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CONGENITAL COXA VARA AND PERTHES' DISEASE

By

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Congenital coxa vara is a rare condition of unknown etiology. In a recent survey from two large clinics in North America, the present author reported that only 15 cases were registered over a period of 20 years (1).

Of these 15 cases, 7 were boys and 8 girls. The right side was affected in 9, the left in 2, and both sides in 4.

The etiology, as already mentioned, is unknown, but in the above survey it is suggested that probably the most acceptable theory is a *disturbance of the vascular supply of the femoral neck during embryonic life. J. F. Brailsford* gives as his personal view the theory that it is caused by a *dysostosis,* —a developmental defect in the zone of ossification (2).

The diagnosis must in most cases be based on the radiographic examination. Clinically, there is a history that the child has limped sice it began to walk. The limp is similar to that seen in congenital luxation of the hip, but clinical examination shows that there is no telescoping and the head is in the acetabulum.

In the radiographs the most important criteria for the differential diagnosis are the appearance and direction of the epiphyseal line. In congenital coxa vara the line is vertical and is often divided in its lower part to form an inverted Y (1).

Halfdan Sundt, in his paper on the "Malum Coxae Calvé-Legg-Perthes", describes the same criteria when he writes of "the vertical, broad and often fork like divided epiphyseal

line", and later on he writes of the "lower corner of the neck being rounded off" (3).

This appearance is very different from that seen in rachitic coxa vara, where the epiphyseal line is oblique and has an edge-like prominence of the lower corner of the femoral-like neck.

Perthes' disease, or coxa plana, occurs rather frequently in boys, and its pathological and clinical picture is much better known. Here I will concentrate on the changes seen in this condition in the neck of the femur, distal to the epiphyseal line.

According to *Sundt*'s description the changes in the neck in Perthes disease are found *in the upper corner.* There are osteoporosis and a number of osteochondritic foci surrounded by a sclerotic zone.

The appearance of the neck changes; it becomes broad and clumsy, but the *direction* of the epiphyseal line does not change—especially it does not become more vertical; rather, it appears to go the other way and, in fact, the result is a slight displacement of the head in a *valgus* position. This is due to the broad and deformed shape of the neck.

In *Sundt*'s paper a wide variety of changes is described, and of particular interest for this article is his statement that Perthes' disease has been observed to develop in cases of pre-existing congenital coxa vara. It has not been specified that the osteochondritic changes tend to appear in the *neck* in pre-disposed cases of congenital coxa vara. In only one case were there changes compatible with Perthes' disease in both head and neck. (3).

After this introduction, a case is presented which has been observed in Farsund Hospital for 9 years, from 1940 to 1949, when the final treatment was begun.

L.B. was aged 7 years when he was admitted to hospital on 2.26.40 with a history of limping on the R. lower limb since he began to walk. The right leg appeared to be shorter than the left one. He never had any pain or discomfort, and had therefore never been examined by a

doctor until now, when he had had an accident on a sledging hill. He received a blow on his right hip and a scalp wound, which was sutured. He was then sent to hospital for radiography of his hip.

Clinically, there was marked shortening of the R. leg with a high trochanteric prominence. There was no pain on movement, but there was limitation of internal rotation, abduction and flexion. Otherwise, nothing of importance was found.

The radiographs of 2.26.40 are shown in fig. 1. There is a R. coxa vara with osteochondritic changes in the upper corner of the neck; the left hip appears to be normal.

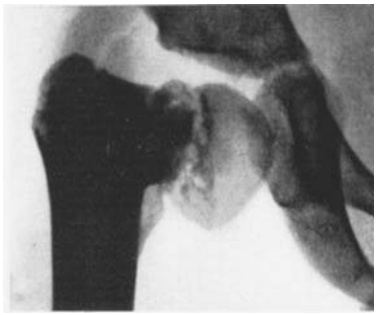

Fig. 1.

The epiphyseal line is vertical on the affected side; it is wide, and the lower corner of the neck is rounded-off.

The diagnosis made at this time was that of an atypical case of Perthes' disease, drill holes were made through the neck into the head, apparently without any beneficial effect, as can be seen from the following radiographs. These show that the coxa vara persisted, or rather progressed, and led to the development of a deformed head, which gradually became displaced distally in relation to the neck, with a wide and open epiphyseal line. Osteochondritic changes are seen in the upper corner of the neck; these gradually disappeared, but in its course the condition followed the typical pattern of Perthes' disease figs. 2 and 3).

The patient returned for periodical examination and radiography until March 1943, when, due to the circumstances in Norway at that time, he disappeared.

He was not seen again until February 1949, when he was re-admitted to the Hospital, suffering from drug poisoning. On this occassion he was re-examined for his hip condition with the following report:

"The patient is a well-developed boy aged 15 years, with an obvious

limp on the right side, where the trochanteric prominence is high. There is a positive Trendelenburg sign with shortening and atrophy of the R. lower limb and marked limitation of abduction, internal rotation and flexion at the hip."

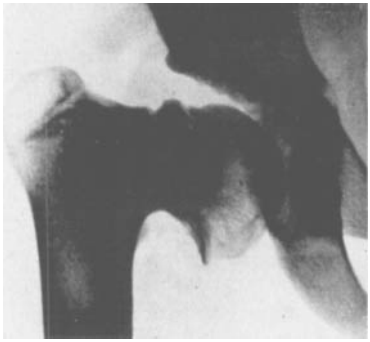

Fig. 2.

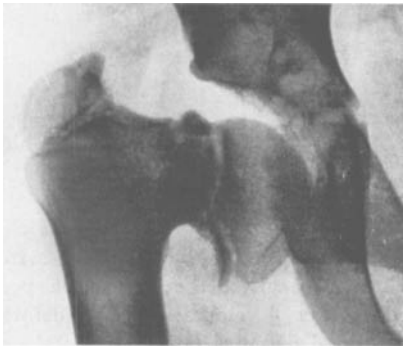

Fig. 3.

Radiopraphy on 2.12.49 showed a wide space at the site of the epiphyseal line, with a high position of the neck and major trochanter. The head was deformed, with a protruding lower lip, but the articular surface and space were well preserved. The changes seen earlier in the upper corner of the neck had completely disappeared (fig. 4).

The patient was treated in bed with moderate traction on the R. lower limb. Repeated radiography during this period showed the develop-

ment of a pseudarthrosis at the site of the epiphyseal line, where there was definite movement between the head and neck.

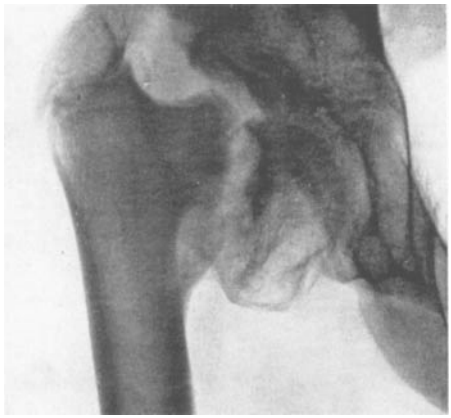

Fig. 4.

From the patient's story, the clinical examination and the radiographic appearance, it was concluded that the original diagnosis only covered one of the conditions present. The diagnosis was revised to one of congenital coxa vara with secondary changes in the neck, identical with those seen in Perthes' disease.

The unilateral involvement and the radiographic appearance both fail to conform to the picture of rachitic coxa vara.

The treatment of congenital coxa vara has been discussed, and it has been said that subtrochanteric osteotomy, with abduction of the distal fragment, is the procedure of choice. By this operation one can obtain a more oblique direction of the epiphyseal line with a tendency to "impaction of the fragments", which should promote healing and prevent pseudarthrosis. This treatment often promotes healing in children, but in the present case there was already a well-established pseudarthrosis, and we could not expect any beneficial effect from it alone.

An operation was therefore performed in two stages. At operation we found the pathology we expected.

The following operation was performed under general anaesthesia, by the author, on 23.3.49.

1. *Resection of the pseudarthrosis—Bone transplantation—Screwing.*

Using a Smith-Petersen incision, the R. hip was exposed and the joint was opened anteriorly. As seen on the radiographs there was a pseudarthrosis with articular surfaces covered by smooth cartilage on both ends. The pseudarthrosis was open on the proximal-anterior surface, but covered on the distal-posterior surface by strong ligamentous bands. On movement the head glided on the rounded surface of the neck.

The pseudarthrosis was resected, and bone chips from the ilium were plugged into the open space between the denuded ends of the head and neck. The head was then fixed by a vitallium screw through the long lower edge of the neck into its central part. The fixation appeared to be firm when tested by movement at the joint. The wound was closed, and light traction was again applied.

5 weeks after the first operation, there was evidence of union at the site of the resection and the position was unchanged (fig. 5).

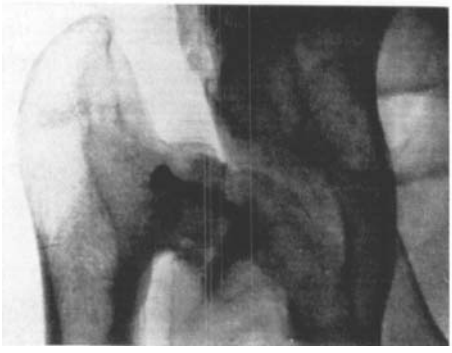

Fig. 5.

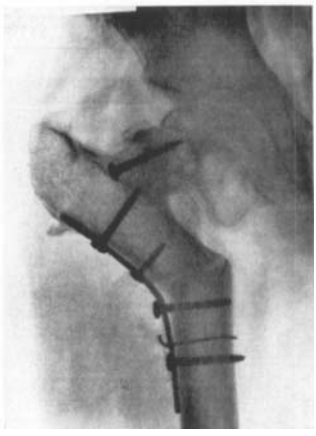

Fig. 6.

At a second operation the following procedure was completed:

2. *Subtrochanteric Osteotomy-Osteofixation-Hip spica.*

A lateral (Badgley) incision was made, starting 1 inch behind the anterior superior spine and curving down behind the trochanteric prominence and along the posterior aspect of the femur, which was exposed just below the lesser trochanter. At this site an osteotomy was per-

formed with removal of a triangular piece of bone with its base laterally. By this procedure the distal fragment was opposed to the proximal in marked abduction; an angulated Soyland's plate was then fixed to the

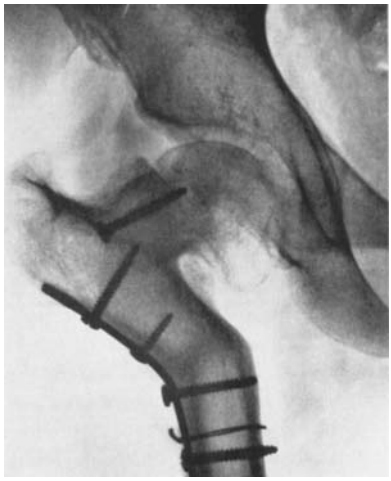

Fig. 7 a.

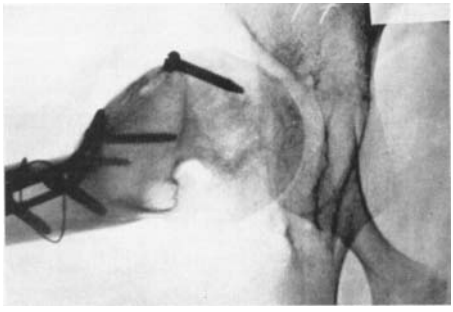

Fig. 7 b.

fragments which were in close contact with each other. After closure of the wound a half hip spica was applied.

There were no complications after either operation and the cast was bivalved after 8 weeks, when there was radiographic evidence of union (fig. 6).

During the following 4 weeks the patient was doing quadriceps-exercises, and flexion of the knee was allowed. After this period the pro-

gramme was intensified, and 13 weeks after the second stage he was up on crutches with a good range of movement at the hip.

4 months after the second stage operation he was re-examined. At this time he was ambulatory. He was still using a stick and walking with a slight limp. He did not notice any pathological movements, and spontaneously remarked that its absence felt very peculiar, as he had been used to it for so many years. There was no pain or feeling of instability in the R. lower limb.

Clinical examination gave the following report:

Trendelenburg's sign: Neg.

There is ½ inch real and apparent shortening of the R. lower limb.

There is 2 inches atrophy of the leg and 1 inch of the R. calf.

Range of R. hip movements: No contracture. Flexion 135° (155°). Abduction 20° (30°). Adduction 30° (25°). Internal rotation 20° (25°). External rotation 30° (30°).

Thus, only the slight limitation of flexion and abduction are of practical importance.

Radiography on 9.1.49 (fig 7) showed sound union at both sites, with a good static position. No evidence of secondary changes in the head as yet, but for this reason the patient will have to be kept under close observation and radiographic control for a long period.

The case is considered to be of considerable diagnostic and therapeutic interest and it is therefore presented at this early post-operative stage.

SUMMARY

A case of congenital coxa vara complicated by osteochondritic changes in the neck identical with those occurring in Perthes' disease is reported with a complete radiographic follow-up. The case has been followed for 8 years; during this time a pseudarthrosis developed in the neck and was treated by a two-stage operation. First, the pseudarthrosis was resected, bone chips were transplanted from the ilium, and the head and neck were fixed with a screw. Later, a subtrochanteric osteotomy was performed, with a resulting good position of the femoral neck.

RESUME

Un cas de coxa congénital compliqué d'altérations ostéochondritiques dans le col fémoral, identiques à celles apparais-

sant dans la maladie de Perthes est rapporté avec une série complète de radiographies. Le cas a été suivi pendant 8 ans; pendant cette période une pseudarthrose s'est dévelopée dans le col et a été traitée par une opération en deux phases. Dans la première, résection de la pseudarthrose; des fragments de l'os iliaque ont été transplantés la tête et le col ont été fixés au moyen d'une vis. Plus tard une ostéotomie subtrochantérique a été pratiquée et il en est résultê une bonne position du col fémoral.

ZUSAMMENFASSUNG

Ein Fall von angeborener coxa vara mit komplizierenden osteochondritischen Veränderungen in collum femoris, identisch mit den Veränderungen bei Perthes'scher Krankheit, wird mitgeteilt zugleich mit einer kompletten röntgenologischen Verfolgung des Falles. Die Beobachtung wurde 8 Jahre hindurch fortgesetzt. Während dieser Zeit entwickelte sich eine Pseudartthrose im Femurhals, die mit Operation in Zwei Sitzungen behandelt wurde. Zuerts wurde die Pseudarthrose reseziert und gleichzeitig Knochenstückchen transplantiert und eine Schraubenfixation vorgenommen. Später wurde eine subtrochantäre Osteotomie ausgeführt, die in einer guten Position des Femurhalses resultierte.

LITERATURE

1. The Journal of Bone and Joint Surgery. Vol. 31-A, No. 1, p. 115.
2. The Radiology of Bones and Joints. James F. Brailsford. p. 257.
3. Examinations of Malum Coxae Calve-Legg-Perthes. Dr. Halfdan Sundt. P. 86 and 159.