CONGENITAL SYPHILIS.¹

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It has been truly said that whoever knows syphilis, knows the half of medicine. I believe this to apply almost as much to children as to adults, for in my experience there is probably not an organ or tissue in the child's body which may not be affected by the spirochæte of syphilis or by the syphilitic toxin.

The Hospital for Sick Children, Great Ormond Street, holds the unique position of being the only Children's Hospital in London which is included in the Venereal Diseases scheme of the London County Council and the Ministry of Health. The result is that quite a large number of cases of congenital syphilis gravitate to the Hospital, and patients come from as far afield as Aylesbury for their weekly injections.

By the courtesy of my colleagues at the Hospital, I see all, and treat most of, the cases which come there, and as a result I have seen, probably, upwards of 1,500 cases during the past fifteen years, and from this large amount of clinical material one has learnt a great deal and some of the more interesting points I am embodying in this article.

The whole subject of congenital syphilis is obviously too big a one to treat in detail during the course of a short paper, and I shall not, therefore, give a systematic account of its symptomatology and treatment, for this can be learned from the textbooks on pædiatrics or general medicine.

Rather do I propose to bring to notice some of the more important points in connection with the mode of transmission, ætiology, and symptoms of the disease, which my experience has shown me are worthy of attention.

First, with regard to the mother's obstetric history. In accordance with the dictum which is sometimes known as Kassowitz's law, that the virus of syphilis becomes progressively more attenuated in the mother, it is generally taught that the usual history is for the mother to have first, perhaps, a period of sterility, then a series of early abortions, later on miscarriages at four to seven months, then a stillborn child at term, and finally a living syphilitic child. Although this sequence of events may rarely happen, it is, in my experience, by no means common, and if one waits before diagnosing a case of congenital syphilis for such a history in the mother, one would undoubtedly miss a large number of cases of the disease. I think in a considerable number of cases one may obtain a history of one or two miscarriages or premature births before the birth of a syphilitic child, but even such a history is not by any means the usual one, for I have on many occasions seen cases where the syphilitic infant is the result of the mother's *first* pregnancy. Possibly the absence of a series of accidents to the mother's pregnancies is to be explained by the fact that so many more fathers are now treated for their primary or secondary syphilis in the many clinics which have been established in recent years throughout the country than was formerly the case. I should like to emphasize this fact, because it is still very generally taught that one need not think of syphilis unless there has been a history of one or more miscarriages before the living child.

Another point of interest is, can syphilis be transmitted to the third generation? In this country it is generally held that syphilis is not transmitted to the third generation

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So also in Germany. In America they do not believe it, but they say it is possible. The French, on the other hand, hold strongly that syphilis is transmissible to the third generation, and I am convinced that it is true, though I admit you cannot prove it scientifically. If a man tells you he never had connection till his marriage, the tendency is not to believe him; it is felt one cannot be certain the man did not have syphilis. I think the father of a child which recently passed through my hands, a definite case, had told me the truth when he said he did not have connection until he was married. The mother's blood was Wassermann positive, the father's blood was negative, which does not help us, because 70 per cent. of the fathers of my cases have given negative Wassermanns. I went into the history of this mother. I asked if her mother was alive, and she said "Yes, she is a housekeeper at Eastbourne." I went down to Eastbourne and called upon her, telling her she had a grandchild who was very ill, and it would help if she gave me some information. I got information, and also some of her blood, and this I took back and tested, and found it was strongly positive. I have now the records of over thirty families in which syphilis was present in the third generation, and it is often severe syphilis, though it may be a mild form.

Coming next to the patient itself; when the classical symptoms of congenital syphilis, namely, the typical rash, snuffles, and hoarse cry, the peculiar *cafe-au-lait* colour, and the epiphysitis are present, there is, of course, no difficulty about making the diagnosis, but it must be remembered that all these symptoms are by no means invariably present in the same infant, and indeed a child may be suffering from congenital syphilis without any of these symptoms being manifested. To give you one or two instances which have occurred recently in my own experience at Great Ormond Street; a child who at the age of 4 weeks was brought into the Hospital on account of pylorospasm, and was consequently failing to thrive. After a couple of months, as the child was not getting on, it was suggested that a blood-test should be taken, and the Wassermann was found to be positive. When the appropriate treatment was given, the child rapidly did well. Contrary to my usual experience, the mother's blood in this case gave a negative Wassermann reaction, and the father's was positive. Another case was as follows: A child about 3 months of age was brought to Hospital with a history of bronchitis at birth, and some snuffling, which lasted about a month. There was no rash or clinical epiphysitis—the child was not thriving, it cried a good deal, and the cry was a hoarse one. There was a history of one miscarriage at 2 months, two years previously. The child attended for three months before a blood-test was taken, which was found to be positive. So also was the X-ray examination of the bones of the limbs, although there had not been any symptoms suggesting epiphysitis.

The so-called epiphysitis of congenital syphilis is an important manifestation, and merits full consideration. The classical clinical description is that given by Parrot, that a child who may already be suffering from snuffles and rash, suddenly becomes paralytic in one or more limbs, the arm or leg hanging helpless, on which account the condition is sometimes known as Parrot's pseudo-paralysis.

An X-ray examination of the child's limbs generally shows that a considerable number of the long bones of the body are affected by a condition of osteo-periostitis, osteo-chondritis, or osteo-myelitis, and as a matter of fact very often the epiphyses show little change. This condition is very much commoner than is generally recognized,

and indeed may often be demonstrated by systematic examination of the bones in congenital syphilis, even in the absence of any clinical manifestations. According to some observers, this osteo-chondritis is present in *every* case of congenital syphilis. Though I have myself not been able to confirm this statement, I agree, nevertheless, that it is present in a very large number of cases, and therefore I think that an X-ray examination of the bones is quite as important as the blood-test, and a negative finding is a strong point against a diagnosis of congenital syphilis.

Wegner, in 1870, was the first to give a detailed description of the changes in the bones in infantile congenital syphilis. He was closely followed by Parrot, who published various papers in 1871-1873, and he is regarded by McLean as having been the first to establish osteo-chondritis as a clinical entity. Wegner regarded it rather as an inflammatory process, hence the name osteo-chondritis, but Parrot denied the inflammatory causation and regarded it as a nutritional disturbance of the bone tissue at the zone of ossification. Parrot was the first to declare that every bone of the skeleton in infantile congenital syphilis is affected, and he also established the fact that the earliest time for the lesions to appear is at about the fifth month of intra-uterine life.

Subsequent clinicians and pathologists have, from time to time, written upon the subject and added details to our knowledge of it, and perhaps the best description, at any rate in the English language, is that by Stafford McLean, which was published in a series of numbers in the *American Journal of Children's Diseases*, from January to June, 1931. I have no space to enter into details, but would refer those of you who are interested in the subject to Dr. McLean's articles, and shall give you just the barest outline of his conclusions, in so far as they are helpful to us.

The most frequent lesion is situated at the junction of the epiphysal cartilage; hence the name epiphysitis, which is still so generally applied to this condition, but which is more appropriately named osteo-chondritis. The X-ray changes include: (1) The deepening of the shadow at the end of the diaphysis, which is sometimes known by the name of metaphysis. This is due to increased calcification along this line. (2) Rarefaction and loss of cortex, particularly at the upper and inner aspects of the tibiae; such sub-metaphysal rarefaction may also be present in other bones, and in other situations than the ends. When this occurs the condition is known as osteomyelitis. (3) Sometimes, also, there is a definite periostitis with the formation of new bone under the periosteum, which gives rise to the condition known as periosteal "cloaking" or "splinting." The affected joints have been known at times to become secondarily affected, and I have myself isolated streptococci and pneumococci on different occasions from the pus aspirated from such joints. Separation of the epiphyses not infrequently appears to have occurred, but it is very rare for the epiphysis actually to be separated from the diaphysis; what more commonly happens is for the epiphysis together with the metaphysis to become separated from the rest of the shaft, owing to a weakening of the trabecular structure of the bone itself at that spot.

Macroscopically, in fatal cases, the areas of rarefaction and osteomyelitis can be distinctly seen, looking rather like pinkish grey connective tissue. As is well known, the epiphysal line is broader than normal, yellowish instead of pink, and feels gritty.

As I have already mentioned, this condition of the bones, in varying degrees of severity, is present in a very large proportion of infants suffering from congenital syphilis. It may be present before birth; on the other hand, it may not develop until

after birth, but it rarely starts after the fourth month of age, which helps to distinguish it from other diseases, e.g., scurvy, rickets and infective osteomyelitis, which generally commence at a rather later age. Moreover, the lesion tends to heal, sometimes even spontaneously and in the absence of any treatment. Lastly, one should add that frequently the condition of the bones may be entirely unsuspected, owing to the absence of any symptoms in connection with the limbs.

The eruption of congenital syphilis may be so characteristic that no one can miss

Rash. it; for example, if it is a pemphigus or a typical copper-coloured rash

over the body, with ulceration of the lips, you cannot make a mistake, but there are other rashes which are more difficult to diagnose; sometimes there is a papular rash, sometimes an ulcerative rash, and these have misled even eminent physicians. Trousseau described a papular rash, and Parrot an ulcerative condition of like character, as syphilitic when it was not; these were varieties of Jacquet's erythema, which may occur as an erythematous condition, or an erythematovesicular or papular, or ulcerative condition. It is not venereal, but is due to some special decomposition of the urine. It is a kind of napkin rash, but it may cause lesions which sometimes look like syphilis. A blood-test is of great help in these doubtful cases.

Another symptom of congenital syphilis which it is important to bring to your

Hydrocephalus. notice is hydrocephalus of mild degree. It is not sufficiently taught

that in a number of cases of congenital syphilis a mild hydrocephalus, due to syphilitic meningitis, may be present. So I advise you, in any case in which the head is moderately enlarged, to get the blood-test done, and if that is negative, even have a lumbar puncture done, because I have seen cases in which the blood has been negative but the cerebrospinal fluid positive. In hydrocephalus there is the overhanging forehead, with an enlargement of the scalp veins, part of the effect of the interference with the circulation inside. I would emphasize the importance of doing a lumbar puncture in every case of congenital syphilis. I saw a child in 1926 in which there was no rash and no snuffles, the reason for bringing it was a "rolling of the eyes." There was slight hydrocephalus, and X-ray showed periostitis. After nine months of treatment her blood Wassermann became negative, but three months later it relapsed again. The cerebrospinal fluid had been negative, but later became positive. It shows that because the spinal fluid was negative it does not follow that neuro-syphilis may not occur subsequently. I could not get the mother's blood negative. I treated her in 1926 to 1931, yet her blood is strongly positive. To me, that points to congenital syphilis in the mother. The mother's father was dead, and I wrote to the asylum, and found that he had had G.P.I. So when she was born, her father was a syphilitic patient.

Jeans and Cooke have brought out a book on syphilis in children, based on 1,100 cases, and they found that 40 per cent. of all their syphilitic infants had positive cerebrospinal fluids, and the important thing is that in 32 per cent. there was no sign of disease of the brain or nervous system; the disease in them was latent, so that in only 8 per cent. of the children was there obvious congenital neuro-syphilis, i.e., fits or hydrocephalus. My own results confirm those of Jeans and Cooke.

The question of the condition of the teeth in congenital syphilis is a very important one. Are Hutchinsonian teeth seen in the deciduous set? In this

Teeth. country we say No, but the French say that even the temporary teeth

may show the typical Hutchinsonian notching, and the case photographed here seems

to bear that out. One case, 2 or 3 years of age, had a big notch, but such cases are very rare. She gave a negative Wassermann, and Mr. Pitts, of the Dental Department at Great Ormond Street, says that was not due to syphilis but to a particular kind of caries or deficient calcification of the tooth; and he says you do not see the Hutchinsonian characteristics in the temporary teeth, but you do sometimes see hypoplastic teeth in a congenitally syphilitic child.

I have one slide which shows what was called a Hutchinsonian tooth, but it is not; it is a physiological notching. The teeth are developed in three segments, and if they do not fuse properly, segmentation and notching ensue. Another slide of a syphilitic patient shows physiological notching, and, in addition, a Hutchinsonian type of tooth. Pitts calls this a "potential notch," and in five years or so there will develop the typical Hutchinsonian tooth.

In one case sent as a congenital syphilitic, the teeth were normal; but I went into this mother's history, and found her blood was strongly Wassermann positive, and three or four years of treatment did not make it negative, so it was certain this mother was a congenital syphilitic. That is one of the points I want to bring out: *that though typical Hutchinsonian teeth are pathognomonic of congenital syphilis, they only occur in 25 per cent. of congenital syphilitics; and there may be mild degrees of Hutchinsonism in another 25 per cent.*

The "screw-driver" type of Hutchinson teeth is also met with; the lower incisor may also show Hutchinsonian characteristics, being narrowed towards the cutting edge, and sometimes there is notching.

The molar may be of the type known as "Moon's molar." Owing to lack of development of the middle segment of the tooth, the four big cusps tend to sink in and produce a dome-shape molar.

With regard to the later symptoms, after the first year there is the *condyloma*, the flat, papular, tumour-like formation which occurs particularly round the anus, though it may be met with on other parts of the body. My experience—and it agrees with that of many authorities—is that condylomata are rare before the end of the first year; so that if you see a lesion which you think may be a condyloma and the child is, say, 3 months old, it is almost certainly not that. In one case seen recently the physician in charge said the condition present was condyloma, but I disagreed because of the patient's age. The Wassermann reaction was negative, but he persisted that it was a condyloma. The mother of the child was also Wassermann negative. Our skin physician was asked to look at it, and he said it was the typical papular type of Jacquet's erythema. It was appropriately treated and the condition cleared up. When mothers bring their children because of condylomata, they think they have piles, but, of course, children do not have piles. Sometimes condylomata are not recognized as such. I once saw a child which had had condylomata for five months, and though it had been taken to two hospitals, the possibility of it being a condyloma had not occurred to them. We scraped it, and found numerous spirochætes in the exuded serum; appropriate treatment was given, and in a short time it had cleared up. As a rule, condylomata do not occur until the patient has passed the first year without treatment.

In the second or third year you find the children may have fits and paralyses. It is

well to do lumbar puncture in all cases, to see if there is positive evidence of syphilis of the central nervous system.

Later still you get the lesions which are known to you all, lesions of bones and joints, though sometimes enlargement of the glands of the neck is the only sign of syphilis. There may be infantilism or mental backwardness, but sometimes there is precocity. Eye and ear lesions also occur. Hæmoglobinuria is nearly always due to congenital syphilis, and it responds very well to treatment; you give a course of arsenic injections, and the mother will bring the child back saying there has been no recurrence of the dark water, and will sometimes add that last week she took him out in the cold, and although he shivered and she expected the patient to pass dark water, he did not do so. These children are very responsive to anti-syphilitic treatment.

You probably know the kind of joints to which the name Clutton has been given. The condition seems to affect the knees particularly, in children of 8 and a little older. Clutton joints are generally stated to be painless, but they are not always painless, and I have seen several such cases in which the wrong diagnosis has been made. For example, one child was kept in bed seven months because of painful knees; it was because "they wanted to protect his heart," as the case was thought to be rheumatic; no blood-test had been done. On subsequent examination the blood was Wassermann positive and the correct diagnosis made. At Great Ormond Street we do a blood-test in every case with enlarged joints. Another child came from a sanatorium with one enlarged and tender knee, which was thought to be a tuberculous condition. Another child was a long time in hospital for a rheumatic heart, until she suddenly developed keratitis; she had the typical syphilitic teeth, which had been missed. I am directing attention to the difficulties you are likely to encounter in your practice.

I have already spoken of the bone affections in infancy; the bones may also be affected in older children. There is a periostitis particularly, and often a gummatous periostitis, which may undergo necrosis. Changes often occur in the tibiæ, causing the curving known as "sabre tibia."

With regard to the eyes, I shall not say much about interstitial keratitis. The age for this is usually stated to be 8 to 10 years, but I have a number of cases in which it has occurred at $2\frac{1}{2}$ and 3 years. One very important point about interstitial keratitis is, that it is a condition which may arise when you think you have cured your patient. Recently I had a patient whose Wassermann was negative eight years, and you might naturally think she was cured, but she came back with interstitial keratitis. It makes you feel despondent that a child who has had a negative Wassermann for eight years should show that the disease has not been eradicated. I do not agree with those who consider that interstitial keratitis does as well treated with mercury as with arsenic. After the latter you never get corneal scars and nebulæ, such as you sometimes see after mercurial treatment alone.

Finally, I should remind you that we now know that the treatment of a syphilitic mother during pregnancy will almost certainly protect her child from being born syphilitic, and it is your duty to impress the importance of treatment upon any pregnant mother whom you know to be syphilitic or who has previously borne a syphilitic infant.