ON CASES DESCRIBED AS 'ACUTE RICKETS'

WHICH ARE PROBABLY

A COMBINATION OF SCURVY AND RICKETS

The Scurvy being an essential and the Rickets a variable element

BY

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This paper is a contribution to the study of a disease occurring in young children, of which several cases have been recorded in recent German and English literature, but of which hitherto, with one exception, no satisfactory account of the morbid anatomy has been given.

I propose (1) to narrate a typical case, (2) to give an analysis of principal symptoms in the recorded cases and in cases which have come under my own observation, (3) to give the results of post-mortem examinations, and (4) to discuss the etiology and affinities of the disease.

(1) Typical case.—A boy, aged fifteen months, was seen in the month of December when the following note describes his condition:—The child has an excessively pale, sallow complexion and is flabby, although he has a moderate covering of fat. There is not sign of nervous disease nor of visceral disease except that the liver is perhaps larger than normal, extending to two fingers' breadth below the thoracic margin as the body lies in the horizontal posture. The bowels have acted one or twice daily; a stool which has been saved is greyish-brown in colour and a little slimy. There has been no vomiting; the tongue is clear. He has cut his two lower incisors; the gums are natural with the exception of a minute erosion in the upper gum opposite the cutting edge of one of the lower teeth. The boy is continually moaning and when approached he screams and still more when he is touched. It is difficult to describe a cry, but it is sufficient to say that in this case it suggested deep-seated pain connected with bones and not brain disease. And it is clear that the dominant symptoms are related to the bones. The boy is rickety; there is some beading of ribs although the thorax is not grooved, his epiphyses are a little enlarged and he has only two teeth, but rickets is not sufficient alone to explain his condition. He lies on his back and scarcely moves the trunk though he frequently turns his head from side to side. Both radii are enlarged at the lower end but the right more so than the left, not only in circumference but in vertical measurement to a slight amount. It,
in fact, suggests a rickety enlargement plus slight thickening extending upwards for perhaps an inch. The child cries wherever he is touched, but the mother has noticed his right wrist notably more tender than the other for a day or two. The skin is pale in the neighbourhood and there is no special heat to be felt. The upper limbs are not bent. The left thigh is kept half flexed. Both the left thigh and leg are slightly swollen so that the contour of the limb is different from natural, assuming in the thigh rather a cylindrical shape. It is of the same colour as the other limb and does not feel hotter than any other part of the body. There is no fluctuation and no sign of effusion in the knee or ankle-joint. The child screams so much that one cannot examine thoroughly, but in spite of his thigh being flexed there does not seem reason to suspect hip-joint trouble.

The epiphyses at the knee and ankle are enlarged.

The right thigh is natural in the sense of there being no swelling. Besides the slight enlargement of epiphyses at the knee there is a tendency to knock-knee on the right side. There is no general swelling of the right leg, but there is a little thickening to be appreciated down the shaft of the right tibia.

There is profuse sweating about the head. There is nothing special about the head itself, except that there is a suspicion of slight thickening of the frontal in front of the fontanelle.

The boy’s temperature taken in the rectum is 101°F., 7 p.m.

The history was as follows:—He was the only child of his parents, a young couple in good circumstances, and living in a healthy dwelling. The father is in good health, but the mother is a thin, poorly developed woman, without, however, any history of particular ailments. The child had been born before term and had in its babyhood occasional stuffiness of the nose, but beyond this there was nothing to suggest congenital syphilis, though it is of course difficult to negative its possibility. During the first six weeks he was said to be a vigorous child. For that period he had his mother’s milk, then it entirely failed, and from that time until I saw him he had been quite deprived of fresh food. At first his diet consisted of Robinson’s grits and Swiss milk, then of baked flour, then of Nestlé’s food, then of Robb’s biscuits, then of Liebig’s extract, and finally of Swiss milk and saccharated lime water. He had been considered a fairly healthy child. He had not suffered from cough; his bowels had acted not more than twice daily, although the nurse admitted that the evacuations were often unduly offensive; he had had scarcely any vomiting. He had cut his first tooth at twelve months and his second at thirteen months, and these were all he had. His mother admitted that he had been always a pale child, and that although his limbs had been fat they had been thick near the joints, also that he had had much head sweating since he was three months old.

The child had been able to sit up well and stand with assistance at thirteen months old. Five weeks ago he ceased to do either, and then it was noticed that the left leg was swollen especially about the ankle. At this time also he became very peevish and would shriek if he were touched, and often even if he were approached. The boy was then taken to a well-known
bone-setter, who informed the parents that one of the bones of the spine
was out, and that an operation was needed to restore it to its proper position.
Five days later the said operation was performed, but as the swelling of the
left lower limb increased, and the right wrist became swollen and the right
hand dropped, and no explanation was forthcoming except the vague
suggestion of possible paralysis, a further opinion was desired, and the child's
condition was found as above described.

As to diagnosis, it was obvious that the child was the subject of rickets
to a moderate degree, and equally obvious that rickets alone was insufficient
to explain all his symptoms.

On the ground of certain post mortems which are subsequently detailed,
the opinion was formed that in this case there was under the periosteum of
the left femur and tibia an effusion of blood, and that the extreme tenseness
of the limb was due to blood extravasation in the deeper muscular layers
with the serum filtered out into the more superficial parts.

It seemed likely that there was also some slight blood effusion
immediately around the shaft of the right tibia and also in the neighbourhood
of the junction of shaft and lower epiphyses of the right radius. The view
held was that the boy was suffering from the supervention of scurvy on
rickets, albeit there was no trace of sponginess of gums. The treatment
suggested was to surround the whole of the left lower limb and the right
leg with wet compresses which had been thoroughly wrung out. These were
to be surrounded with dry cloths rather closely applied, and the compresses
were to be changed every two hours.

A complete change was made in the diet. He was ordered daily the
juice of a quarter pound of raw beef seasoned a little. He was also to take
a pint and a half of cow's milk daily, to which was to be added in his alternate
meals at one time a third part of strained gruel, and at another a third part of
barley water, and finally two teaspoonfuls of orange juice were to be given
daily. It was also ordered that for at least half an hour twice daily the
boy's crib should be placed near the fire, and that the window of his room
should be opened wide and the door likewise, so that he should have a free
play of fresh air around him, precautions being taken about close fitting
garments. The directions were most loyally carried out, and in three days
there was a notable change in the child. The compresses had certainly been
a comfort to the child, so that on the first night of their application he had
been a great deal quieter than before. He had taken the fresh food most
greedily, and it had agreed with him. His tongue was almost clean and a
healthy evacuation had been passed. His rectal temperature had sunk to
99·4°. The left lower limb was less tense and less tender, and the right leg
was better.

After this the improvement was progressive. It was several days before
any of his urine could be saved, then it was found pale, clear, neutral, free
from albumen or excess of phosphates. After about a fortnight the tenseness
of the soft parts of the left lower limb had disappeared, and then it was
easy to appreciate that the left femur was thicker than the right and in
less degree that the left tibia was thicker than the right. The
thickening gradually diminished, but even at the end of six weeks a slight difference could just be felt between the two femora. Before this time the swelling of the lower end of the right radius had also considerably subsided, so that in fact there was no difference between the two wrists. The compresses were omitted at the end of a fortnight from the date when he was first seen, and he began then to make efforts to raise himself and to move his limbs about. A little gentle shampooing of the lower limbs and back with oil, and likewise douches of first tepid and then cold sea water were commenced at the end of a month. No physic was given to him except at first a single, then two teaspoonfuls of cod-liver oil daily, and an occasional powder of rhubarb and soda. Within eight weeks from the date when first seen, the boy, whenever allowed, would get upon his knees and could stand with a little support; he was of a ruddy colour, and his skin and muscles had become quite firm. He was still not allowed to bear the weight of his body on account of the slight knock-knee on the right side, but he was carried out every day.

(2) Analysis of principal symptoms.

The material from which this analysis is made consists of thirty-one cases. Of these nineteen have been already published. The majority of these cases have been recorded in German medical literature under the title of 'Acute Rickets' (Möller, Bohn, Forster, Hirschsprung, Senator, Petrone, Fürst). Some account of the disease is also given by Steiner and Baginsky in their respective treatises. Individual cases are referred to by these authors, but the accounts of them are too brief to admit of incorporation in the accompanying table*. Stiebel's description of acute rickets, which differs somewhat from the clinical picture given in common by the other authors, is also not available for this analysis, because of the absence of cases reported in detail. One case, briefly recorded under the title of infantile scurvy, by Dr. Ingerslev, also belongs to our group, and is, indeed, quoted by Hirschsprung a propos of his own case.

Sir William Jenner in his lectures refers to the occasional abrupt or acute onset of rickets, but does not specifically describe any case like those in question. The first English case on record is in the 'Pathological Transactions' for 1876. It was under the care of Mr. Thomas Smith at the Hospital for Sick Children, and is described under the provisional title of haemorrhagic periostitis.

Dr. Gee, in the 'St. Bartholomew's Hospital Reports' for 1881, has given brief notes of five cases, which he classes under the head of osteal or periosteal cachexia, pointing out the existence of an obscure bone lesion and of a marasmic condition not explicable by rickets or congenital syphilis. It will be found that these cases come within the group now under consideration.

In the 'Lancet' for November, 1878, and again in July, 1882, Dr. W. B. Cheadle has described two cases of the same kind which came under his care, and has referred to others. Dr. Cheadle has given reasons absolutely conclusive, as far as his own examples are concerned (in which spongy gums

*This table has been omitted in this reproduction.
were a marked feature), for the doctrine that the disease in question is a combination of rickets and scurvy; and his papers are in every way the most important contribution to the clinical side of the subject.

A French writer (Montfalcon) in the article on 'Rickets' in the 'Dict. des Sciences Médicales' (1820), gives a short paragraph on the complication of scurvy with rickets. He narrates a case of a girl, aged ten years, who was certainly the subject of scurvy of the gums and skin ecchymoses, and who had some obscure joint (?) or bone (?) affections. It seems extremely probable that the latter were scorbutic also, and the case is of some value in support of the view subsequently maintained in the following paper, that the essential features of acute rickets are truly scorbutic; but it is to be noted that the writer of the article in question gives no reason for the view that his patient was the subject of rickets, and, indeed, throughout his article uses the term rickets in an extremely vague and indeterminate sense. Further, this patient was much older than those (chiefly infants) the reports of which have already been referred to, and for this reason is not suitable for the analysis.

Eleven cases have come under my own care, and one has been communicated to me by Mr. Shoppee, and these with the nineteen before published constitute the material from which the analysis is made.

The invasion of the disease may be gradual or abrupt. After a few days inexplicable fretfulness, or several weeks’ manifestations of pain, without obvious cause, when the limbs are touched, the child is rather suddenly taken off its legs, with more or less swelling of the lower limbs. In twenty-two cases, out of thirty-one the thighs have been attacked, and in the same number (many of them of course the same cases) the legs. In two other cases the lower limbs are spoken of as affected without definite description. In a typical case both lower limbs, though to an unequal degree, are swollen tense, and shining, the skin is generally pale, but may be reddish in colour, not generally hotter to the feel than the rest of the body, sometimes indeed colder. There is a varying amount of oedema, often more over the thigh than the leg or the foot. The tenderness on pressure is extreme; the child cries not only on movement but even on the approach of the nurse. In a severe case there is often continuous moaning as though the child were in constant pain. The child lies sometimes with one or both limbs flexed, but in the more severe cases with the limbs extended, or extended and everted in an immobile condition, which has been called pseudo-paralysis. In less severe cases, even at the outset, and in more severe cases after the partial subsidence of the subcutaneous swelling, it is not difficult to appreciate that there is a cylindrical swelling which envelops the shaft of the femur to a varying degree. The longitudinal extent of this swelling may vary, it may be very slight and be confined to the neighbourhood of the junction region between the shaft and epiphysis, or it may surround the whole shaft. In the most severe cases of all, of which I have seen two examples, soft crepitus was to be obtained just below the hip-joint. This no doubt is a late and very severe phenomenon, but from considerations afterwards to be referred to, I think it probable that it may occur not
unfrequently, and owing to the excessive tenderness of the patient, which interferes with careful examination, may escape detection. In Dr. Fürst's case, after the conclusion of the illness, a sharp angular bending with some thickening was found in both femora though the exact site is not indicated. Nevertheless, the ordinary sequence of events with respect to the thigh is, first swelling of the whole of that part of the limb, then subsidence of the affection of the soft part, allowing the thickening around the bone to be appreciable, then very gradual subsidence of the bone thickening, leaving the limb without deformity. In one of Möller's cases and in Förster's case after the illness a marked increase of the length and growth of the limbs was noticed. This is, however, by no means a constant occurrence.

The affection of the legs was occasionally present without involvement of thighs, but whether it existed alone or in combination with the thigh affection, it was in all the cases in my experience less severe than the thigh affection. Here, also, it may be said, in general terms, that the thickening radiates from the junction areas of shaft and epiphysis. The fibula is always much less affected than the tibia. In one of Möller's cases there was still a little thickening of the bones of the legs when the child died of atrophy, and in Hirschsprung's case there remained at the end of the illness a little "thickening of one ankle." With respect to the upper extremity in two of the cases already referred to, there was soft crepitus obtainable below the knee. Probably also in another case there had been some separation in this junction region, for at the end of the illness the shaft of the tibia was found displaced backwards to a slight extent just below the upper epiphysis. With regard to the leg as with regard to the thigh, the rule is for ultimate resolution to take place without deformity.

Other bones may become affected either simultaneously or more commonly by subsequent involvement. In one of Dr. Gee's cases there was some swelling over both scapulae. The same condition was observed in one of mine, the swelling being much more marked in the right than the left, and affecting especially the infraspinous fossa.

The humerus was affected in nine cases, and the radius and ulna, either separately or conjointly, in twelve. In one case, there was soft crepitus obtainable near the upper end of the humerus on both sides.

In general, it may be stated, with respect to the upper limbs, that the swelling of the soft parts was much less and the thickening along the shafts much more limited in extent. In the majority of cases, indeed, the swelling was limited to the epiphysial region; and it may reasonably be asked how was this to be distinguished clinically from ordinary rickets enlargement of the wrist for example? To this it may be replied, as in the type case, that probably some of the swelling is truly rickets but that there is something over and above the rickety swelling. (1) The appearance of the two wrists is often unlike; the enlargement above one epiphysis, being greater considerably than above the corresponding one on the opposite side; (2) the extreme tenderness is not present in simple rickets; (3) the pseudo-paralysis which often obtains in these cases does not occur in simple rickets; (4) the marked
subsidence in a short time as in the type case is quite unlike the slow retrogression of an ordinary rickety enlargement.

With regard to the cranium, morbid conditions were found either during life or post mortem in eight cases out of thirty-one. The exact amount which belonged to the illness in question is difficult in all cases to assign. Setting aside for the present the marked cranial bosses (Parrot's swellings) which were found, and also the slight thickening near the sutures, it seems probable that some slight swellings on the parietals, which were found post mortem to be due to sub-periosteal haemorrhage, belonged strictly to the illness in question.

Further, there appeared during the illness some swelling of both upper and lower jaws, and some thickening of the zygomatic regions occurred, and likewise considerable tenderness of the occiput followed by some thickening.

Let us consider now the other structures involved in movement. With respect to the joints generally of the upper and lower limbs, although the phrase 'painful joint affection' is more than once employed, there is nothing in any of the reported cases to lead us to believe that there was effusion into the synovial cavities. Senator, Fürst, and others, are most careful in their descriptions to exclude joint effusion. There was certainly no evidence of it in any of my cases.

The muscles generally in the severe cases were more or less wasted. It is difficult to estimate how much of this was due to antecedent rickets, but it is to be observed that several of the children had been in good condition before the illness began. In several the weakness of the back was most remarkable. No lesion of the bones was detected, but the prostration was often quite absolute. In Hirschsprung's case for a time there was great tenderness over the neck.

With respect to the skin and subcutaneous tissue, the anaemia in the severe cases was profound; besides the pallor there was noted sometimes a peculiar sallow, muddy tint in the complexion.

In one of Möller's cases sugillations appeared on one leg, and Hirschsprung says of his that 'in several places the skin was tinged bluish red.'

In one of Dr. Gee's there was an appearance over the sternum like that produced by a 'bruise.'

In one of Dr. Cheadle's there were unhealthy sores on the wrist and finger, and it was stated that a blow on the thigh had left 'a swelling for a considerable time.'

In one of my cases extensive purpura appeared before the bone condition became manifest.

In one of Dr. Gee's and in one of my cases there was ecchymosis into the eyelids, and in the latter case also there was for a few days slight proptosis of one eye. Some conjunctival haemorrhage occurred in Mr. Shoppee's case.
The tenseness of the lower limbs is in the very severe cases very considerable, but it is remarkable how completely as a rule the swelling passes away. In one of my cases there was for a week or more a small, soft, fluctuating area just above the junction of the lower epiphysis of the femur with the shaft on the outer side of the thigh. This also underwent spontaneous absorption. In another case, which I believed to belong to the group, the affection was confined to one thigh. There appeared considerable swelling, which at one place on the outer side of the middle of the thigh gave at length some fluctuation. This was opened by my colleague Mr. Morgan, a little pus escaped, and the small diffuse abscess cavity very rapidly healed, leaving, however, considerable deep-seated thickening and (which could be appreciated after a time) marked thickening around the shaft of the femur. As this case is so exceptional in regard to suppuration, I have thought it wiser not to incorporate it with the others, the more so that as the other limbs were not involved there was no opportunity of comparing their progress to both typical cases, and so render its identity certain. It deserves further investigation whether the subcutaneous swelling may not occasionally in severe cases of the so-called acute rickets undergo a partial suppuration.

In almost every case very severe head sweating is referred to, but in several this symptom had existed from a very early period, and it may be questioned whether it was special to the illness under consideration, although a very striking feature.

Here may be conveniently considered the subject of the body temperature in these cases, and some details must be given because in the reports there is some divergence of results. Senator in discussing acute rickets insists that pyrexia is a constant accompaniment, but this is certainly too sweeping a statement.

The case which best supports this view is that of Dr. Fürst. Observations made every three or four days during a month, recorded some pyrexia lasting through that period. At the onset of the illness the evening temperature was 102.7°, and this was the highest recorded. Within the next five days 101.8° and 101.5° were registered, the first an evening the second a morning temperature. After this no temperature higher than 100.4° was recorded, and at the end of the month it was 99.8°. It is important to bear in mind that in Dr. Fürst’s case during the first eleven days, there was progressive involvement of different limbs, viz. first the left thigh and leg and right leg, then the right forearm, then the right arm, then the left arm and forearm, and that even so late as the twenty-fourth day there was some infiltration and tension about the left arm. After the end of a month no further observations were taken as the child was improving. Six weeks after this a relapse occurred which affected both zygomatic regions and the right thigh—that thigh not having been previously attacked. The temperature rose to 102°, but in a fortnight had become normal again.

Petrone gives the temperature of his case at the outset as 102.9°, but does not mention what it was subsequently. Hirschsprung states that in his case the illness commenced with febrile symptoms, and that there
ON CASES DESCRIBED AS ‘ACUTE RICKETS’ 231

was marked but irregular pyrexia. In his case also there was successive involvement of left shin and foot, then of left forearm and head of humerus, then of right foot, then of upper, then of lower jaw, which afterwards relapsed.

In Thomas Smith’s case the temperature was observed twice daily during the six days that the child was in hospital. The highest temperature was 101.4°. On another occasion it was 101.2° and at other times it ranged between 97.4° and 100.6°.

This case was very severe in regard to bone lesions, but I do not think there was proof of any fresh start of bone-mischief during the week that she was in hospital. Furthermore, the child was suffering from some bronchial catarrh, and post mortem a small patch of consolidation was found.

In the type case I have mentioned that the rectal temperature when I first saw the child was 101°. This was probably the twenty-sixth day of his illness. An intelligent nurse told me that there had been some fever for several days.

Three days later the rectal temperature was 99.4°, and subsequently taken every day was scarcely ever above 99°. There was no reason to suppose that any fresh bone-mischief supervened after the twenty-sixth day. In one of my cases, twenty-two days after the onset, the axillary temperature was 99.8°, and in another, ten days after the onset, rectal temperature was 99.6°. In several others, although the temperature was not taken, it was noted that the skin was cool. Bohn states that in his case there was no fever, but the child was only brought to him one month after the onset.

In some of Möller’s cases there was for a time a little local heat, but he states that there was no fever. In another case the temperature was not elevated at the onset, but at a later period the child was febrile, with some catarrhal conditions. In Dr. Cheadle’s second case the temperature when first seen was 98°, and 99° is the highest degree recorded. In Dr. Cheadle’s first case, during the six weeks that the child was in hospital the axillary temperature was either normal or subnormal, except on the day after admission, when it was 99.5°. When I saw this child in the out-patient room before his admission into the ward, his rectal temperature was 103°. So far as the history could be relied on this was between two and three months after the onset of the illness. In both these cases of Dr. Cheadle’s it is important to note that no new manifestations of bone affection appeared after they came under care, and a definite line of treatment was promptly adopted by him.

To sum up these somewhat divergent results, it is clear that Senator’s statement that pyrexia is a constant accompaniment of the disease cannot be accepted. It is not considerable in amount, nor does it show any special hectic character, and for long periods it may be absent. A careful examination will, I think, leave little doubt that, setting aside some cases of intercurrent catarrhs, the pyrexia, when present, is related to the bone affection, and that, not so much in regard to extent as to the amount of
tension present. Probably, with a fresh involvement of bone, if much stretching of periosteum occurs, there is a little fever, at all events for a short time.

Of the affections of the mucous membranes the most important is that of the gums. In fifteen out of the thirty-one cases there was some morbid condition present. In at least four of these there was sponginess with a tendency to bleed, and some putrid odour. Nine showed varying degrees of mouth affection, from slight swelling confined to the neighbourhood of newly-cut teeth up to general swelling of both gums, and in one case of the lower lip also.

As to the date of appearance of the stomatitis in relation to the swelling of the limbs, in some cases it was antecedent to the limb affection in others it occurred after the limb affection was well established, and in others it was probably simultaneous.

In one of Dr. Gee's cases, and in one of mine, the gums were not swollen, but there were small localized ecchymoses beneath the gum in the situations of the on-coming teeth.

It is very important to note that in six cases it is specially stated that there was no stomatitis. It is almost absolutely certain that in a great many of the other cases stomatitis, or, at least, sponginess, was conspicuously absent; for the reports are given by authors who were fully acquainted with the occasional occurrence of this symptom in this group of cases (Senator, Möller, and others), and it is reasonable to believe that the frequent absence of sponginess of gums is the cause of the scorbutic hypothesis often being dismissed, or not entertained, or regarded as unimportant.

The details with respect to disturbances of the digestive tract are not very complete. In two cases the bone affection appeared to start after a severe attack of diarrhoea. On the other hand, in a great many of the cases the intestinal condition was considered healthy at the time of onset of the acute symptoms, and there was no notable disturbance throughout.

In the ninth week of the illness in one of Möller's cases, severe diarrhoea with some bronchial catarrh occurred. In Hirschprung's case there was for a time obstinate vomiting. This is very exceptional. In the majority of cases the appetite was maintained. In two of my cases children of four years and two years respectively, there was present the greatest antipathy to meat and vegetables, and one of Dr. Cheadle's patients, a boy aged thirteen months, had very great dislike to gravy and potatoes. But in these cases this dislike was long antecedent to the illness in question.

There is very little noted with respect to the liver. Slight enlargement of the spleen and lymphatic glands occurred in Petrone's case. The former I believe, is exceptional, for in several other cases, absence of enlargement of this viscus is specially noted. In one only of my cases was the spleen palpably enlarged in a child in whom there was some suspicion
of congenital syphilis. The spleen continued larger than normal after the bone affection had subsided, and possibly had existed previously.

The accounts of the urine are somewhat scanty. In one of Dr. Gee’s cases there was haematuria for a short time, and this was also noted in Mr. Shoppee’s case. In Dr. Cheadle’s two cases there was for a few days a definite trace of albumin. In one of Möller’s cases and in Mr. Shoppee’s, for a time there was a considerable deposit of uric acid. Both Bohn and Hirschsprung specially note that in their respective cases the urine was normal, being free from albumin or excess of phosphates, and this also was the case in the type case given at the outset of this paper.

There is nothing important to note regarding the heart and lungs.

With respect to the nervous system, extreme fretfulness must be mentioned as a symptom so striking and constant that everybody who has recorded cases has dwelt upon it as something quite remarkable in this affection. In fact this is one of the five constituents, viz. pallor, wasting, immobility, swelling of limbs, and fretfulness, which, with or without swelling of gums, pretty nearly sum up the clinical whole of the so-called acute rickets. This special fretfulness is almost certainly related to the affection of the limbs.

The laryngismus stridulus and fits which occurred during the illness in Dr. Fürst’s case, and the tetany and laryngismus which occurred a short time before the onset of the bone symptoms in one of mine, belonged doubtless to the severe rickety diathesis which both these children presented. No manifestation of this kind occurred in the case of children who had been previously healthy.

There remains to be considered, in this section, the duration of the disease.

Slight differences of reckoning depend on whether the end of the illness is considered to correspond with the entire or partial disappearance of bone thickening, the child being well in other respects. Excluding the comparatively mild cases, there are the rather wide limits for the duration of one month and six months. The duration in the greatest number of cases is between two, and three and a half months.

It will be seen from the table that the disease is a very fatal one; no less than seven out of thirty-one died. It is equally remarkable to note the slow but gradual, and apparently spontaneous, tendency to complete recovery which occurred in others, even when the cachexia had been very profound.

Two of the cases, viz. Bohn’s and Mr. Thos. Smith’s, appeared, from the history, to have passed through similar attacks many months previously.

(3) **Post-mortem appearances.**

Although the German physicians have given such careful accounts of the symptomatology of this disease under the designation of acute rickets, they have no description of the morbid anatomy to offer, and this accounts for their very vague and unsatisfactory explanation of its pathology.
Professor Möller in his second paper describes two cases. One of these is fairly typical. The other was a marasmic child aged fifteen months, who was brought with exophthalmos and sanious discharge from the nostrils, and who died from exhaustion. On post-mortem examination an extensive haematoma was found under the frontal and anterior parts of the parietals, extending down to the orbital plates of the frontal and to the ethmoid and into the orbits. The blood was believed to be derived from the inner surface of the bones of the skull in which no thickening was found, but the vascular canals of the inner surface of the frontal were considered to be wide and the bone itself somewhat porous. No affection of the limb bones either during life or post-mortem was found. The case can therefore in no sense be regarded as agreeing with the typical examples of so-called acute rickets, of which the foregoing analysis has been given, albeit I am not prepared to say that it has no alliance with them.

It must in justice be stated that Förster made the shrewd guess with regard to the affection of the limbs in typical cases, that the essential condition was a sub-periosteal haemorrhage; and Möller, in some parts of his paper, seems to have come very near to that view, but in his summing up he speaks in a vague way of 'acute rickets as being solely a developmental disease in which there is a precipitate and tumultuous bone growth, which at times destroys the organism, killing through exhaustion, but most often is overcome leaving behind it a marked increase in the length of the limbs.'

A much simpler doctrine will I believe be elicited from the study of the clinical phenomena in the light of post-mortem examination.

I have made three autopsies on cases of the disease under consideration. The first was on a child under the care of Mr. Thos. Smith at the Hospital for Sick Children in February, 1875. Mr. Smith has recorded the case in the 'Pathological Transactions,' Vol. xxvii, p. 219, under the title of 'Haemorrhagic Periostitis of the Shafts of several of the Long Bones with Separation of the Epiphyses.'

Although many of the points in the clinical history of this and the two following fatal cases have been embodied in the foregoing part of the paper, yet in order to show the correspondence of certain morbid appearances with the clinical picture of the disease it is necessary that details of the history should be given with each fatal case, even at the expense of repetition.

Mr. Smith's case was a female child aged 1 year 11 months. She had been suckled for three months and subsequently fed on cow's milk and corn flour. There was no reason to think from the history that she had had congenital syphilis; but her rickets had been shown in her not cutting a tooth till she was twelve months old.

When thirteen months old, in April, 1874, her feet, legs, and thighs gradually became swollen and excessively tender, and, according to the mother's statement, 'hung down cold and dead.' This continued so for two months, and then there was slow improvement, so that by summer the swelling and tenderness had gone and the child was considered well, but for diarrhoea to which she had been subject more or less since six months old.
ON CASES DESCRIBED AS 'ACUTE RICKETS'

At Christmas, 1874, the child being then twenty months old, the swelling appeared again in both lower limbs, first in the feet and legs then in the thighs. After a month the swelling of the feet had considerably diminished but that of the thighs had increased. She had sweated a great deal. The bowels were only opened twice or thrice daily and the motions were not offensive.

When admitted on February 26, 1875, that is, two months after the beginning of this second illness, she was an extremely pale and cachectic child. She was rickety with a large fontanelle and beaded ribs. She lay on her back with her lower limbs flaccid, never raising them. There was some oedema, more in the thighs than the legs, and scarcely any in the feet. The skin was natural in colour and not hot to the touch. There was some prominence in the region of the trochanters, the thighs projecting outwards there, more on the right than on the left side. On lifting up the limbs distinct softish crepitus was obtained without the slightest difficulty just below the hip-joint and knee-joint on either side. It was evidently produced below the epiphyses. The hip-joints and knee-joints seemed natural. There was no oedema elsewhere.

I regret not to have recorded a note at the time about the upper limbs. But a note taken a few days afterwards in the post-mortem room about them may be here intercalated, to the effect that there was crepitus obtainable below the head of each humerus but none in connection with the elbows or wrists. None of the epiphyses could be considered very large.

The child coughed a little and there was abundant sharp râle over both backs with some doubtful resonance at the bases. There was nothing abnormal to be detected in the heart or abdominal cavity. The urine could not be saved.

During the week she was in hospital the child lay on her back and whimpered a great deal, and continued extremely marasmic. She died rather suddenly, the cause being not perfectly obvious, but she was very feeble, and had, moreover, a considerable amount of bronchitis. Her temperature in the axilla had been as follows:

February 26.—Evening 101.6°.
27.—M. 100.6°. E. 100.6°.
28.—M. 98°. E. 100°.
March 1.—M. 97.4°. E. 101.2°.
2.—M. 99.6°. E. 98.4°.
3.—M. 100°. E. 98°.
4.—M. 99°.

At the post-mortem examination the lower limbs were first examined.

On the left side the glutaeus maximus was found rather pale, but nothing else was noticed abnormal about it, or about the muscles attached round the head of the femur.

The vastus externus was swollen, pale, and pulpy. A little yellow serum bulged out of the upper part, and on cutting into the muscle some blood was found extravasated into its substance, and the same character applied to the
vastus internus and crureus. The blood was pretty uniformly diffused through the deeper layers, and no coarse laceration was seen.

On making an incision down to the shaft of the femur, the periosteum was found separated from the shaft entirely in a continuous sheet. It was about \( \frac{1}{4} \) inch thick, and was intensely injected all over the inner surface.

The shaft of the femur was separated from the epiphyses, and was almost entirely surrounded by a layer of maroon coloured blood clot, \( \frac{1}{2} \) to \( \frac{3}{4} \) inch thick, which loosely adhered to it, and in fact separated the shaft completely from the periosteum. On removing a little of this clot, the surface of the shaft of the femur was seen to be perfectly smooth. The ends of the shaft were not splintered, but had a 'sugary' surface. There was no lymph or suppuration anywhere.

The abruptness with which the changes were confined to the shaft was very striking.

The hip-joint was natural, and so were the lower epiphysis and knee-joint.

There was no extravasation of blood in the muscles of the leg.

The changes round the tibia were not so extensive as round the femur, but they were of the same character. The periosteum was thickened, vascular, and separated from the upper and lower thirds of the shaft by a thin layer of blood.

In the middle third there was no blood; the periosteum was adherent to the bone, but easily stripped up. The shaft did not, as the femur, lie absolutely separated from the epiphyses, but it was loosened at the upper end.

The left fibula at its upper extremity was natural: there was, however, separation of periosteum and effusion of blood to a slight extent at the junction of the lower epiphysis with the shaft. The ankle-joint was natural.

The right femur presented almost identical appearances with the left. There was a minute spot of blood in the floor of the acetabulum, but nothing else abnormal in the hip-joint.

The right tibia and fibula also presented almost identical characters with the corresponding bones of the left side.

The upper limbs were not examined beyond the ascertaining of the crepitus below the head of each humerus.

The abdominal organs were natural, and so was the heart.

There was some collapse in both lungs, and in part of the lower lobe of the left a small wedge-shaped patch of consolidation, very like a past pulmonary anoxemia, over which was a little velvety lymph.

As in this case, I regret to say I was responsible for the suggestion of the term haemorrhagic periostitis, there can be no impropriety in my now criticizing it.

It was a provisional term employed to designate what seemed a remarkable and exceptional condition. but it would no doubt have been better to have described it as simply sub-periosteal haemorrhage.

For there was, indeed, as is fully stated in the report, no proof of any true inflammatory process, and the appearances were, to a great extent,
explainable by a primary haemorrhage from the periosteum, the cause of that haemorrhage, whether from damaged nutrition of capillaries, altered blood state, or both, being still an open question.

The second post mortem was made on a female child, L. S., act. ten months, who was brought to me as an out-patient in October, 1881.

She had never had breast milk, but had been fed first on condensed milk, then on cow's milk, and then on various foods; at the time when she was brought to the hospital she was taking Anglo-Swiss food. A teaspoonful of cod-liver oil had also been given to her thrice daily for two months. Careful inquiry failed to elicit anything in favour of syphilis, and subsequent examination of the other children was negative in this respect, although they were found extremely rickety. The mother was a delicate woman, but the father was healthy. The house seemed to be wholesome. This child had had much head sweating since she was three months old. Bowels had been constipated until two months ago, when she had a severe attack of diarrhoea, and after this her legs were noticed to be very tender. Three weeks ago her wrists also became very tender. When brought to the hospital she was extremely fretful. She not only screamed directly she was approached, and still more when she was examined, but continually whimpered as though in constant pain. Her axillary temperature was 99·8°. Her skin generally was pale to the last degree. There was ecchymoses in both upper eyelids; also underneath the mucous membrane of the gums in the lower median incisor regions, and also in the lower molar regions separate ecchymoses were present. The child had not cut any teeth. The lower end of each radius was much enlarged, and the left hand hung prone in a condition of pseudo-paralysis. The left thigh was strongly flexed. There was some deep thickening to be felt along the lower third of the shaft of the left femur. The epiphyses of the lower limbs were a little enlarged. She lay on her back and never attempted to move. She was so ill that a complete examination could not be made, but it was ascertained that the liver and spleen were not enlarged. It was not expected that she would live, but the mother was ordered to give her as much raw meat juice as possible, and to continue the cod-liver oil. In a week's time she was brought again, and her condition was not worse, with the exception that there was slight proptosis of the left eyeball, as though there might have been some extravasation into the areolar tissue of the orbit. Eleven days afterwards this had subsided, but the child was paler and feeble, and she died October 29, i.e. three weeks after she had been first seen, the total duration of her illness being about three months.

The post-mortem was made with considerable restrictions. Body wasted.

Cranium.—On the external surface of each parietal, just behind the fontanelle, there was found an area of thin sub-periosteal haemorrhage, about the size of a shilling. The bone underneath it was slightly porous.

Thorax.—The intercostal muscles and some of the other muscles on the thoracic wall, especially the left serratus magnus, of a pale yellowish colour and slightly pulpous consistence as through infiltrated with serum. The periosteum of the ribs extensively detached, thickened, rather vascular, and
slightly granular on the surface towards the rib, from which it was separated by a considerable quantity of flaky, chocolate coloured débris. There was no lymph or pus, and the flaky débris was more like disintegrated blood-clot than anything else. The ribs were extensively bare and white and slightly rough. They were distinctly wasted. What had been taken for 'beads' of the ribs were simply the extremities of the costal cartilages. The bony part of the ribs was much wasted especially along the anterior surface. Thus the anterior extremity of the rib was by no means in complete apposition with the whole of the extremity of the costal cartilage. There was no beading on the inner surface. The ribs were extremely brittle. They contained only thin soft medulla, and when this had escaped the rib was a mere shell. It was a wonder that the ribs had not separated at their junctions with the costal cartilages, or that some of them were not fractured beyond. They could be snapped in two easily. On the parietal pleura of both sides there were numerous petechiae along lines corresponding with the ribs. There was a quantity of blood-stained serum in the left pleura but no lymph. In the middle of the left lung there were two or three very small masses of caseous tubercle and a few gray granulations on the surface in this neighbourhood. There was no disease of the right lung or pleura, and the bronchial glands were healthy. There was no disease of the other viscera.

We were not allowed to examine the lower limbs completely, but some extravasation was found partly into the periosteum near the left crista ili and the deeper part of the muscles attached there, whilst the superficial parts were pale and slightly pulpy. There was also some subperiosteal haemorrhage in the region of the junction of the upper epiphysis of the femur with its shaft.

The third post mortem was also made on a female child, D. L., who was an out-patient of my colleague's, Dr. David Lees, to whom I am indebted for ultimately transferring the case to me.

The child was not regarded as syphilitic by Dr. Lees or myself, but the following points of family history ought to be stated. The first child had been born dead at seven months. The second died aged four months, cause unknown; the third was a full-time healthy child; the fourth was full time and suffered much from spasmodic croup and fits.

The mother stated that she was in good health during pregnancy with the child now in question, who was the fifth. The child was full time, is said to have snuffled occasionally but had no rash. She was suckled exclusively for seven months and not finally weaned till eleven months old. From seven months old, in addition to her mother's milk, she had cow's milk and two teaspoonfuls of Chapman's entire wheaten flour in the milk thrice daily; she never had condensed milk. At the time when the limbs became bad, i.e. at seventeen months old, the mother states that she was giving her the best part of a breakfast-cupful of beef tea fresh-made every day with bread and farinaceous puddings and a little milk, not more than half a pint. No vegetables whatever were given to her. The mother affirms that the child had six teeth at six months. She was, however a weakly child from four
ON CASES DESCRIBED AS 'ACUTE RICKETS'

months onwards. At nine months old she began to suffer from laryngismus, and she was first brought to the hospital on this account when fifteen months old. She was decidedly rickety, had frequent diarrhoea, and when about seventeen months old began to be very tender, especially about the lower limbs.

When she was admitted under my care, December 9, 1881, she had had this extreme tenderness for two months, gradually becoming worse, so that she could not be touched anywhere except above the upper limbs without screaming. She was anaemic and flabby but not wasted, she sweated about the head considerably. The fontanelle measured 2 inches by 1; there was marked beading of the ribs, and the epiphyses of the upper limbs, especially at the wrists, were decidedly enlarged. There was some swelling over each scapula evidently belonging to the bone. On the right side it formed a low rounded tumour over the infraspinous fossa. Both were extremely tender to touch; there was no alteration of the overlying skin. Both thighs were swollen evidently from affection of the bone. The child was too sensitive to be examined thoroughly, but soft crepitus was obtainable above and below both knees. The legs were also tender and swollen, less so than the thighs. The child was ordered beef-juice, mashed potatoes, and one orange daily. On the fourth day after admission she developed a clangy cough, and on the fifth a typical rash of measles, from the supetration of which disease she died on the sixth day.

At the post mortem, examination of the limbs gave the following result:—There was considerable effusion of thin pale yellow serum into the substance of the muscles of the thigh, rendering the superficial layers pulpy. In the deeper layers there was tolerably uniformly disseminated blood-clot. The periosteum was thickened and, except at the upper extremity, separated from the shaft of the bone. Surrounding the shaft, and in some places bridging over the space between it and the displaced periosteum, was a thick sheath of blood-clot. There was no fluid contents in the periosteal sac nor was there any lymph or caseous material.

Along a line about $\frac{1}{2}$ inch. above the junction line of shaft and lower epiphysis the shaft was separated. The separation had taken place through the loose imperfectly ossified material at the end of the shaft. The opposing surfaces were rough but not splintered. There was no callus. The ossifying centre of the lower epiphysis was a great deal of it diffuent. It was larger than natural, in fact, of the diffuse form which Mr. Sutton has recently pointed out as characterizing the ossifying centre of rickety epiphyses. There was no cushion of cartilaginous material, such as is present in the early stage of rickets between the epiphysis and the shaft, but this, I take it, had been replaced by the loose imperfectly ossified material through which the fracture had taken place.

The muscles below the knee also showed the result of extensive blood extravasation in the deeper layers. The periosteum of the right tibia was thickened and vascular; there was blood extravasation between it and the shaft for the whole length, but it was greater in amount at the extremities than in the middle. There was fracture through the loose, imperfectly
ossified, brittle material, about half an inch below the line of junction with the upper epiphysis. The ossifying centre of the upper epiphysis was diffuse, red, and very soft. There was no separation of the lower epiphysis from the shaft. The medulla of the shaft was very soft and red, and the trabecular structure of the shaft broke down very readily; there was no trace of suppuration or caseous material.

The periostea of the right fibula was not actually detached from the shaft but it was very vascular, and there was slight extravasation between it and the bone, especially near the extremities.

The femur and tibia of the left side corresponded closely with those of the right side except that there was less extravasation.

The right scapula had a firm layer of blood-clot on both the ventral aspect and on the infraspinous fossa. That situated on the ventral aspect had led to the complete stripping of the periostea, and the osteogenic power of the periostea was shown by the formation of a thin lamina of osseous material over part of the subjacent blood-clot.

The clot on the infraspinous fossae was also nearly ½ in. thick, but I failed to detect any earthy material in the stripped up periostea.

The right humerus did not present any periostea lesions similar to those described above. It showed many of the features of rickets passing from the first to the second stage.

It is true the typical proliferated cartilaginous zone was no longer obvious as such, but its place was represented by a buff-coloured layer of imperfectly ossified material, and below this there was some loose trabecular bone.

The medulla was very red and soft and the trabecular structure of the shaft loose and scanty. There was no deformity of the shaft.

The radius and ulna were free from extravasation, and presented on section rickety characters very like those of the humerus.

The lumbar vertebrae presented on section a marrow which was unduly red and soft and a trabecular structure which was very easily broken down. The ribs presented characteristic beads which were undergoing partial ossification.

The cranium did not present any bosses, but close to the medio-frontal suture and in front of the fontanelle was slight thickening, which was evidently old.

Some microscopic sections of the shaft of the femur, kindly made for me by Dr. Money, show the periostea vascular and thickened, but I think without cellular infiltration; extensive haemorrhage in the deeper portions and also between the periostea and the bone; considerable absorption of the trabecular structure with large spaces showing in places slightly eroded margins; at the upper extremity rickety ossification.

There is a little to state about the viscera. The lungs showed many patches of collapse and some commencing lobular pneumonia. The bronchial glands were a little enlarged. The liver and heart were healthy. The spleen weighed nine drachms and was firm to the feel. There were some small, flat extravasations of blood under the capsule, and also some patches with a
ON CASES DESCRIBED AS ‘ACUTE RICKETS’ 241

superficial area about the size of a shilling, of extravasation in the substance of the spleen, which, however, appeared to have caused but little laceration. The kidneys and intestines and peritoneum were natural.

We may now sum up the morbid appearances as far as these three cases are concerned, and add these to the general synopsis of symptoms.

Lower limbs.—Muscles: serum in the upper layer, which, probably partly as the result, are pale and slightly pulpy. Deeper layers contain extensively disseminated blood-clot.

Periosteum of femur and tibia thickened, vascular, separated from affected bones in great measure by sheath of blood-clot.

Fracture through loose trabecular structure at extremities (one or both) of shafts of femur and tibia. No callus. The two bony surfaces rough but not splintered.

I would suggest that the subperiosteal blood extravasation is the first event and the fracture the second. The extensive blood extravasation probably interferes with the nutrition of the bone, and thus the very minimum of violence, such as an ordinary movement, may lead to fracture.

The medulla of the shafts soft and red, and the trabecular structure scanty and friable.

Upper limbs.—Extensive blood extravasation under periosteum of both dorsal and ventral surfaces of scapula.

Unfortunately, with respect to the long bones, no complete post-mortem evidence is yet obtained, but there can be little doubt that in some of the cases that recovered there existed for a time a haemorrhagic extravasation in the neighbourhood of the junction of shafts with epiphyses, especially near the wrists.

Ribs.—Extensive separation of periosteum, probably by blood-clot. Ribs wasted, bare, and brittle, with very thin red medulla. Ribs very readily separable from junctions with costal cartilages.

Cranium.—In one case subperiosteal haemorrhages in position where Parrot’s bosses are often found, and inasmuch as the scapula in one case showed new bone formed in the upraised periosteum, it seems possible that bone might also be formed over the subperiosteal haemorrhages on the cranium and give rise to a condition indistinguishable from a cranial boss.

Visceral changes.—Blood-stained serous pleural effusion and petechiae along the parietal pleurae in the second case; haemorrhage under the capsule and into the substance of the spleen in the third, and the small haemorrhagic focus in one lung in the first, though perhaps not of great importance, are interesting from their association with the subperiosteal haemorrhage.

(4) Etiology.

We must now ask what is the etiology of the disease under consideration, how is it to be distinguished from other diseases with which it has points of resemblance, and with what disease known to us has it the closest affinity.

Approximate answers to these questions will be best obtained by reviewing some of the conditions under which the symptoms arise.
ARCHIVES OF DISEASE IN CHILDHOOD

As to age: My earliest case occurred in a child of five months, but Senator has described one in a child aged four months, and Steiner says the disease may occur as early as the fourth month. Of the thirty-one cases analyzed twenty-six belonged to the first two years of life, and five to the second two years of life. Of the twenty-six cases belonging to the first two years, half belonged to the first year and half to the second. Dividing into periods of six months, the greatest number of cases occurred between six months and eighteen months of age, so that as far as our numbers help us the disease would appear to be pre-eminently a disease of the second part of infancy.

The sex is not stated in all the reported cases, but from the data forthcoming there appears to have been twelve males and eleven females, so that sex cannot be considered of any importance.

With respect to period of the year in which the symptoms become manifest,—Hirschsprung asserts that the disease always occurs in the winter months. This is much too sweeping a statement. Out of twenty-eight of which accurate data are given, seventeen occurred in the colder six months and eleven in the warmer six months. Probably a greater preponderance would appear if we had the dates of all the cases.

Although most of the cases belonged to the poor, there were several quite typical amongst children of those who were well to do, and there is nothing of importance to be elicited about the dwellings of the patients, which in some instances at all events were perfectly satisfactory.

It will be convenient to discuss the important question of diet at a later period, but possible hereditary causes may now be considered.

In several the mother was delicate, and in some of the cases other children were rickety in the ordinary sense, but in others the parents were healthy and the other children healthy. It does not appear from any of the histories that any other member of the family in any given case had suffered from the symptoms of so-called acute rickets, at least there is no statement to the effect that more than one member of the family suffered in this way, which is sufficient if not conclusive.

In regard to hereditary syphilis it must be admitted that it is difficult to prove a negative because the so-called acute rickets rarely develops during the period when the early indubitable syphilitic signs are present.

Steiner states that of the ten cases seen by him acute rickets supervened on congenital syphilis in two children aged four months, and that these two children soon died. As no post-mortem account is given of these cases it is open to us to ask whether the disease from which they suffered may not have been the congenital syphilitic affection of the ends of the shafts of the long bones which has been described by Wegner, Parrot, and others.

For, truth to say, this disease has some clinical features not unlike those which I have described as belonging to the so-called acute rickets, and it is desirable here to refer to the similarities and differences between the two.

The junction area between the shaft and epiphyses is specially affected in both, and in the syphilitic affection there may be some accompanying
perichondritis and periostitis, which latter causes a swelling for a varying
distance up the shaft. Also in the syphilitic disease there may be, as I have
several times observed, displacements of epiphysis from shaft. Further,
pseudo-paralysis, which is very common in the syphilitic disease, may occur
especially about the wrists in the cases which occupy our attention. But in
the syphilitic affection the pain and tenderness are not nearly so severe as in
the disease under consideration; they are often, indeed, quite trifling in
amount, whilst in the so-called acute rickets they are more continuous and
more severe than any bone disease of childhood with which I am acquainted.

There is occasionally a concomitant joint-effusion which may be purulent
in the syphilitic affection. I think it is doubtful whether the joints them-
selves are affected in acute rickets. In a typical case of acute rickets in
which the thigh is affected, the involvement of soft parts is more extensive
than is ever met with in the syphilitic affection of the end of the shaft.

Finally, the element of age is of very great value.

Acute rickets, as I have shown, is very rare in early infancy, whilst the
typical congenital syphilitic affection is common under six months, indeed,
under four months, and even occurs in the foetus. But it must be confessed
the only thoroughly satisfactory distinction is one derived from post-mortem
examination. In the syphilitic affection, of which I have examined three
specimens post mortem, and of which specimens have been shown in this
country by Mr. Haward and Dr. Goodhart, the change is mainly, as
M. Parrot has pointed out, an endosteal one, and consists of what he calls
a gelatiniform transformation of the ossiform material which exists as the
extremity of the shaft. It is quite different in character from the massive
proliferation of cartilaginous material found in an ordinary case of the first
stage of rickets, which material forms a large cushion between the shaft and
the epiphysis.

In the syphilitic affection, along the gelatinous softening, there may be
a varying amount of concomitant perichondritis and periostitis, which latter,
as I have said, may extend up the shaft for a varying amount and be followed
by an osseous deposit. But I have not seen in the syphilitic affection the
extensive separation of periosteum and shaft by a mass of blood-clot such
as occurs in the specimens of so-called acute rickets now under considera-
tion, and such a striking condition has not been described by Wegner or Parrot
who have both examined a great many cases.

But the affection of the ends of the shafts is by no means the only form
of syphilitic affection of the long bones. Cases may be seen in older children
where several long bones are thickened along the greater part of the shaft
with firm solid deposit. This condition may last for months and slowly
clear up. How is this to be distinguished clinically from acute rickets? I
can only say that I have never seen a syphilitic case of this character lying
prostrate with the pain, tenderness, and cachexia comparable with that we
have already considered, but that in fact the general suffering and the
progress are quite different.
To return to the cases analyzed in this paper. In the greater number, as I have said, the early indubitable signs of congenital syphilis are no longer capable of being brought into evidence on account of the age of the children, but are there any other signs available which might help us in this direction? In no less than eight cases either the facial or cranial bones were affected, and in two if not three there were bosses on the frontals or parietales of the kind described by M. Parrot as characteristic of congenital syphilis. Now, although it is certainly the fact that many unquestionable syphilitic infants develop these bosses, especially if they be also rickety, yet in our present state of knowledge it seems to me premature to regard them as decisive of the question of congenital syphilis in the absence of other signs.

In one of my cases, along with these cranial bosses had marked splenic enlargement, and both these conditions persisted after the affection of the limbs and the sponginess of the gums had subsided, not under mercurial or iodide treatment, but under raw-meat juice and vegetables. In this case I have unfortunately lost the early history; the conclusion I had formed was that the child was possibly syphilitic, but that this had nothing to do with the condition of the limbs.

The early infantile history of one of the three cases of which post-mortem notes have been given, was compatible with congenital syphilis, but there was nothing conclusively syphilitic found post mortem.

It must be remembered that although the effects of syphilis may last for a considerable time, and seem to have a very special incidence upon and proneness to relapse in the osseous system, yet that they may also very rapidly pass away. Thus I made a post-mortem examination on a child aged ten months, who died of acute tuberculosis without any syphilitic lesion, and whom I had had under observation when it was an infant with severe congenital syphilis. There must be many parallel experiences.

To sum up these observations it may be stated—(1) that of the cases recorded in the great majority there was no conclusive proof that congenital syphilis was actively present; (2) that in several there was nothing in the previous history to justify the view that congenital syphilis had been present in early infancy; (3) that even in those in whom syphilis had possibly existed it would not necessarily follow that the symptoms of acute rickets had any connection whatever with the infantile syphilis; (4) that it seems possible that two cases, briefly mentioned by Steiner, of children four months old, who were the subjects of congenital syphilis, and considered by him to be also suffering from acute rickets, were really the subjects of the special syphilitic affection of the ends of the shafts of the long bones which presents considerable resemblances to the so-called acute rickets.

We must now consider the relation of the disease in question to the ordinary form of rickets with which we are so familiar.

Was rickets present at all in the ordinary acceptation of the term? In nine cases the details are not sufficient to allow us to give a definite reply. Of the twenty-two remaining cases reference to the table will, I think, show
that in at least three the signs or ordinary rickets were very pronounced indeed, in seven moderately well marked, and in nine slight.

In at least three we are justified in saying that there was no rickets, and amongst the nineteen slight cases the evidence often amounts to nothing more than slight beading of ribs.

Hirschsprung asserts that the disease has always appeared in formerly healthy children. This is another of the sweeping statements of a very able observer. But, although the statement is not accurate, yet it is remarkable that many of the children before the sudden onset of the disease were considered to be in fair general nutrition.

In Dr. Fürst's case, after the subsidence of the acute symptoms, marked bendings of the femora were found which had not occurred before the illness and which he considers established the truly rickety nature of the illness.

I believe that even that condition is susceptible of another explanation, but whether that be so or not let it be noted that in several of the recorded cases the recovery was absolute without deformity within a period of three or four months, which is quite unlike the ordinary course of rickets.

We are confronted then by two difficulties:—1st. The complex of symptoms which we have described may occur in a child in whom the ordinary signs of rickets are practically nil; and 2nd. very severe cases of rickets, in the ordinary sense, may run their course without presenting the complex of symptoms which we have described.

There is little wonder then that the German writers have found it difficult to 'dovetail' the so-called acute rickets with ordinary rickets, the more so that it is admitted that acute rickets so called is quite a different thing from severe or aggravated rickets.

I do not think that anybody will maintain that the subperiosteal haemorrhage in the cases of which the post mortems have been given (and which also I assume to have been present in the cases analyzed) is a feature of rickets as such.

I showed the specimens to Sir William Jenner, who told me that, in respect to the striking feature, viz. the subperiosteal haemorrhage, he had never seen it in ordinary rickets. There is also no description of such a condition in Guérin's account of rickets.

With respect to other diseases, I have already shown that, clinically and anatomically, the one in question differs from acute periostitis single or multiple.

The partial death of the shafts in severe cases is, I think, sufficiently explained by the mechanical interference with efficient vascular supply by the extravasated blood clot. In the cases which recover, the thickening of the bone shafts, which may remain for a considerable period may, I think, be explained by the osteogenic power of the upraised periosteam.

With what disease can we connect this non-inflammatory, sub-periosteal haemorrhage, associated with blood in the deeper muscular layers and serum in the superficial layers? Blood has been found in the joints in haemophilia, but it is not described as occurring under the periosteam in that disease.
In the group of cases analyzed there is, I think, no proof of effusion into the joints, although once or twice the term 'painful joint affection' is used. In a great many, at all events, there is no sign of effusion. But certainly the clinical history of our group is very different from that of one of the arthritic attacks of haemophilia.

It is a question how far purpura ought to be ranked as a separate disease, and whether it would not be desirable to consider it rather as a symptom occurring in many diseases. Although suggestions occurred a few times in our present group it is interesting to note that only in one case are small, numerous, spotty ecchymoses in the skin recorded. Without attempting to define purpura haemorrhagica, I may mention that in a few cases (to which for want of a better this term might have been applied) I have seen general painful swelling of the leg occur along with the appearance of purpuric spots in that region. This painful swelling has, however, so far as I have seen, only lasted a few days, and thus differs considerably from what obtains from our present group.

Although it would be absurd to limit the possible occurrence of subperiosteal haemorrhage to any one malady, I believe it will be found, on analysis of the cases before us, that they approximate more closely to scurvy than to any other disease with which we are acquainted. Let us consider the parallelism first along anatomical lines.

The painful brawny induration of the lower limbs in adult scurvy was shown by Lind to be due to blood extravasation in the bellies of the muscles and serum in the tunica adiposa (p. 496, 3rd edit.) He often found the blood extravasation most extensive in the deeper layers, and lying on the periosteum; and once he discovered lying in spoonfuls beneath the periosteum.

Dr. Budd, in his article on 'Scurvy,' in Tweedie's 'System of Medicine,' describes a post-mortem on one case in which, although there was no swelling of the calf, there was a node-like swelling over one tibia, and on cutting down upon it there was found a thin layer of blood under the fascia and a solid clot of chocolate colour a line or two in thickness for a length of six or seven inches under the periosteum, the periosteum itself being thickened and infiltrated with blood in this region. Other sub-periosteal haemorrhages were found on one femur, one fibula, the opposite tibia, and the upper and lower jaws.

There are some older observations which are still more interesting. In the year 1699 M. Poupart made some dissections of scorbutic bodies in the Hospital of St. Lewis, at Paris. Amongst his remarks is the following, quoted by Lind:——In some, when moved, he heard a small grating of the bones. Upon opening those bodies the epiphyses were found entirely separated from the bones, which by rubbing against each other occasioned this noise. All the young persons under eighteen had in some degree their epiphyses separated, and 'in some' he says 'we perceived a small low noise when they breathed,' and in them the cartilages of the sternum were found separated from the bony part of the ribs.
ON CASES DESCRIBED AS 'ACUTE RICKETS'

He further describes a condition of rib very like that to which I have referred in my second post mortem.

There is another observation by Dr. Godechen, a Russian physician, which is quoted by Budd, and is very important. In a case of scurvy, separation of the ribs from the costal cartilages and fractures of ribs near their anterior extremities occurred, without violence, whilst the patient was in the hospital.

Without laying too much stress upon it. I may refer to the blood-stained effusion in one pleura in my second post mortem as being comparable with the condition of the pleura found in some of the fatal cases of adult scurvy.

Having shown, then, a certain anatomical resemblance between our group of cases and adult scurvy, let us work back along the clinical lines and see how far they also run parallel.

The order of appearance of symptoms, as set down by Lind in his own words, is, first 'a change of colour of the face, from the natural and usual look, to a pale and bloated complexion with a listlessness to action.' The second symptom is a stiffness and feebleness of the knees upon using exercise, and the third is the swelling of the gums.

In our group of cases the change of colour of the face is most striking. In a typical case it is not simple pallor but pallor of a somewhat dirty sallow tint. The general prostration is quite as marked a symptom. With regard to the swelling of the limbs during life, though it is true it is not so brawny in our cases as in the adults yet there are many similarities. The distribution is singularly parallel. As pointed out by Lind, there may be in the adult only a single swelling, but more commonly the swelling is bilateral or indeed multiple; and this obtains also with regard to our cases. In both alike the lower limbs are in the majority of cases affected and in the greatest severity.

There are parallels also in distribution, to which I have already referred in the occasional involvement of the ribs and of the upper and lower jaws.

With respect to the gums, we have, it would seem, at first a remarkable divergence between some of our cases and the typical adult scurvy.

In fifteen cases out of thirty-one it is noted that the gums were affected. In a few of these the swelling was obvious and characteristic, as in Dr. Cheadle's two cases, especially the first, from which there was much bleeding. Also in the case of Dr. Ingerslev the sponginess of gums was accompanied by a carrion-like odour, which was no doubt sufficiently suggestive to him of the true affinities of the malady. But in several others the swelling was very slight, and in fact only consisted in small localized ecchymoses in the sites to be occupied by the coming teeth. In six cases it is specifically stated that there was no stomatitis, and in ten it is not mentioned as being present.

The question of scurvy was indeed considered at the time of making the post mortem of Mr. Thomas Smith's case, and dismissed on account of the absence of any swelling of gums.
But even in adults I have, since making the above post mortem, learned that the absence of swelling of the gums does not negative scorvy. Dr. Ralfe has informed me that in crews suffering from scurvy there have been well accredited cases of men who have had all the other symptoms of profound cachexia, etc., but without the spongy gums; and this view is also expressed by Dr. Buzzard in his article on scurvy in 'Reynolds' System,' and in his definition he implies that sponginess of gums is not absolutely essential. I believe it has been observed that if a man who has lost all his teeth gets scurvy subsequently, sponginess of gums does not occur. Sir James Paget has told me that it is almost impossible to salivate a patient who has lost his teeth, and the difficulty of inducing sponginess of gums in young infants by the administration of mercury is well known. It is interesting to note in our own group that where no eruption of teeth had occurred no sponginess occurred; that the sponginess when present was chiefly in the neighbourhood of teeth that had been cut, and that the small submucous ecchymoses when present were above the sites of the oncoming teeth. I submit, then, that this divergence is not sufficient to disprove the identification of the so-called acute rickets and scurvy. The history of the study of disease has led us to discredit universal propositions in medicine and to doubt the existence of an absolutely pathognomonic sign, that is to say, of a sign which is present in every case of a given disease and never present in any other disease.

To those who will be willing to admit that the cases with limb affection and spongy gums were truly scorbutic, whilst denying the scorbutic character of the cases of limb affection without spongy gums, I can only reply that in every other symptom several of these cases were as nearly as possible identical.

With respect to pyrexia there is another seeming divergence. I have already pointed out the inaccuracy of Senator's statement that in these cases some fever is an invariable and characteristic symptom, and I have suggested that the fever, when present and not due to intercurrent ailments, may perhaps be proportionate to the tension of the haemorrhagic effusion under the tight periosteum. The bones have not been carefully examined in a sufficient number of cases in adult scurvy to admit of a dogmatic statement, but it seems probable that in them the periosteum is not so extensively involved as in these children's cases; that in fact the blood extravasation and serous exudation are more superficial and may give rise to less tension. On the other hand, although scurvy in adults is generally an apyrexial disease, it must be remembered that, in the words of Dr. Budd, occasionally we find the skin hot and the pulse attaining or even exceeding the rate of 120 in the minute. In these cases, Dr. Budd remarks, the swellings are exquisitely tender and the slightest movement of the limbs occasions great suffering.

Thus, even in this respect, I think we may establish a certain parallelism.

There are many other parallelisms to be drawn from a further analysis, but we must no longer defer the consideration of diet as an etiological factor
of these cases. The great difficulty in this part of the inquiry is the paucity of information in many of the reports as to the quantity and quality of the food which was being taken at the time of the onset of the acute symptoms.

First, with respect to breast milk. Several of these thirty-one children had previously been suckled for varying periods, but with the exception of a dubious statement by Møller about his third case, I think none of them are recorded as being at the breast at the period of onset of the acute symptoms. Steiner, however, in his brief account of the disease, though he speaks of its onset as being generally after weaning, uses the phrase that it may even appear during lactation. I venture to suggest that these exceptional cases coming on during lactation may have been not so-called acute rickets, but examples of the congenital syphilitic bone disease which may appear in infants at the breast, if the subjects of syphilis.

In the cases at present under review, then, we have to deal with children fed at the time and for a varying period previously by hand, and it is of the greatest importance to see if there was any other point in common with respect to the food.

First, it will be found that certainly five, probably six, and perhaps more, were taking cow’s milk at the time of onset. On further investigation, we find that in one case the quantity was extremely small, in another not more than half a pint daily; in a third it was two pints in twenty-four hours, with an equal quantity of water, the child suffering the while from considerable diarrhoea. We have no information about the quantity in the other cases nor indeed of the quality, which is probably important.

It is clear that the use of beef tea was not adequate to prevent the appearance of the disease, for in three cases, perhaps four, this food was being given at the time of onset.

It is very important to ascertain whether the affection ever appears whilst a child is taking raw-meat juice. The only case bearing on it is Förster’s, but it is not explicit enough.

A child of eleven to twelve months old had been breast-fed for three months, then had had cow’s milk, Liebig’s soup, flesh broth, eggs, scraped meat, etc., and had suffered from jaundice and diarrhoea. But it is unfortunately not clear from the account of the case what was the exact diet at the time of the onset. This is important, because for several months the child had been well nourished in spite of jaundice and diarrhoea; and Förser notes that the evidence of rickets was very slight.

Two of my cases had hysterical objections to meat and meat and vegetables respectively. One of them cried and even vomited when I had a plate of meat set before him. Also in two cases of scurvy in older children not included in this analysis I have observed the same curious dislike. One of these latter children absolutely screamed whenever any vegetables were placed on the table anywhere near to her, and refused any kind of food offered to her if the spoon had been previously used for vegetables.
I cannot find that it is stated in any of these cases that at the time of onset fresh vegetables formed a part of the diet.

The most important fact is that in at least seven there was absolutely no fresh food given. Thus Nestlé's food made with water, Ridge's food made with water, mealy foods, exclusively amylaceous food, and Anglo-Swiss food are examples.* In some of the cases the food and hygienic conditions are said to be satisfactory, but as the details are not given we cannot discuss them.

What light does the result of treatment throw upon the disease? Möller's cases convinced him that anti-phlogistic remedies were distinctly injurious, and in Dr. Fürst's case they seem not to have been followed by any benefit.

Great influence is attributed by Bohn and Hirschsprung to the return of spring and the possibility of getting the child out, and this is parallel to the experience with regard to adult scurvy.

With regard to antiscorbutics, they appear to have been given in some of the German cases without, in the opinion of the authors, leading to obvious benefit.

I venture to suggest that, before arriving at a definite conclusion, it is necessary to know the exact period of the disease at which they are given. When the marasmus is very profound indeed, it is, perhaps, too much to expect an immediate improvement, or, perhaps, any improvement at all. And, moreover, when there is much sub-periosteal haemorrhage, it must take a considerable period before absorption can possibly be completed, and the bone return to a normal state. But in Dr. Cheadle's two cases, and in seven of my own, the improvement was perfectly obvious and most striking in those which could be personally supervised, and in which treatment could be persevered in, in spite, as sometimes happened, of objections on the part of the child.

In Ingerslev's case, it is noted that no treatment, including antiscorbutics, led to the slightest improvement until the spring came, and the child was able to eat garden cress. This is parallel with an interesting observation recorded by Dr. de Mertans in the 'Philosophical Transactions' for 1778. This physician, who was attached to the Foundling Hospital at St. Petersburg, had been accustomed to treat many severe cases of scurvy in children, especially in the winter and spring, and a very fatal disease he sometimes found it. Experience taught him that if the cases came under his care early in the disease vegetable soups succeeded very well, so that three or four weeks were generally sufficient for cure. But in one winter the outbreak was particularly severe and resisted his ordinary treatment, and he then found that the most stubborn cases yielded only when he gave them raw vegetables as well as vegetable soups ('Phil. Trans.,' vol. lxviii, p. 676).

* To which I may add, from two subsequent cases not included in this analysis, Savory and Moore's Food and Neave's Food.
Reverting to our cases it is clear that they differ very much in severity. Those in which the cachexia is very profound often end fatally, just as in adult scurvy. Nevertheless, a careful perusal, especially of the German cases, convinces me that the disease in question often tends towards a slow, but ultimately complete, recovery, and this independent of any special treatment.

Let us turn once more for a parallel in adult scurvy to Lind, whose work is such a masterpiece, not only of learning, but of accurate and candid observation. Lind’s arguments for the employment of fresh vegetables in scurvy are unanswerable, but it is interesting to read in the postscript to his third edition, concerning certain cases that he had carefully watched, that ‘strict abstinence from the fruits of the earth was continued long enough to convince me that the disease would often, from various circumstances, take a favourable turn, which cannot be ascribed to any diet, medicine, or regimen whatever.’

It may very properly be asked why, if it be true that these cases are mainly produced by a faulty diet, are they not more frequently seen, since a faulty dietary must obtain in London and other large towns to an extreme degree, especially amongst the poor?

A complete answer to this question cannot be given. But first, probably minor, degrees of scurvy are not so rare as might be thought.

It is possible that some slight cases, and even severe cases of the bone affection, are dismissed as ordinary rickets, with an excess of tenderness and fretfulness. Probably, also, in this affection as in others, idiosyncrasy plays a part; and we have to remember that in adults the scurvy-producing diet may be in use for a considerable time before the disease is precipitated, so to speak, by some additional, often unknown, depressant agency.

A valuable remark of Dr. Cheadle’s may, however, be referred to in regard to the reason why scurvy does not more often occur amongst the children of the London poor. A bread and butter diet, with the exclusion, or extremely meagre supply, of milk is common enough, and is probably responsible for a great deal of rickets, but poor children are often saved from scurvy by the common use of potatoes. If potatoes are excluded and only the bread and butter diet given scurvy, sooner or later, is exceedingly likely to manifest itself.

To sum up this paper, I will submit that (1) the characteristic symptoms of the so-called acute rickets, viz. the special limb affection and the cachexia, with or without sponginess of gums, are not due to rickets at all but are truly scorbatic.

(2) That the anatomical basis of the limb affection is sub-periosteal haemorrhage, and that this haemorrhage probably accounts for some of the anaemia.

(3) That the disease may occur in rickety children, and perhaps in them more readily than in non-rickety children, but that the amount of rickets may be almost nil.
(4) That although the disease tends spontaneously in many cases towards a slow but complete recovery, marked improvement often follows a vigorous and especially an early antiscorbutic treatment.

(5) That the treatment recommended is—locally, during the acute stage wet compresses and avoidance of movement, at a later period careful shampooing and douches; internally, the use of raw-meat juice, fresh milk, and orange juice, or of some other fresh raw vegetable, and from the first the access of as much free air as is possible.

(6) That the use of the term acute rickets should be abolished for these cases, and that of infantile scurvy substituted, the special note of which, as distinguished from adult scurvy, being the greater incidence of the disease on the bones.

(7) That with regard to the hand feeding of infants it seems probable that the so-called ‘infant foods’ cannot be trusted as sole aliment for any lengthened period, however useful they may be as adjuncts.

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