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THE STAGES OF DEVELOPMENT OF THE CARTILAGINOUS FOCI IN DYSCHONDROPLASIA (OLLIER'S DISEASE)

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Radiographs of the extremities of a boy with Ollier's disease taken at intervals of two and a half years were published by the present author in 1947. The observations made in this case, and a thorough study of the literature on the subject, led to the conclusion that small and middle-sized cartilaginous foci occurring in the long bones in dyschondroplasia develop in the manner shown schematically in Figure 1.

Ollier explained the histogenesis of the cartilaginous foci in the disease bearing his name in the following way: "The disease is characterized by the irregularity and delay of ossification of the intermediary cartilages. The cartilage intended for the growth in length of the bones does not pass through the normal ossification process. It preserves its structure and persists in the form of more or less regular cartilaginous masses which often need a very long time for transformation into bone tissue." The correctness of this view has generally not been doubted by other authors. However, the phenomenon illustrated in Figure 1 is difficult to explain on the basis of the common conception that the epiphyseal plate normally grows in diameter by apposition of cells from a perichondrium at the periphery.

In 1949 *A. Langenskiöld & Edgren* reported results of experiments in which limited portions of epiphyseal plates in rabbits had been injured by heavy doses of roentgen rays. The experiments supported the view that in normal epiphyseal cartilage, interstitial growth of the layer called "the layer of the reserve cells" causes an expansion of this layer and a successive displacement of its cells to the periphery in relation to the bony epiphysis and the cartilage cell columns. Ac-

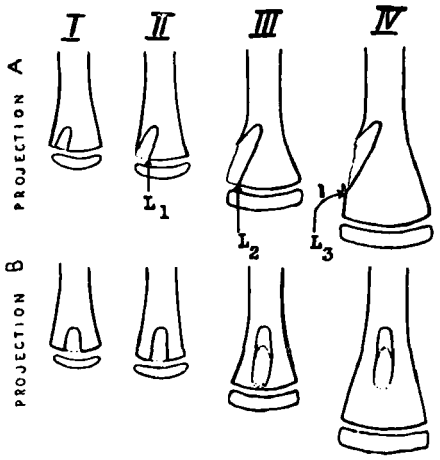


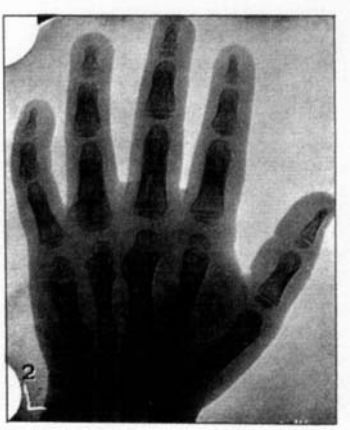
Figure 1. Picture published by A. Langenskiöld in 1947. Ollier's disease. Schematic drawing of the development of a metaphyseal focus into a diaphyseal one with growth of the bone. Stages I–IV in two projections perpendicular to one another.

cording to several classical authors (*Ranvier, Carey, Policard*) a continuous supply of cells takes place at the periphery from the cartilage for the growth in length of the osteogenic layer of the diaphyseal periosteum. The view that the epiphyseal plate grows in diameter by interstitial growth of the layer of the reserve cells and that the ossification groove (encoche d'ossification) derives its cells from the cartilage tissue would be fully in accordance with the phenomenon illustrated in Figure 1.

The question how the epiphyseal plate grows in diameter is still being discussed in recent literature (*Rigal, Solomon*). Recent investigations concerning the fate of radioactive sulphate (^{35}S-sulphate) bound to the ground substance of epiphyseal cartilage carried out by *Langenskiöld, Rytömaa & Videman* strongly support the view that the epiphyseal plate grows in diameter by interstitial expansion of the layer of the reserve cells.

Dyschondroplasia is a rare disease and radiographs illustrating the development of the cartilaginous foci in this condition during skeletal growth are very seldom seen. Figures 2–4 show radiographs of the hands of a child with dyschondroplasia. It was possible to follow the development of typical longitudinal cartilaginous foci in the bones of the hands from the age of three years to the age of nine. Figures 5 and 6 are contour drawings of the radiographs seen in Figures 2–4.

Figure 2. Radiograph of the left hand of a child with dyschondroplasia. Age three years.



ure 3. Radiographs of the right hand the child who's left hand is seen in ure 2. Age three years.

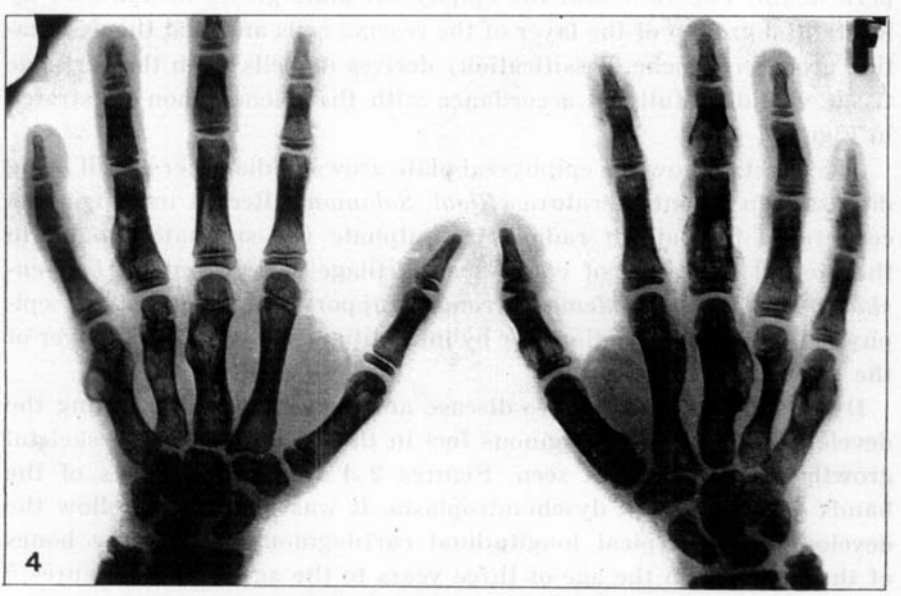


Figure 4. Radiograph of the same hands as seen in Figures 2 and 3. Age nine years.

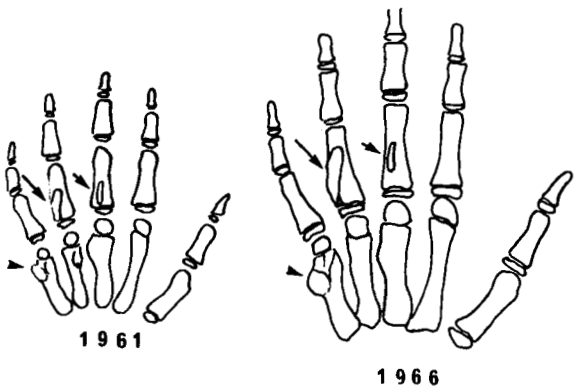


Figure 5. Contour drawings of the left hand as seen in Figure 2 and Figure 4. Arrows indicate three different foci as they appeared in 1961 and 1966.

The radiographic findings in this case confirm the former observations illustrated schematically in Figure 1.

In 1966 *Solomon* stated that serial radiographs of four patients with dyschondroplasia observed by him did not confirm the findings of the present author published in 1947. In the case illustrated by *Solomon,* there was a large cartilaginous focus in the lower end of the femur and severe disturbance of growth. It is obvious that the development of a metaphyseal focus into a diaphyseal one in dyschondroplasia can take place only when there are possibilities for the continued growth of the bone in question. This means that the phenomenon oc-

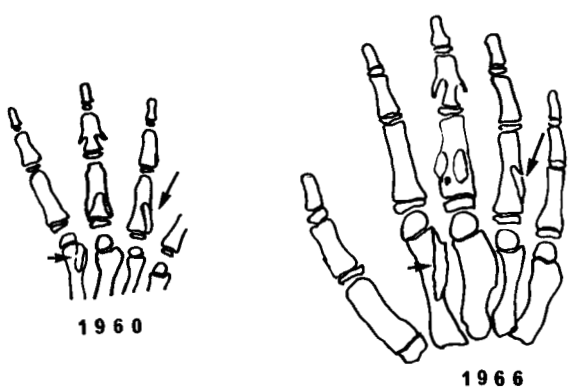


Figure 6. Contour drawings of the right hand as seen in Figure 2 and Figure 4. Arrows indicate two different foci as they appeared in 1960 and 1966. Compare with Figure 1.

Figure 7. Picture published by Speiser in 1925. The microphotograph shows a section of the peripheral part of the basis of the second phalanx of a big toe of a young child with enchondromatosis. The epiphyseal cartilage (Ep) is seen in the left lower corner. The periosteum (P) is seen to the right. The arrow indicates the limit between normal epiphyseal cartilage and a chondroma. This limit continues into the metaphysis as a borderline between enchondroma tissue and metaphyseal bone.

curs especially when a focus is small or of middle size. However, even the displacement of the base of a small focus from the epiphyseal plate to the surface of the bone is difficult to explain if it is assumed that the plate grows in diameter by apposition from the periphery.

In 1925, *Speiser* was able to carry out a thorough microscopic study of the entire skeleton of a child who died from anemia caused by enchondromatosis. Figure 7 shows one of *Speiser*'s illustrations in which a part of the epiphyseal plate of the second phalanx of a big toe and an enchondroma occupying the peripheral part of the plate is seen. In 1949, the present author stated that the "oblique lines" in the metaphyses in Ollier's disease are traces left behind by the limits

between normal and pathological cartilage in the epiphyseal plate. In Figure 7 the arrow indicates such a limit between normal cartilage and chondroma tissue in the epiphyseal plate continuing into the metaphysis as a borderline between the tumor tissue and normal metaphyseal bone.

SUMMARY

The characteristic manner of development of cartilaginous foci in dyschondroplasia described by the author in 1947 (Figure 1) could be confirmed in a child with typical multiple cartilaginous foci in the hands. Recent research concerning the fate of radioactive sulphate (^{35}S-sulphate) in epiphyseal cartilage (*Langenskiöld, Rytömaa & Videman)* has brougt new support to the author's conception of the pathogenesis of dyschondroplasia.

RESUME

La manière caractéristique du développement de foyers cartilagineux dans la dyschondroplasie décrite par l'auteur en 1947 (Figure 1) a été confirmée chez un enfant des foyers cartilagineux multiples typiques dans les mains. Des recherches récentes concernant l'accumulation du sulfate radioactif (^{35}S-sulfate) dans le cartilage épiphysaire (*Langenskiöld, Rytömaa & Videman*) a apporté un nouveau point d'appui à la conception de l'autheur sur la pathogénèse de la dyschondroplasie.

ZUSAMMENFASSUNG

Die characteristische Entwicklung von den bei Dyschondroplasie auftretenden Knorpelherden, die vom Verfasser in 1947 (Figure 1) beschrieben wurde, konnte bei einem Kind mit typischen multiplen Knorpelherden in den Händen bestätigt werden. Neue Untersuchungen über das Verhalten von radioaktivem Sulphat (^{35}S-Sulphat) im Epiphysenknorpel (*Langenskiöld, Rytömaa & Videman*) stützen die Auffassung des Verfassers von der Pathogenese der Krankheit.

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