(FROM QUEEN LOUISE'S CHILDREN'S HOSPITAL, COPENHAGEN)

MULTIPLE CHRONIC DISEASE OF THE JOINTS IN CHILDHOOD[1])

BY

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Our knowledge of the nature of the so-called *Still's disease* is still lacking elucidation in so many points that any casuistic communication should be taken interest in. Moreover do the cases which I have had the opportunity to observe and to treat in several respects display considerable deviations from the usual facies morborum.

Before I explain my own cases I would draw the attention to a paper by Prof. *Hirschsprung* which he published already in 1901 in the »Hospitalstidende« under the title of »Multiple Chronic Infectious Disease of the Joints in Childhood« and in which he describes no less than 8 cases of this strange disease, all the cases having been observed by him in Queen Louise's Children's Hospital. Unfortunately it seems as if this paper, which in reality is a classical one, has been allowed to pass rather unnoticed; at any rate it is never mentioned by the authors who have pursued the same subject more recently.

My own material comprises 5 cases in all, and we at once see the peculiarity that whilst two of the children have a typical intermittent pyrexia the three others have not had any pyrexia at all at any juncture of their disease. Granted that it is the same disease in all five cases, and there is hardly any reason to

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doubt it, it seems as if we must distinguish between two different types, a febrile and an afebrile type.

The febrile type, the facies morbi of which corresponds very nearly to the one described by *Still,* comprises the following two cases:

Case record No. 1. Astrid D., born on July 14th, 1904, who had been in good health previously — apart from the ordinary diseases of childhood — *was suddenly taken ill* in Oct. 1911 (*at the age of* 7) with *fever and painful swelling of wrists and ankle-joints.* It was only natural that the disease was regarded as febris rheumatica, and it was treated at home with confinement to bed and salicyl; as this was of no avail phenacetin, antipyrin, pyramidon etc. were tried but did not produce any effect. After 3 months' painful confinement she was admitted to this hospital in January, 1912, where she remained until May, 1913.

During this stay in hospital for nearly 16 months she afforded a typical example of Still's disease: *symmetrical, periarticular swelling* of most of the joints of the extremities, of the mandibular joints and of the cervical collumn; no exudation in the joints, no involvement of cartilage and no destruction of bony structures but gradually shrivelling and to some extent marked contractures. *Intermittent pyrexia,* often up to 40° (104 Fahr.), associated with *sweating,* marked decrease of general health, a rather considerable emaciation, and a yellowish pale, *cachectic* complexion. Moreover *enlargement of the glands of the peripheral glandular system,* also of the cubital glands, and now and then a slight *enlargement of the spleen.* All through the period of disease the liver was swollen to a rather great extent. Apart from simple anæmia examination of the blood showed nothing abnormal, and the blood was found to be sterile on culture. On stethosc. cordis hæmic murmurs were heard at the base and on a single occasion some friction-murmurs (pericarditis?) but never any signs of valvular disease. The urine normal. Pirquet and Wassermann negative. Add to this that a marked *muscular atrophy* rapidly occurred and we have a typical picture of Still's disease. The course, too, was quite typical with alternating good and bad periods apparently without demonstrable causes and at

any rate quite independent of the various lines of treatment which were followed and which partly consisted of hot soap baths and hot-air treatment and partly of the administration of numerous drugs: salicyl, aspirin, diplosal, quinine, arsenic, atophan, collargol etc. Days would come suddenly on which she was able to sit up without difficulty and even make attempts at walking, but recrudescences with pyrexia, painful enlargement of the joints and complete immobility would occur as suddenly.

She was discharged in rather a desolate condition but after the course of 2 years the acute attacks ceased spontaneously. Now her general health improved rapidly, but on account of severe contractures and deformities of knee- and ankle-joints various orthopedicosurgical interferences were made during the following years resulting in what may be called an almost complete recovery, although she has some difficulty in walking.

Case record No. 2. Helene B., born on March 9th, 1925, who had been in good health previously, was suddenly taken ill in February 1928 (nearly *3 years old*) with *fever and painful swelling of wrists and ankle-joints.* The disease was regarded as febris rheumatica and was treated accordingly at home. As the treatment, however, was of no effect she was admitted to this hospital in May 1928 and is still here.

During this stay in hospital for nearly 2 years she afforded a typical example of Still's disease — just as the first patient; *symmetrical, periarticular swelling* of most of the joints of the extremities and of the cervical column but not of the mandibular joints; no exudation in the joints, no involvement of the articular cartilages and no destruction of the bony structures of the joints. Very marked *muscular atrophy. Enlargement* of the *peripheral glands,* especially of the axillary and cubital glands; but *never any enlargement of spleen* or liver (see fig. 1 and fig. 2). *Intermittent pyrexia* with severe decrease of general health, marked *sweating,* emaciation, and yellowish pale *cachectic* complexion. On examination the blood showed a marked anæmia (40 % Leitz) but the blood picture was normal and the blood appeared to be sterile on culture. Stethosc. cordis revealed hæmic

murmurs at the base but never any signs of endo- or pericarditis. Pirquet and Wassermann negative. The urine normal. X-ray examination of all the articulations and adjoining bones reveals a very considerable *bony atrophy* (halisteresis) everywhere; moreover a considerable swelling of the capsular ligaments is

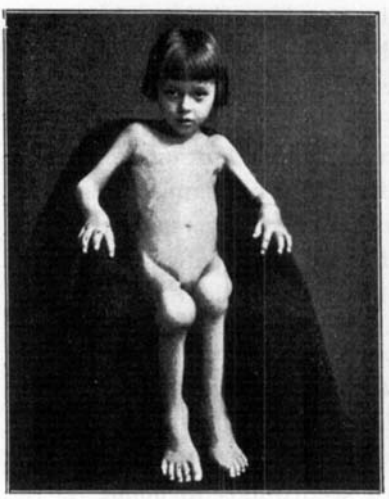

Fig. 1.

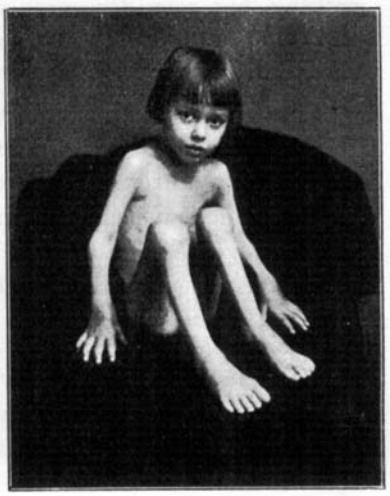

Fig. 2.

Helene (Case record No. 2).

seen and in the case of some of the joints there is some narrowing of the articular cavities but in no case signs of destruction of the surfaces of the joints.

The course of the disease has also been typical so far, long febrile periods alternating with shorter good periods. In the febrile periods she is very prostrate; the highly swollen joints are then very tender and she lies motionless and crouched in her bed with strongly flexed knee- and hip-joints. During such a period with much tenderness and marked flexion especially of the left hip the occurrence of a *dislocation* was discovered, the femur having been moved about 3 cm. in an upward direction with the caput resting on the upper edge of the acetabulum, which otherwise did not display anything of interest. During the afebrile periods she rallies, eats much, increases her weight,

gets a healthy complexion, and the swelling of both joints and glands decreases considerably. She is then able to move about a good deal in her bed, and once she even flounced so much that her right leg was caught between the rungs of the bed which caused an *infraction* of the right femur which, however, was healed in a strikingly short time. On another occasion she got an infraction of her left humerus by a similar accident — in both cases a consequence of the deficient quantity of lime-salts of the bones characteristic of this disease.

This alternation between good and bad periods always arises quite *spontaneously* and at any rate irrespective of the various lines of treatment followed. *Intercurrent infections* have been of a varying effect: thus, in a period during which she had measles and whoopingcough and therefore was completely isolated and under no special treatment, we saw that her condition improved strikingly whereas grippe-infections always caused a considerable aggravation.

All drugs hitherto administered (salicyl, iodine, melubrin etc.) have been of no effect whatever. In March—April, 1929, *Walbum's metal-salt* treatment was tried, 14 injections (from 0,20—0,40 ccm.) in all being given, but they, too, were of no effect whatever.

During the apyretic periods *light massage was given and cautious attempts at redressments* were made but often the latter had to be discontinued owing to pain and tenderness.

It is still too early to say anything about her future, but when the febrile attacks cease I will perhaps try the treatment with fibrolysin recommended by Frölich.

The afebrile type comprises the following three cases:

Case record No. 3. Florry P., born at term after normal labour on January 10th, 1918, breast-fed, in completely good health, came on well and was able to sit up at the age of 6 months. *Already at the age of 7 months her disease began*, her mother noticing that her *right third finger was swelling.* The practitioner who was called in though the case was one of »spina ventosa« and sent the child to the Finsen Light Institute where

she got 30 light treatments which, however, did not bring about even the slightest improvement. When the child was about 1 year old her mother noticed that both knee- and ankle-joints were swollen and that the child winced when somebody touched the swollen joints; some weeks after the left elbow-joint also became swollen and tender and the child was then admitted to this hospital on May 3rd, 1919. During the 9 months of her disease the child has been completely well and lively apart from the said swelling of the joints and it should be noted that there were *not the slightest signs of fever.* Status præsens on admission: she is of a healthy, normal appearance and in a good state of nutrition; is not exhausted. There is a *typical, deforming swelling* of either knee-joint, either ankle-joint, of the left elbow-joint, and of the first interphalangeal joint of the right third finger; no exudate in the joints and the swelling does only involve the capsular elements and the periarticular soft parts. Either knee-joint is fixed at an angle of about 90° and attempts at movements cause pain; passive movements of the other joints can be carried out almost to the normal extent but they are accompanied by pain. X-ray examination shows a *marked bony atrophy* but otherwise nothing abnormal; there is no narrowing of the articular cavities and there are no signs of involvement of the surfaces of the joints.

Thus the picture presented by the joints involved completely resembled the joint affections seen in the true Still's disease. But on the other hand the rest of the examination showed that there was *no enlargement of the spleen, no enlargement of the peripheral glands, no fever and no cachexia.* Examination of the blood showed a simple anæmia; the blood picture was normal. The heart displayed nothing abnormal. Pirquet and Wassermann negative. After observation for about a fortnight *light massage treatment and redressment exercises* of the joints involved were commenced. During her stay in hospital no other joints were attacked and the swelling and tenderness of the affected joints gradually decreased. In the month of July she got a catarrhal infection which, however, was very mild and lasted a few days; there was a slight rise of temperature associated

with this infection, but otherwise *there was not the slightest sign of fever at any juncture of the disease.* After 4 months of treatment she was able to rise to her feet and to stand when supported. On Oct. 1st, 1919, she was discharged for continued treatment at home with massage and passive exercises and when, at the beginning of October, 1920, she was brought to see us she had *recovered completely:* the swelling of the joints had disappeared entirely and all the joints could be moved freely. *Her disease had then lasted for 2 years.* Apart from massage and kinetotherapy no treatment, especially no drugs, had been used.

Case record No. 4. Jens J., born on Oct. 9th, 1927, previously in good health until, in Oct. 1929 (*at the age of 2 years*), he got a *tender swelling of the left knee-joint;* a trauma was said to have preceded the occurrence of the swelling. He was treated at home by confinement to bed and fomentations, the disease being regarded as incipient tuberculosis. About 1 month later the left wrist and the right ankle-joint became swollen and as it was regarded now as a rheumatic affection a salicyl treatment was instituted but was of no effect whatever. Later on either knee-joint was involved and as the boy gradually felt more and more sick, became emaciated and pale he was admitted to this hospital on May 24th, 1930, having been ill for 8 months. I add that during his stay at home his temperature had been measured frequently and that there had *never been any fever.* Status præsens on admission: a lean, pale, somewhat *cachectic* looking boy. Hæmoglobin 68 %; normal blood picture. Stethosc. cordis displays nothing abnormal. *No enlargement of the spleen. No enlargement of the peripheral glands. No fever* during his stay here. Pirquet and Wassermann negative. There is a *typical, deforming, periarticular swelling* of both knee-joints which are kept strongly flexed and are very tender when attempts at movements are made; moreover painful swelling of either wrist, of the right ankle-joint and of the first interphalangeal joint of the left third finger. No exudate in the joints. X-ray examination reveals *a marked lime-atrophy,* but otherwise nothing abnormal. *Moderate amyotrophia.*

When he had been to bed for 2 weeks and still was apyretic,

and as the joints involved were less tender *light massage and kinetotherapy* were commenced, but it is, of course, impossible to say how this case wil develop.

Case record No. 5. Børge B., born on Dec. 31st, 1913, previously in good health, was *rather suddenly taken ill* at the beginning of January 1917 (*at the age of 3 years*), his *left knee* —

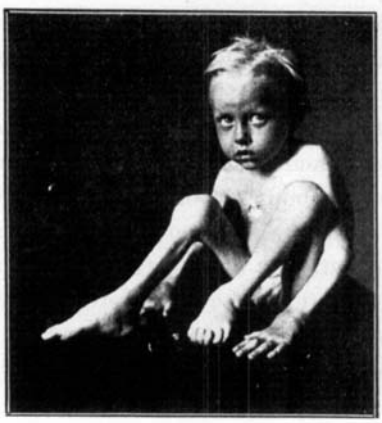

Fig. 3.

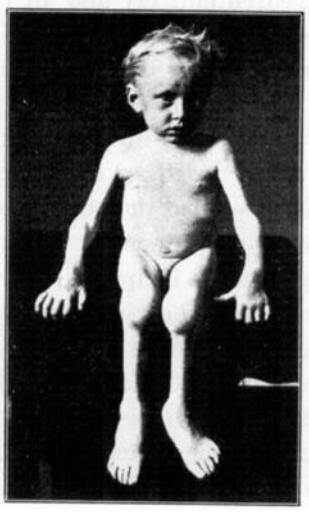

Fig. 4.

Case record No. 5. Børge. May 1917.

without any trauma or other known accidental cause — swelling very much in the course of a few days and becoming very tender and painful and incapable of being extended; *there was no fever.* About a fortnight later his right knee-joint became swollen in exactly the same manner, also without fever, and after the course of further 3 weeks either wrist began to swell for which reason he was admitted to this hospital in February 1917.

During the first months after admission the disease was aggravated and successively attacked the ankle-joints, the elbow-joints, most of the finger-joints and the cervical column, only the hip-, shoulder- and mandibular joints being intact at last.

The joints attacked all showed the *characteristic, deforming swelling of the capsule and of the periarticular tissues* without exudate in the joints and without destruction of the cartilaginous and bony structures of the joints (see fig. 3 and fig. 4). This was also confirmed by repeated X-ray examinations showing *marked bony atrophy* but otherwise nothing abnormal. During the first months of the disease all the articulations involved were so tender and painful that he was lying quite motionless and cried out on the slightest touch. Most of the joints were in a position of medium flexion, but a marked contracture of either knee-joint with a tendency to subluxation soon developed. Simultaneous with this quite symmetrical joint affection a very considerable *muscular atrophy* developed, causing the swollen joints to look imposing as if they were larger than they really were.

During the whole of this period his general health was extremely bad. He had no appetite, was whimpering, profusely *sweating,* became anæmic, *cachectic,* and rather emaciated. *But there has never been any fever during the whole course of the disease.* The remaining part of the objective examination showed normal conditions; I want especially to emphasize that there was *neither enlargement of the spleen nor of the peripheral glands;* stethosc. cordis was constantly normal. The examination of the blood showed a rather considerable anæmia (60 %), but the blood picture was normal and the blood proved to be sterile on culture. Pirquet and Wassermann were negative.

This painful condition lasted for more than 6 months and it was not until the end of Oct. 1917 that his condition improved so much that a *light massage* treatment could be commenced. Two months later, i. e. about 1 year after the occurrence of the disease, he was able for the first time to sit up in a chair and pain and tenderness had now subsided so much that we ventured to begin with *manual redressments* of the contracted joints. There was, however, little progress to begin with and only 6 months later did he begin to crawl. At the end of Aug. 1918 he began to walk in a gocart and from that time he made comparatively rapid and steady advances (see fig. 5).

The most persistent contracture was the flexion contracture of the knee-joints, especially that of the left knee, but *active exercises on a velocipede* were of an excellent effect. At the end of 1918 he began to walk alone, although with great difficulty.

I add that during the whole of his stay in hospital he did not get any medicinal treatment.

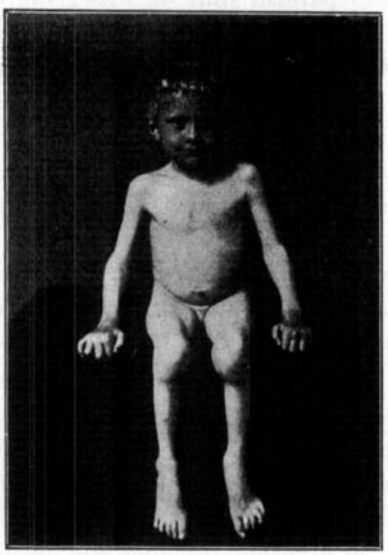

Fig. 5.
Case record No. 5. Børge.
September 1918.

In January 1919 he got a whooping-cough and was discharged for this reason but re-admitted in March 1919 for continued treatment. The whooping-cough had no influence whatever upon his joint affections. During the following months massage and kinetotherapy were employed causing the condition to improve constantly. The swelling of the various joints subsided more and more, his general health became excellent, his weight increased and he ran about all day long without any stick. He was discharged at the beginning of July 1919 but was enjoined to

continue with massage and manual redressments, especially because there still remained a tendency to contracture of the left knee.

When we saw him in December 1919 there was still a slight swelling of the capsular ligaments of the knee-joints (see fig. 6) but it disappeared completely in the course of the following 6 months.

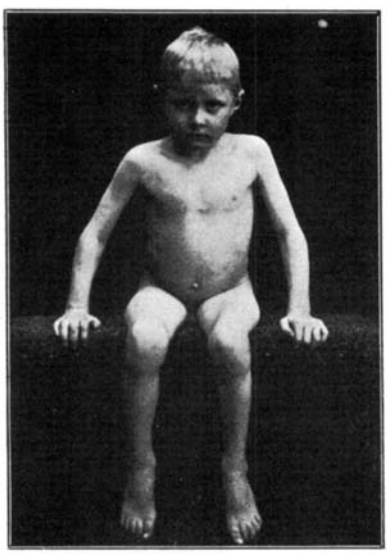

Fig. 6.
Case record No. 5. Børge.
December 1919.

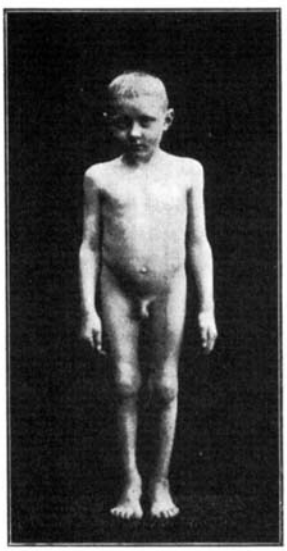

Fig. 7.
Case record No. 5. Børge.
January 1921.

When we saw him in January 1921, i. e. *4 years after the occurrence of the disease,* he was *completely restored* (see fig. 7).

I have reproduced these case records so much in detail because I want my readers to judge for themselves when interpreting the nature of the cases, and in order to do so one must be in possession of the most important documents of the case. A consequence hereof is, however, that, owing to the space allotted to

me, I must refrain from entering into a closer examination of the numerous problems of this strange disease.

On comparison of my three last cases, and especially on comparison of case record No. 5 with the two first records, *the symmetrical joint swelling, the halisteresis, and the muscular atrophy* speak very much in favour of the view that we are confronted with the same disease in all the cases, the more so as the photos of the two children resemble each other very closely. But how can we explain that there is *intermittent fever* and *enlargement of glands and spleen* in one group of cases, whilst these symptoms are about absent in the other group?

Looking at my first two cases one can easily endorse the theory that the disease is due to a *chronic infection.* But if attention is given exclusively to the last three cases it is difficult to believe in an infection and more natural to suppose that there is a *neurogenous origin* thus rather referring the disease to the progressive muscular dystrophias. It goes without saying, however, that there cannot exist two so highly varying etiologies of the same disease. Of recent years many authors have spoken in favour of referring the disease to disorders of the endocrinous system. Quite apart from the fact that the internal secretion has had to bear the brunt so often in cases where we have no certain knowledge, I am not in the least inclined to advocate this theory but I admit openly that — having come to know the two different types of the disease — I am at a loss with regard to the etiology of the disease.

With regard to *treatment,* too, my two groups of the disease resemble each other in so far as they are equally refractory to drugs.

Dr. *Nic. Johannsen,* in the Acta pædiatrica (vol. II, page 362), has given a survey of the various lines of treatment suggested from time to time. I can supplement this with several others, e. g. injections of sulphur, injections of metal salts, thyroidin and other opotherapy, tonsillectomies, radium drinking courses, fibrolysin, cholin etc., but it holds good of all these therapeutic measures that, in the hands of the various authors, they have given very unstable and highly varying results. There-

fore I believe that for the time being we must maintain, that we do not know any certain *causal* therapy.

According to my experience the lines of treatment should be as follows:

So long as the joints are tender and painful and so long as febrile attacks continue to occur the child should be kept *quietly in bed*, care being taken only to keep the extremities in as favourable a position as possible in order to counteract contractures and luxations. If the temperatures are very high and if the child is much distressed owing to pain a *merely symptomatic* administration of aspirin, cibalgin and the like may be used.

Only when the disease is supposed to have subsided — which very often will last several years — should *massage, manual redressments, and kinetotherapy* be commenced; but the greatest care should be taken, as rough and brutal methods may do much harm, partly by causing recrudescences of the joint affections, partly by causing infractions or fractures. On the whole massage treatment demands much patience; it should be borne in mind that definitive restoration in many cases only occurs after several years of treatment. It will appear among other things from case record No. 5 that *active therapeutic exercise*, too, may be very beneficial.

As to *surgical interference* one ought to be very cautious, partly because the disease itself has a tendency to spontaneous recovery and partly because fractures may easily occur owing to the halisteresis, as it is seen for instance in one of *Hirschsprung*'s cases.

For the sake of completeness I add that the cachexia and anæmia which nearly always occur should be counteracted by roborant food, iron, arsenic and the like.