

THE ULTIMATE INVALIDISM IN A CASE OF STILL'S DISEASE

BY

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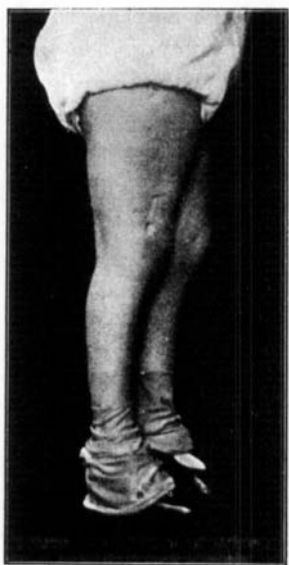


Fig. 1.
Jan. 1931.

On page 327 of the present issue of the *Acta orthopaedica scandinavica* Prof. Monrad, Copenhagen, communicates 5 cases of multiple chronic disease of the joints in childhood. The first of these cases was treated later on in this clinic for contractures and ankyloses, and as the writer still has connection with the patient a description will here be given of the ultimate sequelæ of her severe joint affection, which — as stated by Prof. Monrad — must be considered quite a typical case of the so-called Still's disease.

The patient came to be examined (by Prof. Slomann) at the clinic for the first time in 1917 having been free from acute attacks during the last 2 years, this juncture, therefore, being considered a favourable one for surgical correction of the contractures. There came, however, a period with mild pain and fever for which reason the patient was only admitted on Oct. 3rd, 1918, at the age of 14. Her general health was satisfactory at this time. Menses from the age of about 12½ years. Apart from the above transient febrile period with pain in the shoulder

regions she had been free from acute joint attacks for about 4 years. She had been able to walk a little by means of a crutch.

The condition of the joints was as follows:

Right upper extremity: completely free movements of the shoulder-joint. Elbow: flexion to 55° , extension to 135° . Pronation-supination of the forearm limited by about 15° in either direction. The wrist cannot be dorsiflexed past a position in which the hand is on the same straight line as the arm; the volar flexion is unimpaired. The movements of the thumb are unimpaired. The remaining 4 fingers are capable of moving freely in the metacarpophalangeal joints; II finger shows bony ankylosis in slight flexion of either interphalangeal joint, III finger bony ankylosis of the 1st interphalangeal joint, the distal joint being mobile, IV finger bony ankylosis of either interphalangeal joint, and V finger all joints mobile.

Left upper extremity: free movements of the shoulder-joint. The elbow: flexion to about 75° , extension to about 150° . The pronation-supination of the forearm is limited by about 20° in either direction. The wrist: bony ankylosis in volar flexion of about 25° . The fingers display bony ankylosis of the same joints as the right hand, apart from the fact that there is some mobility here of the distal interphalangeal joint of the II finger and of the proximal interphalangeal joint of the IV finger.

Of the movement of the head rotation to the *right* is somewhat (10 — 15°) restricted, the same applies to reclinatioin but bending forward is natural. The movements of the mandibular joint are somewhat restricted, the patient only being able to open the mouth to 3,5 cm's distance between the teeth.

The mobility of the spinal column is little restricted. There is a rather marked lumbar lordosis (the remnants of a flexion contracture of the hips).

Right lower extremity: apart from a slight extension defect (secondary?) the *hip* can be moved freely. The knee can be extended to 140 — 145° and flexed to 115 — 120° . There is complete bony ankylosis of the joints of the foot in plantar flexion of about 30° , no varus or valgus position. The tarso-metatarsal joints are ankylotic. The metatarso-phalangeal joints of the

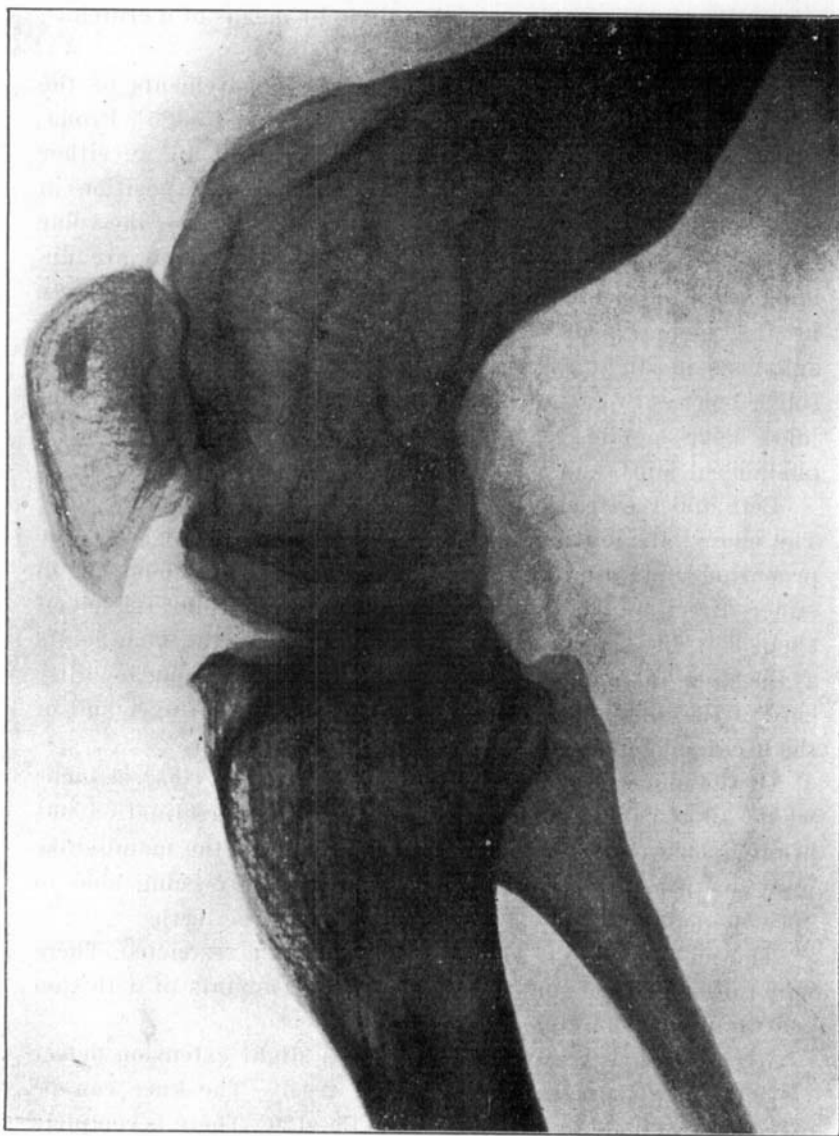


Fig. 2.
Right knee 10 years after the para-articular osteotomia femoris.

toes can be moved freely, but there is bony ankylosis in slight flexion of *all* the interphalangeal joints.

Left lower extremity: there is a slight extension defect of the hip, just as on the right side. The knee-joint can be extended to $145-150^{\circ}$ and flexed to $80-85^{\circ}$. The ankyloses of the joints of foot and toes correspond to those on the right side, but besides

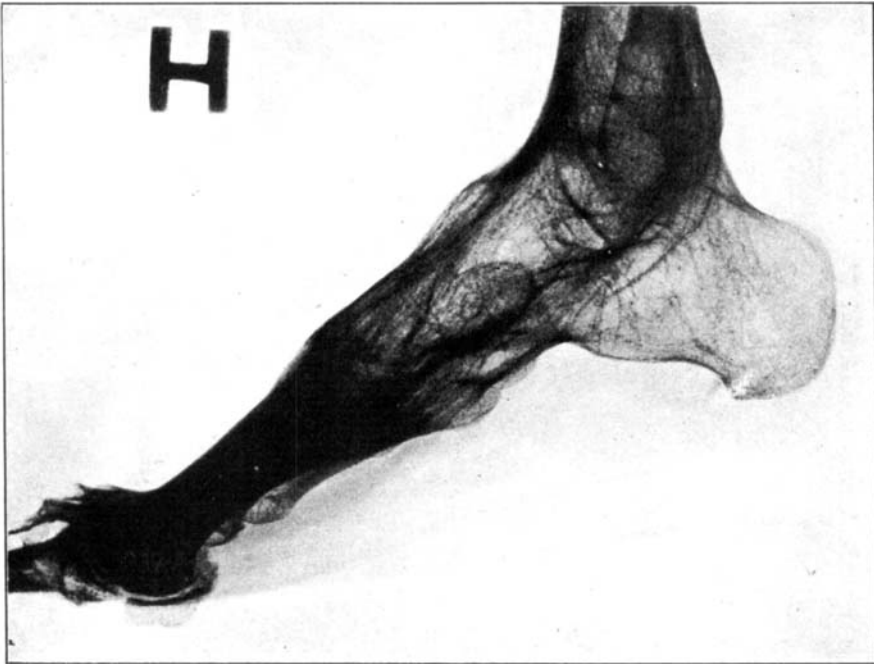


Fig. 3.
Right foot.

being plantiflexed the foot is in a slight varus position. The interphalangeal joint of the great toe and the distal interphalangeal joint of the V toe are not ankylotic in this foot.

On Oct. 11th, 1918, para-articular osteotomy femoris was performed at the *right* knee with a view to straightening the flexion.

On Nov. 6th, 1918, osteotomy at the *left* ankle-joint was per-

formed with partial extirpation of the astragalus and lengthening of the triceps tendon with a view to correcting the equinus position and, simultaneously, para-articular osteotomia femoris at the left knee in order to correct the flexion there.

On Dec. 6th, 1918, osteotomia of the ankylotic right ankle-joint and lengthening of the triceps, in order to correct the equinus position there, were performed.

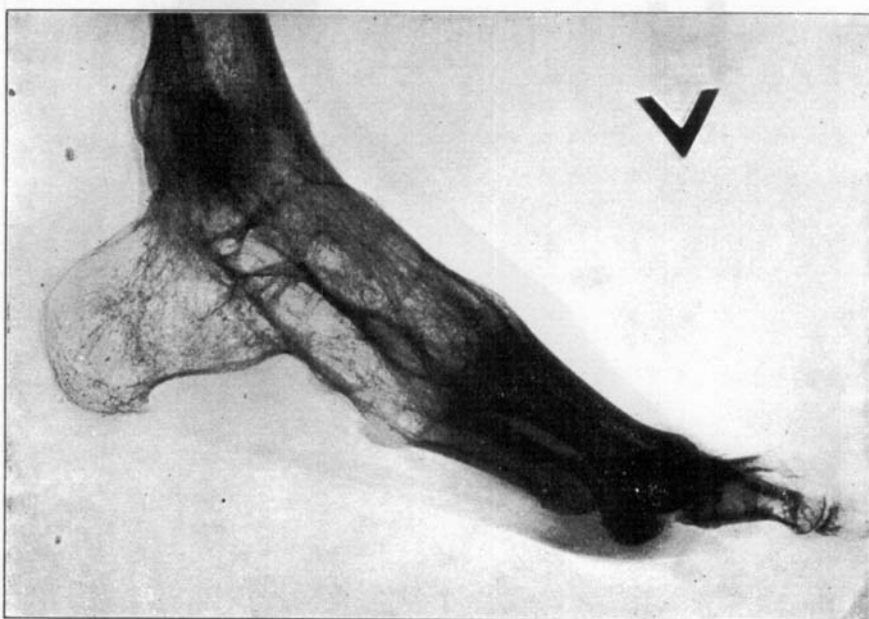


Fig. 4.
Left foot.

All the osteotomias healed in a satisfactory position, the patient maintained her former mobility of the knee-joints, movements of the knees, however, being commenced in a position of complete extension (see fig. 1 and fig. 2). The ankle-joints were ankylotic in slight plantar flexion. She began to walk on Jan. 16th, 1919.

Up to Nov. 1919 the plantar flexion became more marked than was desirable for which reason a bloodless correction

(osteoclasia) was performed on Nov. 21st, 1919. The position was good afterwards.

On April 19th, 1923, an osteotomy was performed with a view to straightening the bony ankylosis of the left wrist, which was in a volar flexion of about 25°, the hand thus coming on the same straight line as the forearm.

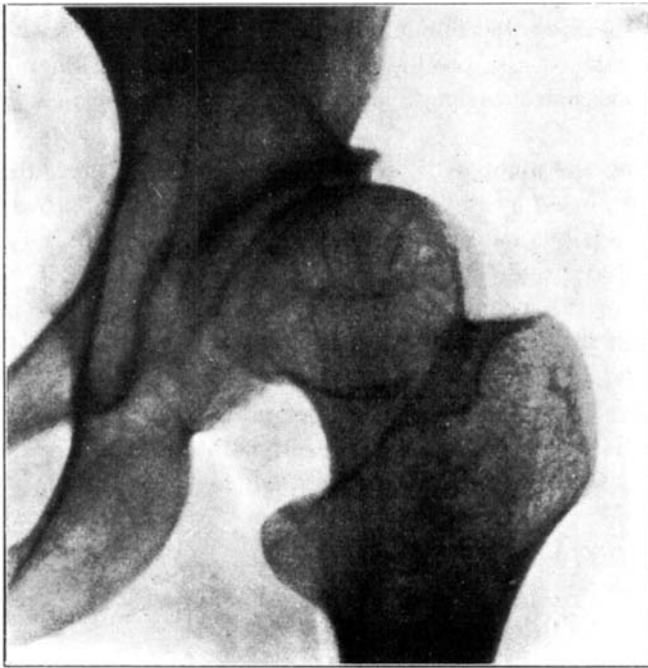


Fig. 5.
Left hip joint.

Since the operations on the lower extremities the patient has been able to walk without a stick, although with some difficulty and with some claudication. She has been travelling a good deal and was able to manage for herself abroad for long periods. During the last few years she has even been riding a good deal. —

From an orthopedic point of view this strange and rare in-

fantile multiple joint affection, which has been termed Still's disease, is mainly of interest in that it has a marked tendency to end in bony ankyloses. The skiagrams of the feet reproduced in Fig. 3 & 4 show the complete obliteration of all the articular lines from the metatarso-phalangeal joints to the crus. The almost complete exemption of shoulder- and hip-joints (see fig. 5) is also a strange but very pleasant feature of the facies morbi. If these joints were involved the ultimate invalidism would be of an especially terrible nature, whilst a patient as the one described here still is able to manage for herself during life despite extensive cosmetic and considerable functional inconvenience.

During the orthopedic after-treatment the greatest difficulties were caused by the feet, which were entirely rigid owing to the complete bony ankylosis of either talo-crural joint and of all the tarsal and tarso-metatarsal joints. In order to distribute the pressure of the weight fairly evenly the patient has had to use insets, but in making the latter it was impossible to have them fitted with sufficient exactness by the ordinary technique used in making insets by means of plaster of Paris casts. *Leather insets* adjusted by the writer himself with a shoemaker's knife under repeated weighting attempts had to be employed.



Fig. 6.