FROM THE ORTHOPAEDIC CLINIC OF KAROLINSKA INSTITUTET, STOCKHOLM
(CHIEF: PROFESSOR PATRIK HAGLUND)

A FEW CASES OF OSTITIS DEFORMANS (PAGET) OF THE TIBIA

BY
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CASE I. Foundry man, aged 65. Admitted Feb. 3, 1931 to St. Erik's Hospital, Stockholm, for right-sided inguinal hernia, which was operated upon. In so doing a well-marked ensiform bowing of the right tibia was observed.

At the age of 30 the patient had already noticed his right leg beginning to bend in an outward-forward direction. This bending gradually increased, although it did not inconvenience mained unchanged during the last few years; a slight "creepy" the patient to any appreciable extent. The deformity had refeeling along the leg and above all a progressive sensation of unsteadiness in walking had forced him, however, to use a stick. The patient himself thought that the bowing of the leg was due to his resting a great deal on his right knee during his work as a foundry man. Denied lues. An X-ray examination had been made in 1920, when the tibia was described as being the seat of chronic osteomyelitis (fig. 1).

Condition Feb. 1931. — Powerfully built man. Heart normal. Bloodpressure 160/180 mm. No decided rigidity of radial arteries.

Wassermann's reaction negative. Sedimentation rate in blood 3 mm./hour. Blood calcium 9.6 mgm %.

No palpable thyroidal or parathyroidal tumours.

Well-marked ensiform bowing of the right leg with the convexity forwards-outwards (fig. 2 and 3). The tibia can be

felt to be much thickened. Circumference of leg 3 cm. greater than of the left leg. When the patient stands with his heels together the distance between the inner femoral condyles measures 5 cm. The right knee shows normal outlines; no effusion. Distinct crepitations in both knees, on flexion as well as

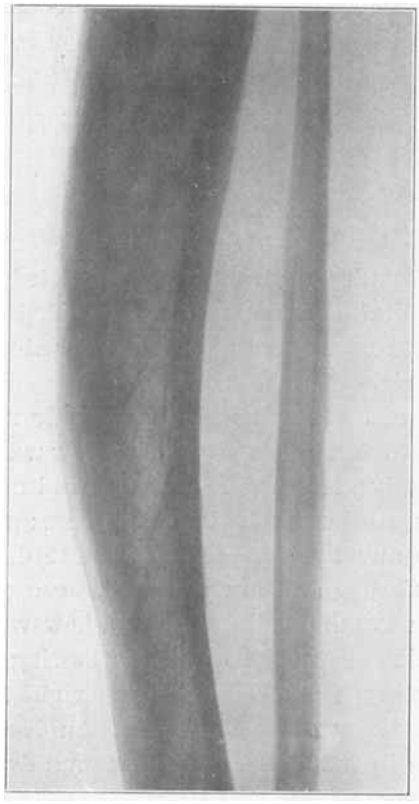

Fig. 1.

extension. Flexion of normal extent. Extension limited by 15° —20°. With the knee extended there is considerable lateral mobility.

X-ray examination (fig. 4—7). — Right tibia: well-marked ensiform bowing. The normal bone structure of the tibia has almost entirely disappeared and been replaced by a new patho-

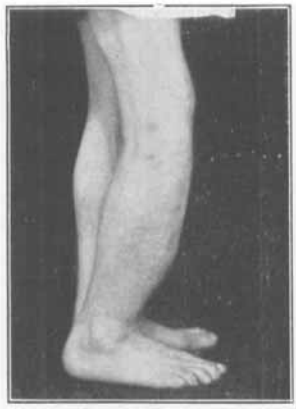

Fig. 2.

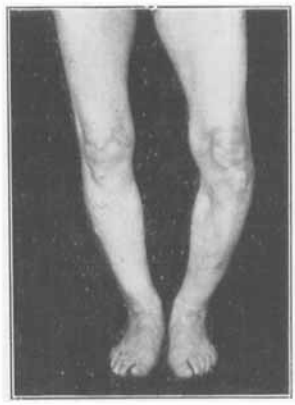

Fig. 3.

Fig. 4.

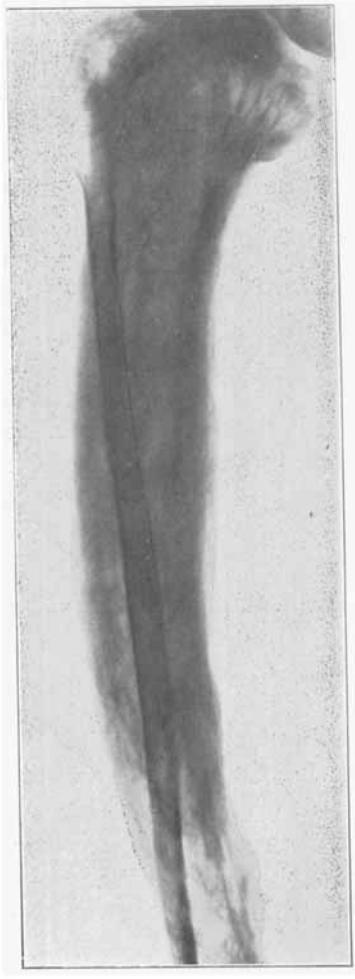

Fig. 5.

logically changed tissue. The cortex thus consists of broad longitudinal bars between which can be seen some elongated interspaces. The spongious structure, too, has almost entirely disappeared. Instead can be seen some rarefied areas of larger or smaller size which must be caused by destructions and which

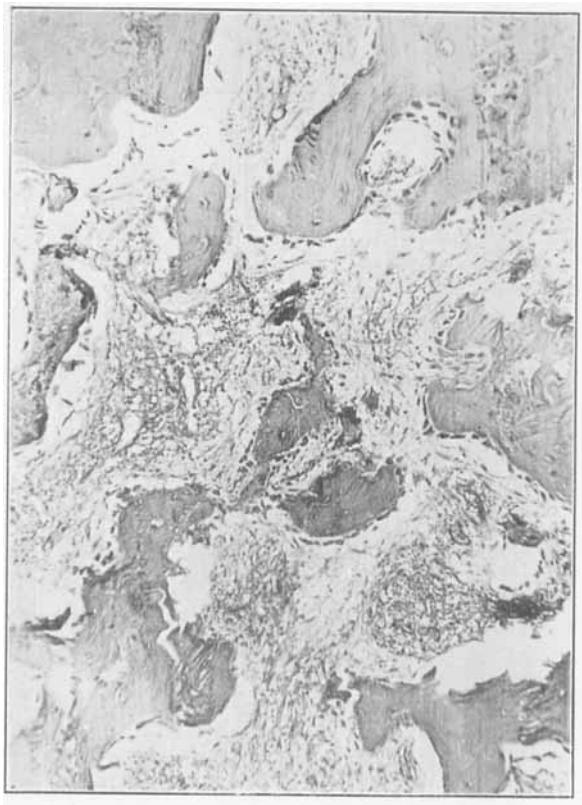

Fig. 12.

are surrounded on all sides by bone of irregular structure, showing considerable sclerosis. The fibula is perfectly normal. The deformity of the right tibia just described is in all probability occasioned by ostitis deformans. With a view to finding out whether similar changes were present elsewhere in the body, radiograms were taken of pelvis, skull, right shoulder-

joint, left leg and right foot; all, however, were found to be normal.

I subsequently excised a portion from the right tibia for biopsical examination. The bone was very hard. I excised a piece measuring 2 cm. in length, 1 cm. in width and 3 cm. in

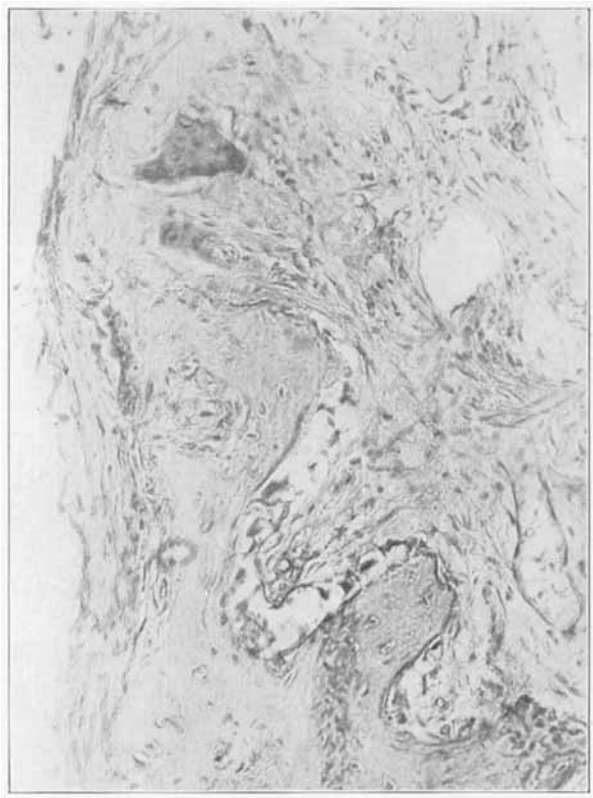

Fig. 13.

depth without coming across any marrow cavity. Macroscopically the periosteum was normal; the cortex was irregularly porous.

Microscopic sections (fig. 12 and 13) show spongious bone with coarse irregular bars within which the bone corpuscles are not arranged in any definite manner. The calcium content

of the bone seems on the whole to be normal; only here and there it appears in some sort of mosaic pattern. Some of these osseous bars are bordered by a row of epithelioid cells — so-called osteoblasts. Here and there in cavitations in the borders of the osseous bars are seen large multinuclear cells — osteo-

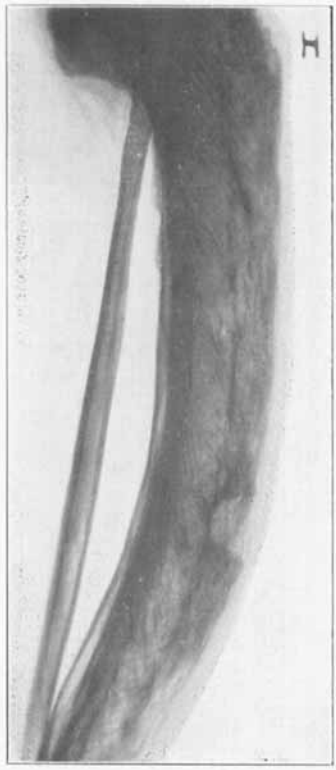

Fig. 6.

Fig. 7.

clasts. The bone-marrow is fibrous, fairly rich in cells and with blood-filled vessels. There are no signs of any inflammatory process. The changes agree with the appearance of ostitis deformans.

No treatment.

In December 1931 the patient came up again to be seen. His symptoms were very much the same; his walking, however,

was somewhat more unsteady. Renewed X-ray examination (9 months after biopsical excision). The defect (fig. 6) could very clearly be seen, from which one could gather that the pathological bone formation had been arrested and that the process was in course of healing.

The patient was recommended to wear a jointed knee-cap to give the knee sufficient stability.

On comparing the X-ray pictures of 1920 and 1931 we find that the deformity has increased during this period, at the same time as the bone has become more sclerotic, from which it would be inferred that the process has passed over from the osteoid hyperosteitic stage to the sclerotic stage of healing.

The relatively slight arthritic trouble with the knee show of course that in spite of extensive pathological changes in the upper end of the tibia (fig. 7 and 10), the cartilage has nevertheless remained untouched by the diseased process.

CASE II. Female, aged 70. The patient says she has always been slightly bow-legged but has never herself thought of the legs being dissimilar. She is unable to say when the left leg began to get crooked. During the last few years she is said to have been unsteady on her left knee. She is really seeking medical advice now because "other people" have told her that she is walking crookedly on her left leg. Has always been in good health. Lues denied. Having been doing a charwoman's work she has been obliged to kneel a great deal.

Condition Dec. 1931. — Heart normal. Blood-pressure 165/90 mm. Radial arteries somewhat rigid. Wassermann's reaction negative. Sedimentation rate in blood 4 mm./hour. Blood calcium 10.2 mgm %.

There is an ensiform curvature of the left leg (fig. 8 and 9), the most marked curve being just below the knee (fig. 9 and 10). The leg is on the whole much thickened. The circumference of the left calf is 3 cm. more than the right. Extension in

the knee limited by 10°; considerable lateral mobility. Moderate crepitations. Distance between knees 15 cm.

X-ray examination shows a typical appearance of ostitis deformans. The whole tibia is involved in the process — most marked above, fading away downwards. Fibula unaffected. No other localised foci in the skeleton. The appearance agrees well with that of Case I.

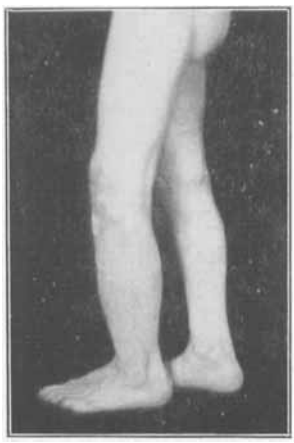

Fig. 8.

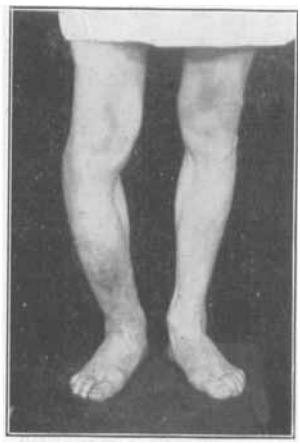

Fig. 9.

Treatment. — A jointed knee-cap is advised, but the lady is too coquettish to wish to wear a thing like that, wherefore for the time being she is to try with a strong elastic knee-cap.

CASE III. Female, aged 61. Four years ago the patient was caught with sudden aching pains in her left leg, with blue discoloration and swelling. Her doctor diagnosed "blood stasis". Ever since then the leg has been thicker than normal and begun to bend forwards-outwards and the patient has suffered severe aching pains from it. The mobility in the left knee is restricted. Slight limitation of flexion. Extreme flexion painful.

X-ray examination (fig. 11) shows the appearance of a re-

latively early process of ostitis deformans where normal bone structure still remains here and there.

Treatment. — The patient is advised medication for six

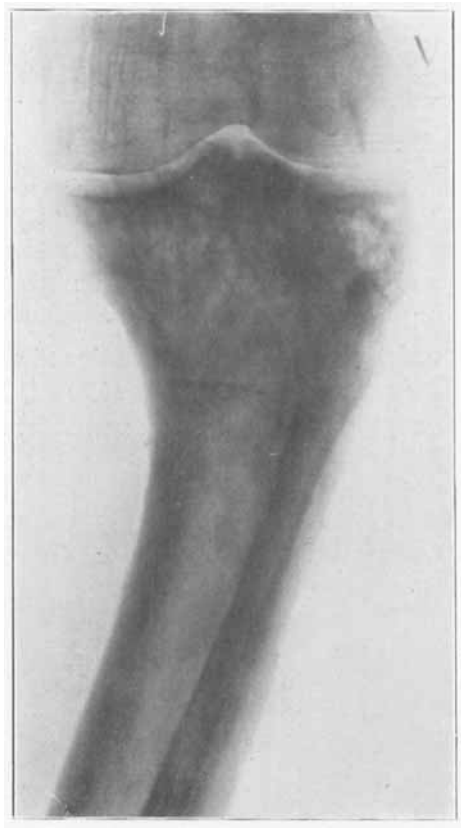

Fig. 10.

months (potassium iodide and calcium) later to return for renewed X-ray examination.

This exceedingly characteristic bone disease of advanced age has been given a great number of different names in the literature. *Ostitis deformans* tells a great deal about the disease.

Let us therefore stick to this simple name — but by all means adding Paget to honour the man who was the first to describe a case of that nature. It was in November 1876 that Paget told the London Medical-Surgical Society about an uncommon bone

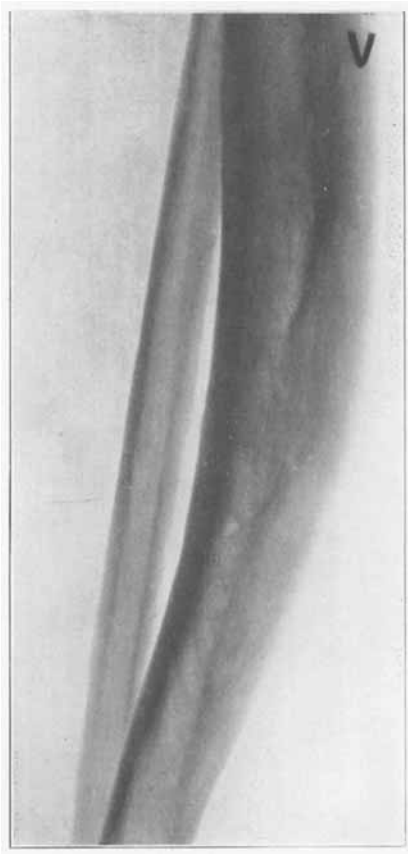

Fig. 11.

disease in an old man. The course of disease was quite characteristic of ostitis deformans.

Ostitis deformans is a typical disease clinically observed in persons over the age of 50. It may begin earlier, however, at the age of 30—40, but as it takes a very chronic course and

gives rise to very slight symptoms to start with, it is not generally detected until after the age of 50. It is somewhat more frequent in men than women. The Germanic race is more often affected than the Roman. Patho-anatomically, ostitis deformans is a very common disease. *Schmorl* has shown on an extensive case material that no less than 4 per cent of people above the age of 50 have some signs of ostitis deformans in their bony system. Clinically, however, the condition is relatively rare. The commonest localisations that can be clinically observed are found in the tibia, skull and spine.

People do not of course consult a doctor until they detect something amiss or feel ill. These localisations cause most of the trouble. The very common kyphosis that comes with advanced age is often due to ostitis deformans in one or several vertebrae. When in an early stage of ostitis deformans the vertebrae become decalcified and less resistive to external influence, they become readily compressed in the thoracic region through gravitation, particularly in the fore part of the vertebral bodies, whereby kyphosis arises.

The changes taking place in the skull cause this to increase in circumference. Particularly is this the case with the transverse diameter. The inner diameters of the skull remain practically the same.

Perhaps the most characteristic seat of ostitis deformans is the tibia. The sharp anterior crest becomes rounded. The whole leg below the knee curves in a forward-outward direction. The tibia becomes greatly thickened, particularly its uppermost and middle parts. It is not only a question, however, of a thickening but also of an elongation of the bone, as is clear from the X-ray picture (fig. 4). The fibula is seen there to be perfectly intact and appears like the string on a bow while the tibia is much thickened, elongated and curved in a swordlike manner. If the radiograms are examined (fig. 10) we shall find a very pronounced varus deformity of the tibia just below the knee and a forward curve lower down. These bends pass over into one another in such a way that the result is the characteristic ensiform bone with curves forward-outwards.

Several factors are involved in the development of this deformity, in the first place the diseased process itself and, secondly, the great mechanical strain through gravitation. The disease commences with an often very slow process of softening and decalcification of the bone. By rapid activity of the osteoclasts at the beginning of the disease the leg may diminish in volume; but what usually happens is that together with this softening process a regenerative new bone formation takes place which soon predominates. The result of this is often a considerable increase in the volume of the cortex and a reduction of the marrow cavity. The newly-formed bone is of an osteoid and soft type, poor in calcium. It is during these two first stages of the diseased process that deformities arise. Gradually the calcium content in the bone is increased and the gravitation deformities are arrested. The calcium content may frequently become so much increased as to give rise to a sclerotic form of bone. This sclerosis is to be regarded as the healing stage of the process. From the radiograms it is possible, therefore, to deduce the stage of disease for the time being, which is of course a matter of great importance for the treatment.

We distinguish between two forms of ostitis deformans — a localised and a generalised form. One is about as common as the other. The localised form is the most benign of the two.

On determining the blood calcium in the cases described above this proved fully normal — 9.6, 10.2 and 12.2 mgm %. Ostitis fibrosa (*Recklinghausen*), on the other hand, always shows an increased blood calcium — up to 20 mgm % — due to hyperfunction of the parathyroid glands. The glands are enlarged through tumours (adenoma).

Schmorl has examined, patho-anatomically, a very large number of cases and points out that ostitis deformans has a typical "mosaic" structure, histologically, which is not found in any other bone disease. Softening of normal bone takes place through the activity of osteoclasts, while the osteoblasts are responsible for the regeneration. The absorptive spaces appear under the microscope as "putty-spots" or in longitudinal sections as "putty-lines". It is these "spots" and "lines" that, if

pervading the normal bony bars quite irregularly, give to the structure the characteristic appearance of mosaic.

The aetiology of ostitis deformans is quite unknown. Some authors maintain that some specific infection (lues, tbc) must be the cause of this bone disease. Some again consider ostemyelitis to be the primary cause; this is, however, contradicted by the microscopical examination.

Ostitis fibrosa (*Recklinghausen*) is often confused with *ostitis deformans* (*Paget*). They are, however, two quite separate conditions. The former has not the bone structure characteristic of the latter.

Ostitis deformans being a phenomenon of advanced age, one would feel prone to connect it with arteriosclerosis. There is evidence to show, however, that the cases in question have no more widespread arteriosclerosis than others of the same age. The bloodpressure is also about normal.

Treatment. — As already mentioned, the symptoms are rather insignificant. As to deformities it is only those of the lower extremities that sometimes may become sufficiently well-marked to call for operative treatment — as in the cases described below. Most orthopaedic specialists and surgeons warn against osteotomy on account of poor osseous union; others again like *Trinca* maintain that fractures in cases of ostitis deformans unite much quicker than usual. *Schnek* has described a case of traumatic fracture of the femur which united quite normally with good callus formation in the course of 8 weeks. *Korzeborn* described a case in which the deformities of the lower extremities — thigh as well as leg — were so marked that they crossed each other in walking which, naturally, was thereby greatly hampered. In spite of the risk of pseudarthrosis he performed a subtrochanteric osteotomy, first on the one limb which united in 6 weeks, and then on the other limb with equally good result. To get rid of the varus deformities of the tibiae K. subsequently performed an osteotomy on both sides, similarly with quite a normal progress of union. During the first stages of the healing process when we have to deal with a soft, osteoid bone poor in calcium that is unable to stand the

weight of the body, a jointed knee-cap may come in handy. The great difficulty, however, is to get these early cases for treatment and to get the patients to wear some support during the many years required for the bone to become sclerosed and regain its normal stability. Some supporting appliance is also indicated in these cases to counteract the feeling of insecurity of the knee of which these patients often complain, and to steady the "wobbling" joint due to gravitation in varus position. With regard to medication, no drug has as yet proved better than any other. Some calcium preparation, however, can do no harm to aid calcification.

SUMMARY

Three cases of quite typical osteitis deformans (Paget) are recorded, solely localized in tibiæ, all fairly old patients in whom the course of the disease has been very chronic, and who suffered only rather slight subjective disability in comparison with the great morbid changes on the X-ray pictures. The curving of the bones increased throughout the whole of the osteoporotic stage but ceased in the sclerotic stage. Ætiology unknown. The phosphor and lime content of the blood was found to be quite normal. Despite the great pathological changes in tibiæ the cartilage in knee and ankle was found to be in no worse condition than was consistent with the age of the patients. As a general rule treatment will not be necessary.

RÉSUMÉ

On communique 3 cas tout à fait typiques d'osteitis deformans (Paget) localisés seulement jusqu'au tibia. Il s'agit de malades très âgés awec cours de maladie absolument chronique, et très peu de gênes subjectives en comparaison des grandes altérations trouvées à la radiographie. Les recourbements osseux augmentent tout le temps au stade ostéoporotique mais s'arrêteut au stade sclérotique. Etiologie inconnue.

Le contenu de phosphore et de chaux du sang est complétement normal.

Malgré les grandes altérations pathologiques au tibia, le cartilage articulaire des genoux et des pieds n'était pas en plus mauvais état que ne le comporte ordinairement l'âge des malades. Dans ces cas aucun traitement ne s'exige en général.

ZUSAMMENFASSUNG

Drei Fälle von ganz typischer Osteitis deformans (Paget) nur in der tibia lokalisiert werden mitgeteilt. Es handelt sich um recht alte Patienten mit einem sehr kronischem Krankheitsverlauf und ziemlich kleinen subjektiven Beschwerden in Verhältnis zu den grossen krankhaften Veränderungen auf den Röntgenbildern. Die Krümmungen der Knochen werden während des osteoporatorischen Stadiums fortwährend vermehrt, aber kommen in sklerotischen Stadium zum Stillstand. Die Aetiologie unbekannt. Der Fosfor- und Kalkgehalt des Blutes war vollständig normal. Trotz der grossen patologischen Veränderungen in der tibia war der Gelenksknorpel im Knie- und Fussgelenk nicht schlechter als dem Alter der Patienten entsprechend. Irgendwelche Behandlung wird im Allgemeinen nicht notwendig sein.

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