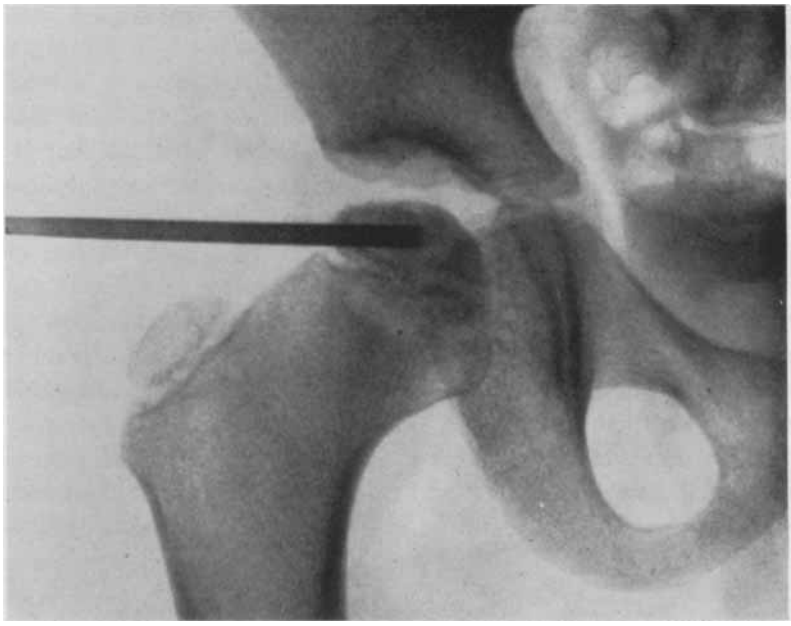

Fig. 1. The puncture needle is drilled into the epiphysis from the lateral side. The picture was taken immediately before the needle with the inlying preparation was withdrawn.

The bone is then decalcified in a solution consisting of:

Alcohol 95 %	150 ml
Chromic acid 0.5 %	150 ml
Nitric acid 10 %	200 ml

After this the preparation was embedded in paraffin and series of sections were cut lengthways. These have been stained partly with hematoxylin-eosin, partly in accordance with van Gieson. Some of the preparations were lime-stained in accordance with Bock's method.

As will be seen, this examination was only undertaken in order to discover the histological conditions of the bone epiphysis and, to some extent, also of the joint cartilage, because it is my opinion that the essential processes take place in the epiphysis. It had also been valuable to study the metaphysial changes and the changes there might be in the epiphysial cartilage, but I have refrained from extending my investigations also to these regions because I was unwilling to subject the patient to traumatizing more than was absolutely necessary.

Naturally a preparation of this size does not give a satisfactory idea about the appearance of the whole epiphysis in each individual case, but the sum total of the observations ought to give a picture of the conditions of the disease during the different stages.

The morphological changes manifested during the histological examinations have been put up in tables and graded 0 ; + ; ++ ; +++ , where 0 signifies no manifestation, + a sparse manifestation, ++ a moderate manifestation and +++ a profuse manifestation of the morphological elements in question. This grading is naturally not absolute, but is only an endeavour to show the relative manifestation of the morphological structures.

After the puncture, one of the patients showed signs of coxitis with a rise in temperature and increased pains and restricted mobility in the hip-joint. The exudate in the joint was slightly cloudy, but cultivation showed no growth of bacteriae. The symptoms receded rapidly with sulfa and penicillin treatment, and there was never any manifestation of roentgenological signs of coxitis.

Otherwise none of the patients showed any reaction after puncturing, nor did it influence the clinical and roentgenological course, as I was able to convince myself of by following up the cases for at least 4 years after the examination.

CHAPTER 5

Results

The initial stage

A. Bony changes

Fourteen biopsies were undertaken in this stage. Prior to the puncture, the patients suffered from the trouble for periods varying from half a month to ten months with an average of 3.5 months. The onset of the trouble has been counted from the time when the child first complained about having pains in the hip or when it was noticed that it limped.

In the majority of cases the bone, in the initial stage, has been soft, which could be noticed from the small resistance it offered, when the puncture needle was bored in. In only two cases has the bone been hard, and in a further two cases one encountered alternating soft and hard parts while drilling. This variation in the hardness of the bone might be explained by the degree of decomposition within the bony tissue. This is also confirmed by the macroscopic appearance of the removed preparation. The soft bone had the appearance of being a greyish-white, soft, granular mass. And the harder preparations, too, have had a greyish-white appearance that diverged largely from the red-brown colour of normal cancellous bone.

The histological picture in this stage is dominated by a marked necrosis of the bone. In some cases it has proved difficult to decide whether the bone is vital or not. When evaluating I have resorted to the usual criteria of necrosis of the bone as propounded by, among others, AXHAUSEN and BERGMANN (1937), and mainly judged the bone from the conditions of the nuclei. Therefore I have considered a bone as being necrotic when the nuclei have manifested pyknosis, karyorrhexis or total karyolysis. I have not made any endeavours to differentiate between necrobiosis and necrosis because, whithout any definite border, the latter arises from the former and forms the final stage of the dying off of the tissue.

Necrosis of the bone was manifested in all cases belonging to this group, and in the majority of the preparations it has been of high degree. In 11 cases it has been classified as $+++$, in 2 cases as $++$, and in one case only as $+$. The bone in the preparations gives the impression of being quite dead. In some preparations one can still see preserved trabeculae, but the

Table II. Showing age, sex, duration of trouble of the cases and bone-changes in the initial stage

Case No.	45	24	1	9	21	29	35	38	39	41	25	3	20	34
Age in years	9	9 1/2	5	4 1/2	4	8	7 1/2	5	4	6	4 1/2	6 1/2	4	4 1/2
Sex	♂	♂	♂	♂	♀	♂	♂	♂	♀	♂	♂	♂	♀	♂
Trouble in months	1/2	1	2	2	2	2	3	3	3 1/2	4	5	6	6	10
Bone necrosis	+++	+	+++	++	+++	+++	+++	+++	+++	+++	+++	+++	++	+++
Normal bone	0	++	0	+	0	0	0	0	0	0	0	0	++	0
Normal bone marrow	0	0	0	0	0	0	0	0	0	++	0	0	0	0
New-formed bone	0	0	0	0	0	0	0	0	0	++	0	0	0	0
Connective tissue	0	0	0	0	0	+	0	0	0	++	0	0	0	0
Vessels	0	0	0	0	0	0	0	0	0	+	0	0	0	0
Osteoblasts	0	0	0	0	0	0	0	0	0	+	0	0	0	0
Giant cells	0	0	0	0	0	0	0	0	0	0	0	0	0	0
Haemorrhage	0	0	0	++	0	+	0	0	0	+	+	0	0	0
Lymphocytes	0	+	0	0	0	0	+	0	0	++	0	0	0	0
Leucocytes	0	0	0	0	0	0	0	0	0	0	0	0	0	0

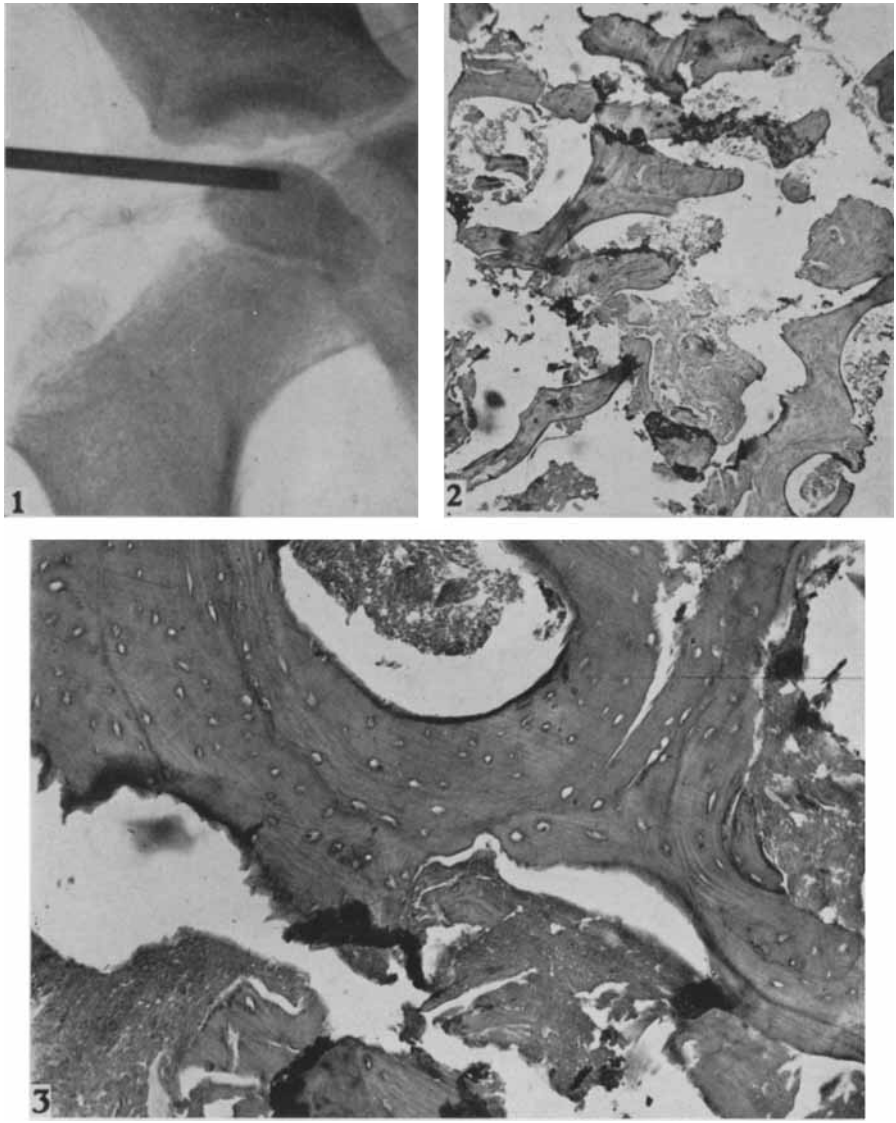

Fig. 2. Case 1. The initial stage.

1. Roentgen: The epiphysis is condensed. No deformation.
2. Microphotograph. (×35). The trabeculae are broken. The marrow space is filled with an amorphous mass consisting of necrotic marrow and bony fragments. No visible normal marrow. There are no signs of reaction in the bone.
3. (×100). The bone is totally necrotic with empty lacunae. The dark lines in the trabeculae are contour lines denoting that the bone has been rebuilt before necrosis became total.

nuclei are pyknotic or entirely absent (see fig. 4), for which reason it must here be a question of necrotic bone as well. In some places the trabeculae are broken off and split up into small fragments. The marrow spaces, in so far as they are preserved, are filled with a structureless mass consisting of necrotic bone marrow and small pieces of bone without nuclei. This amor-

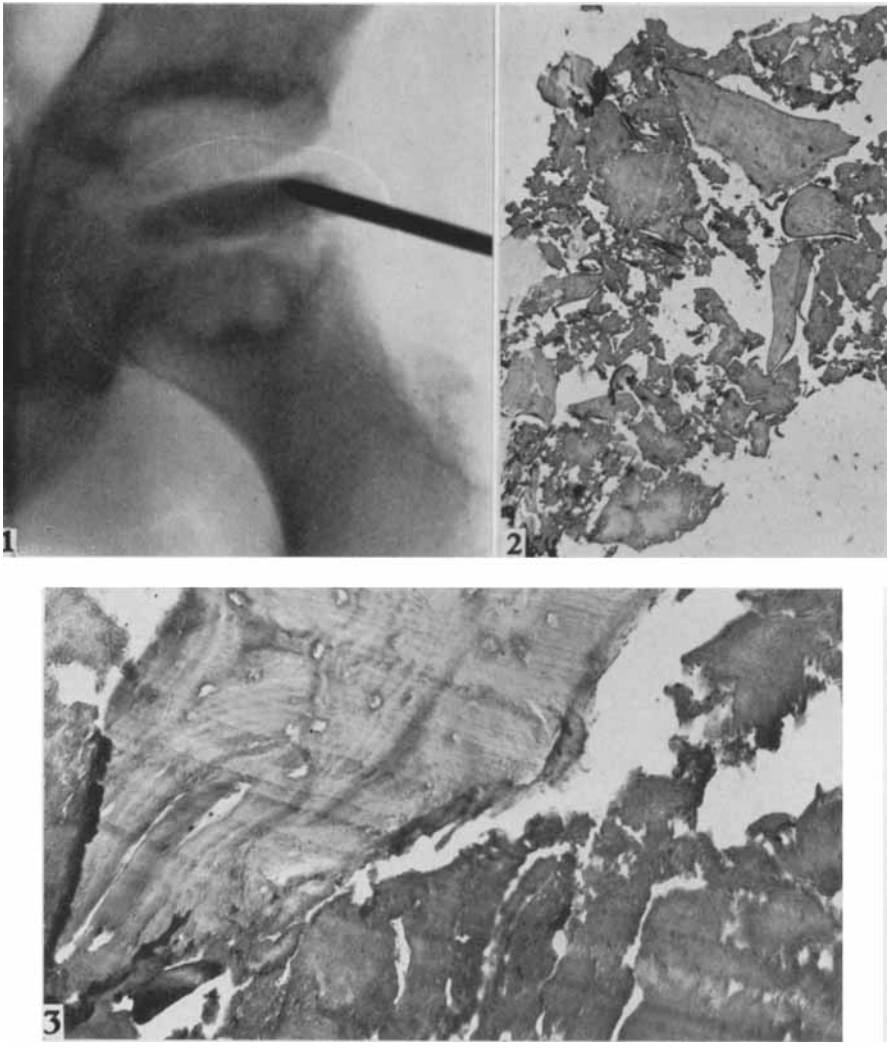

Fig. 3. Case 3. The initial stage.

1. Roentgen: The epiphysis is markedly dense and compressed.
2. Microphotograph. (×35). The bone is totally necrotic with crushed trabeculae. No bone marrow is visible. No signs of reaction.
3. (×200). The nuclei are pyknotic or completely absent. On some places there are contour lines. Amorphous, necrotic masses surround the trabeculae.

phous mass has been called by German authors (Axhausen and Bergmann, 1937, and Haslhofer, 1937, and others) "Trümmermehl", and is thought to arise from the fact that the trabeculae, under the influence of loading and movement, have been pressed into one another and ground to bits until they have become a fine-grained pulp.

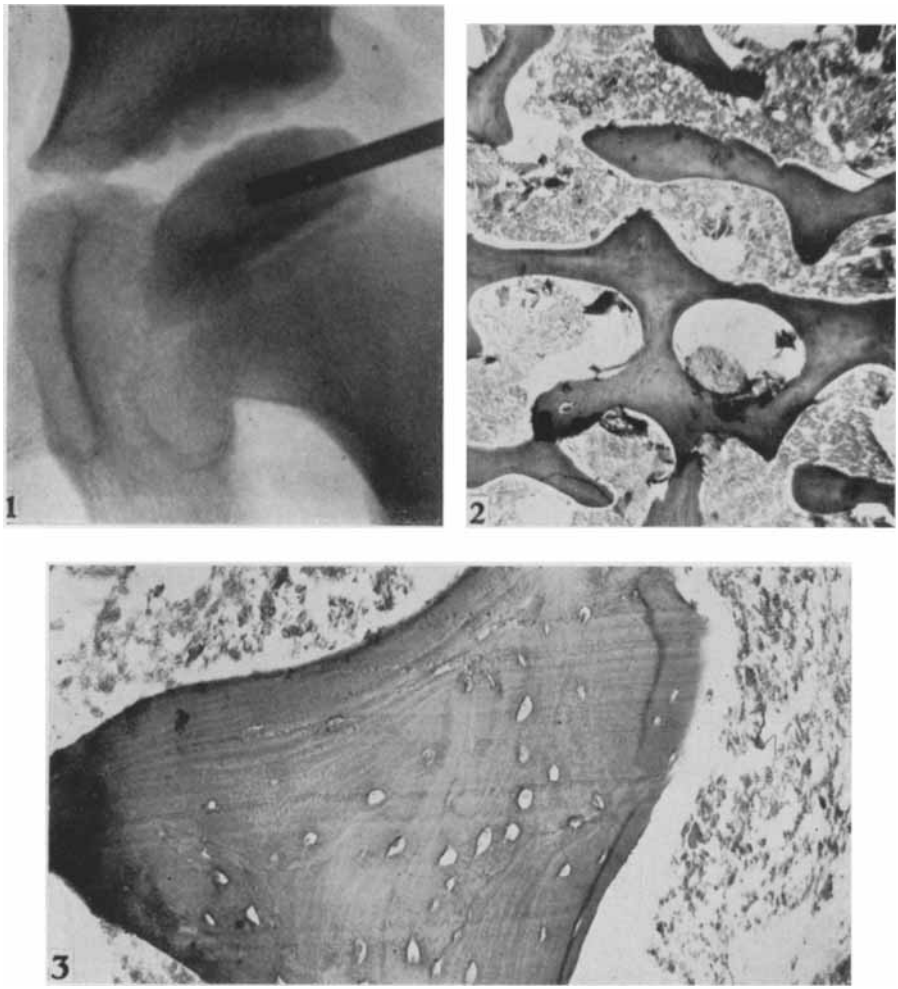

Fig. 4. Case 45. The initial stage.

1. Roentgen: The epiphysis is more dense than normal, and insignificantly compressed.
2. Microphotograph. (×35). The trabeculae are partially retained, but the marrow spaces are filled with a necrotic mass consisting of dead marrow and small pieces of dead bone (Trümmermehl). No signs of reaction.
3. (×200). The nuclei are pyknotic or completely absent. Individual contour lines observed in the bone. Total necrosis.

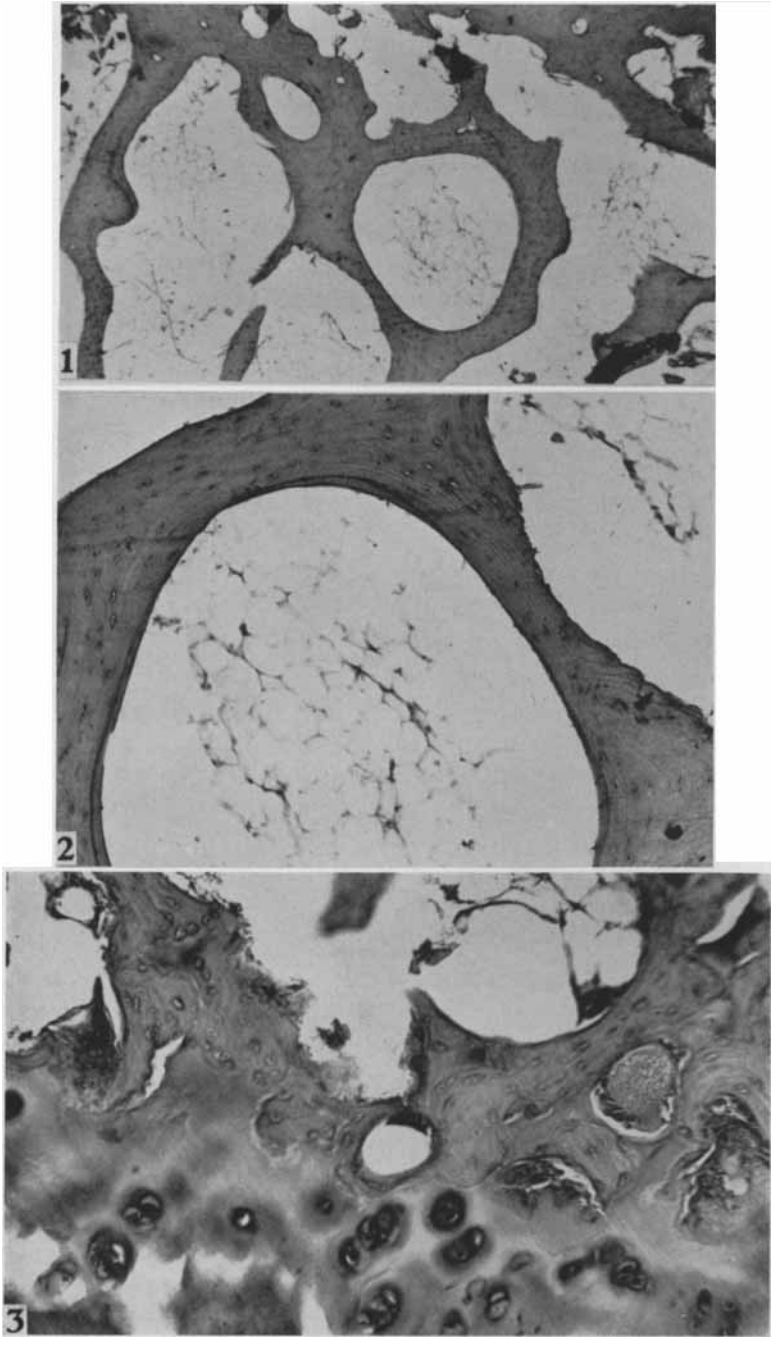

Fig. 5.

For the most part the fat marrow is absent. Only in one case was there any unchanged marrow left. In 3 cases one can still see trabeculae of vital appearance, but the marrow has necrosed which would indicate that the marrow is more sensitive to influences that cause necrosis, and so shows signs of being devitalized before the bone itself.

Practically speaking there are no signs of bone regeneration. A tissue, resembling connective tissue, with vessels has only been manifest in 2 cases and osteoblasts in one case only. There were no giant cells at all among the cases falling under this group. As a consequence of there not being any regenerative tissue, there is neither hardly any newly formed bone. Bone of this nature was only found in 1 case which is the only one in this group that shows any degree of regeneration worth mentioning. Haemorrhage was manifest in 4 cases, but in all cases it was of an insignificant amount. In the table, 1 case was marked ++ while the remaining 3 cases were marked +. In the other cases no blood corpuscles were found in the preparations. Whether this haemorrhage is old or recent cannot be decided by means of the technique used for fixing and staining.

There have been no signs of inflammation. A few lymphocytes were manifest in 3 cases, but not in any greater numbers than would correspond to the scanty haemorrhage. Leucocytes were not found in any of the cases.

B. Cartilaginous changes

Larger or smaller parts of the joint cartilage have accompanied the preparations in 12 out of the 14 cases in the initial stage, and then always the inner part of the cartilage with the ossification zone. The histological picture in the basal layer of the cartilage shows large cells arranged in columns at right angles to the cartilage-bone border as a sign of an increased cartilaginous activity in this layer. Beyond this comes the enchondrally formed bone forming, so to say, edge seams in the concavity of which the primary marrow cavity comes up. In preparations having a more tangential sectional direction one can see the marrow cavity like a small insula surrounded by a ring of enchondrally formed bone. This ossification zone is an expression of the fact that the epiphysis in children also grows out from the joint cartilage.

Fig. 5. Normal hip. ♂ 8 years. Preparation taken out with the puncture needle.

1. Microphotograph. (×35). The bone forms slender trabeculae with wide marrow spaces containing fat marrow.
2. (×100). The trabeculae have well stained nuclei.
3. Joint cartilage and bone. (×100). A clear enchondral ossification is in progress. The cells in the inner part of the basal layer are bladder-shaped. Vascular faucets rise up in the cartilage, around which there are bone deposits. The trabeculae nearest to the cartilage contain remnants of cartilage.

As a rule, the bone necrosis does not reach further than to the inner margin of the zone of ossification, but in some of the cases it was noticed that the enchrondrally formed bone had also become necrosed. This would indicate that necrosis begins in the inner part of the epiphysis and gradually spreads towards the periphery, at least as far as concerns that part of the periphery that is covered by the joint cartilage.

Of the authors who have reported histological changes in coxa-plana, ROCKEMER (1927), among others, states that there were no changes in the joint cartilage, while RIEDEL (1922), KONJETZNY (1934), GALL and BENNET (1942), HAYTHORN (1949), to mention some, have shown definite changes also in the joint cartilage.

In 6 of the cases in hand, there has been a manifestation of changes in the joint cartilage occurring in the initial stage. These changes are somewhat varying from case to case and will be an expression for a more or less pronounced devitalization of the inner layer of the cartilage. The cartilage looks lighter than is normal. As a rule there is no columnar arrangement of the cells, they lie more scattered than is usual and the nuclei are often smaller than those lying in the exterior parts of the cartilaginous layer, here and there they are totally absent. See figs 6 and 7.

As mentioned, these changes occur in the basal layer of the cartilage, and only in those places where the bone necrosis reaches so far out that it also embraces the enchondrally formed bone; there is, then, reason to suppose that the necrosis occurring in the bone and the changes in the cartilage have got the same cause.

C. Discussion

When studying the literature on coxa-plana, one finds that the authors, who have given histological descriptions of the disease, give accounts of pictures that only partly coincide with those that were manifest in the present investigation of this stage. Practically all have shown bone necrosis, but these have more or less faded into the background of the changes of another, and then usually, reparative character that has been shown. Such descriptions have been made, among others, by PHEMISTER (1921), RIEDEL (1922), HEITZMAN and ENGEL (1923), AXHAUSEN (1923), ZEMANSKY (1928), LIPPMAN (1929), FERGUSON and HOWORTH (1934), KONJETZNY (1934), GALL and BENNET (1942) and HAYTHORN (1949). The reason for the diverging picture described by these authors might be sought in the fact that their cases represented later stages of the disease and that the cases were not described against a background of the roentgenological changes.

It will be seen from my investigations that necrosis of the bone is the dominating histo-pathological change in the initial stage. Here and there

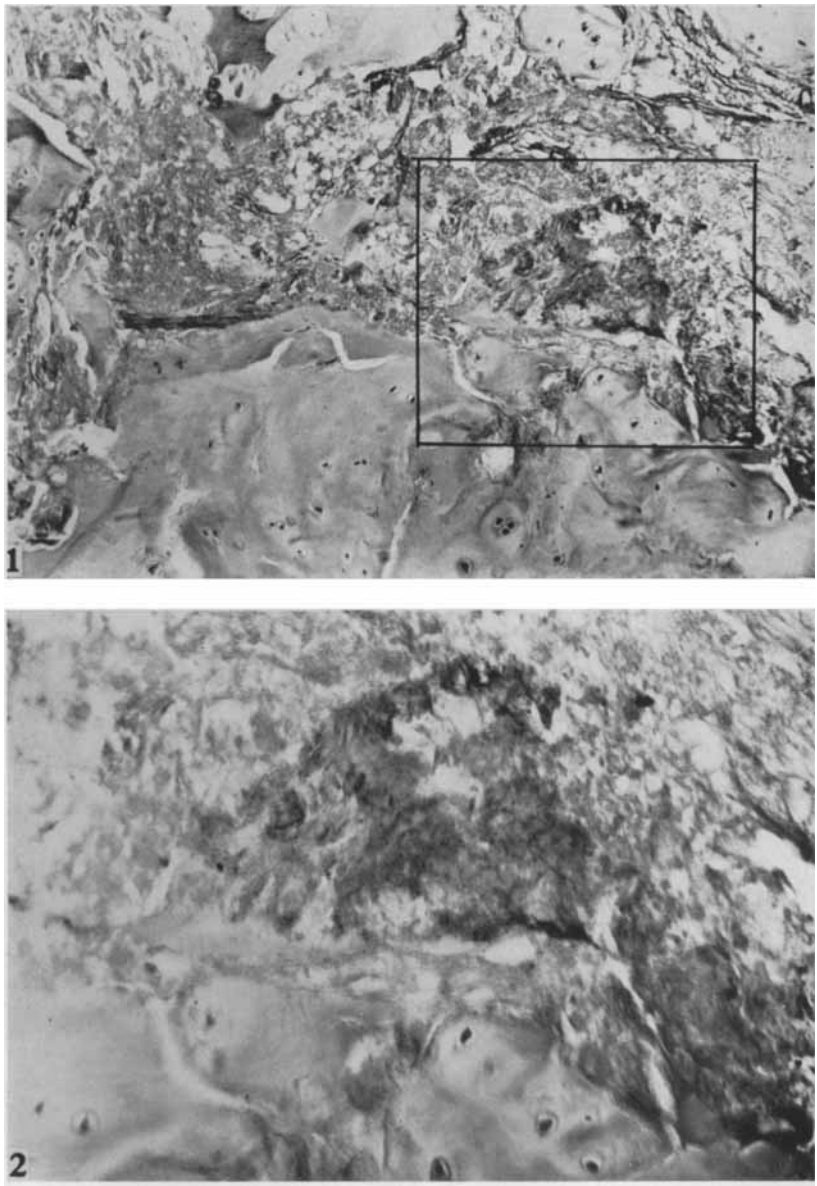

Fig. 6. Case 21. The initial stage.

1. Microphotograph. (×100). Right at the bottom of the picture, joint cartilage of normal appearance. Further up the nuclei become pyknotic and disappear entirely. Higher up in the picture the cartilage becomes a thready mass without nuclei. Absence of ossification zone.
2. The squared-off area (×200).

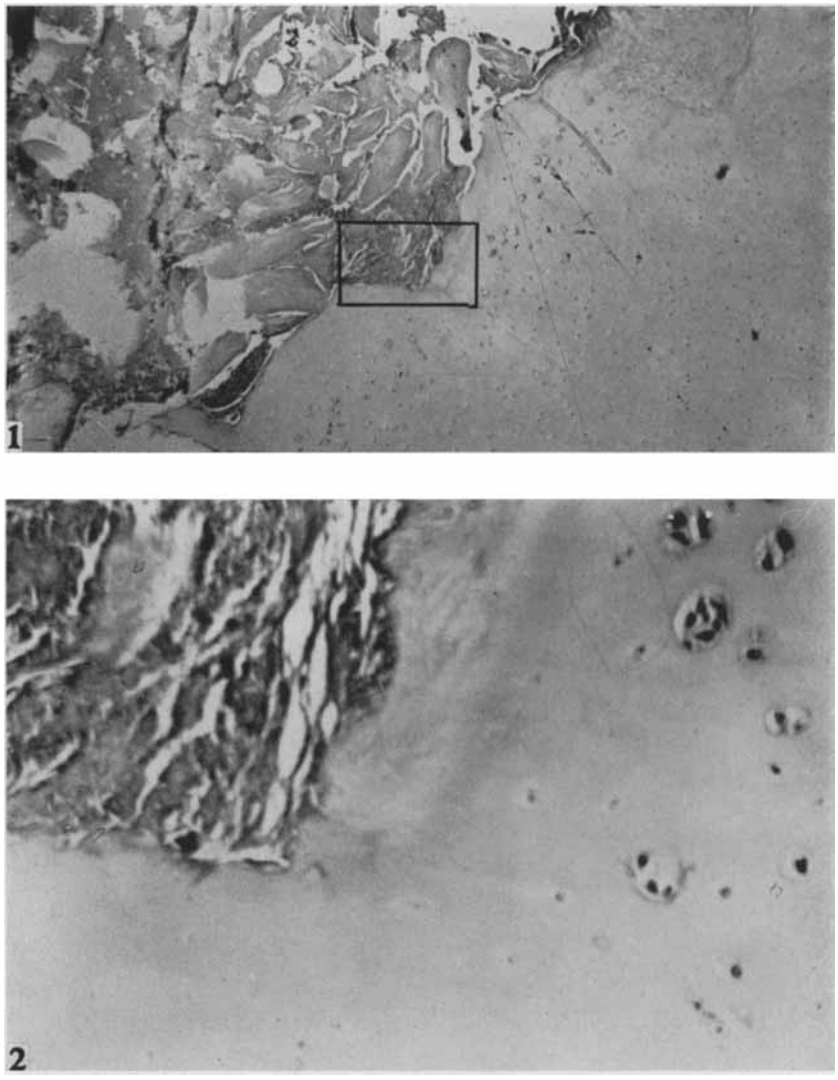

Fig. 7. Case 34. The initial stage.

1. Microphotograph. (×35). To the left of the figure, necrotic bone. Necrosis embraces the ossification zone as well.
2. The squared-off area (×200). The innermost layer of the joint cartilage is markedly devitalized. The nuclei are practically absent.

there are remnants of bone that is still vital, while the signs of bone regeneration are practically speaking absent.

If, when looking at Table II, one studies the duration of trouble, one will see that 6 of the 14 cases were troubled for only 2 months or even shorter,

one case for half a month only. One does not know exactly how long it takes for a necrosis of the bone to develop, and the time will depend on the nature and degree of the damaging influence, the vascularization of the bone and the age of the individual. One knows from traumatology, however, that it usually takes a considerably longer time, after a fracture, before roentgenological signs of bone necrosis show up, for which reason one is entitled to draw the conclusion from the information given in the present investigations about the duration of the trouble that necroses are developed prior to the patient's feeling any subjective trouble or showing any objective sign in the form of limping or restricted mobility.

WALDENSTRÖM (1934) has another opinion and considers founding himself on his detailed clinical and roentgenological studies, that it is probable that necroses give trouble right from the beginning, and that the roentgen picture, at that time, could still be normal.

RIEDEL (1922) describes the disease as a purely productive process resembling most nearly osteitis fibrosa localisata, even though he mentions the occurrence of bone necrosis. The explanation of this seems to be the one referred to above, that the case was in the reparative stage. From the presence of lymphocytes, plasma cells, leucocytes and eosinophils in the marrow spaces Riedel draws the conclusion that the disease is caused by an infection. This opinion has been shared by numerous authors including PERTHES (1910), EDEN (1911), NIEBER (1916), PHEMISTER (1921), chiefly on the basis of clinical and histological investigations. The cultivation of resection material carried out by PHEMISTER (1921) and HAYTHORN (1949) has, however, not given any growth of bacteriae.

The clinical course, the conditions of temperature and sedimentation rate do not indicate that the disease is of an infectious nature, and my own investigations point definitely against inflammation as being the cause of the changes in coxa-plana but, instead, show, as before mentioned, that the main change is the bone necrosis.

As for the explanation of the causes of the necrosis, our knowledge is still very limited. BENTZON (1926) carried out experiments on animals by injecting alcohol around the collum femoris in an endeavour to produce the same change as manifested in coxa-plana in human beings. These experiments were then extended by RANDLÖV-MADSEN (1949), but the pictures that were published do not resemble those which I found, at least not in the earliest stage of the disease.

ROCKEMER (1927) and LERICHE (1934) thought, as did the two previous authors, that the changes were due to hyperaemia.

Most of the other investigators are inclined to consider that the necrosis is a consequence of ischaemia, but no convincing proof of this opinion has been given. Neither do my investigations give any definite clarity on this

point. There are, however, a number of circumstances that would indicate that the necroses are ischaemic in origin. The total necrosis in the microscopic sections and the necrotic bone's macroscopic grey-white granular appearance indicate a poor or even a totally ceased blood circulation within the affected area, and the absence of vessels and corpuscles in the section also points in the same direction.

The investigations give no indication as to what the ischaemia would be due to.

AXHAUSEN and BERGMANN (1937) drew attention to the fact that the increased roentgen opacity of the epiphysis in the initial stage might be due to the fact that the necrotic trabeculae are pressed into each other through overloading. It would seem from my preparations that a compression of the trabeculae actually takes place, but how can one explain this compression in the early cases where one has an obvious increase in roentgen opacity but where apparently the epiphysis still has retained its shape?

AITKEN (1947) considered that the epiphysial opacity was only apparent and due to a decalcification of the bordering bony parts, especially the metaphysis, but a comparison of the roentgen pictures of both sides in unilateral cases shows that the affected epiphysis is actually more opaque and the normal bony structure obliterated.

The precipitation of lime salts in the necrotic area should produce an increased roentgen opacity, but as the preparations in the preceeding investigations were decalcified, they unfortunately do not give any enlightenment on this point.

The early symptom described by WALDENSTRÖM (1934), which he calls "the sub-chondral thinning", seems to me as being discussible as an explanation of the roentgen opacity in early cases, having my own investigations as a background. Waldenström observed in the anterior superior part of the epiphysis a thinner, streak-shaped formation separated from the joint cartilage by only a thin layer of bone, and supposed that this thinning had been caused by a resorption of the bony tissue. In my cases there are no signs, at this stage, of resorption. Instead, the thinning zones might be thought to originate in the following manner. As previously mentioned, bone necrosis leaves the ossification zone intact for a long time, while the underlying bone decomposes. The enchondrally formed, still vital bone hangs fast intimately to the basal layer of the cartilage. When subjected to loading the necrotic bone is compressed, but the cartilage, which in this age is thick and elastic, coupled with its attached ossification zone springs back into position when the pressure is removed, whereby a gap arises which, in all probability, is filled up with tissual fluid. In this way one might find an explanation for both the "sub-chondral thinning" and for the circumstance

that one gets an increased roentgen opacity in the epiphysis with a retention of the outer contours of the bone. The investigations, however, do not give any definite proof that this is really so.

The fragmentation stage

A. Bony changes

In this stage the material comprises 15 biopsies with a period of trouble for the patients lasting from 2 to 20 months and an average time of trouble of 6.5 months.

In 14 cases there are notes about the hardness of the bone at the time of puncturing. In 6 cases the bone was soft, in 5 it was hard, and in 3 cases the needle encountered alternatively hard and soft parts. On an average the bone in the cases belonging to this group seemed harder and firmer than it was in the previous group. The macroscopic appearance was also somewhat divergent. In those cases where the bone was quite soft it had the same greyish appearance as was manifest in the initial stage, but in other cases the colour was more of a brown-red tinge which was reminiscent of the colour in normal cancellous bone.

Microscopically, the bone necroses take a rather prominent place even if they are not so pronounced as in the initial stage. In 3 cases there have been no necroses, in 5 cases they have, in accordance with the signs given in Table III, been marked +, in 2 cases ++, and in 5 cases +++.

The microscopic picture does not show the uniformly dead appearance that was the case in the foregoing stage.

Normal bone occurs to about the same extent as in the initial stage, while normal bone marrow is present somewhat more profusely and can be manifested in 9 out of 15 cases.

That which especially characterizes the microscopic picture in the fragmentation stage is the profuse occurrence of the morphological elements which indicate a regeneration of the decomposed bone. One can see, in all cases except one, a tissue that is profuse in cells and resembling connective tissue which, in more or less broad formations, grows into the necrotic bone. This tissue, which is profusely vascularized in most cases, is probably not connective tissue in the real meaning of the word, but rather a tissue, the cells of which, according to WILTON (1937), are determined to become bone, and should perhaps therefore sooner be called bone of low-grade maturity. For practical reasons of language it is referred to as connective tissue in this study. As far as the presence of this tissue is concerned, it has been marked in the table as follows: 7 cases as +, 6 cases as ++, and 1 case as +++.

Table III. Showing age, sex, duration of trouble of the cases and bone-changes in the fragmentation stage

Caso No.	13	42	43	44	47	28	8	15	19	31	36	5	48	6	40
Age in years	3	4 1/2	4 1/2	4 1/2	7	7	4	5	7	6	3 1/2	6 1/2	5 1/2	6 1/2	4 1/2
Sex	♀	♀	♂	♂	♂	♂	♀	♂	♂	♀	♂	♂	♀	♂	♂
Trouble in months	2	2	4	4	4	5 1/2	6	7	7	7	7	7 1/2	8 1/2	9	20
Bone necrosis	+	+	+++	0	++	+	+++	+++	0	+	+	+++	+++	++	0
Normal bone	+	++	0	+++	0	0	0	0	++	0	+	0	0	0	+++
Normal bone marrow	0	++	0	+	++	0	0	0	+	0	+	0	0	0	++
New-formed bone	0	+	0	0	++	+++	+	0	0	++	+	0	0	++	0
Connective tissue	+	+	+	+	++	+++	+	++	+	++	++	0	++	++	+
Vessels	+	+	0	0	+	+	+	+	+	++	++	0	++	++	0
Osteoblasts	+	0	0	0	+	+	+	0	0	0	0	0	0	+++	0
Giant cells	0	0	0	0	0	+	+	++	0	0	0	0	+	++	0
Haemorrhage	++	+	+	0	+	+	+	+++	+	+	+	+	++	+++	++
Lymphocytes	+	+	0	0	+	0	0	++	+	++	+	0	+	+	++
Leucocytes	0	0	0	0	0	0	0	+	0	0	0	+	0	0	0

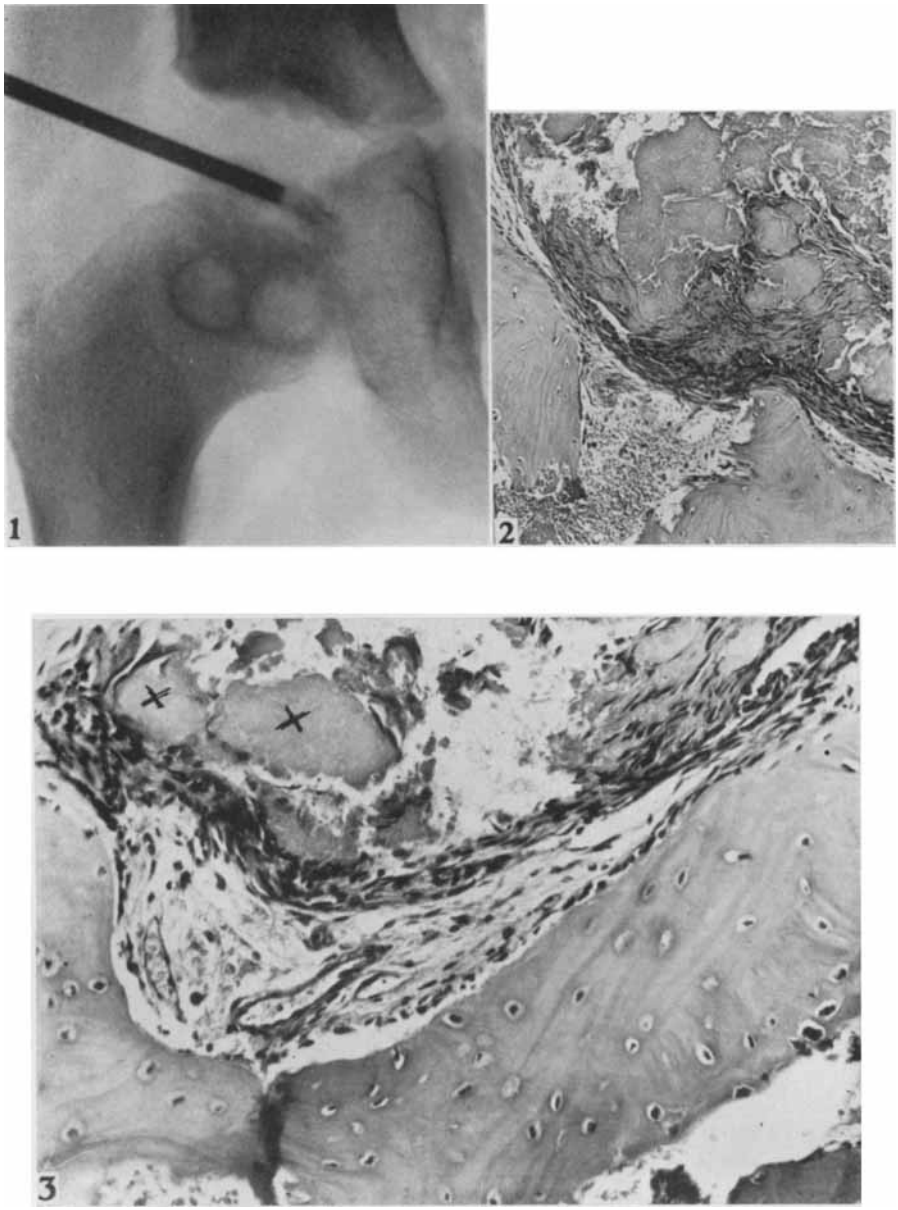

Fig. 8. Case 6. The fragmentation stage.

1. Roentgen. The epiphysis is markedly shrunken and fragmented.
2. Microphotograph. (×100). Above, to the right of the picture, necrotic bone into which grows a connective tissue that is rich in cells. Below to the left, newly formed bone.
3. (×200). The connective tissue is profuse in vessels. In the lower part of the picture it goes over to immature bone with large, well-stained nuclei.
×) = remnants of necrotic bone.

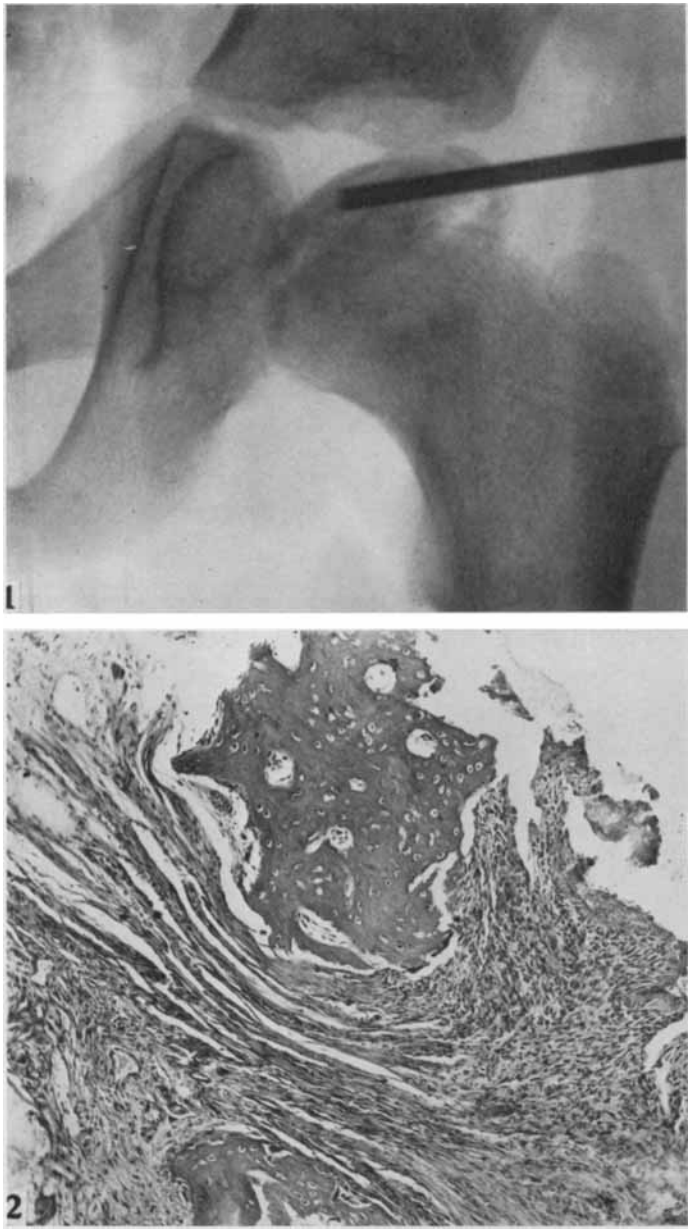

Fig. 9. Case 28. The fragmentation stage.

1. Roentgen. Moderate compression of the epiphysis. Marked fragmentation.
2. Microphotograph. (×100). A connective tissue rich in vessels, going over (at the top of the picture) to immature bone with closely lying, well stained nuclei. The round, light parts in the bone are the beginning of marrow spaces.

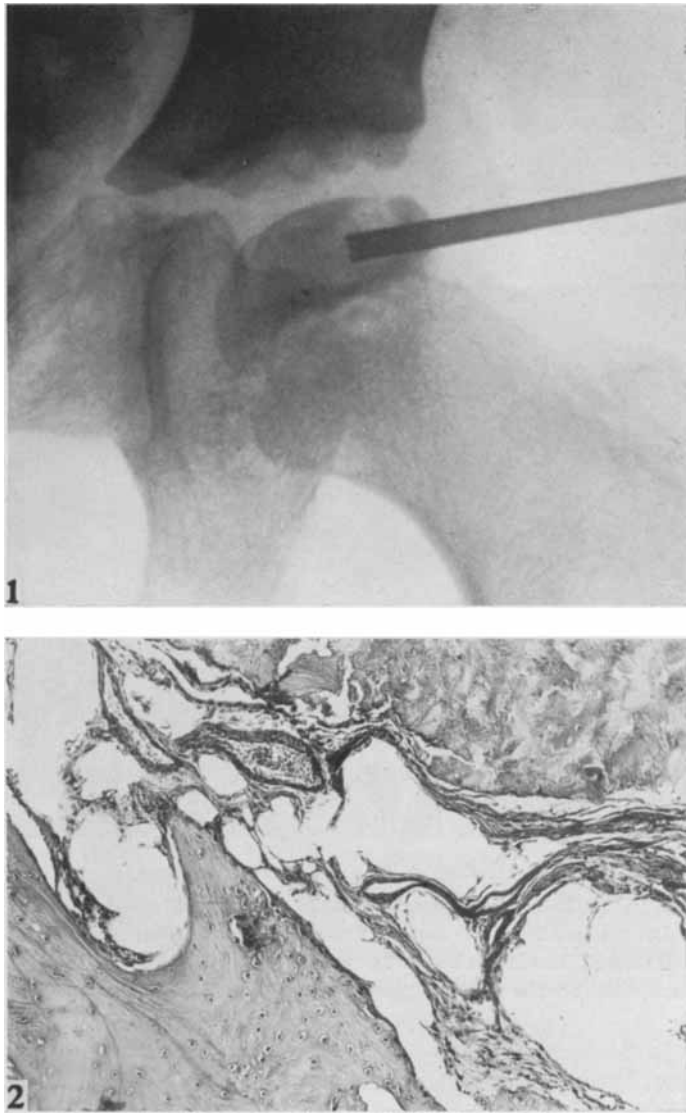

Fig. 10. Case 47. The fragmentation stage.

1. Roentgen. Slight flattening of the epiphysis. Initial fragmentation.
2. Microphotograph. (×100). To the right at the top of the picture, necrotic bone. After this a layer of connective tissue rich in vessels going over to newly formed bone where the nuclei begin to arrange themselves in the direction of the lamellae.

Here and there this connective tissue changes its character. The cells become larger and darker and the intercellular substance partially loses its fibrillar structure and becomes more homogeneous (osteoid). This will have

to be regarded as a process of maturity in the osteogenetic tissue. See fig. 11. In 5 cases in the fragmentation stage the ingrowing regenerative tissue contains giant cells, the presence of which in 2 cases is marked ++, and in 3 cases +. These giant cells are rather large, polynuclear and of the type, according to DIETRICH (1935) and BROMAN and HÄGGQUIST (1928), usually seen in bone decomposition. In individual places one can see giant cells arranged like osteoclasts in depressions in the necrotic trabeculae.

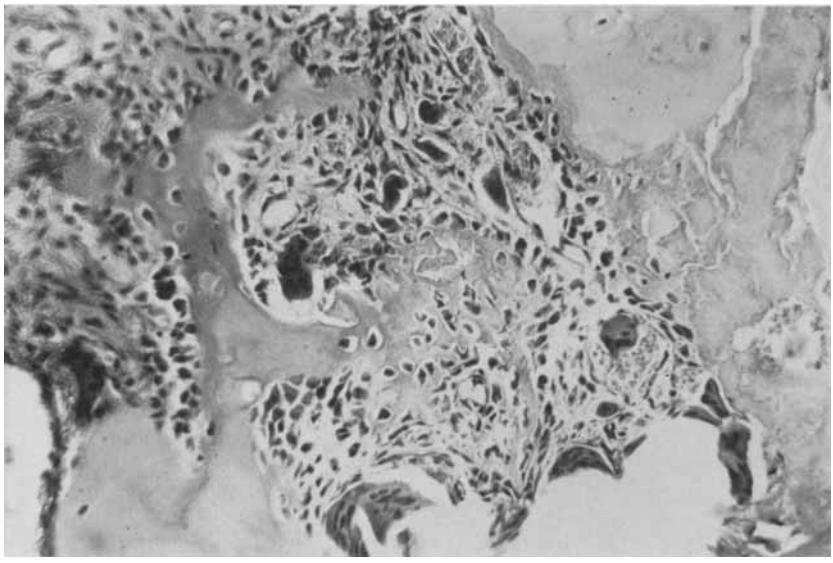

Fig. 11. Case 6. The fragmentation stage.
Microphotograph. (×200). The reparative tissue has here attained a higher degree of maturity which appears from the fact that the fibrillar structure is about to disappear. A trabecula is formed (to the left and in the centre of the picture). The large, dark formations are giant cells. To the extreme right, necrotic cartilage and bone.

In 5 cases there was a clear manifestation of osteoblasts. They have a typical appearance with large, dark nuclei and lie in rows like necklaces around the edges of the connective tissue, thus forming regular osteoblast seams. See fig. 12.

Outside the connective tissue and the osteoblast layer there comes a tissue with rather large and highly coloured nuclei. The intercellular substance is homogeneous and calcified. Thus it is a question of a newly formed, calcified bony tissue. In the majority of cases one can trace the beginning of a lamellar disposition, but the lamellae are coarser and thicker than in ordinary cancellous bone in the ages under discussion. One can observe how the cells begin to arrange themselves in the lengthwise direction of the

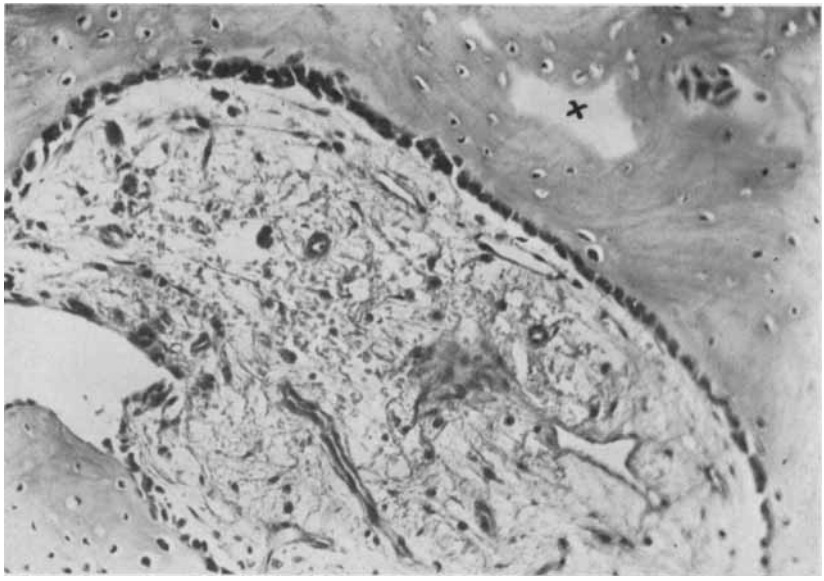

Fig. 12. Case 6. The fragmentation stage.

Microphotograph. (×200). Between the connective tissue rich in vessels and the newly formed bone lies a layer of osteoblasts.

×) = Cartilaginous remnants from the enchondrally formed bone.

lamellae. Here and there in this unmatured bone there are round areas filled with a loose tissue denoting the beginning of a marrow cavity. See figs. 8, 9, and 10.

In a few places in the newly formed bone one can see small cartilaginous insulae, the presence of which is accounted for by the fact that the enchondrally formed bone, which often contains such small cartilaginous insulae, has become necrosed and is supplemented by the newly formed bone that then encompasses the cartilaginous remnants.

This newly formed bone, as expected, was manifest in all of the cases where there were osteoblasts, but even without such the unmature bone makes its appearance and then as a direct development of the connective tissue. In Table III the appearance of the newly formed bone has been classified as +++ in 1 case, ++ in 3 cases, and + in a further 3 cases.

As previously mentioned, there was only a most insignificant manifestation of haemorrhage in the initial stage. In this connection the fragmentation stage shows conditions of an entirely different character as there was haemorrhage in all cases except one. It is relatively profuse and the occurrence has been marked, in 2 cases with +++, in 3 cases with ++ and in 9 cases with +. In preparations that are treated in the manner done here, it is difficult to determine whether the corpuscles are fresh or of an older date.

As far as one is able to judge them, however, they give the impression of being fresh as they show no signs of shrinkage and there is no sediment of blood pigment. It should be pointed out in this connection that in many of the preparations there is a sediment of a fine-grained, black substance at the border of the preparation which is somewhat reminiscent of pigment. However, it is definitely not a question of pigment but of impurities from the walls of the puncture needle caused by the sterilization of the needle. Nor is there any reason why there should be any sedimentation of blood pigment only at the borders of the preparations. The haemorrhage observed is probably not spontaneous but brought about during the puncture, and it has been included in the report only in order to show further that, in the fragmentation stage, one has to deal with a powerfully vascularized tissue.

Lymphocytes and leucocytes are also represented in this stage, the former in 10 cases and the latter in 2 cases. Naturally it has not been possible to make a differential count of them, but, in estimation, they would seem to occur in the same proportions as in an ordinary haemorrhage. There were no signs of inflammatory or infectious changes manifest in this stage either.

B. Cartilaginous changes

In 11 of the 15 cases in the fragmentation stage, a piece of the joint cartilage came away during punctation, and in 5 of these the cartilage manifested changes. In similarity with the foregoing stage, the cartilaginous changes are localized to the basal layer and only occur where the bone necrosis reaches right out to the enchondral ossification zone. As was the case in the foregoing stage they are not very characteristic and are reminiscent of the changes shown there. Thus one sees changes in the nuclei in the form of pyknosis and karyolysis. In some cases there is a manifestation of clear fibrillary structure in the intercellular substance combined with a disappearance of the columnar arrangement of the cartilaginous cells. These changes, in similarity with the foregoing stage, may be regarded as being an expression for a more or less pronounced devitalization of the cartilage within the regions in question. In some places there are slit formations in the cartilage, and these slits are sometimes filled with blood corpuscles.

C. Discussion

The histological picture in the fragmentation stage in the present investigations coincides better with the descriptions one comes across in the literature than was the case in the initial stage. The changing picture described by the authors previously referred to makes it probable that these cases have belonged either to the fragmentation stage or, perhaps, some later stages.

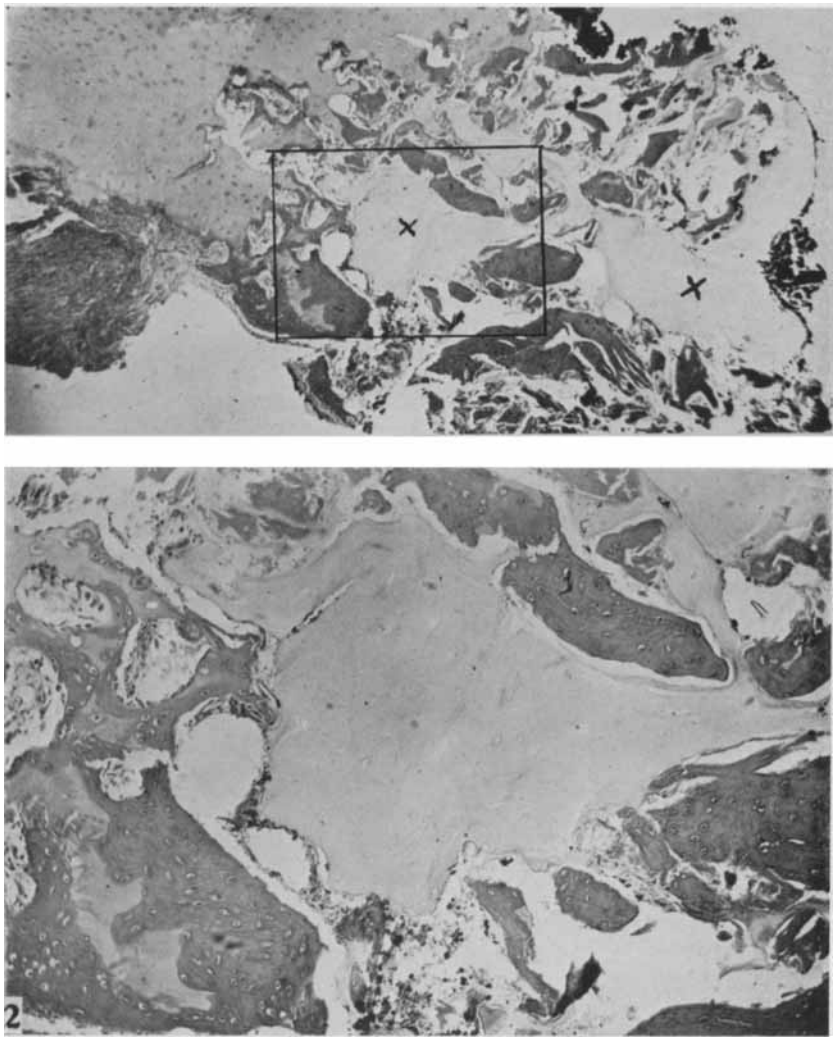

Fig. 13. Case 28. The fragmentation stage.

1. Microphotograph (×35). High up to the left of the picture, normal joint cartilage. The bone is in the upper part of the picture necrotic, in the lower part mainly vital.
2. The squared-off area (×100). On the left one sees how an enchondral bone formation takes place from the joint cartilage.
   ×) = necrotic cartilage without nuclei.

As mentioned before, most of them have, in addition to bone necrosis, even described changes of a reparative nature such as connective tissue that is rich in vessels, osteoblasts and giant cells.

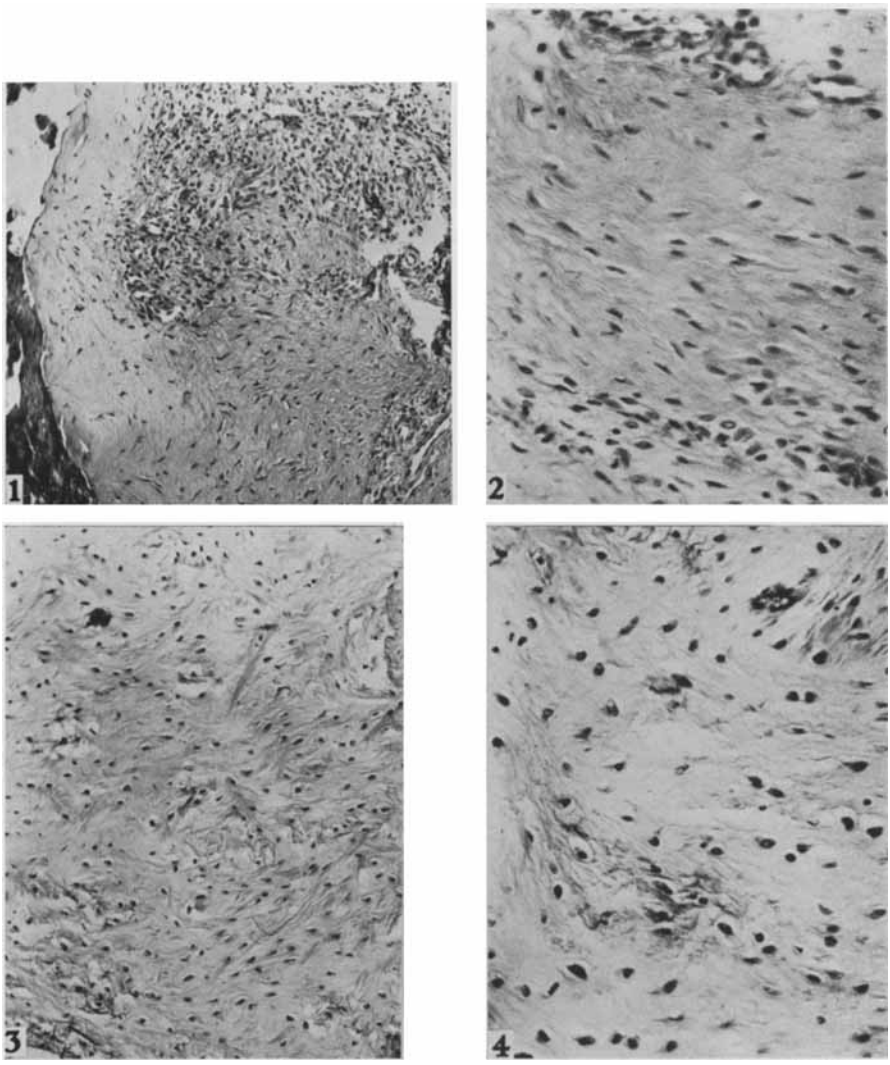

Fig. 14. Case 48. The fragmentation stage.

1. Microphotograph (×100). To the left of the picture, necrotic bone which embraces the ossification zone. The cartilage within is supplied by a tissue that is rich in cells and vessels that might be bone of a very low degree of maturity.
2. The same as 1, (×200).
3. (×100). Further out (towards the joint-surface), cartilaginous cells remain but they are smaller than normal. The intercellular substance is not homogeneous but has become thready denoting degenerative processes in the cartilage, by which the latter has got an embrynoal appearance.
4. The same as 3, (×200).

It would seem as if a number of authors consider that the fragmentation stage represents the culminating point of the disease, in doing which they base their opinion on the pronounced roentgenological changes in this stage with a high grade decomposition of the epiphysis. It appears from the present investigations, however, that the culminating point has been passed and that the fragmentation stage is one in which a pronounced reparation of the previously necrosed bone has taken place.

HEITZMANN and ENGEL, AXHAUSEN (1923), GALL and BENNETT (1942) as well as BERNBECK (1947) noted in their cases a connective tissue that grew into the necrotic bone, and considered that it came from the metaphysis or the metaphysial periosteum and penetrated the epiphysial cartilage usually, in accordance with their information, in the angle between the epiphysial and joint cartilage in order to spread itself in the epiphysis. Their explanation seems correct to me as only the metaphysis and its periosteum will contain the elements necessary for osteogenesis.

The question is, then, which histological tissues correspond to the different parts in the epiphysis on the roentgen picture. It will be quite obvious that the opaque parts without any visible bony structure correspond to remnants of necrosed bone, especially if one bears in mind the histological picture in the initial stage where the bone has this opaque appearance roentgenologically.

On the other hand the question of the lighter areas in the roentgen picture is more difficult to answer. It seems scarcely probable that they should be cavities, as in that case the epiphysis ought, in its entirety, to show a pronounced compression, which is not the case. Therefore the lighter areas must consist of a tissue that does not give roentgen shadows, that is to say, that is not calcified. In the given place, then, one only has two tissues to choose from, either cartilage or the ingrowing reparative tissue. RIEDEL (1922), KONJETZNY and WALDENSTRÖM (1934) give a description of cartilaginous insulae, in their preparations, inside the partially necrosed and partially regenerated epiphysis. In my own material there was no manifestation of cartilage within the epiphysis with the exception of the previously described small cartilaginous remnants within the enchondrally formed trabeculae. These cartilaginous remnants appear in microscopic enlargement only and so cannot give any lighter areas on the roentgen picture. In my preparations practically all the cases manifested an ingrowing regenerative tissue, and in 7 cases there is a newly-formed immature bone. Naturally the regenerative tissue is not calcified and the newly-formed bone ought to be able to show every grade of calcification including the most insignificant. It therefore seems most probable that it is these tissues that give rise to the lighter areas in the roentgen pictures.

The reparation stage

A. Bony changes

In this stage, fifteen punctures were made on patients who had been troubled between 12 and 45 months. The average time of trouble was 22 months. In only 9 cases are there any notes about the hardness of the bone when punetured, but of these, 4 are noted as having hard bone, 3 alternatingly hard and soft and only 2 completely soft bone. It will thus seem as if the bone in this stage would be harder than in the fragmentation stage. Unfortunately there are no notes about the macroscopic appearance of the bone in this group.

There is still a manifestation of bone necroses in the reparation stage, but they are decidedly less pronounced than in the fragmentation stage. Of the 15 cases, necroses were manifest in 7 cases only and the occurrence in the individual cases was considerably less than in the foregoing stage (see Table IV). In 5 cases they have been noted with +, in 1 case with ++ and in 1 further case with +++.

In this group the duration of the trouble shows a greater variation than in both of the foregoing, and from the table it will be seen that of the 6 cases, whose time of trouble has been longer than the average of 22 months, necroses occurred only in 1 case. From this it will be seen that the necroses diminish the longer time elapses after the beginning of the affliction, a circumstance which one had reason to expect.

On the other hand, normal bone occurs in considerably greater amounts than in the foregoing stage, i. e., in 10 out of the 15 cases and, in 6 cases, the occurrence has been marked by +++, and in the other 4 by ++. From the table it will be seen that the normal bone increases in the same proportion as the necroses diminish and that in the 6 cases who have been troubled for a longer time than 22 months, there was a profuse manifestation of normal bone in all cases except one.

In all cases with normal bone there has also been a manifestation of normal bone marrow although in relatively small quantities due, in all probability, to the fact that the fine marrow structure was partly destroyed during the technical production of the preparation.

The presence of newly formed bone was considerably less than in the foregoing group and was manifested in 4 cases only. It appears approximately in the same amounts as the necrosed bone and is found, among the cases with the longest time of trouble, in only 1 case, and then in moderation. Otherwise the occurrence has been marked +++ in 2 cases, and + in one case.

In 10 cases the regenerative tissue that resembles connective tissue has been manifest. In the latest cases it occurred in very meagre quantities, in the earlier ones it was somewhat more profuse.

Table IV. Showing age, sex, duration of trouble of the cases and bone-changes in the reparation stage

Case No.	30	7	33	10	2	16	11	14	46	12	4	22	23	32	49
Age in years	7	$6^1/_2$	$7^1/_2$	$6^1/_2$	$7^1/_2$	6	$7^1/_2$	$8^1/_2$	8	$5^1/_2$	11	5	$7^1/_2$	$8^1/_2$	11
Sex	♂	♂	♂	♂	♂	♂	♂	♂	♂	♂	♂	♂	♂	♂	♂
Trouble in months	12	14	14	15	16	17	18	19	20	23	24	28	30	34	45
Bone necrosis	+	+++	0	+	0	+	0	+	+	0	++	0	0	0	0
Normal bone	0	0	++	0	+++	0	++	++	++	+++	+++	0	+++	+++	+++
Normal bone marrow	0	0	++	0	+++	+	+	+	+	++	++	0	++	++	+
New-formed bone	+++	0	0	0	0	+++	0	+	0	0	0	++	0	0	0
Connective tissue	+++	0	+	++	+	++	0	+	0	0	+	+	+	+	0
Vessels	+++	0	+	+	++	++	0	+	0	0	+	+	+	+	0
Osteoblasts	++	0	0	0	0	+	0	0	0	0	0	+	0	0	0
Giant cells	++	0	0	0	0	+	0	0	0	0	0	0	0	0	0
Haemorrhage	++	++	0	+	0	+++	0	0	++	0	+++	0	0	+++	+
Lymphocytes	0	0	0	0	0	++	0	0	0	0	+++	0	0	0	0
Leucocytes	0	0	0	0	0	0	0	0	0	0	+	0	0	0	0

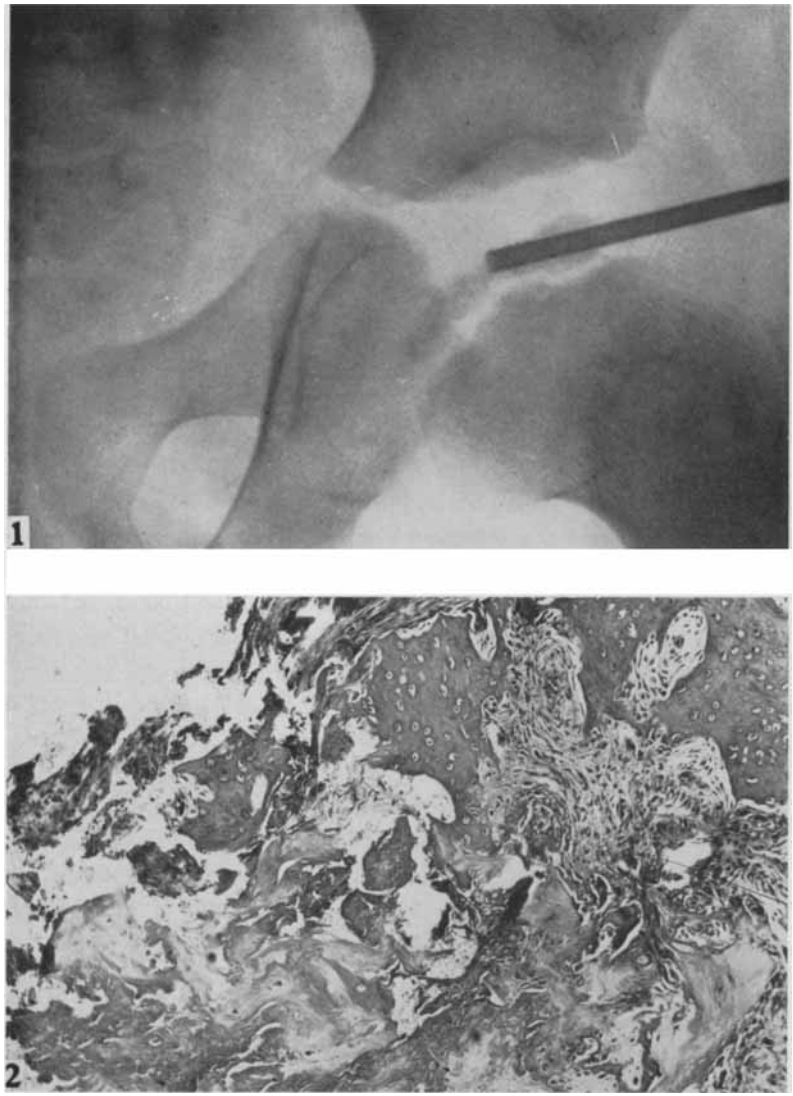

Fig. 15. Case 30. The reparation stage.

1. Roentgen. Pronounced fragmentation. Normal bony structure begins to manifest itself in the epiphysis.
2. Microphotograph. (×100). Osteonecrosis still remains (at the bottom of the picture to the left), but a marked regeneration is in progress. Newly formed bone on the right top of the picture.

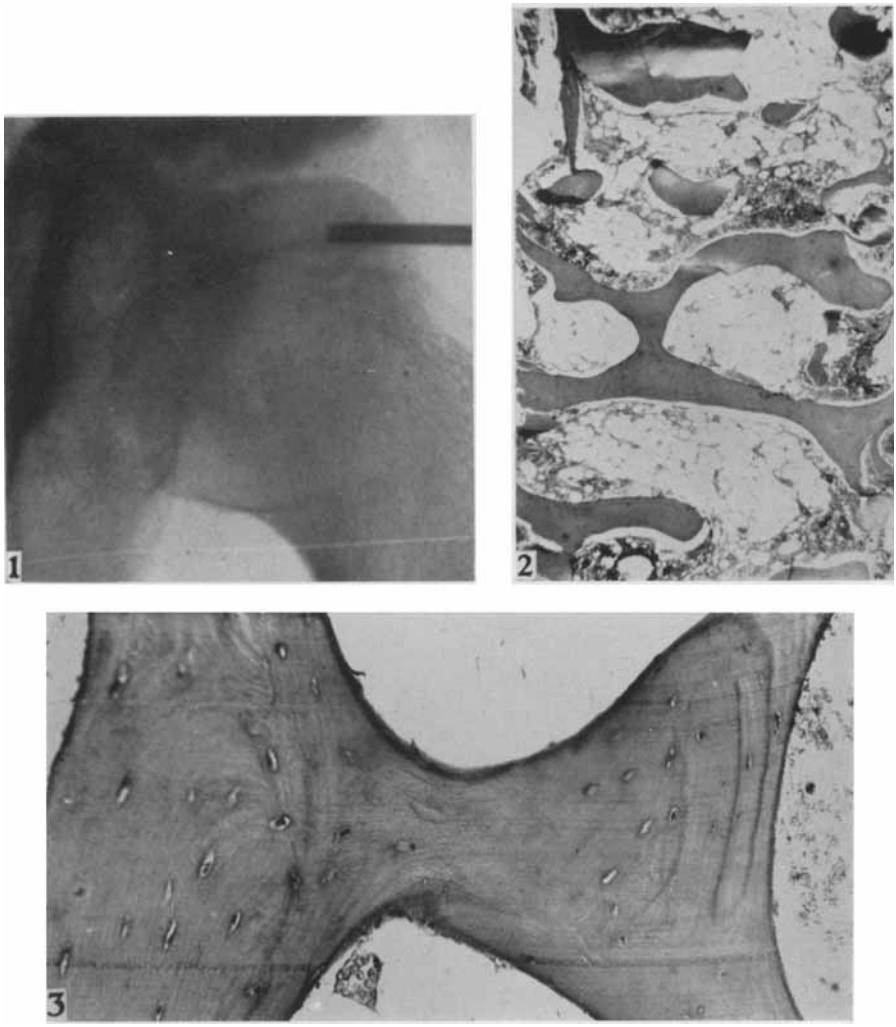

Fig. 16. Case 4. The reparation stage.

1. Roentgen. The epiphysis is somewhat flattened but has mainly normal bony structure.
2. Microphotograph. (×35). The bone is normal in appearance and forms slender trabeculae. The marrow spaces are filled with fat marrow. No signs of necrosis. No signs of reaction.
3. (×200). The bone contains well-stained nuclei arranged lengthwise to the trabeculae.

Osteoblasts were manifest in 3 cases and giant cells in 2, but only in meagre quantities, thus palpably less than in the foregoing stage.

Even the frequency of haemorrhages has been less in this group, and haemorrhages have occurred in 8 cases which were denoted profuse in 3, moderate in 3, and scanty in 2 cases. Lymphocytes were observed in 2 cases,

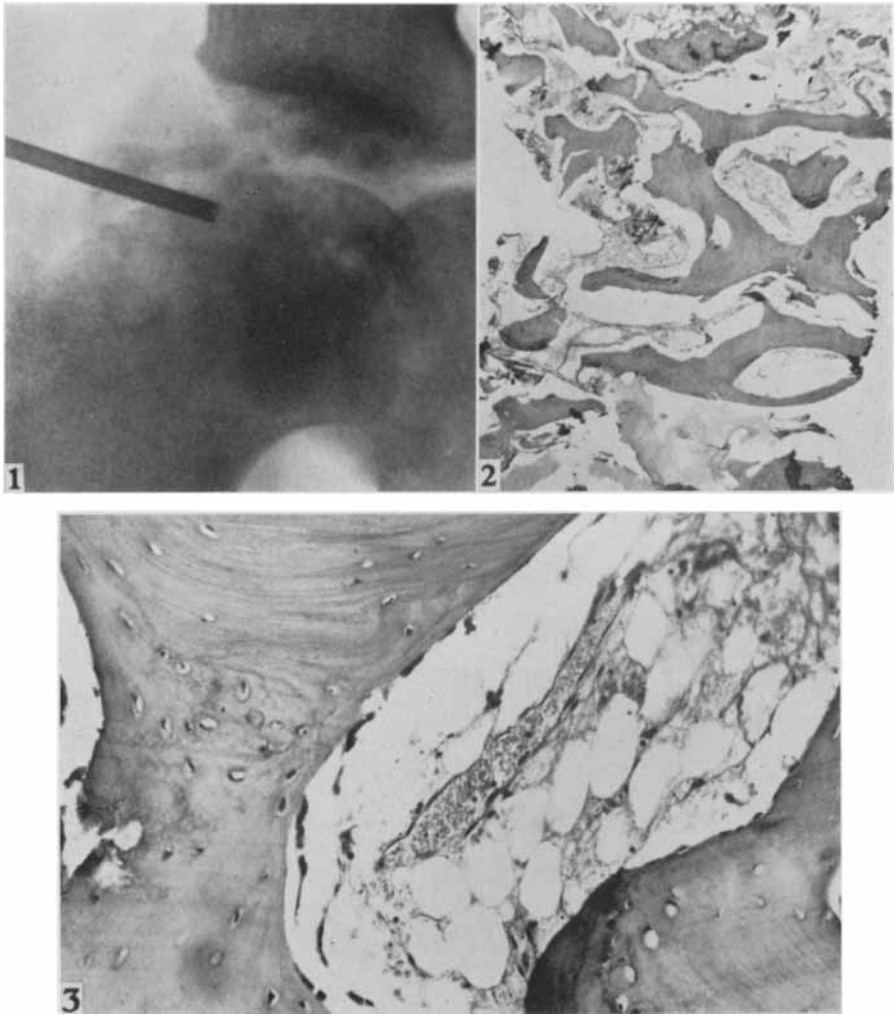

Fig. 17. Case 32. The reparation stage.

1. Roentgen. On the whole the epiphysis has a normal bony structure. Fragmentation remaining in parts.
2. Microphotograp. (×35). The bone forms slender trabeculae with fat marrow in the mesh spaces.
3. (×200). The trabeculae are normal in appearance with well-stained nuclei. In the marrow there is a vessel.

and leucocytes only in 1 case. They have been manifested only in such preparations where there has been a profuse haemorrhage, and their numbers seem to correspond only to the normal content in the blood. Thus, there have not been any signs of inflammation in this stage either.

B. Cartilaginous changes

In 4 of the 14 cases in the reparation stage where cartilage accompanied the preparation, there was a manifestation of changes in the cartilage. These changes, in similarity with the foregoing stages, have been limited to the basal layer of the joint cartilage. And as was the case with the earlier stages, they will be an expression for a lowered vitality of the cartilage and have approximately the same histological appearance as previously described. Within some regions, and as a consequence of degeneration, the cartilage has assumed an almost embryonal appearance with a clear fibrillar structure in the intercellular substance. In other places the vitality of the cartilage has decreased further and here the nuclei show pronounced degenerative changes. In some places they are pyknotic, in other places they are clogged together into large, dark lumps, and, again, in other places they are totally absent. Within these areas showing nuclear changes, the intercellular substance is paler and lacks fibrillar structures. See fig. 18.

C. Discussion

From a histological point of view, there is not the same kind of difference between the fragmentation and reparation stages as there is between the initial and fragmentation stages. In both of the first-mentioned stages the changes are the same. It is only the degree that varies, in so much as in the reparation stage there is a pronounced decrease in the bone necrosis and an equally pronounced increase in normal bone and normal bone marrow. At the same time there is a decrease in the regenerative tissues.

When giving an account of the material in chapter 3, it was pointed out that it was not possible to obtain any preparations from patients in the definite roentgenological stage, and yet, on the base of what has been said above, one could perhaps make certain suppositions in regard to the histological picture in the definite stage. And since the reparation stage manifests an histological picture that is characterized by a decrease in necroses and reparative processes in the epiphysis and by a successive increase of normal bone, and, further, since relapses of the disease are unknown, and, again, since the roentgen picture shows, as the patient grows older, more and more normal bony structure, there is reason to suppose that the reparation of the previously necrotic bone continues, so that a normal cancellous bone occurs in the definite stage.

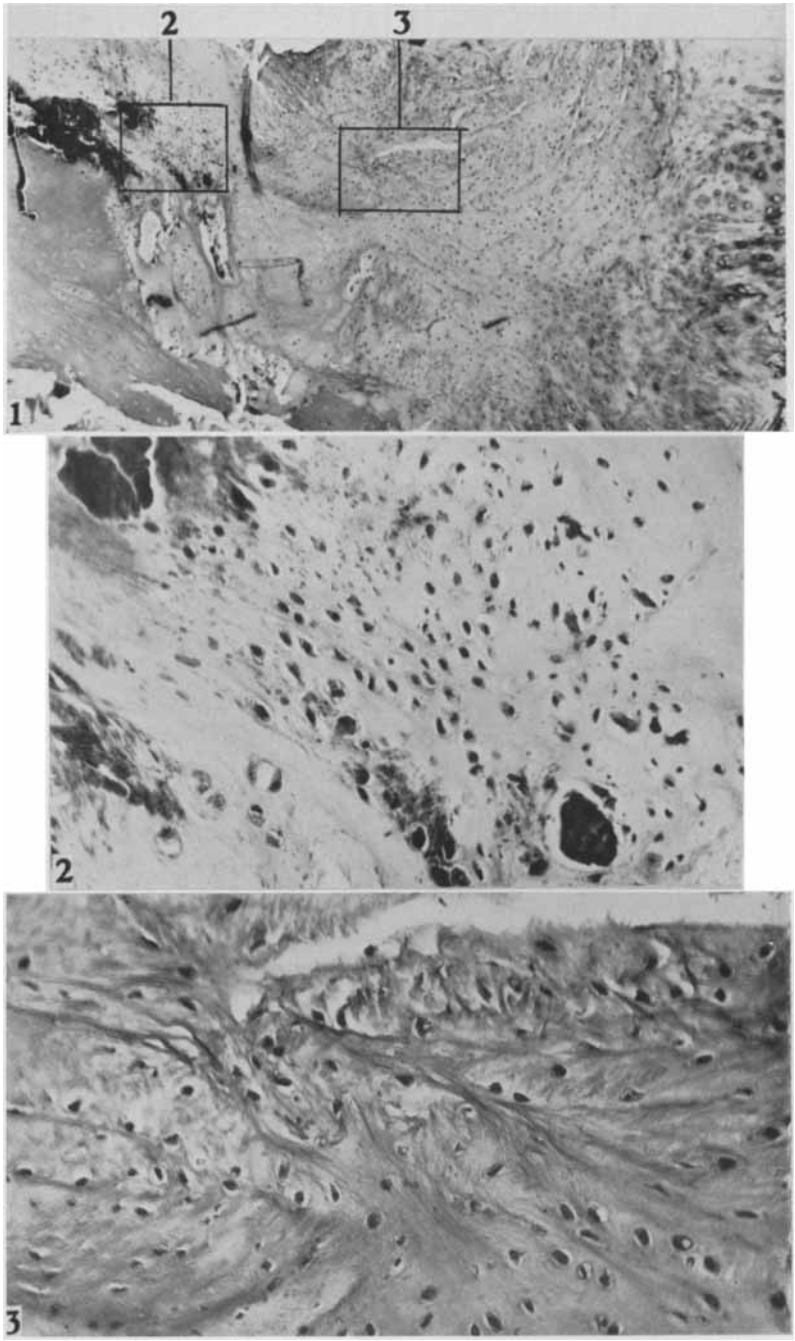

Fig. 18.

CHAPTER 6

Conclusions

From the present investigations the following conclusions can be drawn as to the relation between the roentgen picture and the histological changes occurring in the different stages of coxa-plana.

The roentgenological initial stage corresponds histologically to a high-grade necrosis in the epiphysis of both bone and bone marrow.

The diffuse bone structure appears to be caused by the fact that the marrow spaces, as far as they have been retained, are filled with necrosed marrow and pulverized, necrotic bony masses.

It is possible that the subchondral thinning arises because the necrotic bone is compressed, through loading or muscular tension, after which the elastic joint cartilage and a thin layer of the enchondrally formed bone spring back and leave a gap in the epiphysis. If this is so, it offers an explanation as to why one gets an increased opacity of the epiphysis before the outer contours even are changed.

In the majority of cases in the initial stage the bone is softer than normal.

As a rule there are no histological signs of reparation, for which reason this stage represents the culminating point of the disease.

Because of the macroscopic and microscopic appearance of the bone, as well as the absence of haemorrhages, there is reason to suppose that the necrosis is of an ischaemic nature.

Cartilaginous changes of a degenerative nature occur. They are localized to the basal layer of the joint cartilage and only manifest themselves in the places where the bone necrosis reaches as far as the cartilage-bone margin, indicating that the cause of bone necrosis and cartilaginous change is the same.

Fig. 18. Case 46. The reparation stage.

1. Joint cartilage. (×35). At both ends of the picture the cartilage is normal, but, in the centre, shows pronounced changes.
2. (×200). The nuclei are partially pyknotic, and partially merging together into dark lumps.
3. (×200). The nuclei are irregular in size and shape. The intercellular substance shows clear thready structure.

The fragmentation stage in not the most destructive phase of the disease. On the contrary it is a reparation stage where the opaque parts in the roentgen picture correspond histologically to necroses, and where the lighter areas correspond to an ingrowing reparative tissue and a newly formed bone that has not yet calcified at all, or only partially.

In the fragmentation stage the epiphysis is somewhat harder than in the foregoing stage.

Cartilaginous changes of the same nature as in the initial stage also occur in the fragmentation stage.

The roentgenological reparation stage is also histologically a reparation stage and differs from the foregoing stage only as far as the degree of the changes is concerned. The necroses diminish and normal bone appears instead. The regenerative tissue decreases which, in the roentgen picture, finds expression in the disappearance of the lighter areas.

The bone in the epiphysis seems to be somewhat harder than it is in the fragmentation stage.

Also in the reparation stage there are cartilaginous changes having the same localization and the same characteristics as in the foregoing stage.

With regard to the definite stage, the histological investigations do not give reason for drawing any definite conclusions, but, from the changes in the fragmentation and reparation stages, one seems justified to come to the conclusion that the definite stage constitutes a return to normal bone in the epiphysis.

The investigations will entitle one to suppose that, as a rule, subjective trouble does not manifest itself until the necroses are fairly pronounced. For in the initial stage one comes across cases that have had their trouble for only 2 months, or even for a shorter time, and in which, to judge from the degree of the changes, there is reason to suppose that they have been present for a considerably longer time.

Finally, the histological investigations of normal hips show that the growth of the epiphysis not only takes place from the epiphysial cartilage but also from the joint cartilage, a circumstance which, as it seems, has not been given sufficient attention to in the literature.

PART II

ARTHROGRAPHIC EXAMINATIONS

Arthrographic Examinations

As the ordinary roentgen examinations of joints only give us an idea about the condition of the roentgen-opaque parts, i. e., the skeleton, investigators have long tried different methods of injecting a contrast medium into the joint cavity by which the roentgenogram will also show the organs that are not roentgen-opaque. In this way one has arrived at chiefly two different methods with regard to the contrast medium. When using a negative contrast, a gaseous substance, usually air, is blown into the joint whereby the air layer appears on the roentgenogram like a darker streak. As a rule this method does not give so distinct a picture as is the case when a positive contrast medium is used. This consists of an easy-flowing substance which gives a strong shadow on the roentgen picture, and which rapidly mixes with the synovia. This substance must not irritate the joint and should be rapidly absorbed by the body without giving any reactions. There are many such media available and arthrography nowadays is carried out in most roentgen departments.

It is chiefly the knee-joint that is subjected to arthrographic examination, although most joints can be examined by this method provided they are neither too difficult of access nor too small.

As for hip-joints the method has been used especially when studying congenital dislocation and, in this sphere, SEVERIN (1941) has given a very detailed report on the conditions prevailing in the normal and the dislocated hip-joints of children as presented by arthrographic examination.

Severin has given certain details in the arthrographic picture of the normal hips of children which are characteristic.

The zona orbicularis, the circular, reinforcing ligament in the capsule, presents a ring-shaped contrast defect around the medial part of the collum femoris.

Laterally to the zona orbicularis there is a contrast ring around the collum caused by the fact that the joint capsule has bulged outwardly at this place before becoming fastened to the linea intertrochanterica anteriorly and the lateral part of the collum posteriorly.

The ligamentum transversum acetabuli which, on the inferior side of the joint, stretches between both shafts of the facies lunata, gives a rounded

bulging-in in the contrast medium in the inferior angle between the head and the socket.

Both medially and laterally to the ligamentum transversum the capsule bulges out and, here, one gets two pools of contrast.

The limbus fibrocartilagineus, which forms the free edge of the socket, is seen in the superior part as a wedge-shaped formation with its point lying laterally above the head, and separated from this and from the joint capsule, emanating above the limbus, by two thin contrast streaks.

In good pictures the joint cavity shows up as a thin line.

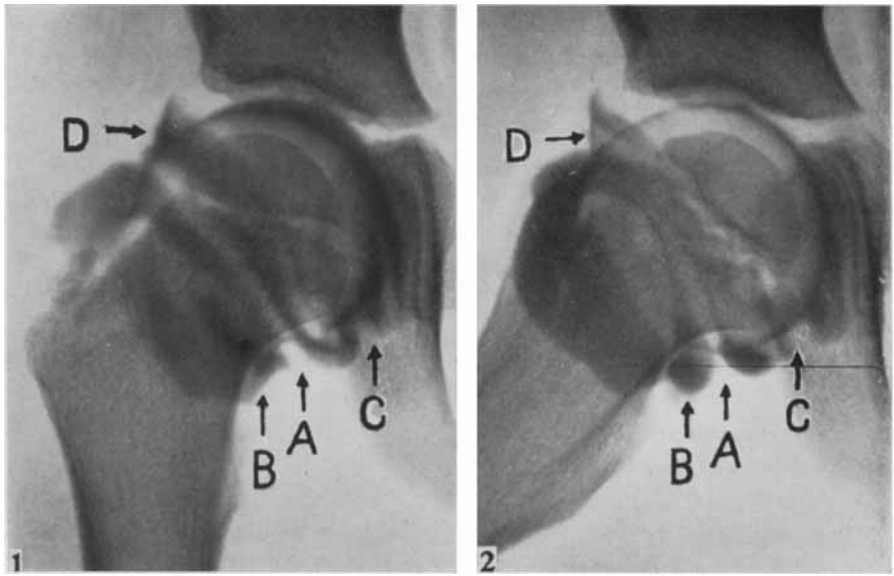

Fig. 19. Normal hip. ♂ 6 years.

A. Zona orbicularis.
B. Contrast ring distal to the zona orbicularis.
C. Contrast defect caused by the ligamentum transversum acetabuli.
D. Limbus fibrocartilagineus.

It is to be supposed that, to a certain extent, arthrography has been carried out also in cases of coxa-plana, since the method of examination ought to give better information about the conditions of the articulating surfaces than ordinary roentgen examinations do. Publications concerning this subject, however, are very scanty.

Waldenström (1938) and Bernbeck (1950) have both undertaken contrast examinations of coxa-plana hips with the primary intention of elucidating the increased distance between the head and the socket to which

already at an early date Waldenström drew the attention as being an early and constant sign in coxa-plana.

However, I have never come across any series of arthrographies in this disease published in the literature, for which reason I have thought it justified to complement the histological examinations with an arthrographic study, as mentioned above, in order, primarily, to study the shape of the cartilaginous head during the different roentgenologic stages.

CHAPTER 7

Material

The arthrographies have been carried out on patients who have been admitted for treatment into the Orthopedic Clinic of the Karolinska Institutet. All in all, 40 children have been examined. The youngest was 4 years and the oldest 13 years at the time of the examination. Sex classification was 34 boys and 6 girls. 34 cases manifested unilateral changes and 6 were bilateral. Of the unilateral cases, one was examined twice during different roentgenological stages of the disease. Of the bilateral cases, 3 were arthographed on both sides and 3 were examined on one side only, due to the fact that the examination was unsuccessful on the other side.

Of the unilateral cases 26 were also examined on the healthy side in order to obtain a normal material for making comparisons. The reason why all of the unilateral cases were not examined on both sides is that, the arthrography having failed for one or other reason on the healthy side, one did not wish to repeat it in order not to prolong anaesthetizing unnecessarily.

The material then comprises 44 arthrograms of coxa-plana hips and 26 arthrograms of healthy hips.

There has been no other selection of the cases than that one has tried to get approximately the same amount from each roentgen stage. I have only managed to get 4 cases in a definite stage due, partly, to the fact that these children only come for treatment very rarely and, partly, to the fact that the parents have not been willing to agree to an examination when once the treatment has been completed.

The examination was carried out between October 1948 and January 1952, but all the cases that were treated during this time are naturally not included; only those are taken up in which the parents consented to the examination.

CHAPTER 8

Technique and Sources of Error

As a rule, the technique described by, amongst others, Severin has been used. The hip-joint is punctured with a somewhat cross-ground needle right from the front and laterally to the femoral vessels, the position of which is determined by palpation of the artery's pulse. After this the needle is introduced through the soft parts in front of the joint and through the joint capsule in the direction of the upper part of the head. By simultaneously rotating the hip carefully back and forth one can feel a slight scratching when the needle comes into contact with the joint cartilage and the injection can then be easily made especially if, during the injection, the hip is flexed some few degrees so that the joint capsule slackens on the front side.

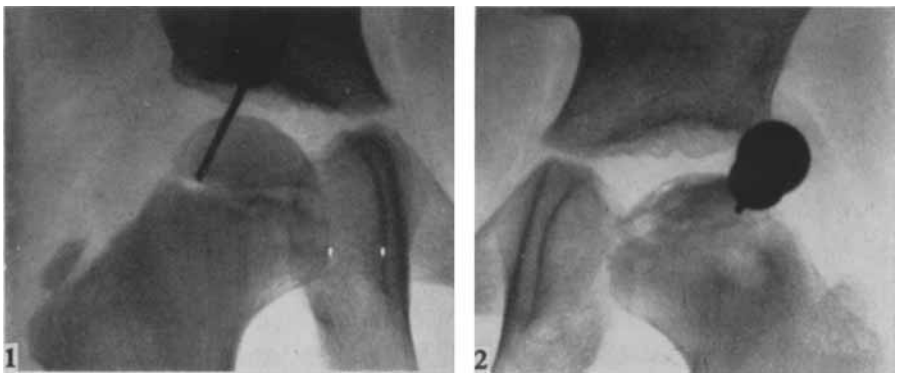

Fig. 20.
Puncture from in front for contrast filling of hip-joint.

In one or two cases the injection has been given from the lateral side of the joint in which case one enters immediately above the point of the trochanter major. This technique strikes me as being more difficult because one has to pass through a considerably thicker layer of soft parts before reaching the capsule, thus necessitating the use of longer needles than when making the puncture from the front.

In a further few cases the puncture was made from underneath the joint. In this case the hip must be flexed to 90 degrees besides being as markedly

abducted as possible. The needle is introduced below the original tendons of the adductor muscles, approximately between the tuberculum pubicum and the tuber ischii, 1 cm laterally to the inferior branch of the os pubis, in the direction of the incisura acetabuli, that is to say, approximately 40 degrees dorsally to and some degrees medially to it. The needle encounters only very slight resistance and one usually feels quite clearly when the point passes the joint capsule. To be on the safe side, one should introduce the needle a further 0.5 cm before giving the injection.

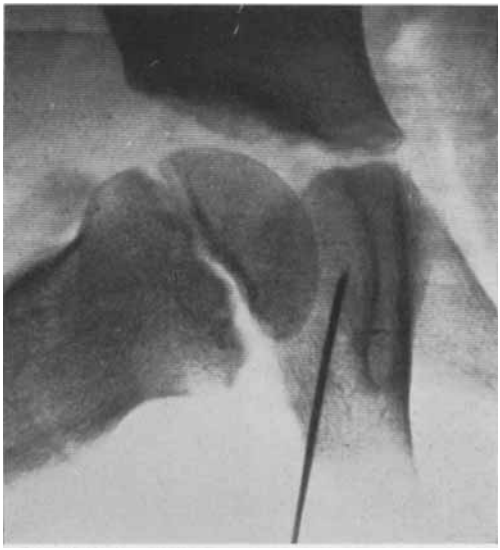

Fig. 21.
Puncture from below for contrast filling of hip-joint.

The advantage attaching to this method is that the incisura acetabuli offers a fairly wide opening to the joint socket. Theoretically, of course, there is a certain risk of damaging the arteria acetabuli with the point of the needle, but, in all probability the vessel lies in such loose tissue that it can give way to the point. In any case none of the children on whom the method was used have shown any signs of haemorrhage.

Puncture from below seems to be best suited for small children in whom the hip can be abducted to the greatest degree.

35 % Umbradil (Astra), diluted with equal parts of normal saline solution, was used as contrast medium, and 3—5 ml, depending on the size of the child, were injected. No damaging effects from the contrast medium was noticed in any of the cases.

On older children arthrography can be carried out under local anaesthesia,

but children are easily frightened and contract their hip muscles making it difficult to inject the contrast medium into the joint. For this reason the examinations, with only one or two exceptions, have all been carried out under general anaesthesia.

The roentgen pictures have been made partly as ordinary frontal pictures and partly in Lauenstein's position. In addition pictures have been taken of the hip in different positions of rotation in several cases.

Measuring Technique

In order to get some idea about the results of the treatment of coxa-plana, EYRE-BROOK (1936) made some measurements on roentgen pictures in two series of patients that had been treated in different ways. He measured the height and breadth of the epiphysis. The relation between the height and breadth multiplied by 100, he called the epiphysial index and used it as a measure of deformation when evaluating the results of the treatment. This index was normally for children under the age of 7 years: 45—55; for children over the age of 7 it was 35—45.

SJÖVALL (1942) used the measuring technique of Eyre-Brooks for a follow-up examination of material from two Swedish hospitals.

HEYMAN and HERNDON (1950) used four quotients in their measurements:

I. The epiphysial quotient = the height of the epiphysial line: the breadth of the epiphysis.

II. The head-neck quotient = the distance from the linea intertrochanterica through the centre of the collum to the top of the epiphysis: the breadth of the collum at its narrowest point.

III. The acetabular quotient = the depth of the socket: the breadth of the socket.

IV. The acetabulum-head quotient = the breadth of that part of the epiphysis which is covered by the socket: the total breadth of the epiphysis.

All quotients were calculated in percentage of the normal hip, and the authors used an average evaluation of all 4 quotients as an expression for the deformation of the head.

All of these measurements refer to the shape of the bony head. I have not come across any reports about suchlike measurements of arthrographs in the literature. All attempts to use any of these methods for measuring the cartilaginous head when making arthrographic examinations were not very successful and so another method had to be tried.

It turned out that the deformed head in the arthrographic picture corresponded approximately to something slightly more than half of an ellipse with the greatest breadth corresponding to the large axis of the ellipse, and

I have made use of this fact when measuring the cartilaginous head. The distances that are measured are, on one hand, the greatest breadth, obtained by measuring the greatest distance between the tangent points of two parallel lines, on the other, the right angular distance from the centre point of the line, which connects the tangent points, to the joint surface of the head. The distance between the tangent points is determined by means of a sliding rule having straight and parallel shafts which are set so that when the sliding rule is laid upon the roentgen picture the shafts meet the inner margin of the contrast layer.

The relation between the height and half of the greatest breadth has been called by me caput index. This index is measured on the roentgen picture both frontally and in Lauenstein's position.

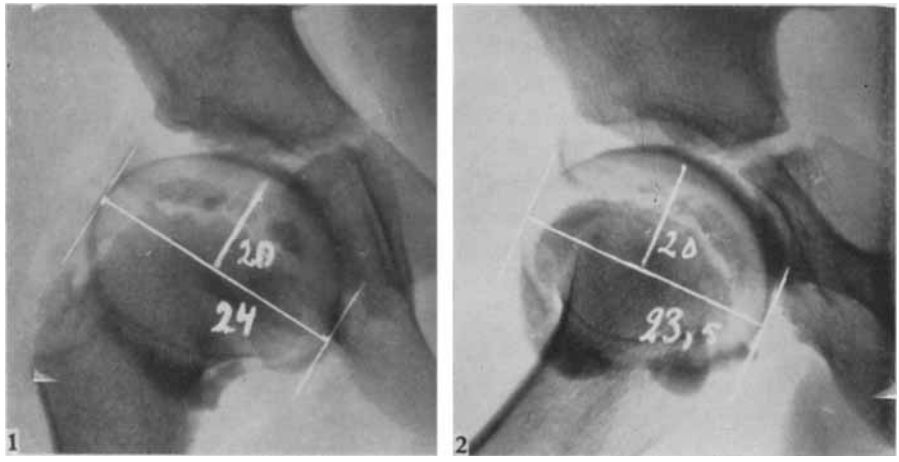

Fig. 22.

The figure shows how the caput-index is determined. The greatest breadth is measured between the tangential points of two parallel lines. From the centre point of the line that connects these two points, the height is measured at right angles to this line.

Sources of Error

The measurements on arthrograms, as measurements made on roentgen pictures on the whole, contain some sources of error that must be taken into consideration when evaluating. The contrast lies like a veil round the free part of the head, and if the contrast medium is too dense or if too much of it is injected, not only will the margins be diffuse, but some of the details might even get lost.

If the leg is exposed to traction there will arise an increased space between the head and the socket where the contrast medium collects so that one gets

a thicker contrast calotte that diffuses the margins and causes the visible cartilaginous layer on the caput to become thinner.

There is always a certain diffusion of the margin between the contrast and the cartilage, but the error of measurement arising from this usually never exceeds 0.5 mm, which, having regard to the relatively great number in question, will not be of any real significance.

In some of the cases the amount of contrast medium that has come into the joint has been somewhat too little so that the margins have been unclear in parts and have had to be filled in with pencil, but not even in such cases will the error in measurement be more than 0.5 mm.

Should one wish to compare the size of the head on both sides in the same case, or compare different patients, the error of measurement can, on the contrary, be significant. And even if one always has the same distance between the roentgen tube and the film, the distance between the hip-joint and the film or the tube can vary considerably depending on the fatness of the patient and the status and position of the pelvis, whereby the roentgen enlargement renders the absolute value that has been measured very uncertain.

I have therefore not tried to give any absolute values; instead, I am anxious to emphasize that it is only a question of relative values that give the relation between two distances in a given plane.

CHAPTER 9

Results

Normal Hips

The normal joint head, according to the authors who have taken measurements from post-mortem specimens (v. LANZ and WACHSMUTH, 1938), is spherical with a slight flattening at the place where the ligamentum teres is affixed, and Severin states that, in arthrographic pictures, the joint head surface should appear to the naked eye as being spherical.

In order, if possible, to have this statement confirmed in terms of figures, the normal hips in my material have been measured in the same manner as coxa-plana hips, with indication of the caput index.

In all cases the head has appeared spherical to the naked eye, and in most cases it has been possible to show those details regarding the deposition of the contrast medium which Severin gives as being normal.

In certain cases there is a manifestation of a slight increase in the breadth of the contrast layer between the medial part of the head and the socket. This is most usual in those cases where the injected amount of contrast medium was profuse and, according to Severin, belongs to the normal variations.

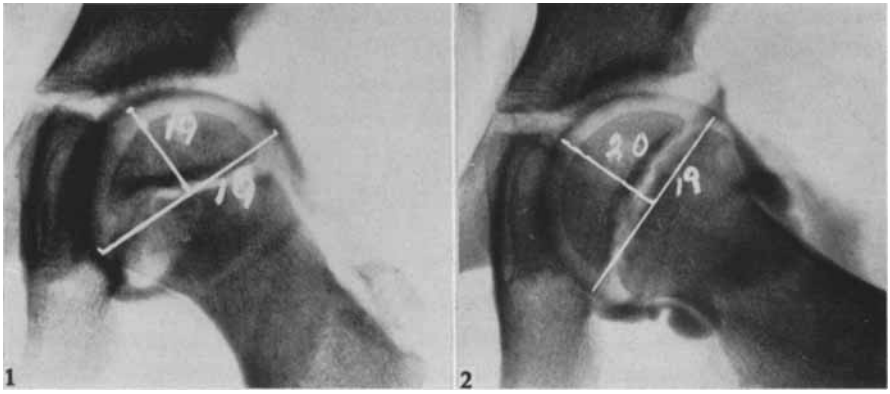

Fig. 23. No. 3147/45 ♂ 6.5 years. Normal hip.
To the naked eye the joint-head seems spherical. The caput-index is 1,000 frontally, and 1,052 laterally.

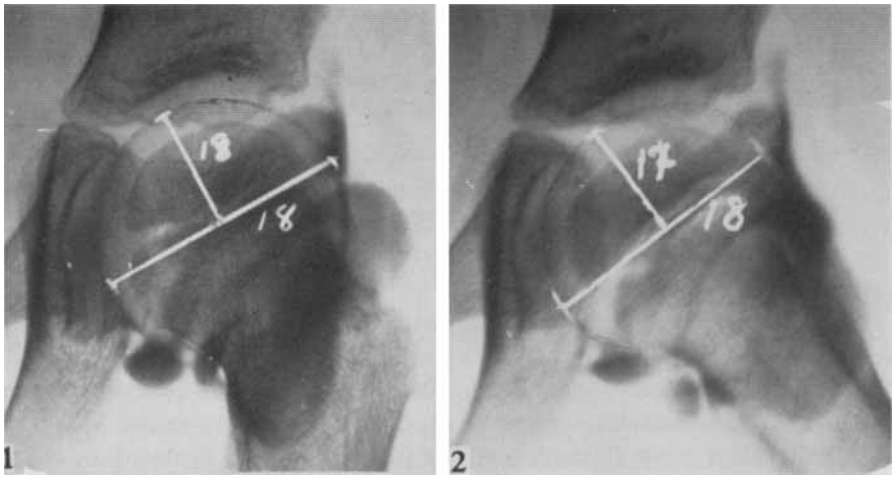

Fig. 24. No. 3276/48 ♂ 8.5 years. Normal hip.

To the naked eye the joint-head seems spherical. The caput-index is 1,000 frontally and 0.944 laterally.

When making measurements one finds that the caput index shows numerous small variations, and that these variations are characterized by the previously mentioned uncertainty when measuring and not by the variations in the shape of the head. The index lies about 1, and the average value for all cases is 0.990 in frontal pictures and 1.003 in the Lauenstein picture. (See figs 23 and 24.) The values will be the same no matter how one places the axes within that part of the caput which is covered by the contrast medium, and the measured distances are thus radii of a circle. As the index is similar in both frontal and Lauenstein pictures, which largely represent two planes standing at right angles to each other, it would seem permissible to draw the conclusion that the normal head is spherical also when it is measured in accordance with this method.

It will be seen from Table V that the distances measured are, in certain cases, different in the frontal and lateral pictures. This does not mean that the head has larger dimensions in the other plane but is due to the fact that the distance of the caput from the film has changed so that the roentgen enlargement appears.

Coxa-plana Hips

Before beginning to describe the arthrographic pictures in the various stages of coxa-plana, I would like to point out that no regard has been taken as to whether the patients have received any treatment, or of what nature the treatment, if any, may have been, but that the aim of the examination is only to try to arrive at a comparison between the arthrograms and the ordinary roentgenograms in the different stages of the disease.

Table V. Showing age and sex of the cases and the shape of the head in the normal material

Case No.	1792/49	2354/47	6212/51	3069/49	3349/51	5266/49	5041/49	5004/48	2519/49	3741/45	6175/51	3386/50	6143/51
Sex	♂	♂	♀	♂	♂	♂	♂	♂	♂	♂	♂	♂	♂
Age in years	4	5	5	6	6	6 1/2	6 1/2	6 1/2	6 1/2	6 1/2	7	7	7 1/2
Shape of the head — Frontal — Measures in mm	17/17	19/19	19/19	19/19	20/20	16/17	21/20	19/20	19/20	19/19	21/21	18/19	20/20
Shape of the head — Frontal — Index	1.000	1.000	1.000	1.000	1.000	0.941	1.050	0.950	0.950	1.000	1.000	0.947	1.000
Shape of the head — Lauenstein — Measures in mm	17/17	19/18	20/19	19/19	19/19.5	17/17	21/21	20/20	19/20	20/19	21/22	20.5/19.5	19/19
Shape of the head — Lauenstein — Index	1.000	1.056	1.053	1.000	0.974	1.000	1.000	1.000	0.950	1.053	0.955	1.051	1.000

Case No.	4842/48	3276/48	2029/49	4583/41	6981/47	184/50	3516/47	179/40	6797/51	4218/48	2801/49	5704/45	1549/49
Sex	♂	♂	♂	♂	♂	♂	♂	♂	♂	♀	♂	♂	♀
Age in years	8	8 1/2	8 1/2	8 1/2	8 1/2	9	9	9 1/2	9 1/2	10	10 1/2	11	13
Shape of the head — Frontal — Measures in mm	23/23	18/18	20/20	20/21	22/22	26/25	22/22	21/21	23/23	23/23	20/21	24/25	24/24
Shape of the head — Frontal — Index	1.000	1.000	1.000	0.952	1.000	1.040	1.000	1.000	1.000	1.000	0.952	0.960	1.000
Shape of the head — Lauenstein — Measures in mm	24/24	17/18	18/19	21/21	23/21.5	25/24	22/22	21/21	23/23	22/23	23/22	24/24	24/24
Shape of the head — Lauenstein — Index	1.000	0.944	0.947	1.000	1.070	1.042	1.000	1.000	1.000	0.957	1.045	1.000	1.000
Average of index: Frontal: 0.990; Lauenstein: 1.003													

The Initial Stage

As a rule there is no great deformation of the bone epiphysis at this stage, so there is scarcely any reason for anticipating that the cartilaginous head should be deformed. The arthrographic examinations showed, indeed, that this was not so to any large extent. In any case the cartilage surface in both planes is evenly rounded and devoid of any unevennesses. (See figs 25 and 26.)

Table VI shows that 4 cases have a caput index of 1.000, while a further 4 cases have an index which is larger than 0.920. 3 cases lie immediately below 0.900, while one case manifests an index as low as 0.740. In all cases it is a question of the index in the frontal plane. In most of the cases the index in the lateral plane is somewhat higher. The average value for the whole group is 0.932 for the frontal plane and 0.964 for the lateral plane.

Table VI. Showing age sex, and duration of trouble of the cases and the shape of the head in the initial stage

Case No.			6212 /51	3733 /48	5041 /49	6143 /51	6086 /48	4835 /49	2029 /49	5266 /49	1549 /49	5245 /49	4266 /46
Sex			♀	♀	♂	♂	♀	♂	♂	♂	♀	♂	♂
Age in years			5	6	$6^1/_2$	$7^1/_2$	$4^1/_2$	$7^1/_2$	$6^1/_2$	$6^1/_2$	13	7	11
Trouble in months			2	2	2	2	$2^1/_2$	$2^1/_2$	3	3	5	12	18
Shape of the head	Frontal	Measures in mm	19/19	20/20	19/20	20/20	14/15	21/23.5	20/20	16/18	20/27	19/20.5	24/26
		Index	1.000	1.000	0.950	1.000	0.933	0.894	1.000	0.889	0.741	0.927	0.923
	Lauenstein	Measures in mm	20/19	21/20	21/21	19/20	16/17	22/23	19/19.5	19/19	19/24.5	19/20	25/26
		Index	1.053	1.050	1.000	0.950	0.941	0.957	0.974	1.000	0.776	0.950	0.962
Average of index: Frontal 0.932; Lauenstein 0.964.													

Further, the table shows that the cases that have the shortest duration of trouble have the greatest index, that is to say, that the shape of the head mostly resembles that of the normal head, whereas the cases that have been troubled for a longer time have a lower index as a sign of the beginning of a deformation of the cartilaginous head. However, as previously mentioned, this was more pronounced in only one case.

It has been pointed out by Waldenström amongst others that already in the early stage of coxa-plana, the head can be palpably enlarged in relation to the healthy side.

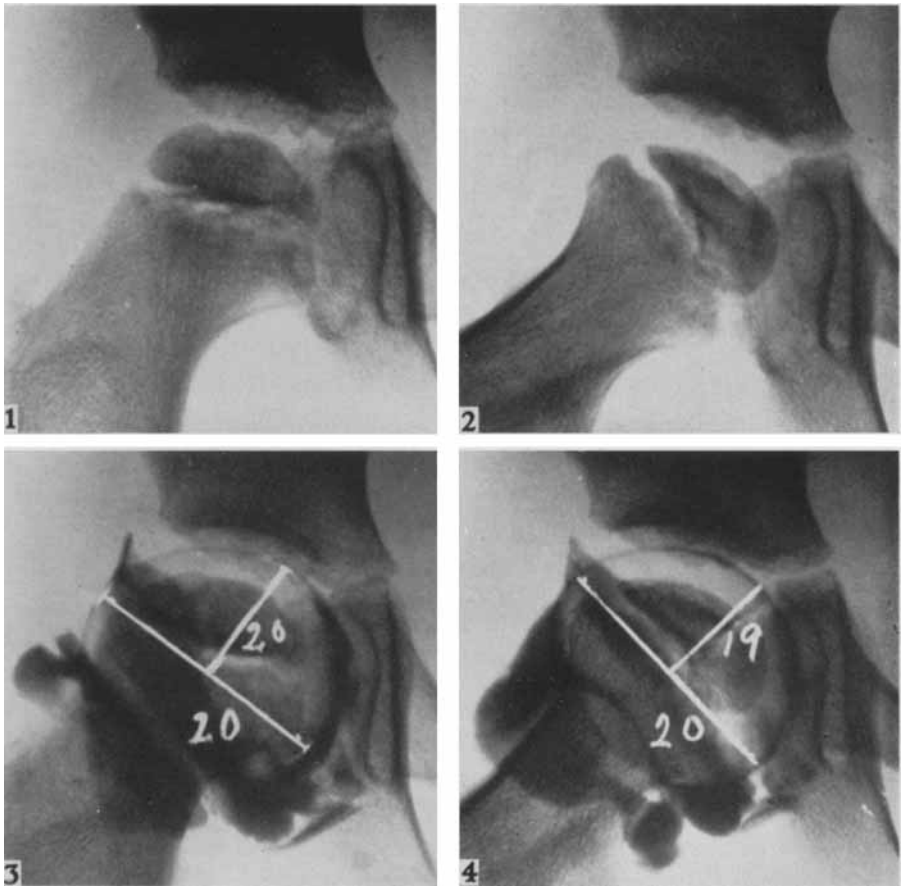

Fig. 25. 6143/51. ♂ 7.5 years. The initial stage.

Roentgen. The epiphysis is condensed and slightly flattened. In Lauenstein's position the subchondral thinning appears.

Arthrography: The joint-head is spherical in appearance, the joint cartilage is of even thickness. The caput-index is 1.000 frontally and 0.950 laterally.

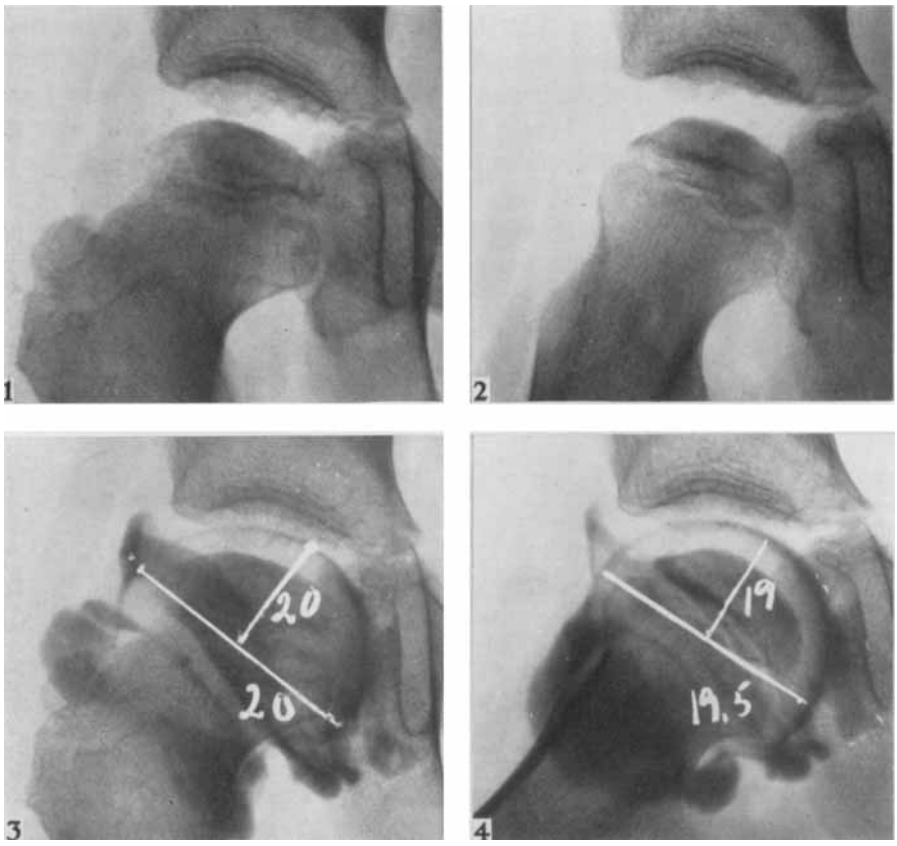

Fig. 26. No. 2029/49. ♂ 6.5 years. The initial stage.
Roentgen: The epiphysis is condensed and slightly flattened.
Arthrography: To the naked eye the joint-head appears spherical. The joint cartilage is of uniform thickness throughout.
The caput-index is 1.000 frontally and 0.974 laterally.

In order to get some idea as to whether this is due to a veritable enlargement of the head, I have compared both sides in individual cases where arthrograms have been taken of both hips. It was not possible, however, to discover any definitely measurable difference. Naturally there was a certain difference in the absolute measurement to be observed in some of the cases, but this difference will be due to variations in the position of both hips together with consequential roentgen enlargements. When looking at the pictures with the naked eye, one does not get the impression either that the affected side is so enlarged that the enlargement could be determined by palpation from the external skin. The palpatory enlargement in early cases is most probably to be explained by the fact that the soft parts around the hips

are swollen. In later stages accompanied by severe deformation of the head there are naturally cases where the saggital axis increases to such a high degree that it is provable by means of palpation.

It has not been possible either to prove any increase in the thickness of the joint cartilage.

The Fragmentation Stage

In this phase of the disease, the bone epiphysis often manifests great changes in its shape with increasing flattening and markedly irregular borders. From the appearance of ordinary roentgen pictures of this stage, one might get the idea that the surface of the joint also showed the same irregularities.

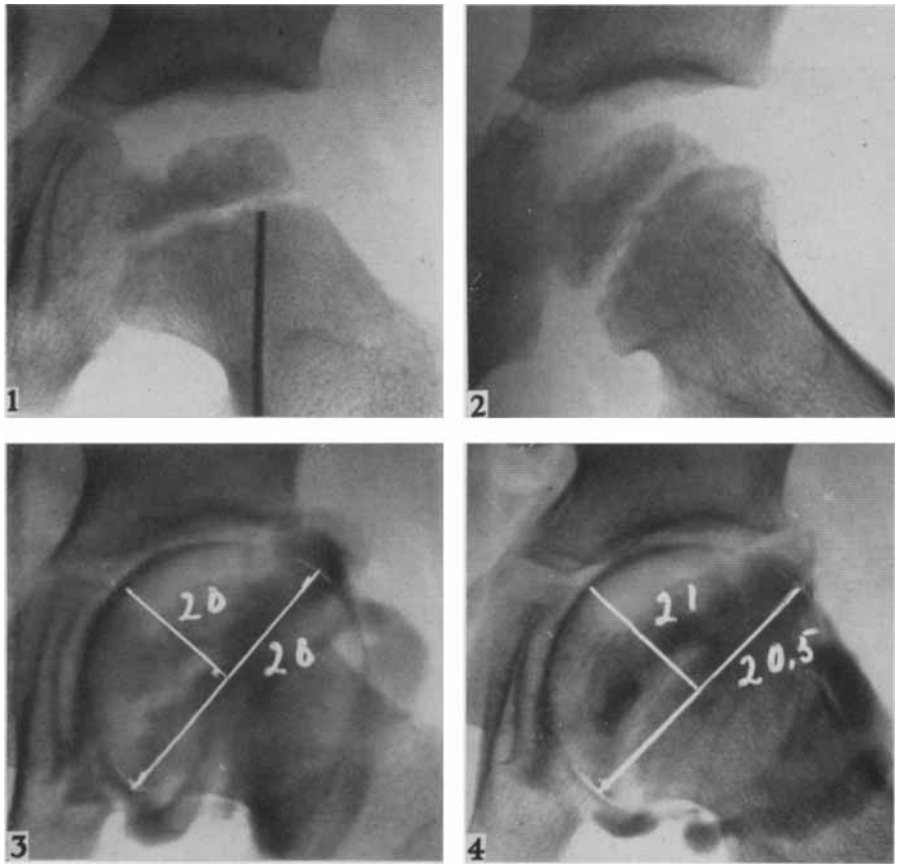

Fig. 27. No. 4016/48. ♂ 4.5 years. The fragmentation stage.

Roentgen: The epiphysis is condensed, somewhat shrunken and fragmentated. (The injection needle is seen frontally.)

Arthrography: To the naked eye the joint-head is spherical in shape. The joint cartilage appears to be thickened on the place where the epiphysis has shrunk.

The caput-index is 1.000 frontally and 1.024 laterally.

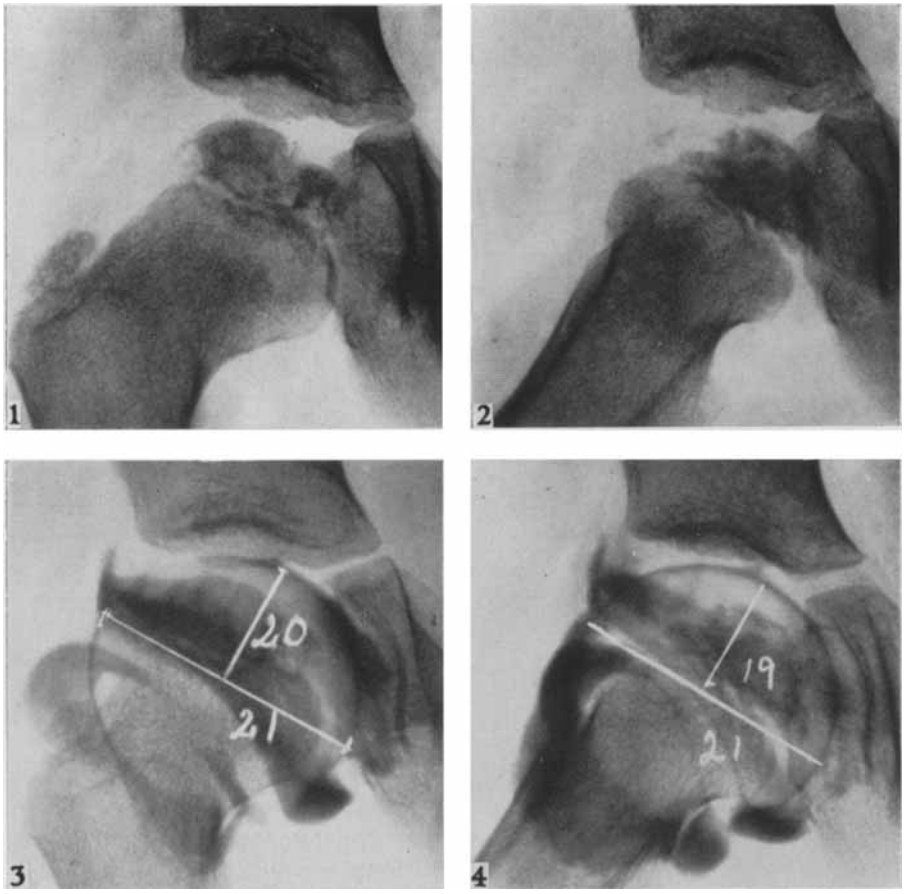

Fig. 28. No. 3276/48. ♂ 8.5 years. The fragmentation stage.
Roentgen. The epiphysis is condensed and fragmentated. Somewhat shrunken.
Arthrography: The joint-head looks somewhat deformed to the naked eye, but the surface of the joint is evenly rounded. The joint cartilage appears to be thickened on the places where the epiphysis is shrunken.
The caput-index is 0.952 frontally and 0.904 laterally.

However, the arthrographs showed that the contour of the surface of the joint does not follow that of the epiphysis, but that it is evenly rounded even in such places where the bone shows depressions. (See figs 27 and 28.)

For this reason one gets in these places at times a marked increase of the layers that are not roentgen-opaque.

When discussing the histological changes in the fragmentation stage, it was pointed out that in no case was cartilage manifest in the preparations, within the epiphysis, but it was surmised that lighter areas here are due to a non-calcified regeneration tissue.

One must then ask oneself whether this regeneration tissue fills up the space right out to the inner margin of a joint cartilage of normal and equal thickness, or whether a veritable increase in the cartilaginous layer has taken place.

It seems scarcely probable that the osteoid tissue would accumulate into one more or less thick layer around the bone epiphysis. In that case it should have been manifest during the histological examinations.

On the other hand, Riedel, Konjetzny and Waldenström have given a description of how the joint cartilage grows down into the partially necrotic epiphysis. This will only happen in such places where the enchondral ossification is disturbed, which, as previously shown, takes place within the area where bone necrosis embraces the ossification zone. It might be thought that here the cartilage, which is preponderantly vital, grows down into the epiphysis when the equilibrium between cartilage proliferation and ossification is disturbed. That the cartilage, as shown from the histological examinations, manifests changes of a degenerative nature in its innermost layer in these places, need not necessarily gainsay this supposition.

It would thus seem as if the joint cartilage in this stage, in the places where the bone subsides, increases in thickness in order to retain the spherical shape of the head as long as possible.

Those parts that are not roentgen-opaque should thus, in their outermost parts, correspond to proliferous joint cartilage, and, further in, to a non-calcified osteoid tissue. But the roentgen picture gives us no idea as to where these two tissues meet.

As previously mentioned, the surface of the joint in the fragmentation stage is evenly rounded, but there is a certain deformation of the cartilaginous head as will be seen from Table VII.

In five cases the deformation is insignificant, having an index that lies over 0.900 in the frontal plane. In six cases the index lies between 0.800 and 0.900, in one case the figure is 0.782 and for another it is as low as 0.652; all values refer to the frontal plane.

The average value for the whole group is 0.860 in the frontal plane, and 0.896 in the lateral plane.

Thus, in this stage the deformation of the cartilaginous head is not as great as one might believe from the appearance of the epiphysis.

The Reparation Stage

As previously pointed out, this stage is characterized histologically by a progressive regeneration, and roentgenologically by a normal bone structure beginning to make itself manifest simultaneously as the borders of the bone epiphysis become more even.

Table VII. Showing age, sex and duration of trouble of the cases and the shape of the head in the fragmentation stage

Case No.			3069 /49	6175 /51	1792 /49	4583 /41	4016 /48	2354 /47	2801 /49	2519 /49	3733 /48	2044 /47	5004 /48	4842 /48	3276 /48
Sex			♂	♂	♂	♂	♂	♂	♂	♂	♀	♂	♂	♂	♂
Age in years			6	7	4	$8^1/_2$	$4^1/_2$	5	$10^1/_2$	$6^1/_2$	6	$8^1/_2$	$6^1/_2$	8	$8^1/_2$
Trouble in months			$2^1/_2$	3	$3^1/_2$	$3^1/_2$	5	5	8	9	10	11	12	12	12
Shape of the head	Frontal	Measures in mm	18/21	21/22	17/18	15/23	20/20	17/21	20/24	18/23	21/23	20/25	21/24	20/25	20/21
		Index	0.857	0.955	0.944	0.652	1.000	0.810	0.833	0.783	0.913	0.800	0.875	0.800	0.952
	Lauenstein	Measures in mm	19/21	21/22	17.5/18.5	18/22.5	21/20.5	19/20	21/24	20/23	21/22	20/25	19.5/22.5	19/24	19/21
		Index	0.905	0.955	0.946	0.800	1.024	0.950	0.875	0.870	0.955	0.800	0.867	0.792	0.905
Avcrage of index: Frontal 0.860; Lauenstein 0.896.															

One has found in the arthrograms of the majority of the cases an evenly rounded joint surface, but there are exceptions where the joint surfaces show a flattening-out corresponding to the depressions in the epiphysis.

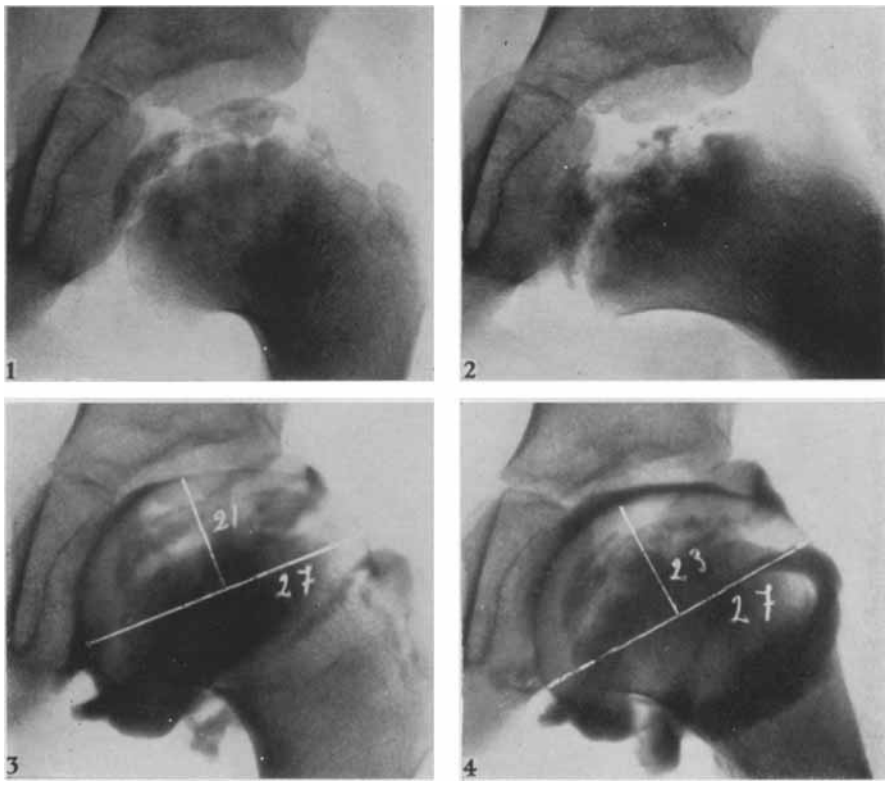

Fig. 29. No. 2044/47. ♂ 8.5 years. The reparation stage.
Roentgen. The epiphysis is considerably flattened and still fragmentated. Normal bony structure begins to appear.
Arthrography: The joint-head is deformed. The thickness of the cartilage is more uniform than in the fragmentation stage.
The caput-index is 0.777 frontally and 0.851 laterally.

On the whole, the cartilaginous head begins to assume the same shape as the bone epiphysis, the joint cartilage does not manifest such large variations in thickness as is the case in the previous stage, but the deformation of the cartilaginous head is more marked. (See figs 29 and 30.)

Of the 16 cases included in this group there were only 2 that had a frontal index greater than 0.900, 4 cases lay over 0.800, 7 lay between 0.800 and 0.700, one case had an index of 0.689 and 2 cases had as low an index as 0.520 and 0.516 respectively.

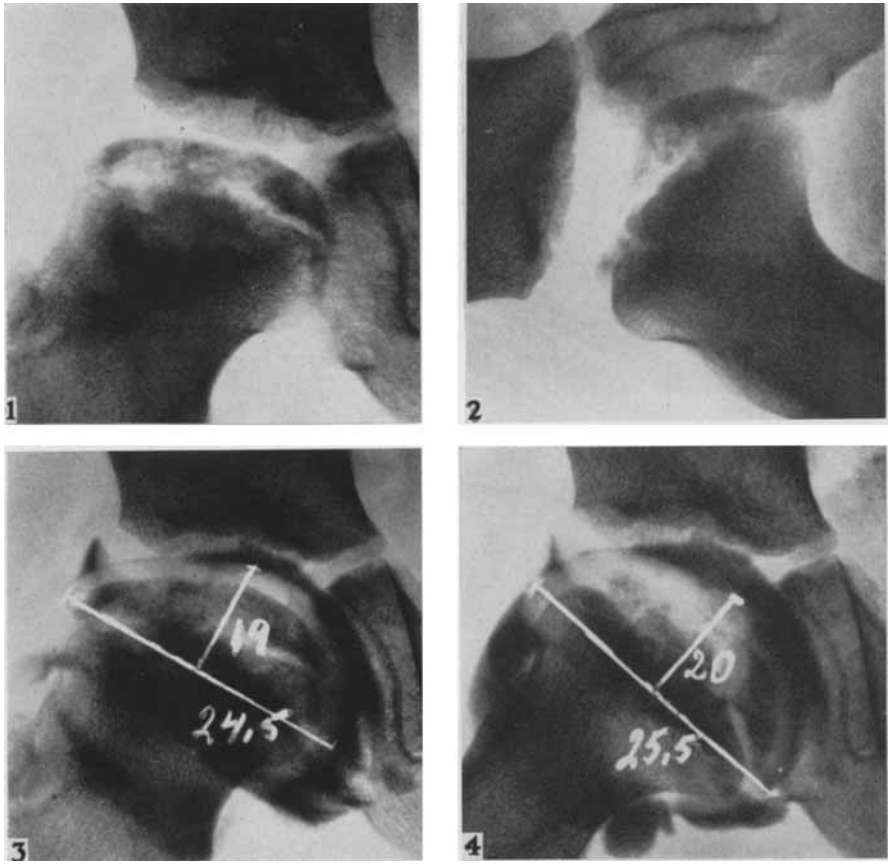

Fig. 30. No. 3386/50. ♂ 7 years. The reparation stage.
Roentgen. The epiphysis is markedly flattened and there are partly remnants of fragmantation. Bone structure begins to be normal.
Arthrography: The joint-head is markedly deformed.
The caput-index is 0.775 frontally and 0.784 laterally.

The average value for the group is 0.766 in the frontal plane and 0.841 in the lateral plane.

It will be seen from the tables and pictures that in those cases that showed deformations, the index is greater in the lateral picture than in the frontal one. As they represent two planes at right angles to one another, one can draw certain conclusions in regard to the shape of the cartilaginous head.

The head is not evenly flattened like the cap of a mushroom but is drawn out at the breadth like an egg or a roller with rounded-off ends. This has already been pointed out by Waldenström after studying ordinary roentgenograms, and the arthrographic examinations thus give clear verification of his conception being correct even in regard to the total head.

Table VIII. Showing age, sex and duration of trouble of the cases and the shape of the head in the reparation stage

Case No.			6797 /51	3349 /51	179 /40	3908 /48	6859 /48	6981 /47	1196 /51	4499 /47	2044 /47	3741 /45	3516 /47	3386 /50	3591 /46	184 /50	5704 /45	755 /44
Sex			♂	♂	♂	♂	♂	♂	♂	♀	♂	♂	♂	♂	♂	♂	♂	♂
Age in years			$9^1/_2$	6	$9^1/_2$	$4^1/_2$	8	6	$7^1/_2$	$5^1/_2$	$8^1/_2$	5	9	7	6	9	11	$8^1/_2$
Trouble in months			$14^1/_2$	$16^1/_2$	17	20	21	22	23	25	27	28	28	30	32	48	55	62
Shape of the head	Frontal	Measures in mm	27/28	18/25	13/25	16/19	20/25	19/24	21/24	18/24	21/27	20/24	21/27	19/24.5	20/28	16/31	28/31	20/29
		Index	0.964	0.720	0.520	0.842	0.800	0.792	0.875	0.750	0.778	0.833	0.778	0.776	0.714	0.516	0.903	0.690
	Lauenstein	Measures in mm	25/25	20/24	23/26	17/19.5	21/25	19/23	21/23	20/23	23/27	20/23.5	22/26	20/25.5	20/27.5	18/29	29/31	23/29
		Index	1.000	0.833	0.885	0.872	0.840	0.826	0.913	0.870	0.852	0.851	0.846	0.784	0.727	0.621	0.935	0.793
Average of index: Frontal 0.766; Lauenstein 0.841.																		

Naturally the shape varies rather considerably from case to case, but the basic shape is as described above.

The reason why the head assumes just this shape will have to be sought in the mechanical conditions of the joint. Loading and muscular tension bring pressure to bear upon the head in the direction of the body's length. During the period when the epiphysis is malleable, it is pressed out in the direction where the socket is unable to prevent its deviation, that is to say, laterally. The movements in the joint which mainly consist of flexion and extension, cause the head to get the roller or ovoid shape already described. That the joint cartilage endeavours, in the early stages, to retain the spherical shape cannot prevent this deformation as the deforming pressure is probably transmitted through the cartilage.

It was previously mentioned that the joint cartilage in the reparation stage begins to assume a more equal thickness. This is probably explained by the fact that the reparative processes within the epiphysis now begin to reach its surface, whereby an ossification begins to manifest itself once more, and a balance between cartilage proliferation and ossification arises.

The Definite Stage

This group is too small to allow of any conclusions to be drawn, but, nevertheless, one can get some idea about the shape of the head in this stage from the 4 cases that can be found in Table IX.

Table IX. Showing age, sex and duration of trouble of the cases and the shape of the head in the definite stage

Case No.			4218/48	4266/46	4686/45	6981/47
Sex			♀	♂	♂	♂
Age in years			10	11	11	8½
Trouble in months			29	45	48	55
Shape of the head	Frontal	Measures in mm	25/31	24/30	20/31	22/25
		Index	0.806	0.800	0.645	0.880
	Lauenstein	Measures in mm	27/32	25/28.5	24/30	22/24
		Index	0.843	0.877	0.800	0.916
Average of index: Frontal 0.782; Lauenstein 0.854.						

The deformation is approximately the same as that in the foregoing group with a caput index in the frontal plane averaging 0.782, and in the lateral plane 0.854.

The average value of the index is thus somewhat greater in the definite stage, but this can probably be explained by the fact that the cases are so few in number.

It would seem, then, as if some essential deformation of the cartilaginous head had not taken place in the definite stage but that the head had obtained its final shape during the reparation stage. Naturally, in this case, I disregard the change in the shape of the head that occurs later on in life as a consequence of deformative arthrosis.

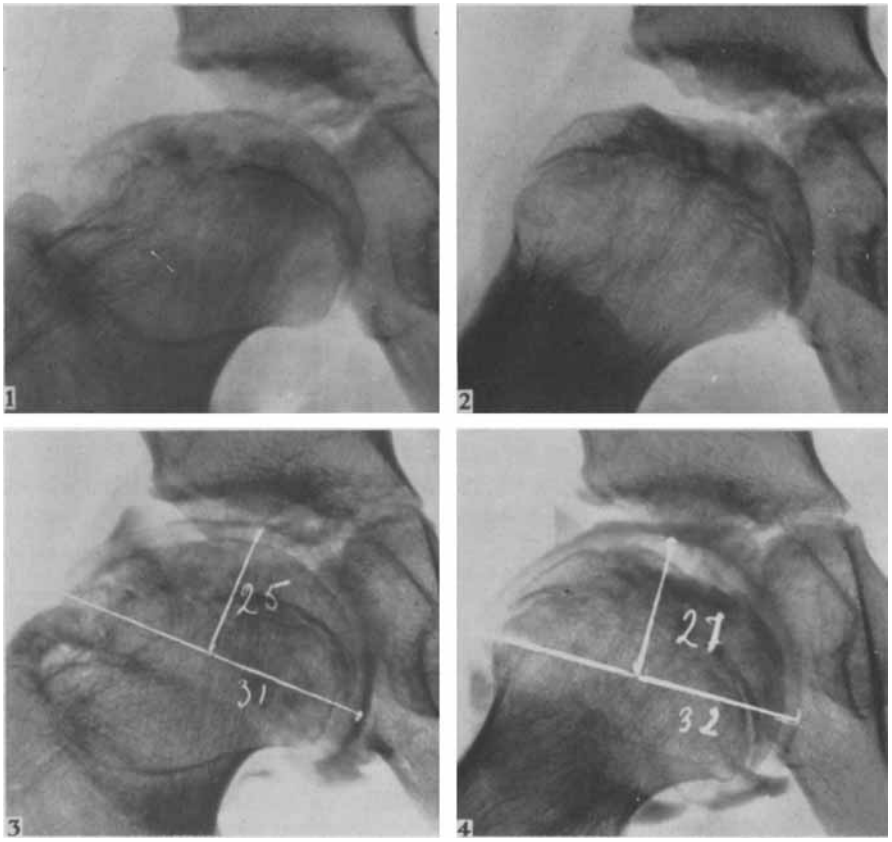

Fig. 31. No. 4218/48. ♀. 10 years. The definite stage.

Roentgen. The epiphysis is flattened but has even contours. Bone structure normal. Arthrography: The joint-head is deformed to about the same degree as the epiphysis. The joint cartilage is relatively thin but of even thickness.
The caput-index is 0.806 frontally and 0.843 laterally.

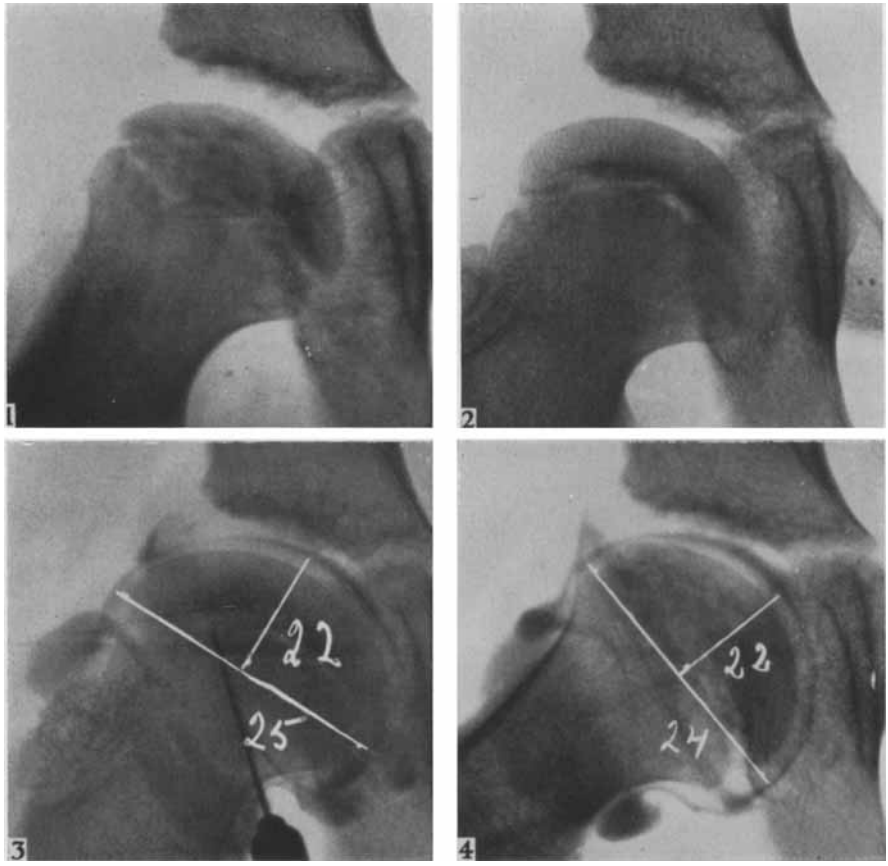

Fig. 32. No. 6981/47. ♂ 8.5 years. The definite stage.
Roentgen: The epiphysis is flattened. The contours are even. Bony structure normal. Arthrography: The joint-head is moderately deformed. The joint cartilage of uniform thickness.
The caput-index is 0.880 frontally and 0.916 laterally.

In this stage the joint cartilage shows no alternating thicker and thinner parts, but covers the bone as an equally thick layer of approximately that thickness which one finds in the normal hips of corresponding ages.

The Increased Head-socket Distance

This symptom was first described by Waldenström in 1934 as an early and constant sign in coxa-plana. He then measured the distance from the inner edge of the epiphysis to the so-called lachrymal figure.

Waldenström was of the opinion that the symptom arose by the head decreasing its volume whereby the intra-articular pressure fell. This would

bring about an increased flow of blood to the soft parts in the fossa acetabuli which swell and press the head laterally.

BERNBECK (1947, 1950), who has also studied this symptom, thinks that the lateral displacement of the head is due to a thickening of the joint cartilage.

Theoretically one can imagine three reasons for the increased head-socket distance in early cases, and these reasons, which also Bernbeck discusses, are:

1. A large exudate which exercises so great a tension on the joint capsule that it seems to pull the head with it laterally.
2. A thickening of the joint cartilage.
3. Swelling of the soft parts in the fossa acetabuli.

1. It is probable that an exudate is manifest in the joint in the majority of cases. In some cases Waldenström succeeded in sucking out exudate, and amongst my own cases, it has sometimes been possible, when puncturing the epiphysis or during arthrography, to draw off synovial fluid, but it has only occurred in small quantities so that one has not had the impression that it was subjected to pressure, which should be the case if the exudate had caused so much tension on the joint capsule that there was a lateral displacement of the head. After the exudate had been sucked out, the head ought to have glided back towards the base of the socket and the distance decreased, but this did not take place in those cases where the synovial fluid had been tapped out.

If an exudate had caused a marked tension of the joint, the head should have, so to say, swum around the closed space made by the joint capsule and, when the patient was in a state of general anaesthesia and of complete relaxation, one should have been able to push it in again towards the base of the socket. I have made experiments with this and, on anaesthetized children, I have exercised strong pressure by hand on the trochanter in the direction of the base of the socket, but this has not decreased the head-socket distance.

It may therefore be considered proved that the cause of the increased head-socket distance is not the presence of exudate in the joint.

2. Bernbeck founds his opinion that the increased head-socket distance is due to a thickening of the cartilage on NUSSBAUM's (1923) experimental tests on animals as well as on the results of his own investigations.

In rabbits, Nussbaum was successful in bringing about changes resembling coxa-plana in man by severing the vessels that supply the superior femoral epiphysis. In some cases that were followed up by roentgen examinations he obtained, as a first sign, an increase in the breadth of the joint cavity and, after making microscopic examinations, he came to the conclusion that this was due solely to a thickening of the joint cartilage.

Bernbeck has made some swelling experiments on the cartilage and found that, under certain conditions, it can increase its volume considerably. Further, he has made arthrographs of coxa-plana hips and found that there was a thickening of the joint cartilage.

As previously mentioned, there is an increase in the thickness of cartilage manifest in certain cases in my own material, but this only occurs in such places where the bone epiphysis has shrunk and where there is a compensatory growth of the cartilage in order, as far as possible, to retain the spherical shape of the cartilaginous head.

In order to judge the general thickness of the cartilage one ought not to begin with such cases, but only with those in the very earliest initial stages where there has been no marked change in the bone epiphysis, more especially because the increased head-socket distance is an early and constant symptom.

In these early cases it has not been possible, as far as the present arthrographic pictures are concerned, to show any definitely measurable thickening of the joint cartilage on the affected side and the examinations can therefore not give any support to the conception that the increased head-socket distance would be due to an increase in the thickness of the joint cartilage.

3. Thus, the thickness of the cartilage is not increased generally, and when one measures the distance from the surface of the cartilage to the base of the socket on the arthrogram, one finds that this is greater on the affected side. Therefore there must be something that presses the head outwards.

In all the cases where a sufficient filling-out of the joint socket with contrast medium has been obtained, we find an interposition of a contrast layer between the head and the base of the socket. This layer is crescent-shaped having its base on the inferior margin of the socket and its laterally-directed point upwards. The broader the layer is at the base, the higher up stretches its point. (See fig. 33.)

As Severin has pointed out, this contrast layer can also occur in normal hips, and in the present material it is also found in a few cases, but only in such where one has so strong a contrast filling that the vacuum in the joint is considerably decreased so that the head can leave the socket base. However, it will never be so broad in normal hips as it is in coxa-plana hips and it will disappear when the head presses against the socket, which is not the case with changed hips. Besides it is tighter in normal hips. (See fig. 33.)

One sometimes sees on normal pictures, just above the defect in the contrast medium given by the ligamentum transversum and in connection with it, a small triangular defect in the contrast which Severin has proved to be the origin of the ligamentum teres. In coxa-plana hips this defect is much larger and can, in the most technically successful cases, form a wedge-shaped defect which breaks the contrast layer around the joint cartilage of

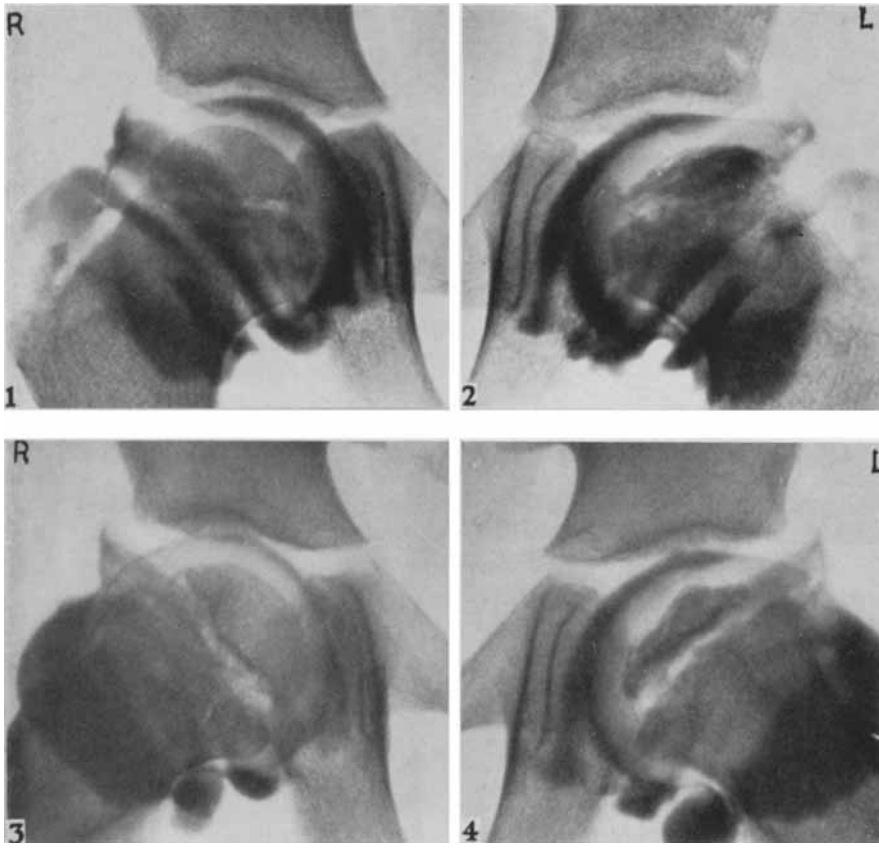

Fig. 33. No. 3069/49. ♂ 6 years. Right hip normal. Coxa-plana on left side in the fragmentation stage.

In frontal aspect can be seen on both sides a contrast-layer between the head and the medial side of the socket, but the contrast-layer is considerably broader on the affected side. In Lauenstein's position this contrast-layer almost entirely disappears from the healthy side, while it remains on the affected side. The soft parts in the fossa acetabuli and the ligamentum teres show up as a wedge-shaped defect in the contrast. This contrast defect becomes less in height in Lauenstein's position.

the head, immediately below the place for the fovea centralis capitis in a direction obliquely upwards and laterally. (See fig. 33).

The upper part of this defect can scarcely be caused by anything else but a thickened ligamentum teres which, in a suitable projection, breaks the contrast layer where the ligament lies against the surface of the head immediately below its insertion in the fovea centralis.

That it is a question of the ligamentum teres is further supported by the circumstance that the upper part of the defect disappears in Lauenstein's posi-

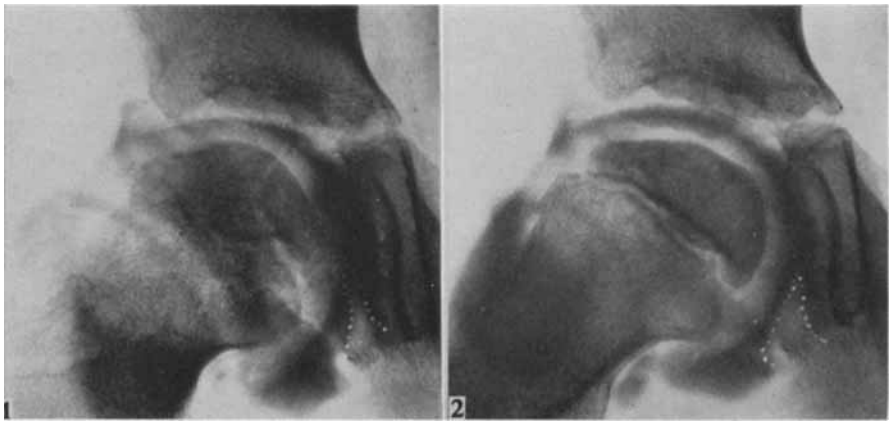

Fig. 34. No. 4835/49. ♂ 7.5 years. The initial stage.

A fairly broad contrast-layer is interposed between the head and the medial part of the socket. It does not disappear in Lauenstein's position. For the sake of clarity the crescent-shaped defect in the contrast is marked with dotted lines.

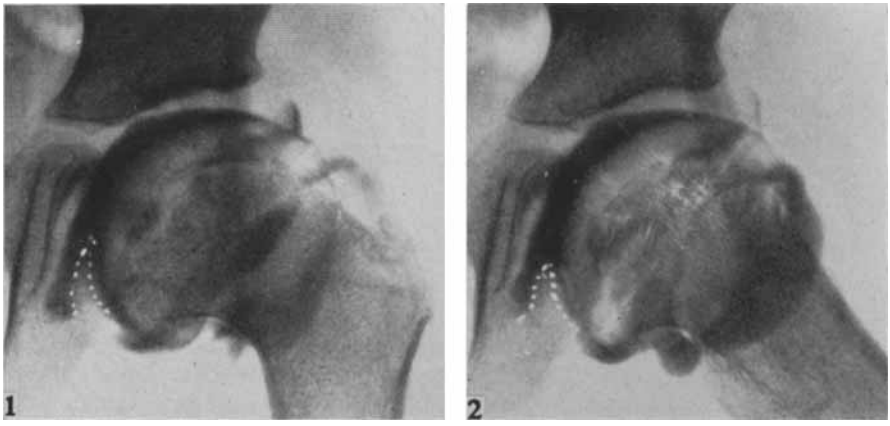

Fig. 35. No. 3908/48. ♂ 4.5 years. The reparation stage.

A broad contrast-layer that does not disappear in Lauenstein's position, is visible between the head and the bottom of the socket. The crescent-shaped defect in the contrast (made more clear with dotted lines), diminishes in height in the lateral picture.

tion when, as is known, the ligament is stretched and its insertion is carried ventrally and, thus, is not touched by the tangential beams, which is a presupposition for its being able to appear during arthrography. In the side pictures, then, the wedge-shape defect is shortened, but it retains its breadth at the base.

It is scarcely probable that the ligamentum teres alone, even if it is thickened, can give so broad a contrast defect as that which is discussed

here, and one may therefore presume that the soft parts in the fossa acetabuli are swollen, too, and contributory to causing the displacement in the contrast. According to v. Lanz and Wachsmuth, the fossa acetabuli is filled out by a pad consisting of fat and connective tissue, and an inflammatory swelling of this tissue and of the ligamentum will be the cause of the above mentioned changes.

Howorth (1948), after having operated a large number by dissecting and drilling, showed that the soft parts in the joint, especially the synovialis, are very inflammatorily thickened, and he presumes that such is also the case with the soft parts in the fossa acetabuli, through which the head would be pressed laterally.

My arthrographic examinations thus give support to Waldenström's and Howorth's opinion that a swelling of the pad of the soft parts in the fossa acetabuli displaces the head laterally and show that the thickening of the ligamentum teres also takes part in the origin of the increased head-socket distance in early cases without any special deforming of the head. In older cases with a high-grade deformation one cannot deny the possibility that an incongruence in the joint surfaces causes a displacement of the head.

CHAPTER 10

Conclusions

The following conclusions may be drawn from these arthrographic examinations:

The normal hip-joint head in children, which, on being measured on preparations, has shown itself as being spherical, also appears as being spherical on the arthrograms, and not only to the naked eye but also from the measurements that have been made on the pictures.

In the initial stage of coxa-plana, which is histologically characterized by a massive bone necrosis without signs of reparation, the head retains, on the whole, its spherical form. There was no manifestation of any thickening of the joint cartilage, and the palpable enlargement of the head which can often be manifested clinically will not be due to a real enlargement of the head but to swelling of the soft parts in these regions.

The high-degree deformation of the bone epiphysis, most often encountered in the fragmentation stage, does not correspond to an equal high-degree deformation of the cartilaginous head. On the contrary, the deviations from the spherical form are relatively insignificant and this will be due to a compensatory thickening of the joint cartilage in the place where the bone has shrunk.

It is only in the reparation stage, which both roentgenologically and histologically is characterized by a return to the normal structure, that a more pronounced deformation of the cartilaginous head takes place. The difference in the thickness of the cartilage in different parts of the head levels up, and the cartilaginous head assumes, on the whole, the same form as that of the epiphysis and, because of the mechanical conditions in the joint, resembles approximately an ellipsoid in frontal position.

In the definite stage the final healing of the bone changes takes place and the cartilage resumes its normal thickness, but no other change in the shape of the head seems to take place.

The increased head-socket distance is most probably due, not to a general thickening of the joint cartilage, but to an inflammatory swelling of the soft parts in the fossa acetabuli and of the ligamentum teres which presses the head laterally.

PART III
ASPECTS ON THE TREATMENT

Aspects on the Treatment

The most important principle in our therapeutic measures is to attack the basic causes of a complaint. Unfortunately, however, it is not possible to follow this principle in regard to coxa-plana because, as yet, we are not cognizant of the innermost nature of the disease. Amongst other things, it has been held that the palpable changes would be due to hormonal disturbances or to avitaminosis, yet it has not been possible to produce any definite proof that this is so, and the substitution therapy which, arising from this assumption, has been resorted to has not given any convincing results.

As we are not yet in possession of any causal therapy, we are still recommended to treat the affliction purely symptomatically and, in this respect, the conservative therapy has been the prevailing one. It is not possible here to deal more closely with all the different suggestions for treatment that have been recommended in the profuse literature dealing with coxa-plana, or to make reference to the results that have been achieved by one or other methods of treatment.

However, we can say, summarily, that the leading principle for the conservative therapy has been confinement to bed, sometimes coupled with extension treatment when there are pains and contracture in the hip-joint, besides avoiding any loading as long as no reliable signs of healing could be traced on the roentgen picture.

Already at an early date, however, there were those who were in favour of a more active operative treatment. One of these was Phemister who, in 1921, gave a report on a case which he had operated on by exposing the joint, chiselling out a hole on the anterior side of the head, and curetting the epiphysis. The case healed quickly and he recommended that the operation should be performed at an early stage.

Cordes (1930) also recommended an operation that ought to consist of creating a communication between the metaphysis and the epiphysis, and Gill (1940) was of the opinion that a drilling operation was useful only if it was carried out in the early stage. In 1934, Ferguson and Howorth had operated on 15 hips by dissecting the joint and drilling up into the epiphysis. Their results were good, and the time of healing was shortened considerably.

In 1948, Howorth gave an account of 50 cases that had been operated on in a similar way with the same good results, and the following year (1949), Haythorn gave his previously referred description of 33 cases that had been operated on by curetting the epiphysis.

PITZEN (1951) and HAUBERG and MATTHIASH (1952), have used instead a three-edged collum nail that was hammered near to or through the epiphysial cartilage in order to bring about a more rapid healing.

The object of our treatment of coxa-plana ought to be twofold, on one hand to retain the spherical shape of the head as far as possible, on the other to bring about as rapid a healing of the bone as possible.

The arthrographic examinations have shown that the joint head, in the initial stage, shows much smaller deviations from the normal in regard to the shape, and it would therefore be desirable to have a treatment that prevented further changes in the shape.

The histological examinations show that the epiphysis, in the same stage, is the seat of a pronounced necrosis that stretches over a considerable time before any signs of reparation set in. During this time, the epiphysis seems to be fairly soft and therefore sensitive to influences of a form-changing nature. Therefore, during the treatment, it is essential, as soon as possible during the initial stage, to start a rapid bone regeneration in order, if possible, to prevent the epiphysis from shrinking. By beginning early with a lengthy unloading, the conservative treatment can certainly give a better final results as far as the shape of the head is concerned than if no treatment at all is given or the treatment is carried out inconsequently, but, as far as one can judge, it cannot accelerate healing: the disease passes through the various stages independent of the conservative steps that have been resorted to.

That it takes so long in untreated or conservatively treated cases, before one gets any signs of an initial healing in the epiphysis might be explained by the fact that the epiphysis is enclosed on all sides by a rather thick cartilaginous covering through which it takes a long time for the regenerative tissues to penetrate. The treatment should therefore be directed to breaking through this cartilaginous barrier, and this can only be done by establishing a communication between the metaphysis, that contains the reparative tissues, and the necrotic epiphysis.

The method of operative treatment, given by Ferguson, Howorth and Haythorn, and which I myself have used in 3 cases, consisting of an exposing of the hip-joint and drilling through the epiphysial cartilage, seems to be a somewhat unnecessarily great operation for the patient. The most important thing, as previously mentioned, is to perforate the epiphysial cartilage and establish a communication between the metaphysis and the epiphysis, and this it will seem possible to do more simply by just drilling from the external part

of the trochanter up through the collum femoris into the epiphysis, or by using Pitzen's method of nailing whereby, in accordance with the discussion above, the point of the nail should penetrate the epiphysial cartilage.

But whichever of these operative methods is chosen, the operation should be performed early in the initial stage. In the fragmentation stage, as the histological examinations show, the reparation of the bone has already begun, and the operative treatment will probably not give equally good results.

Summary

Ever since 1909, when coxa-plana was first described, it has been the object of investigation by a number of authors. Consequently the disease is well known from certain aspects while from others it is not yet completely elucidated. The disease is clinically and roentgenologically well known, and as far as concerns its pathological anatomy, most investigators are unanimous that the essential changes are due to osteonecrosis. On the other hand there are no investigations that describe the histo-pathologic changes against a background of roentgen pictures. Nor are there any series examinations that would elucidate the conditions of the joint cartilage and the "cartilaginous head".

The investigation at hand has the intention of answering two main questions.

1. *Which histological changes correspond to the different roentgen stages?*
2. *What is the appearance of the "cartilaginous head" in the same stages?*

Waldenström's classification has been largely used when determining the roentgenological stages. Thus the initial stage embraces those cases where the roentgen picture shows an increased head-socket distance, subchondral thinning together with a dense and, sometimes, shrunken epiphysis, but where the epiphysis is still homogeneous and no thinning zones can be seen.

The fragmentation stage comprises those cases where the epiphysis shows alternating condensed and thinned parts, but where normal bony structure has not yet begun to return.

The reparation stage includes those cases where, in the roentgen picture, one sees a reappearance of normal bony structure together with a disappearance of the condensed areas.

For the sake of simplicity, Waldenström's two last stages have been joined into one and called the definitive stage.

Part. I. Histological Examinations

The histological examinations have been made from preparations taken from biopsies on patients that were admitted for treatment to the Orthopedic Clinic of the Karolinska Institutet. The material comprises 34 patients on whom 44 biopsies have been made. It was not possible to get any preparations from patients in the definite stage because the parents were unwilling to agree to have the examinations made after completion of the treatment.

The normal material consisted of preparations obtained by autopsy on children within the age-group where coxa-plana was manifest.

The biopsies were carried out by means of a hollow needle having an inner diameter of 2 mm.

Initial Stage

14 biopsies were undertaken in this stage. In most cases the bone in the epiphysis was soft when punctured.

The histological picture is dominated by a pronounced necrosis of both the bone and the bone marrow. The trabeculae are often crushed to small fragments and the marrow spaces are, in so far as they are intact, filled with an amorphous mass consisting of necrotic bone marrow and small fragments of necrosed bone. In this stage there is no sign whatever of bone regeneration. Haemorrhage was sparsely manifest and there were no signs of any inflammatory processes.

The joint cartilage also manifested changes in some cases, indicating a markedly lowered vitality of the cartilage. These changes are localized to the basal layer of the cartilage and appear in the places where osteonecrosis reaches out to the cartilage—bone margin, denoting that the cause of the osteonecrosis and the cartilaginous changes is the same.

The Fragmentation Stage

In this stage 15 biopsies were carried out. When punctured the bone was somewhat harder in this stage than in the foregoing.

Also in the fragmentation stage the osteonecrosis remains, but the histological picture is dominated by pronounced signs of regeneration of the necrotic epiphysis. A tissue resembling connective tissue, profuse in cells and rich in vessels, grows into the necrotic masses which are resorbed and supplemented by a newly formed, immature bone which, in places, begins to assume a clear lamellar structure. Giant cells and osteoblasts occur within the regenerative tissue. There is a greater number of blood corpuscles in the preparations than in the foregoing stage.

Cartilaginous changes of approximately the same nature as in the foregoing stage were manifest in the fragmentation stage.

The Reparation Stage

15 biopsies were carried out in the reparation stage. The bone, when punctured, was somewhat harder than in the foregoing stage.

The histological picture is characterized by a reduction of the necrosed bone and of the regenerative tissue, which are replaced by normal cancellous bone.

In this stage there are still cartilaginous changes of the same nature and the same localization as in the *foregoing* stages.

The following *conclusions* may be drawn from the histological examinations.

The initial stage is the culminating point of the disease and is recognized by, practically speaking, a total necrosis of the epiphysis. The diffuse bone structure in the roentgen picture seems to be due to the marrow spaces being filled with necrotic bone masses.

On account of the macroscopic and microscopic appearance of the bone together with the absence of haemorrhages, there is reason to suppose that the necrosis is of ischaemic character.

It is possible that the roentgenological symptom which is called the subchondral thinning arises in this manner: the necrotic bone becomes compressed when loaded or subject to muscular tension, after which the elastic joint cartilage and a thin layer of adherent bone spring back and leave a slit-shaped space in the epiphysis. In such a case this might explain why one gets an increased density of the epiphysis before the outer contours are altered.

The fragmentation stage is a reparation stage where the dense parts in the roentgen picture correspond histologically to necroses and the lighter areas to an ingrowing reparative tissue and a newly formed bone that is not at all, or not yet fully, calcified.

The roentgenological reparation stage is, also histologically, a reparation stage, and differs only in regard to the degree of the changes from the fragmentation stage.

The histological examinations give no reason for any definite conclusions in regard to the definite stage, but, from the changes in the fragmentation and reparation stages, one might perhaps conclude that the definite stage constitutes a return to normal bone in the epiphysis.

Part. II. Arthrographic Investigations

The arthrographic investigations have been carried out on patients who have been admitted to the Orthopedic Clinic of the Karolinska Institutet. The material comprises 44 arthrographs of coxa-plana hips, and 26 arthrographs of normal hips (the healthy side of unilateral cases). The shape of the head of the joint has been determined with the aid of an index called the caput-index which is the relation between the height and half of the greatest breadth in the arthrographic picture. This index, in regard to normal hips, has proved to lie round about 1, indicating that the caput femoris in children is spherical even when presented arthrographically.

The cases have been classified in accordance with the same roentgenological

principles as described previously. The result of the investigation can be summarized in the following way.

The initial stage. The head of the joint here retains its spherical shape in the majority of cases.

The fragmentation stage manifests very insignificant deviations from the normal shape of the head, despite that the bone epiphysis is often markedly shrunken and fragmented. In the arthrographic picture this seems to be explained by the fact that the joint cartilage increases in thickness in the places where the epiphysis has shrunk.

The reparation stage. It is only in this stage that a clear deformation of the head of the joint is manifest, and the latter begins to assume the same shape that the epiphysis has.

The definite stage is characterized by the circumstance that the joint cartilage regains its normal thickness. No further deformation of the head of the joint seems to have taken place during this stage.

The increased head-socket distance. This early symptom is discussed and is considered, to judge from the arthrographic findings, to be due to the soft parts in the fossa acetabuli and the ligamentum teres being the seat of an inflammatory swelling which displaces the head laterally.

Part. III. Aspects on the Treatment

The results of conservative treatment, to judge from information given in the literature, seem to be somewhat uncertain, and conservative treatment will not accelerate the curing of the disease.

Operative treatment might shorten the duration of the disease and bring about a cure with less deformation of the head provided that it is resorted to at the very earliest stage of the disease. It should consist of establishing a communication between the living bone in the collum femoris and the necrotic epiphysis in order to bring about a rapid ingrowth of reparative tissue.

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