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THE DEVELOPMENT OF MULTIPLE CARTILAGINOUS EXOSTOSES

By

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In two articles published in 1947 and 1949 (*Langenskiöld & Edgren*) the present author discussed the pathogenesis of multiple cartilaginous exostoses on the basis of clinical radiological studies and of experiments concerning bone growth after X-ray injury to localized portions of epiphyseal plates.

It seems clear that the pathogenesis of cartilaginous exostoses cannot be understood without knowledge of the course of the normal histogenesis in the transition zone beween the epiphyseal plate and the osteogenic layer of the diaphyseal periosteum. This zone has been called the ossification groove ("encoche d'ossification") of Ranvier. The origin of the cells of the ossification groove and the question of how the epiphyseal cartilage grows in diameter are still subject to discussion in the literature (*Rigal; Solomon; Langenskiöld, Rytömaa & Videman*). Furthermore, as very few authors interested in the pathogenesis of cartilaginous exostoses have had an opportunity to follow the development of exostoses in their early stages by repeated radiography, the pathogenesis of this hereditary disease is still considered obscure.

On the basis of observations reported in 1947 and 1949 the present author assumed that cartilaginous exostoses may arise in the following way: limited portions of the undifferentiated cell layer, which is displaced by growth from the interior of the epiphyseal cartilage to the surface of the bone, do not as would normally be the case, give rise to cells of an osteoblastic nature. These pathological portions of tissue retain their chondrogenic properties giving rise to a cartilaginous layer on the surface of the bone which forms the outermost zone of the

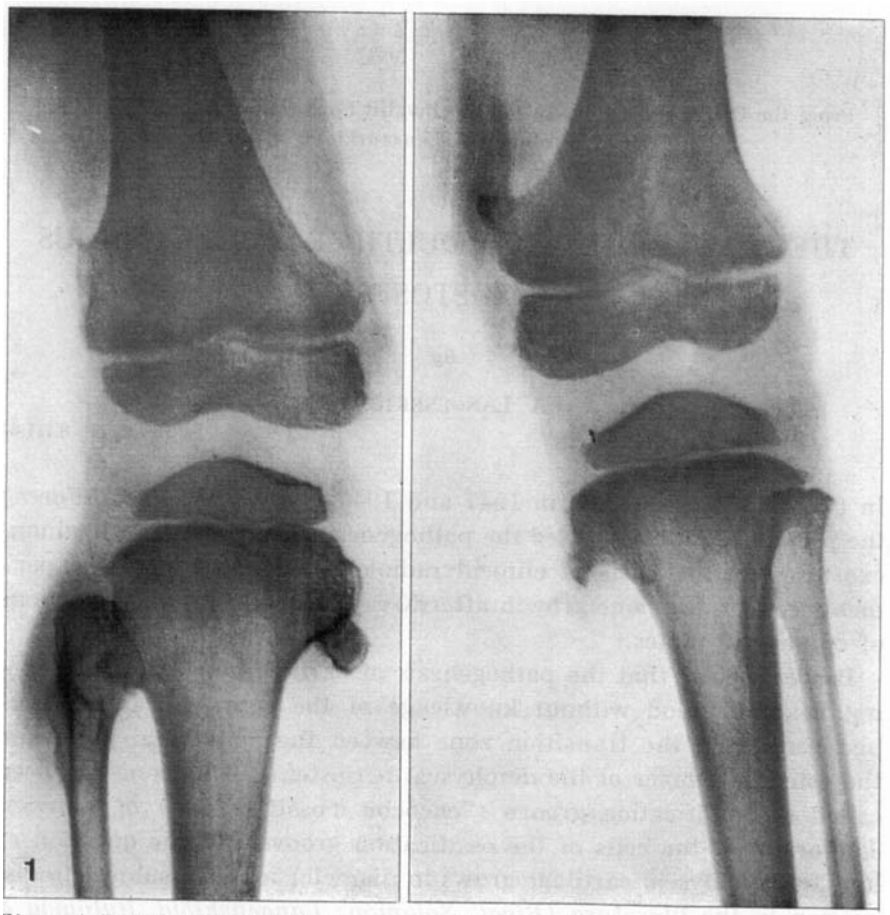

Figure 1. Radiograph of the knees of a child with multiple cartilaginous exostoses. Age four years.

exostosis. Owing to this local disturbance of the differentiation of cells, the formation of an osteogenic layer fails to occur in the corresponding places, as also do the processes which are dependent on such an osteogenic layer, the formation of periosteal bone and tubulation and modelling of bone.

A thorough search in 1946 and 1947 of the literature and of the archives of radiological departments of the university hospitals in Helsinki and Stockholm did not give a possibility to judge, from serial radiographs, the course of the development of cartilaginous exostoses. In 1950, *Lacroix* showed that the place of attachment of a pedunculated exostosis to the bone is displaced, by remodelling, towards the

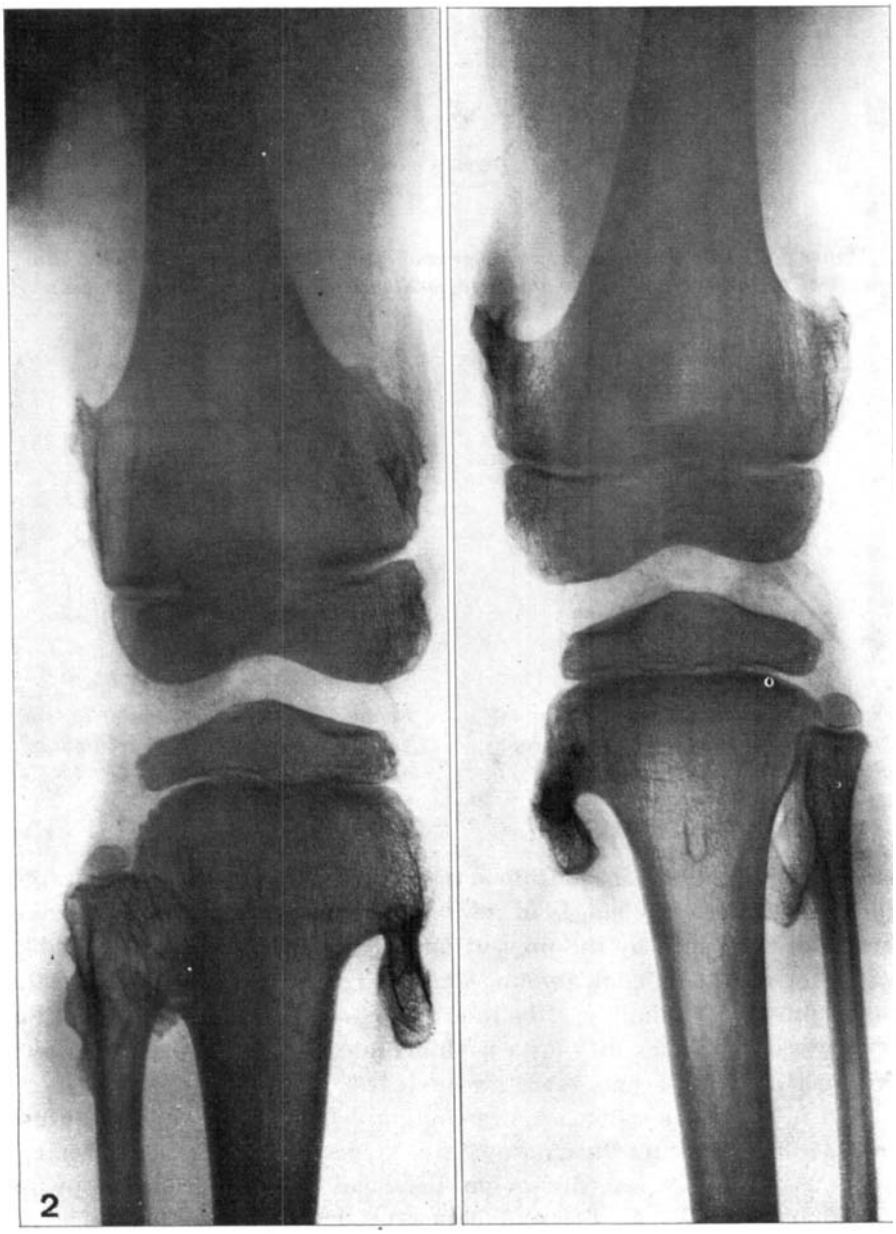

Figure 2. Radiograph of the same knees as seen in Figure 1. Age six years.

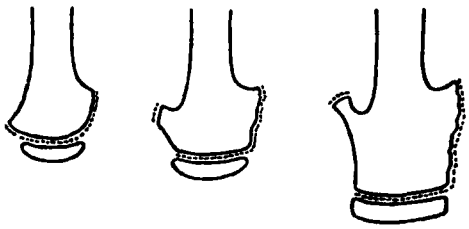

Figure 3. Picture published by A. Langenskiöld in 1947. Schematic drawing illustrating the development of multiple cartilaginous exostoses (three stages).

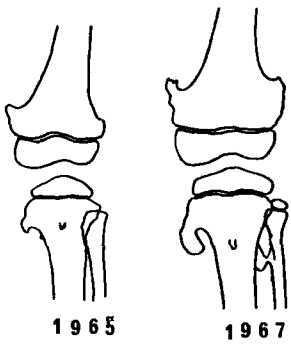

Figure 4. Contour drawings of the left knee region as seen in the radiographs af Figures 1 and 2. To the left 1965, to the right 1967.

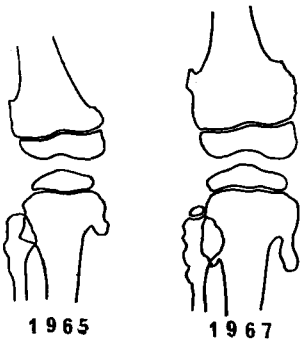

Figure 5. Contour drawings of the right knee region as seen in the radiographs of Figures 1 and 2. To the left 1965, to the right 1967.

end of the bone with its continued growth. However, serial radiographs illustrating the early stages of development of pedunculated exostoses have not been seen by the present author in the literature. The publication of radiographs of a young child (Figures 1 and 2) showing the development of small cartilaginous exostoses situated close to the epiphyseal cartilages into large pedunculated or broadbased exostoses within two years, seems therefore justified.

Figure 3 shows a schematic drawing published by the present author in 1947. The drawing illustrating three stages of development of cartilaginous exostoses was drawn on the basis of radiographs showing different exostoses at different stages of development. The observations made in 1947 are now completed by the radiographs seen in Figures 1 and 2. Figure 4 shows contour drawings of the radiographs of the left knee region in 1965 and 1967 (Figures 1 and 2) and Figure 5, the corresponding drawings of the right knee region. Figure 6 shows

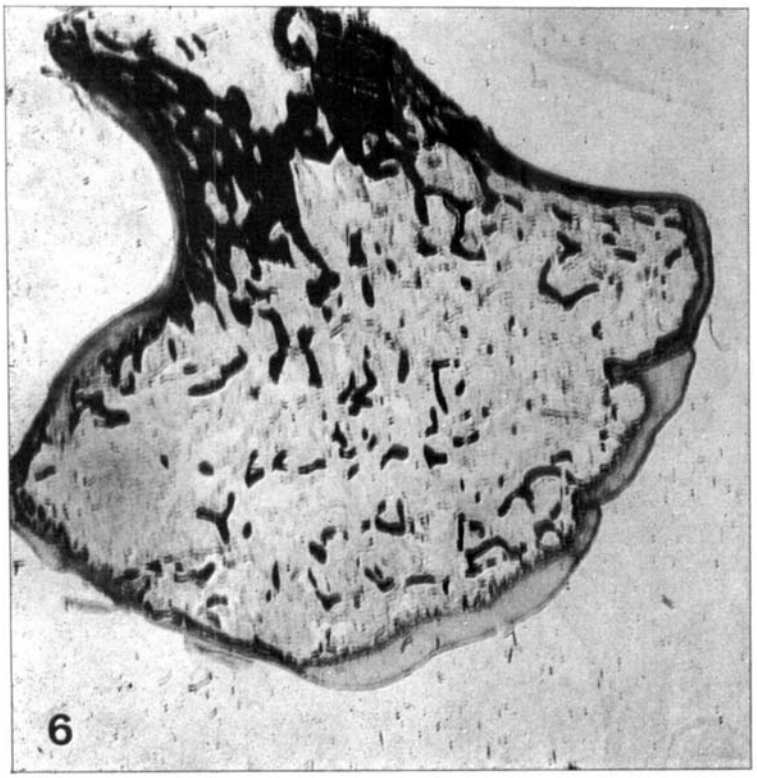

Figure 6. Photomicrograph of a section taken in the frontal plane of the pedunculated exostosis of the left tibia seen in Figure 2. The exostosis was removed soon after the radiograph had been taken. The top of the exostosis (at the bottom of the picture) is covered by a layer of cartilage.

a microscopical section of the top of the pedunculated exostosis of the left tibia as seen in Figure 2.

In the literature there is a microphotograph of a cartilaginous exostosis in an early stage of development, which is probably unique (Figure 7). *Leveuf* (1946) took a biopsy of a cartilaginous existosis of a growing child and included in the same section the adjacent part of the epiphyseal plate and a portion of the bony epiphysis (Figure 7 *A*). In Figure 7 *B* the topography of the cartilaginous layer covering the exostosis in relation to the epiphyseal cartilage and to the ossification groove is clearly seen. This exostosis is in a similar stage of development as the exostosis of the left tibia seen in Figure 1. This would mean that Figure 6 would correspond to a later stage of development of an exostosis similar to that illustrated by Figure 7 *B*.

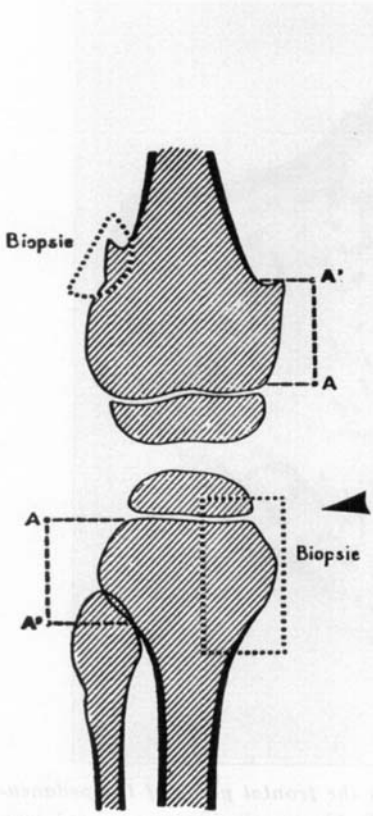

Figure 7 A

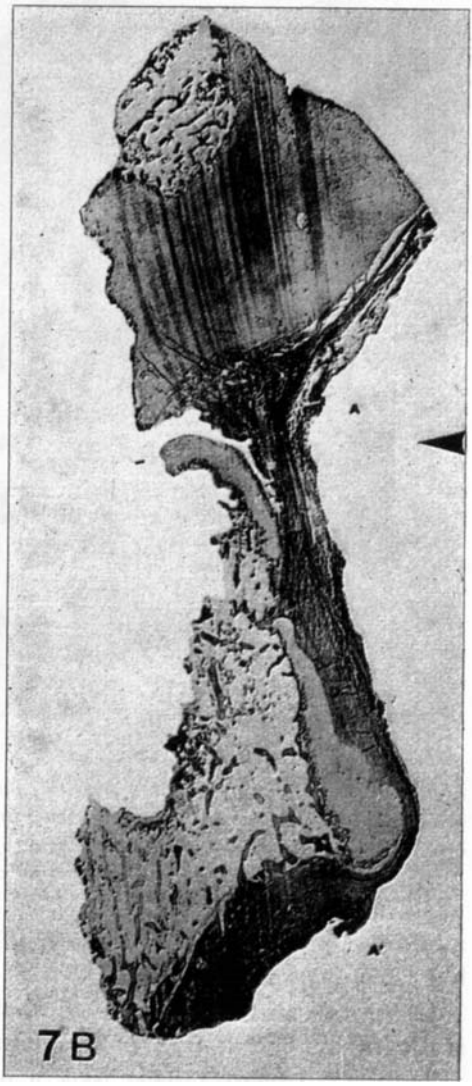

Figure 7. A and B are both pictures published by Leveuf in 1946. A. A contour drawing of the knee region of a child with multiple cartilaginous exostoses aged seven years. For biopsy a section was taken from the tibia in the frontal plane. The section included the exostosis, the peripheral part of the epiphyseal plate and a part of the epiphyseal nucleus. B. Microphotograph of the section described under A. The cartilaginous layer covering the exostosis and the epiphyseal plate merge, as do the perichondrium covering the cartilaginous layer and the tissue of the ossification groove ("encoche d'ossification").

Today the possibilities of differentiation of the different kinds of cells occurring in the skeleton are better known than in 1947. *Holtrop* stated in 1966 that cartilage cells may differentiate into connective tissue (perichondrium or periosteum) and then form bone afterwards. She concluded that to the list of possible precursors of bone cells the cartilage cell should be added. This means that the existence of a layer of undifferentiated connective cells in the epiphyseal plate is not a prerequisite for understanding the conception of the pathogenesis of cartilaginous exostoses held by the present author.

SUMMARY

In a child with multiple cartilaginous exostoses the development of small exostoses in the neighbourhood of the epiphyseal cartilages at the age of four years into large pedunculated or broad-based exostoses at the age of six years could be followed by radiography. The observation is in accordance with findings reported by the author in 1947 and 1949.

RESUME

Chez un enfant ayant de multiples exostoses cartilagineuses, le développement de petites exostoses dans la proximité des cartilages épiphysaires à l'âge de 4 ans en grandes exostoses pédonculaires ou à large base à l'âge de 6 ans a pu être suivi sur des radiographies. L'observation concorde avec les trouvailles rapportées par l'auteur en 1947 et 1949.

ZUSAMMENFASSUNG

Bei einem Kind mit multiplen kartilaginären Exostosen konnte die Entwicklung von kleinen in der Nähe vom Epiphysenknorpel gelegenen Exostosen im Alter von vier Jahren zu grossen gestielten und breitbasigen Exostosen im Alter von sechs Jahren roentgenologisch gefolgt werden.

Die Observation bestätigt vom Verfasser im Jahre 1947 veröffentlichte Befunde.

REFERENCES

Holtrop, M. (1965) The origin of bone cells in endochondral ossification. *Calcified Tissues 1965*, p. 32, Springer, Berlin.

Lacroix, P. (1950) Contribution a l'étude de la maladie exostosante. Sur la migration des exostoses au course de la croissance. *Rev. d'orthop.* **36**, 20.

Langenskiöld, A. (1947) Normal and pathological bone growth in the light of the development of cartilaginous foci in chondrodysplasia. *Acta chir. scand.* **95**, 367.

Langenskiöld, A. & Edgren, W. (1949) Imitation of chondrodysplasia by localized roentgen ray injury.—An experimental study of bone growth. *Acta chir. scand.* **99**, 353.

Langenskiöld, A., Rytömaa, T. & Videman, T. (1967) An autoradiographic study with ^{35}S-sulphate on the growth in diameter of epiphyseal cartilage in rabbits. *Acta orthop. scand.* In press. Supplementum.

Leveuf, J. (1946) Le problème de la croissance de l'os en longeur étudié a la lumière de la maladie exostosique. *Rev. d'orthop.* **32**, 5.

Rigal, W. M. (1962) The use of tritiated thymidine in studies of chondrogenesis. Radioisotopes and bone. p. 197. Blackwell, Oxford.

Solomon, L. (1966) Diametric growth of the epiphyseal plate. *J. Bone Jt Surg.* **48**-B, 170.