

disease of a syphilitic nature is shown to have occurred in one or both of the parents, as indeed is also the fact of coexisting manifestations in other parts of the body of the parents or child—manifestations which are peculiar to syphilis and not to struma or other diatheses, such as peculiar eruptions, erosive ulcerations, nodes, and fissures. To which may be added also the fact that it is the eldest child, or the one born next subsequent to the infection of the parents which is markedly predisposed to be affected, the frequency of the attack and the force of the symptoms decreasing in the later-born children, and, finally, the peculiar physiognomy.

Such evidence as this, and much more of a similar character, has led syphilographers, notably Mr. Hutchinson, to believe and to declare that these ocular troubles, when occurring in young persons, are almost always the result of an hereditary taint due to a specific virus—a conclusion most important in a clinical point of view, as upon it the proper treatment depends.

## CHAPTER XXV.

## AFFECTIONS OF THE EAR.

WITHIN the last few years much light has been thrown upon this subject by a number of observers, among whom may be mentioned Gruber,<sup>1</sup> of Vienna; Schwartz,<sup>2</sup> of Halle; Stöhr,<sup>3</sup> of Würzburg; and Roosa,<sup>4</sup> Buck,<sup>5</sup> and Sexton,<sup>6</sup> of New York, and this chapter is, for the most part, a compilation of their labors.

It may be remarked at the outset, that cases of syphilitic disease of the ear, or those recognized as such, are rare. Thus Buck has met with only 30, out of a total of 3976 cases of ear affections, or a little over three-quarters of one per cent., but, as stated by him, the actual percentage is probably much larger, owing to the difficulty of recognizing the syphilitic element, and the tendency of patients to conceal the fact that they have had this disease. There are, indeed, in most cases, no absolute diagnostic symptoms which enable us to distinguish an affection of the ear dependent upon syphilis from one due to a non-specific cause.

EXTERNAL EAR.—The only instance, so far as I am aware, of the occurrence of a chancre upon the external ear, is reported by Alb. Hulot.<sup>7</sup> There is no reason, however, except the less frequent exposure, why chancres should not be as frequent here as on other portions of the external integument. This region is not unfrequently the seat of secondary manifestations. Syphilitic papules are met with in the post-auricular angle and upon the lobule of the ear, while the macular syphilide is most frequent on those portions supported by cartilage, as the fossa navicularis and the concha.

With patients in the early secondary stage, we often find impacted cerumen, not directly due to the action of the syphilitic virus, but consequent upon the well-known changes in the activity of the glandular apparatus of the skin, which obtains generally at this period. This fact was mentioned by Astruc as early as 1740.

The most frequent syphilitic manifestation, however, in the external auditory canal, consists of broad condylomata (mucous patches),

<sup>1</sup> Ueber Syphilis des Gehörorgans, Wien. med. Presse, 1870, 1, 3, 6, 10.

<sup>2</sup> Arch. f. Ohrenh., Würzb., 1870, 130, 134, 135.

<sup>3</sup> Arch. f. Ohrenh., Würzb., Bd. v., s. 139.

<sup>4</sup> Syphilitic Affections of the Ear, Am. J. Syph. and Derm., N. Y., 1871, p. 97. Also Treatise on the Ear, 4th ed., 1878, p. 521.

<sup>5</sup> Am. J. Otol., N. Y., vol. i., p. 25.

<sup>6</sup> The Sudden Deafness of Syphilis, Am. J. M. Sci., Phila., July, 1879.

<sup>7</sup> Ann. de dermat. et syph., Paris, t. x., 1879, p. 47.

which here find a fertile soil for development on account of the rich supply of glands and papillæ and the vascularity of the part. In the external portion of this passage, they are usually isolated, but further inwards they are multiple and often increase to such an extent as to fill up the canal with the vegetations or papillomata springing from their surface, and they may completely hide the drum. They commence with an ill-defined swelling of the deeply reddened skin, followed by the discharge of a sero-purulent fluid which gradually uplifts the epidermis. The appearance is now that of ordinary otitis externa, and a mistake in the diagnosis may be more readily made as the pain is usually severe. The simple form of papule such as is met with upon the body, is never seen in the external meatus, nor is any scaly eruption, although the latter is not rare on the auricle. Condylomata may also be developed on the drum and simulate acute or chronic inflammation of this membrane, especially as they may become ulcerated and give rise to perforation.

Again, in syphilitic subjects, the auricle and the walls of the external meatus may be the seat of ulcerations, usually rounded in form, which are very painful and obstinate. In some cases they appear to commence as circumscribed inflammations, which do not disappear after the evacuation of the contained pus, as do ordinary abscesses, but take on the ulcerative process. Their surface becomes covered with a diphtheritic secretion; their margins may extend, and the patient be subjected for a long time to great suffering, in spite of the most energetic caustic treatment. Similar ulcerations may arise from gummy deposits in the cellular tissue, the cartilages, or bones, more frequently in the cartilages. These deposits are for a time free from pain, and may be absorbed under anti-syphilitic treatment, but, in other cases, they suppurate and form ulcers of the character described.

Finally, among syphilitic affections of the external ear, we have to mention the affections of the bones, as hyperostosis and exostosis. Gruber has seen a number of such cases, coincident with nodes in other regions. They appear as circumscribed swellings, with rather more elevation than is common to nodes of this size. They are often multiple and not unfrequently seated near the drum, so as to cut off the view of this membrane. They may attain such a size as to interfere with the entrance of sound waves and thus impair the hearing, but such instances are rare. They are seldom painful. Gruber states that they are sometimes associated with similar formations in the bony portion of the Eustachian tube, where they may cause marked narrowing or even complete stenosis.

MIDDLE EAR.—Of all portions of the auditory apparatus, this is the most frequently affected by syphilis, in consequence of its intimate connection with the nose and fauces, where syphilitic lesions are so common.

Mucous patches may form in the Eustachian tube or upon the walls of the middle ear, and either disappear under treatment, or terminate in ulceration, destroying the tissues to a greater or less extent. Gruber states that when situated upon the membrane covering either of the fenestræ, and especially when situated upon the internal aspect of the drum, they are liable to excite very severe pain, which, unlike the pain of ordinary otitis media, does not subside upon perforation of the membrana tympani, but persists until the ulceration is checked.

Although these statements with regard to mucous patches in the Eustachian tube and middle ear emanate from so distinguished an authority as Gruber, yet, when we recall the inaccessibility to observation of the parts said to be involved, their very explicitness cannot but cast a shadow of doubt upon their value in the mind of the reader.

It is stated that syphilitic disease of the middle ear is still more frequently due to the extension of inflammation and ulceration from the nose and pharynx. That inflammation may thus extend along a continuous surface, there can be no question. Whether actual ulceration may extend from the fauces to the tympanum is not improbable, but we know of no autopsy in which the fact has been established. Gruber believes in such extension of the ulceration and describes its progress as follows: The opening of the Eustachian tube is of course first attacked, but the ulceration may proceed to the destruction of the greater portion of the organ of hearing. So long as the ulceration is confined to the Eustachian tube, the patient merely suffers from hardness of hearing, abnormal sounds in the ear, and a sensation of tension or fulness, but as soon as the middle ear is invaded severe pain sets in. In these cases of exulceration of the mucous membrane of the middle ear, changes may be observed in the drum itself. More or less of its brilliancy is lost; its surface becomes uneven and injected; its whole substance may become infiltrated, so that the position of the handle of the malleus can only be recognized by the injected vessels overlying it. Perforation may also occur.

The sequelæ of syphilitic disease of the middle ear are apparently the same as when the origin of the trouble was not specific, but no disease of the middle ear of simple origin ever leaves the patient in such a state of absolute deafness.

Among the sequelæ are noted opacities or destruction of the drum, varying in extent; loosening of the ossicula from their attachments, loss or impairment of the membranes covering the fenestræ, and caries of the temporal bone or of the ossicula. As in ordinary suppurative otitis media, the cells of the mastoid process may be invaded. This may occur without previous perforation of the drum and hence without discharge from the ear, so that the affection of the mastoid may erroneously be regarded as primary. Suppurative inflammation of the middle ear caused by syphilis is usually chronic, but the Eusta-

chian tube may be invaded by a sudden and severe attack from the fauces, resulting in stricture or complete closure.

Gruber also ascribes certain cases of otitis media hypertrophica to syphilis, in which are found not only thickening of the lining membrane, but also membranous bands and growths (polypi), or, again, hyperplasia of the bony tissue sometimes affecting the ossicles, either wholly and symmetrically or partially. The hammer of the malleus is especially liable to be thus attacked. Isolated outgrowths of bony tissue, varying in size, are also met with on the walls of the middle ear and in the bony portion of the tuba. The impairment of hearing which these changes may produce is evident.

We have omitted to mention one not infrequent cause of deafness, occurring in patients in the tertiary stage of syphilis, who have had gummata of the soft palate with the destruction of the soft parts that so often follows. As is well known, in these cases, the remains of the soft palate often become adherent to the posterior and lateral walls of the pharynx, whereby the pharyngeal opening of the Eustachian tubes are closed by a mechanical obstruction, and greater or less deafness ensues, which is irremediable. Frequent instances of this kind have come under my observation.

INTERNAL EAR.—We know nothing of syphilitic affections of the internal ear, although various conjectures have been advanced as absolute knowledge by some writers. It is not unreasonable to suppose that, when the tympanum is the seat of decided inflammatory action, there may be more or less hyperæmia or even extravasation of blood in the internal ear. This is asserted by Gruber, who also states that any long-continued interference with the conveyance of sound may cause atrophy of the auditory nerve, "as shown by microscopic examination after death."

#### SUDDEN DEAFNESS PRODUCED BY SYPHILIS.

Under this somewhat obscure heading, it is intended to include a certain class of cases, in which sudden deafness occurs apparently as the result of syphilis, but the pathology of which is not known with absolute certainty.

These cases may occur at any period of secondary syphilis, but are most common within the first three or four years. The attacks are usually preceded by a state of hyperæmia of the drums, either from cold or from sympathy with the mouth or throat, thus inviting, as it were, an invasion of the drum by the specific affection. They are characterized by their sudden occurrence and by the extreme amount of deafness. Both ears are usually affected simultaneously, but not always. The attack is not attended with pain, but there is often a feeling of fulness in the ear, and vertigo, especially on stooping or rising up suddenly, and staggering of the gait are not uncommon. Abnormal sounds in the ear are a troublesome symptom. The patient

can hear his own voice and also the vibrations of the tuning-fork placed upon the skull. Dr. Sexton calls attention to a phenomenon, which he says has not before been mentioned, viz., the high pitch of all sounds heard under certain conditions. Thus, a player on the cornet or violin, the latter resting the base of his instrument upon his neck beneath the jaw, will hear and play his notes higher than they are. He says that patients have told him that the heavy concussion of a loaded truck passing over the pavement, or the rumbling of the trains of the elevated railroad, produced a painfully high-pitched sound like a whistle.

Physical examination throws but little light on these cases. There is commonly little, if any, affection of the fauces. The Eustachian tube is open, as shown by inflation of the tympanum by either of the ordinary methods. There is no evidence of any collection of fluid in the middle ear. Dr. Sexton punctured the drum in several instances and found the cavity empty. Upon examination with the aural speculum, the external meatus usually contains a certain amount of a tenacious substance which does not appear to be either wax or exfoliated epidermis. It is not unreasonable to suppose that the same exudation takes place in the middle ear. The drum-head is somewhat opaque, only slightly, if at all, injected, and lustreless; and it is often wrinkled about the short process of the malleus, in the antero-superior quadrant,—changes pointing to disease of its internal layer.

This class of cases was first clearly described by Sir William Wilde,<sup>1</sup> under the name of "syphilitic meningitis." Some recent authorities place the seat of this disease in the labyrinth, and Dr. Roosa especially goes still further, and locates it in the cochlea. It is difficult, however, to understand, if such be the case, how patients are able to hear their own voices. Deafness to external sounds but the preservation of autophony, would seem to point to a defect in the conductive apparatus in the ear. We are, therefore, inclined to adopt the following conclusions of Dr. Sexton as an approximation, to say the least, to the true pathology of this affection.

"1. Syphilitic affections of the ears causing sudden deafness, would seem to be induced by a pre-existing hyperæmia of the ears excited by an intercurrent attack of aural mucous catarrh.

"2. This affection speedily causes a disarrangement of the integrity of the chain of ossicles, most likely at the malleo-incudal joint, and probably in some instances at the incudo-stapedial joint, or both of these. The movements of the stapes in the oval window are also likely to be interfered with. The two first-mentioned conditions serve to explain the noises in the ears and the autophony; the last-mentioned condition will increase the anomalies of hearing.

"3. The affection does not depend on anomalies of any portion of the labyrinth, although the latter, of course, is liable to invasion from syphilis, with the nature of which we are as yet unfamiliar."

<sup>1</sup> Practical Observations on Aural Surgery.

These cases of sudden deafness, in the absence of pain, are commonly seen by the surgeon some time after their occurrence, when they are usually found to be incurable. When seen early, very large doses of the iodide of potassium internally and the use of mercurial inunction give some promise of relief and perhaps of cure, and even at a later period the patient should have the benefit of a trial of these remedies.

#### DEAFNESS DUE TO SYPHILITIC AFFECTIONS OF THE BRAIN.

Syphilitic affections of the brain, occurring in such situations as are in direct or indirect relation with the auditory nerves, may, as would be expected, give rise to subjective symptoms in the auditory apparatus. The negative result on inspection of the ear, and the absence of any symptoms referable to the ear itself, will lead us to ascribe the deafness to this cause.

Schwartz considers the following symptoms as characteristic of syphilitic affections of the ear: the trouble is always in both ears and commences several months after the outbreak of other syphilitic manifestations; nocturnal pains in the temporal bones; rapid impairment of hearing, and, finally, early impairment of the transmission of sound through the bones of the head.

#### DISEASES OF THE EAR IN THE SUBJECTS OF CONGENITAL SYPHILIS.

Mr. Jonathan Hutchinson,<sup>1</sup> in 1863, called attention to the frequent occurrence of deafness in the subjects of inherited syphilis, without any adequate changes in the external parts or in the membrana tympani to account for the same. Mr. Hutchinson states that the age at which deafness is most liable to come on appears to be about the same as that at which interstitial keratitis is most frequent, *i. e.*, from five years before puberty to five years after that period. In nearly all the cases reported by him, the loss of hearing affected both ears; in the majority the patients were utterly deaf, and in most of the others the loss of hearing had advanced to a very considerable degree. In most of them there had been some otorrhœa, but of only a mild character.

Mr. Hutchinson adds: "It will be seen that all of the cases in which the ears were inspected go to support the belief that the deafness of syphilitic children is due either to disease of the nerve itself, or to some changes in non-accessible parts of the auditory apparatus. Its symmetry would point to a central cause. In none were there found adequate changes in the membrana tympani, although in none was that membrane quite normal. In all the Eustachian tubes were pervious; my belief therefore is, that the deafness was due either to

<sup>1</sup> Clinical Memoir on certain Diseases of the Eye and Ear, consequent on Inherited Syphilis, London, 1863, p. 174.

disease of the nerves or of their distribution in the labyrinth. The cases constitute the analogues of syphilitic retinitis and of white atrophy of the optic nerves. The prognosis is very unfavorable. From six months to a year would appear to be the usual time required for the completion of the process and the entire abolition of the function."

Dr. Dalby,<sup>1</sup> aural surgeon to St. George's Hospital, states that, next to scarlet fever, inherited syphilis may be regarded as the most fruitful cause of deaf-mutism, as it occurs in children who are born with good hearing power. The patient usually becomes deaf in early childhood—after he begins to talk—or between this period and puberty. With Mr. Hutchinson, he regards the disease as chiefly a nervous one.

<sup>1</sup> Lancet, London, January 22, 1876.