

ORIGINAL ARTICLES.

HEREDITARY SYPHILIS.

Presented to the Section on Diseases of Children, at the Forty-ninth Annual Meeting of the American Medical Association, held at Denver, Colo. June 7-10, 1898.

BY L. DUNCAN BULKLEY, A.M., M.D.

PHYSICIAN TO THE NEW YORK SKIN AND CANCER HOSPITAL; ATTENDING PHYSICIAN TO THE SKIN DEPARTMENT, RANDALL'S ISLAND; CONSULTING PHYSICIAN TO THE NEW YORK HOSPITAL, ETC. NEW YORK CITY.

This topic is one of great practical interest. Fournier has wisely remarked that "nothing is so dangerous to its surroundings as a syphilitic infant,"¹ and it can not be without interest and value to discuss a subject so vitally connected with the welfare of many of those whom we are called upon to treat. The subject of hereditary syphilis is such a vast one that it is difficult to determine just what points to present, and each aspect is capable of development far beyond the proper limits of these introductory remarks.

First, it may be said that our consideration will be confined to *hereditary* syphilis, as distinguished from the broader subject of *infantile* syphilis; for the latter properly includes that acquired after birth in innumerable ways, such as in nursing, feeding, vaccination, circumcision, and many others, and even a primary sore acquired while passing through the mother's parts.

It may be here remarked that Fournier² has made the distinction between syphilis received with conception—that is, when one or both parents were diseased before procreation—and syphilis acquired by the fetus during intra-uterine life, from a subsequent infection of the mother. To the former alone he would apply the term "hereditary" syphilis. He claims that clinically there is a difference to be observed between the ante-conceptual syphilis and that acquired during intra-uterine life; the former is much more fatal to the fetus. In our study we shall not make this discrimination, but for practical purposes consider hereditary as synonymous with congenital syphilis.

Confining our attention then, to hereditary syphilis, or that existing at birth, there are many very interesting points which meet us at the threshold of our study and are not without practical bearing; some of these are still "moot points," and can not be absolutely determined; some lead us so far away from the center point—the syphilitic infant—that they need not be included. We will consider a few of those most vitally related to the subject.

The first question that arises is: How and when does transmission of the disease take place? When the mother is profoundly syphilitic in the early contagious period of the disease, it is not difficult to understand that her offspring should partake of the same. But what are the limitations of time or condition as to the date of the mother's infection? This is a hard question to answer absolutely. Many cases are on record when the mother has acquired the disease one, two, three, five, seven and even eight months after conception, with the result of either abortion or a syphilitic child; I am not aware of any reported cases where infection in the ninth month resulted in a diseased child, and generally it will escape an eighth month infection. Naturally, syphilis acquired before conception will be more likely to cause early abortion; that received later results in a diseased child.

The more difficult question as the direct effect of only paternal transmission of syphilis can not be so easily and certainly dismissed. Many have doubted if the disease could be given by the father alone, without the intermediate infection of the mother, for certain reported experiments have seemed to indicate that the syphilitic virus is not communicated by means of the normal secretions—milk, saliva and semen. On the other hand, clinical evidence is accumulating that many children with syphilitic fathers are born with the disease, while the mothers are free from it—or appear to be free from it, on careful and prolonged medical observation. Some writers have attempted to explain away these cases by the well-known irregularities of syphilis, whereby, especially in women, its course may be peculiar, with prolonged absence of active symptoms. But still other clinical facts are being presented which go far to support the paternal transmission of the disease. Such are the occasional exceptions to Colles' law, in which the mother has been infected by her own child, and also instances of her acquiring a chancre otherwise (after having given birth to the syphilitic baby); both of these show that she has not the disease at the time, and so the infant's infection must have come from the father. Instances are also on record where a woman has borne syphilitic children to syphilitic men, and healthy children to healthy men. The paternal transmission of syphilis must, therefore, be accepted as a fact.

Syphilis is a powerful poison, producing various degrees of vital injury, even to death. It is not necessary to discuss here the nature of the virus, about which very little is positively known; most observers agree that it must be due to a toxin dependent on a micro-organism, which, however, has not as yet been isolated with certainty. The first effect of a full dose of the poison is inhibitive to the powers of procreation, and sterility is the fortunate result. With a slightly less intensity of the virus a non-viability of the product of conception results, and the fetus is thrown off early, even before the third month. With diminished virulence of the poison, or greater vitality of the parents, the life in the uterus will be prolonged, and abortion of a fully-formed and diseased child may take place at any period up to full term; but by far the larger number of miscarriages take place by or before the seventh month. With still less of the poison or greater vital resistance, the child may be born at full term, with or without external manifestations of the disease.

Even if brought into the world alive, the product of syphilitic conception has a relatively weak hold on life. This is instanced in the well-known statistics of the Moscow Hospital, where of 2000 syphilitic children born in eleven years, over 70 per cent. died. Fournier makes the mortality 28 per cent. from exclusive paternal heredity, 60 per cent. from maternal heredity, and 68.5 per cent. from a mixed heredity. Some figures are even more appalling. In Sigmund's wards in Vienna, out of 61 children born syphilitic, 59 died, 13 of them being still-births; of 47 living children only 4 survived more than three months. These figures, however, relate to hospital cases, and were taken some years ago, and it is believed that, with better knowledge of the disease and prophylactic treatment, a very much better showing could now be made; the results are also much better in private practice. But the facts stated serve to show the terrible virulence of the poi-

¹ Fournier: Syphilis et Mariage, p. 211. Paris, 1880.

² Fournier: L'Herédité Syphilitique, Paris, 1891.

son when operating under adverse circumstances, or unchecked, and forcibly point to the necessity of the most extreme care in many directions in connection with syphilis and married life. As such may be enumerated: complete treatment before procreation is attempted; very thorough treatment up to the very hour of confinement, where syphilis exists; the very early recognition and vigorous treatment of the disease in the child; and, moreover, a very guarded prognosis in every direction, whenever there is any syphilitic element in connection with the case.

When the syphilitic child is born alive it may appear profoundly affected by the disease or may have almost or quite the appearance of a healthy child; when born diseased it scarcely survives even many days, death resulting from cachexia and gastro-intestinal trouble, largely dependent on visceral lesions. In the child dying thus early from inherited syphilis very great changes are found in the internal organs, which we have hardly time to mention here. The liver is enlarged and hard, due to fibro-plastic matter, whereby the capillaries are often obliterated, with compression of the cells of the acini and consequent cessation of the secretion of bile. Or there may be other alterations which need not detain us here. The spleen is found to be enormously enlarged, more than double in size, and hard, also with syphilitic endarteritis. The lungs are affected by much the same process, that is, with new formation of connective tissue surrounding the bronchioles and also the capillaries, interfering with circulation and expiration. By this means nodules form in the plugged and distended alveoli, which break down, forming fatty or caseous masses that may involve much of the lung. Almost all of the internal organs, and even the bones, have been found to be affected by more or less similar processes, so that the child born dead with syphilis, or dying soon after, is literally infected throughout with the poison, of which the skin symptoms, which may or may not be prominent, are only a single feature, and that not the most important, except for diagnostic purposes.

If now, the poison is not in excess, or if the vitality is very great, the infant may be born in an apparently healthy condition, and often there may seem to be reason for hope that it will escape the dreaded disease. The period at which syphilitic symptoms manifest themselves varies more or less in different cases, from a few hours or days to several months, the exact distal limit being as yet a "moot question." Most writers believe that one year is the outside possible limit at which the first symptoms may appear, while good observers also claim that it has only developed several years after birth. It is more than likely that in these cases there were earlier manifestations which were overlooked, even as is observed in acquired syphilis.

In the vast majority of instances the disease asserts itself soon after birth. Thus of 249 cases collected by Roger (*Union Méd.*, 1865) in 217 syphilis appeared before the end of the third month, and in only 32 did it appear later. In another series of 105 cases, 10 were attacked by the eighth day, 14 more by the fifteenth day, and 21 more by the thirtieth day, a total of 45 within the first month. Other analyses of cases show about the same result, so that in a large proportion of the cases the disease asserts itself between the third and sixth week after birth—in over three-quarters of the cases within the first three months of life. Beyond six months the proportion is exceedingly small, and

yet it is never safe to give an absolutely favorable prognosis as to escape of the child even until after one year old. When the child is born apparently healthy, there is absolutely no means by which it can be determined whether it will or will not exhibit the disease later, and judgment must be suspended, but the case most carefully watched. Sometimes the skin lesions will be the first evidence of the disease, but more commonly symptoms referring to the nutrition or respiration will be the first to show themselves. The child is found to fail in appetite and strength, not to gain in weight, to be restless at night, and to be peevish and irritable. Soon the child's cry is noticed to be harsh and breathing difficult, and the well-known "snuffles" are noticed, with discharge from the nose; the nostrils become obstructed and it is hard for the child to breathe while nursing. The mouth soon becomes sore, the digestion begins to suffer and the child is really sick. With all this the body loses in weight, the skin becomes sallow and wrinkled, the features are drawn, and there is what is known as the "old man" expression. By this time there is usually more or less of an eruption, generally appearing first about the anus or nates, or about the mouth, and also on the palms and soles, which may become very general, and the syphilis is in its full bloom.

It must be stated, however, that the picture here detailed represents the fullest development of the disease, found in aggravated cases, in poor surroundings and unchecked by treatment, and certain points may be wanting in many cases. Under the best circumstances and with intelligent and proper care, all the symptoms may be very much modified, and the syphilitic child saved much of the injury which the poison is capable of inflicting, even as acquired syphilis in the child or adult may exhibit vastly different phases and degrees of danger.

The skin symptoms observed in hereditary syphilis are essentially similar to those seen in the acquired disease, and need not detain us long. Owing, however, to the delicate structure of the infant skin and its high vascularization, they exhibit some peculiarities. The erythematous eruption, or so-called syphilitic roseola, is that most commonly met with, in the form of oval or round pinkish macules, somewhat resembling measles, which may run together, forming reddened areas of considerable size; when they have lasted a while they more or less darken, giving rise to the so-called "coppery hue," but practically this feature is of much less significance than is popularly supposed. About the mouth this form of eruption sometimes lingers as brownish, scaly patches, and the palms and soles may desquamate.

This early erythematous stage may be rather indefinite and brief, and if the disease is unchecked it may pass into or be followed by another eruption of more solid form; this exhibits soft flat papules, large and small, of a dull red color tending to become coppery, and at first smooth on the surface; later there is more or less tendency to scaling, especially on the palms and soles. There is generally no special arrangement of these papules, although at a later period they may form in more or less circular or crescentic patches. Associated with this form of eruption there occur in parts which are moist what are known as condylomata lata or mucous papules. These are seen most characteristically about the anal and genital region, also about the mouth, and even between the fingers.

Owing to moisture and heat, the papules enlarge and become prominent, the surface softens, and may give off a glairy secretion which is exceedingly contagious. It is from this that many of the cases of innocent infection arise. When these papules occur in quantity about the corners of the mouth they give rise to a destruction of tissue which on healing leaves linear, radiating scars which often prove afterward to be a very valuable diagnostic sign. Within the mouth these mucous patches give off a free secretion which is the source of the greatest danger to those associated with the child.

Vesicular and pustular syphilides are comparatively rare in hereditary syphilis, but bullous eruptions are not very uncommon in severe cases. Bullæ are far more apt to appear on the hands and feet, but may be generalized, and may occur very early or at quite a late date in the disease. They always indicate a serious vital impairment, and when present at birth or developing very early, the case almost always ends fatally. Tubercular lesions are rare in the early history of hereditary syphilis, though they have been observed even in the sixth month. Generally they, together with gummatous lesions, belong to a much later cropping-out of the disease—even many years after birth. This brings us to consider briefly some of the late phenomena of hereditary syphilis.

Remembering what has been stated in regard to the very thorough and complete saturation of the system with the poison *in utero*, as evidenced by the many changes found in all the organs, it is not surprising that the hereditarily syphilitic child, if it escapes the lethal perils of early life, should exhibit changes in its various structures due to the action of the poison, and such is unquestionably the case. It is doubtful if one who has suffered much from the early symptoms of the disease, as indicating a severe infection, is ever so completely cured that there are not somewhere effects of the poison in the system. As the papular lesions about the mouth have left scars which remain for life, so infiltrations and alterations in other structures, when they disappear under treatment or spontaneously, leave marks or alterations of structure other than occur in the child whose development has been normal and not interfered with by such a malign poison. Prominent among these signs of past syphilis stand the changes in the second teeth, to which the name of Mr. Hutchinson of London has been so indissolubly attached. This change in the teeth does not always occur, but when found is certainly a valuable diagnostic aid. It is to be remembered that really only the upper central incisors present truly pathognomonic signs. When characteristically altered, more or less pegged, thickened, and with the curved, horizontal erosion at the end, they are not to be mistaken.

Associated with this change in the teeth there is a tendency to disease in the cornea which Hutchinson has also emphasized as peculiar to hereditary syphilis. This, usually occurring between the ages of three and twenty, results from a more or less diffuse keratitis, which gives rise to a peculiar hazy, ground-glass appearance of the cornea. Happily this condition seems to be getting more rare, owing to good treatment, so that personally I have not met with it for some time, although some years ago I saw it frequently. Ear disease, leading to deafness, also occurs from hereditary syphilis.

Changes in the bones are among the most common

sequelæ of hereditary syphilis, and the flattened forehead with prominence at the sides will often be seen in these cases, in connection with the alterations in the teeth and eyes referred to. The long bones are also very frequently attacked, both in their extremities and shaft. The former, due to an osteochondritis, belongs to the earlier phases of the disease, and by interfering with the nutrition of the bone may produce serious lesions of its structure. Periostitis is commonly of later date, and may cause great deformity and pain; nodes from this cause may often be seen even many years after birth. Dactylitis syphilitica may occur in very young children, or even in early youth; in this there is a general thickening and enlargement of one or more of the phalanges, commonly the proximal, which runs a very slow course, and if injured may break down and ulcerate.

Cutaneous lesions may occur as late manifestations of hereditary syphilis and are mainly tubercular or gummatous. I have seen in a girl 23 years old, exhibiting characteristically pegged and notched teeth and with a well-marked history of hereditary syphilis, ulcerative gummatous disease on the arm, which had always been regarded as lupus, but which yielded rapidly and perfectly to proper specific treatment. Time and space do not permit us to go further into the late manifestations of the disease, such as affections of the nervous system, sexual system, heart, thymus, etc., but it is believed that as research continues it will be found that every portion of the economy is more or less impressed and affected by the profound alterations in nutrition which take place during the active period of the operation of the poison in hereditary syphilis.

We come now to the point of the further transmission of this disease to a succeeding generation. This is a subject which has been hotly discussed, and many have denied its possibility. It would be impossible here to present any of the arguments, but it may be stated that instances are multiplying by reliable observers where such a transmission to a third generation has taken place, and the writer believes that the weight of evidence is now such that it must be accepted as a fact, although such an occurrence is exceedingly rare.

Having now traced the progress of syphilis and its effects from the act of generation to the third generation of transmission, let us very briefly return to the child acutely affected with hereditary syphilis, and consider for a moment its relations to the world.

The already quoted saying of Fournier that "nothing is so dangerous to its surroundings as a syphilitic child," should always be borne in mind in connection with every case of hereditary syphilis. Hundreds and thousands of cases of *innocent syphilis* have had their origin in innocent babes, and too great care can hardly be exercised. Before the danger was fully recognized large epidemics of syphilis have swept through country towns in Europe, while in Russia many villages are said to be almost completely syphilitized, entirely by "family syphilis." I have myself seen most lamentable instances, and not very recently had a grandmother with chancre within the nostril, followed by most severe syphilis with prolonged brain symptoms, who had received the infection from a syphilitic grandchild, which died of the disease.

It is questionable if a child of parents with recent syphilis should ever be wet-nursed. Even if the child exhibit no syphilitic phenomena, there is no certainty

that at some time mucous patches may not develop in the mouth which may infect the nurse. Danger could be avoided only by securing a nurse who was probably immunized by having had syphilis. There would be no danger of adding to the child's syphilis, for it has been pretty conclusively shown that syphilis can not be conveyed by the milk alone.

But even in the artificial feeding and ordinary family care of a syphilitic infant there is danger to those who have not had the disease. Multitudes of cases are on record where the disease has been thus communicated to those around by means of the feeding bottle, from tasting it. I have seen one case where this was believed to be the mode of infection. Time and space forbid our entering further this most interesting field of thought; sufficient to say, the most scrupulous care should be exercised in connection with the hereditarily syphilitic child, that in no way secretions from its lesions should affect others by mediate or immediate contact.

The subject of treatment in connection with hereditary syphilis has several divisions. First we may speak of prophylactic treatment.

The prophylactic treatment relates first, to guarding the prospective child against the dangers threatening from syphilis existing in one or both parents; and, second, to guarding others from the perils incident to the very contagious secretions given forth by the syphilitic child.

Time and space forbid discussion here of the very broad subject of marital syphilis, but it is one which should be very seriously considered by every medical man. The best observers all agree that procreation should not be allowed until after at least two years of active treatment for recent syphilis, and until at least six months have elapsed without treatment and without any manifestation of the disease. When the mother already has active syphilis and has become pregnant, her treatment should be pushed in the most vigorous manner possible. My preference in early syphilis, is for the treatment recommended by Hutchinson, of one-grain tablets of mercury and chalk, given every two hours, to the utmost tolerance of the drug. Eight to ten tablets may be taken daily, and in some cases it is often necessary to double some of the doses, and if necessary to administer opium to prevent purgation. Salivation should not be caused, and to prevent this great care to the mouth may be necessary, with the constant use of chlorate of potash or antiseptic solutions. If there should be intolerance of the stomach, mercurial inunctions should be used, or hypodermic injections of mercury. In somewhat later syphilis, the iodid of potash may also be needed, especially if placental disease is strongly suspected. This very active treatment should be pushed even up to the day of confinement, and as soon thereafter as possible.

Coming now to the prophylactic treatment of the offspring, it may be stated that the hereditarily syphilitic child should be regarded from the first as a dangerous element, even if in apparent good health, for it is never certain how soon syphilitic manifestations may exhibit themselves. It is to be remembered that the coryza may, and usually does, occur before the appearance of skin symptoms, and this secretion from the nasal passages and mouth is intensely virulent, and under proper conditions can give rise to a chancre when least expected. If at all possible, the syphilitic child should be nursed, and by its own mother, as

this affords it the best hope of life and health, as compared with artificial feeding. It should, of course, never be given to a healthy wet-nurse, for, however healthy at the time, mucous lesions may develop at a later period in the mouth, with the almost certain result of a chancre of the breast in the nurse. History is full of records of thousands of instances where this has occurred, and many where veritable epidemics occurred, passing through many individuals by direct or indirect transmission.

If the baby is fed artificially, great care must be exercised that the nursing-bottle and articles connected with feeding, are kept scrupulously clean and away from the possible infection of others. The instances and methods by which brephotrophic syphilis has been communicated would greatly surprise one who has not become familiar with the subject. These can not even be enumerated here, but the list³ amounts to between forty and fifty different categories, relating to almost every possible article of use or means of communication between the syphilitic child and those surrounding it.

The direct constitutional treatment of the hereditarily syphilitic child can be dismissed with few words. When there is strong reason to suspect that the child will develop the disease, a more or less mild treatment should be begun at birth, even before active symptoms manifest themselves, and be continued with judgment until the time of the feared explosion of the disease has passed. With any development of active symptoms the anti-syphilitic treatment should be pushed to all proper bounds.

For the active treatment of the disease no treatment has been devised which is better than inunction, as I believe is universally practiced. The blue ointment, diluted one-half with cold cream, is rubbed over the abdomen and loins, and smeared on a flannel band and worn constantly, fresh ointment being applied once or twice daily, according to the necessities of the case. Grey powder, given every few hours, is also a desirable plan in certain cases, and if there is evidence of intestinal disturbance, the bichlorid of mercury, half a grain to a grain to the pint of water, of which a teaspoonful may be given every few hours. Iodid of potassium is seldom wanted in the early stages.

The greatest care must also be given to the nutrition and general health, and every measure and remedy adopted which will heighten the vitality of the child. Iron is often called for in connection with the mercury, even in very young subjects. Later in the disease, and for the various sequelæ, the treatment is that indicated for acquired syphilis, and need not be detailed here. I may, however, mention that the syrup of the iodid of iron, even pushed in large doses, often yields brilliant results in rebellious cases.

The subject which we have tried to briefly cover is a very large one, and I feel that it has been but imperfectly presented. But if I shall have succeeded in calling attention to any features which may have been forgotten by some, and if a good discussion of the subject shall result, I will be more than satisfied.

4 East Thirty-Seventh Street.

³ L. Duncan Bulkley: Syphilis in the Innocent, p. 19. New York, 1894.

Diagnosis of Sinus Thrombosis.—Voss calls attention to the fact that absence of vein sounds on the diseased side is an aid to the diagnosis, only noted in children to date.—*St. P. Med. Woch.*, October 8.