FROM MARTINA HANSEN'S HOSPITAL, NORWAY
(HEAD: JOHN K. HALD)

POLYOSTOTIC FIBROUS DYSPLASIA

A review of possible treatment

By

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INTRODUCTION

Polyostotic fibrous dysplasia is a relatively rare disease. Amongst the many patients treated for bone cysts (osteitis fibrosa) at Martina Hansens Hospital we have only recorded one case of polyostotic fibrous displasia in the course of twenty years (in three other cases we established the diagnosis *monostotic* fibrous dysplasia). In medical literature the disease is reported mainly in the form of case histories. Most authors have described one or a few cases (*Arnesen* and *Nitter, Bingold, Sante* and colleagues, *Tiitinen, Ulland, Ønne* and more). Others have collected up to 10–12 cases, to illustrate certain characteristics of the disease (*Hobaek, Valls* and *Schajowicz, Russel* and *Chandler, Belaval* and *Schneider*).

The object of this publication is to deal with the relation of the disease to the group *Osteitis fibrosa* and at the same time to inquire into the various possibilities of treating this condition.

DEFINITION AND TERMINOLOGY

We mean by the expression fibrous dysplasia a definite pathologic-anatomical condition of the skeletal system. It has, however, been recognised at the clinic as designating a definite disease or group of diseases. Originally it was introduced by *Lichtenstein* in 1938 to define the fundamental characteristics, that is, the bone changes, of Albright's syndrome. The previous year *Albright and his colleagues* had described a definite disease—in children—with three groups of symptoms:

(1) Cyst-like changes of the skeletal system.
(2) Outbreaks of cutaneous pigmentation.
(3) Pubertas praecox.

Earlier, in 1926, hyperparathyroidism or *osteitis fibrosa cystica* (v. Recklinghausen) was separated from the large case group of fibrous osteitis and regarded as a disease in itself. Correctly enough Albright declared that two of the three patients whose cases were published and discussed by von Recklinghausen in 1891 and who had been given his name had no genuine hyperparathyroidism but an *osteitis fibrosa disseminata*. Finally there is a third recognised type, osteitis fibrosa localisata (benign giant cell tumours).

Even if comprehension is made easier by such distinctions the many fibrous osteitis terms are still confusing. The expression osteitis fibrosa ought to disappear from clinical terminology. It may be reserved, as a descriptive term, for a certain histological reaction of the skeletal system, often independent of the cause producing the disease (Hobaek).

If we take the disease types which are known and mentioned above we then have the following:

1. Hyperparathyroidism (Osteitis fibrosa cystica generalisata or von Recklinghausen's disease).
2. Benign giant cell tumours (Osteitis fibrosa localisata).
3. Albright's syndrome (Osteitis fibrosa disseminata).

Ad 3. When the disease was first recognised it was shown that changes in the skeleton could be found completely identical with those Albright had described, but *without* pubertas praecox and/or without cutaneous pigmentation. These were the changes which Lichtenstein suggested should be called polyostotic fibrous dysplasia. Today we disinguish clinically between the following sub-divisions:

(a) Albright's syndrome (skeletal changes—cutaneous pigmentation and/or pubertas praecox).
(b) Polyostotic fibrous dysplasia (skeletal changes *without* pigmentation and pubertas praecox).
(c) Monostotic fibrous dysplasia (as b, but only with *one* localisation or localised to a single part of the skeleton).

It is not certainly known to what extent there is any principal difference between monostotic and polyostotic fibrous dysplasia and between these and Albright's syndrome (*Valls* and *Schajowicz, Russel* and *Chandler*).

Monostotic fibrous dysplasia is relatively frequent. It is often discovered by chance, for generally it does not reveal symptoms. On X-raying the lungs it is not rare to find it as a limited translucent area on a rib (*Schlumberger*).

Polyostotic fibrous dysplasia occurs most often in girls, tending to set in before puberty. It is above all localised in the long bones, especially on the lower extremities (femur and tibia). The lesion often leads to fractures of these skeletal parts and to deformities. Only the metaphysis and the diaphysis are attacked first of all. The epiphysis is usually not exposed to the pathological changes but *may* occasionally be attacked secondarily. Strikingly, often a unilateral distribution of the skeletal changes is found.

Etiology.

Neither the cause of this disease or of the true Albright syndrome is known. Albright thought that when the disease was localised to several organic systems the origin might be sought in the central nervous system, possibly in the hypothalamus. Others assert that it must be connected with the function of the endocrine glands—a possibe impairment of these—or that it was due to a congenital anomaly of the mesenchyma.

The symptoms with the histological and roentgenological changes are described in detail in earlier publications (*Belaval* and *Schneider, Hobaek, Russell* and *Chandler, Valls* and *Schajowicz, Ønne* and more).

Differential diagnoses.

The following differential diagnoses will be briefly mentioned: Hyperparathyroidism, nevrofibromatosis (v. Recklinghausen), Paget's disease, Ollier's dyschondroplasia, Hand-Schuller-Christian's disease and other lipoid-granulomatoses, osteogenesis imperfecta, multiple myelomas and sarcoidosis.

Treatment.

No causal therapy is known for polyostotic fibrous dysplasia. Medicament treatment and particularly hormone treatment have been attempted, without any certain effect. Because the disease often strikes girls before puberty and has a certain tendency to "burn out" at the onset of puberty, it has been thought that hormone treatment might be indicated. Attempts have also been made to alter the normal relationship between phosphorus and calcium in the blood by administering

aluminium-acetate pr. os. This can have a good effect in various forms of hyperparathyroidism, but is more doubtful in polyostotic fibrous dysplasia (*Helfet*).

Then we have surgical treatment. Attempts have been made to scrape out the pathological tissue of the bone cysts and fill them with autogenous bone transplant in order to increase the stability of the bones. Some reports exist from recent years on more or less successful results of such interventions (*Behrend*). *Adams and colleagues* have operated on three patients by transplanting solid cortical lamellae from the tibia into long bones with extensive cystic changes. The result was more solid bone and thicker cortex but *never completely normal bone structure. Russell* and *Chandler* report that small and limited foci should be scraped clean and filled with bone but that recurrence must be expected. Ønne describes one case in 1953 with an averagely successful result after scraping and packing with bone chips.

We estimate generally that orthopedic interventions are indicated in the following cases:

1. When spontaneous or pathologic fractures occur in the area of the bone cysts. The fractures, however, tend to heal no worse than ordinary fractures. One's aim is often achieved, therefore, with a normal closed reduction. But it may happen that pain persists at the fracture site, and open reduction is thus indicated rather more often than otherwise in these cases with simultaneous scraping of the cysts and bone transplantation.
2. Deformation of the long bones, particularly of the trochanter and neck of femur, either in consequence of repeated fracture or without such fractures being demonstrated. Then the treatment is osteotomy, possibly combined with scraping and bone transplantation.
3. Pain corresponding with the localisation of the bone cysts. Scraping and then packing with bone chips *may* make the pain disappear.

In all cases the result is dependent on what success is obtained with the transplanted bone masses. Some Scandinavian orthopedic surgeons hold the view that recurrence happens easily after local scraping and bone transplantation (Hirsch). We have *not seen in the literature one single case with clinically and roentgenologically complete normality of bone structure in a long bone after scraping and packing with bone chips.* Since this was achieved in our patient—we used homologous cancellous bone for packing—we think that a survey of the case may be interesting.

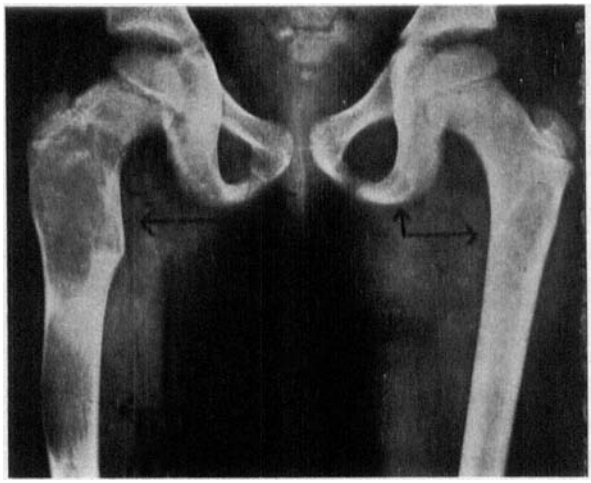


Fig. 1.
Cystic translucent areas in the right trochanter, in the right femur, below the trochanter on the left side, in the os pubis and the os ischii.

CASE HISTORY

A girl aged 8 (T. L. M.) was admitted to Martina Hansens Hospital on 24/4/53. Growth and development were normal as an infant, apart from the fact that the child had difficulty in drinking milk. From 1948 she developed more and more into what is called a difficult child and was treated for this by child psychiatrists. Otherwise there was no special disease in childhood.

On the 13/3/53 she fell and injured her right hip, began to limp and complained about this pain. X-rays in the middle of April, 1953, showed large cysts in the region of the right trochanter and a fracture in a good position. On admittance to the department an obvious muscle atrophy was found in the right lower extremity, but otherwise conditions were normal at an ordinary clinical examination. There were no deformities, either of the upper or the lower extremities, no clinical signs of fracture and good mobility of the various joints of the extremities.

An X-ray of the pelvis and both femurs (Fig. 1) showed a severe cystic area of the whole trochanter mass on the right side, about 5 cm down the thigh bone. The trochanter is broader than on the left side. A fracture line runs along the base of the collum, with slight varus position of the collum in relation to the femoral axis. 5 cm below the point of the trochanter there is a small infraction with an abrupt bend

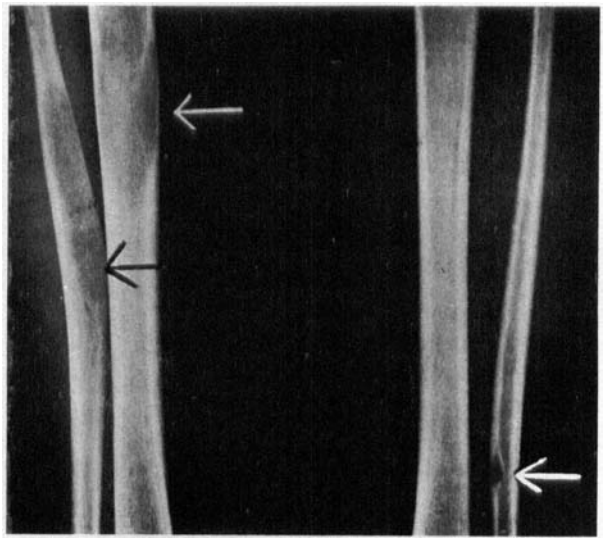


Fig. 2.
Cystic translucent areas in the tibia and fibula.

of the femur so that in all a bayonet position is suggested. Further down the right femur a spool-shaped *formation* of the bone with a central translucent area was found. The cortex medially and laterally is thin. Similar and lesser cystic formations were seen in the right ramus inferior ossis pubis, in the left tuber ischii and immediately below the left trochanter.

X-rays of both knees and calves showed corresponding changes; cystic *formations* in the bones of varying size (see Fig. 2).

X-rays of both hands show a translucent area in the 5th metacarpal on the left side (see Fig. 3).

After X-raying the remainder of the cranium and the skeleton including the spinal column also, no other localisations were found. S. R.: 16 to 22 mm. Repeated tests of calcium, inorganic phosphorus and phosphatase in the blood and urine showed normal values.

We concluded that a polyostotic fibrous dysplasia was present here, but to obtain a clear diagnosis we carried out on 21/5/53 an explorative osteotomy. We removed a section of tumour tissue from just below the right trochanter. Histological diagnosis: typical picture of a polyostotic fibrous dysplasia.

At first the cyst in the region of the right trochanter seemed to be too large to be attacked by radical surgery, scraping and packing with

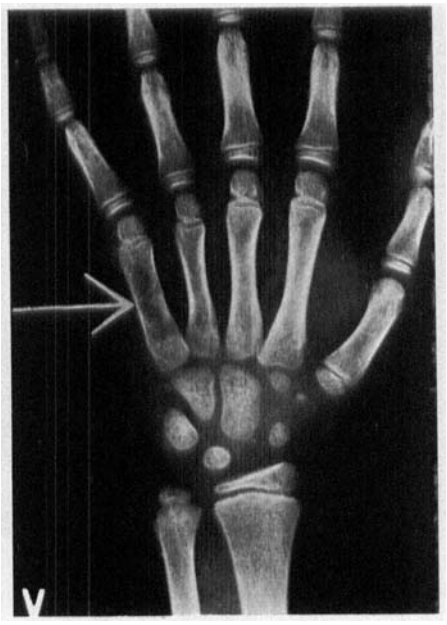


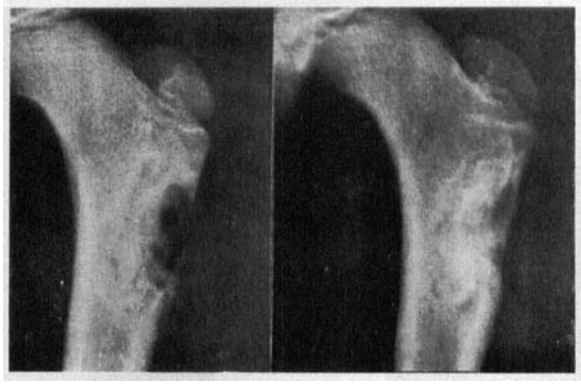
Fig. 3.
Cystic translucent areas in metacarpal 5 on the left hand.

bone-chips, all the more so because the fracture seemed to unite satisfactorily and the position was good. But we had to anticipate the arisal of new fractures at the same site or at any of the other sites with cystic formations in the skeletal system. It seemed to us that we would be justified in undertaking a "test operation" on one of her "silent" localizations, since if the operation succeeded at such a site we had better grounds for what we could offer her later on, surgical intervention possibly being necessary.

On the 26/6/53, therefore, we performed an osteotomy on the upper part of the *left* thigh bone, entering just below the trochanter and scraping out the cyst here with great care. At the end we had thin bleeding cortex on the sides, bone marrow dorsally and cancellous bone upward. After this we packed the hole as well as possible with cancellous bone taken from the *patient's mother* (iliac crest) one hour earlier.

There were no post-operative complications and an X-ray picture some days later showed the operation defect filled with bone (see Fig. 4).

We followed up the patient with regular X-ray checks and clinical



Figs. 4 and 5.
Fig. 4. The cyst in the left femur is scraped clean and filled with bone chips.
Fig. 5. Same as Fig. 4, but three months later.

examinations, and already after three to six months (see Fig. 5 and Fig. 6) initial transformation of the bone masses which we used to fill the hole can be seen.

At the last X-ray check 3½ years after the operation the *bone structure seemed to be completely normal.* (See Fig. 7).

It must be added that regular X-ray checks of the other parts of the skeleton show no change in the cysts as compared with the first X-rays. The fractures in the right trochanter are united, nothing can be seen to remain of these. The cortex of the medial and lateral sides is somewhat thicker than at the first examination but otherwise the defects in the upper end of the femur on the right side are of the same size as before.

In the meantime the patient incurred *no* new fractures.

By this operation we suppose having shown that complete "local healing" of these cysts *can* be achieved by scraping and filling with homologous bone transplant. In those cases where healing has only been "partial" or where—to judge from the literature—recurrence of the cystic development took place, the defects were probably not scraped sufficiently clean of pathologic tissue. We realise very clearly the technical difficulties when large and extensive cysts must be scraped clean and filled with bone transplant (comp. the cyst in the region of the right trochanter in our patient). But we think that it should be possible to "heal" these too, provided that one is efficient, accurate with the scraping and is prepared to devote the time to this which it may require.

Thus we believe that we have a therapy to fall back on if the patient

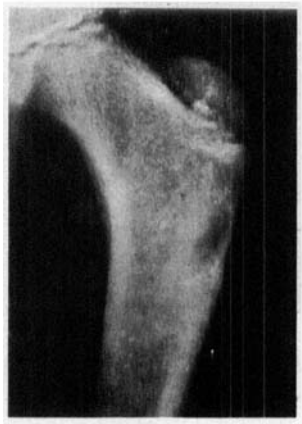


Fig. 6.
Same as Fig. 4, but six months later.

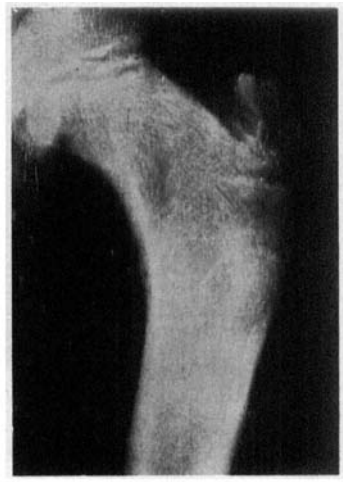


Fig. 7.
The result four years later, compare Fig. 4; almost normal bone structure.

should suffer new fractures, symptoms in the form of pain or increasing deformation of the extremities owing to weight bearing.

SUMMARY

The author discusses the relationship between fibrous dysplasia in the skeletal system and the group osteitis fibrosa and further the mutual relationship between polyostotic fibrous dysplasia, Albright's syndrome and monostotic fibrous dysplasia.

The possibilities of treating this disease are discussed.

Following this a case is reported with a polyostotic fibrous dysplasia (a girl of eight years). A cyst of average size which had not shown symptoms was scraped completely clean and filled with homologous cancellous bone material. Three and a half years later X-rays showed completely normal bone structure at the site operated on.

RESUME

L'auteur discute des rapports entre la dysplasie fibreuse du système osseux et le groupe des ostéites fibreuses, ainsi que le rapport mutuel entre la dysplasie fibreuse polyostotique, le syndrome d'Albright et la dysplasie fibreuse monostotique.

Les possibilités de traitement de cette maladie sont examinés. Un cas de dysplasie fibreuse polyostotique (chez une fillette de 8 ans) est

rapporté. Un kyste de grandeur moyenne qui n'avait pas donné de symptômes a été gratté de manière à être complètement nettoyé puis empli de substance osseuse spongieuse homologue. Trois ans et demi plus tard, un examen aux Rayons X a démontré une structure osseuse entièrement normale à l'endroit opéré.

ZUSAMMENFASSUNG

Der Verfasser bespricht die Beziehung zwischen fibröser Dysplasie des knöchernen Skelettes und der Gruppe der Osteitis fibrosa und weiterhin die gegenseitige Beziehung zwischen polyostotischer fibröser Dysplasie, Albrights Syndrom und monostotischer, fibröser Dysplasie.

Die Möglichkeiten der Behandlung dieser Erkrankungen werden besprochen. Daraufhin wird über einen Fall von polyostotischer, fibröser Dysplasie berichtet (ein 8 Jahre altes Mädchen). Eine Zyste von Durchschnittelgrösse, die keine Symptome hervorrief, wurde vollständig ausgekratzt und mit homologem spongiösem Knochen ausgefüllt. Drei Jahre später zeigte die Röntgenuntersuchung vollständig normale Knochenstruktur an der Operationstelle.

REFERENCES

Adams, C. O., Compere, E. L. and *Jerome, J.:* Surg., Gynecol. and Obst., 1940: 71: 22.
Albright, F., Butler, A. M., Hampton, A. O. and *Smith, P.:* New Eng. Journ. Med., 1937: 216: 727.
Albright, F. and *Reifenstein, E. C.:* The Parathyroid Glands and Metabolic Bone Disease, Ed. I, The Williams & Wilkins Co., Baltimore, 1948.
Arnesen, Hans. Chr. and *Nitter, Lorentz:* Nord. Med., 1951: 45: 735.
Belaval, G. S. and *Schneider, R. W.:* Cleveland Clin. Quart., 1954: 21: 158.
Bingold, A. C.: Brit. J. Surg., 1948: 36: 22.
Helfet, A. J.: Brit. J. Surg., 1940: 27: 651.
Hirsch, Carl: Personlig meddelelse, 1954.
Lichtenstein, L.: Arch. of Surg., 1938: 36: 874.
Lichtenstein, L.: and *Jaffe, H. L.:* Arch. Path., 1942: 33: 777.
Hobæk, Andreas: Acta Radiologica, 1951: 36: 97.
Hobæk, Andreas: Acta Radiologica, 1951: 36: 145.
von Recklinghausen, F. D.: Festschr. z. Rudolf Virchow, Berlin, 1891.
Russell, L. W. and *Chandler, F. A.:* J. Bone and Joint Surg., 1950: 32A: 323.
Sante, L. R., Bauer, W. M. and *O'Brien, R. M.:* Radiology; 1948: 51: 676.
Schlumberger, H. G.: Mil. Surgeon, 1946: 99: 504.
Tiitinen, E.: Annales Chir. et Gyn. Fenniae, 1949: 38: 20.
Ulland, Gunnar: Acta Chir. Scand., 1944: 89: 113.
Valls, J., Polak, M. and *Schjowicz, F.:* J. Bone and Joint Surg., 1950: 32A: 311.
Ønne, Lars: Acta Chir. Scand., 1951: 101: 318.