Owing to the difficulty of obtaining pathological material, and of carrying out the microscopic examination of the auditory apparatus, our knowledge of the pathology of syphilitic diseases of the ear is still very meagre. The importance of the subject is undoubtedly great. Congenital syphilis ranks after epidemic cerebro-spinal meningitis and middle-ear suppuration as the most frequent cause of acquired deaf-mutism. There is reason also to believe that many cases of "congenital" deafmutism are really due to intra-uterine syphilis or to syphilitic changes in the ear, occurring before the child has learned to talk. Statistics as to the frequency of deafness in children suffering from congenital syphilis vary very greatly. Some authorities—Carpenter—put it as high as 60 per cent., others at only 33 per cent.

Yearsley found that 3·5 per cent. of 500 children in the L. C. C. deafmute schools were cases of congenital syphilis, Siebenmann of Basle found that 5·6 per cent. of cases of inherited deafmutism were due to syphilis, while Böck (Muench. med. Wochenschr., 1910, p. 2083) obtained a percentage of 8·1. E. Urbantschitsch has examined the Wassermann reaction in 125 deafmutes and found it positive in 33.

Acquired syphilitic disease of the ear is also of importance, on account of the frequent occurrence of sudden and severe deafness in the secondary and tertiary stages. Further, it has recently been found that one-third of the cases of nerve-deafness of unexplained origin have a positive Wassermann reaction—thus pointing to a probably syphilitic origin.

II. Congenital Syphilis.

A. Pathology.

1. In the Fetus and in Infants.—Baratoux, Panse, Mayer, Asai, Hofer, Ranke, and Gruenberg have microscopically examined the middle and inner ears of syphilitic foetuses and infants. The changes found by these observers may be summarised as follows:—(a) Otitis media is of common occurrence not only in infants but also in children born prematurely. In some cases the infective processes involved the labyrinth by rupture of the annular ligament and consequent invasion of the
vestibule through the oval window. (β) Delayed ossification of the petrous bone, with abnormal marrow spaces. (γ) Intra-uterine meningitis and neuritis of the eighth nerve. (δ) Baratoux alone has noted changes in the vessels, though he also found purulent otitis media and interna. (ε) Hæmorrhages in the middle and inner ear have often been observed, but such findings must be discounted, as they were probably due to suffocation. (ζ) Panse, however, holds that the haemorrhage is of syphilitic origin. Changes in Corti's organ and in other parts of the membranous labyrinth and nerve apparatus, especially in the spiral ganglion, have occasionally been noted, but may in some cases be explained by post-mortem changes. Gruenberg has demonstrated spirochaetes in microscopic sections from the ear of a seven-months' foetus, especially in the cochlear and vestibular nerves, the facial nerve, and in the tympanic and carotid plexuses. Spirochaetes were also found in the neighbourhood of the vessels of the middle ear, as well as in the marrow spaces of the ossicles. There were no spirochaetes in the labyrinth or in the nerve endings in the inner ear. Alexander states that the most severe forms of ear syphilis occur in intra-uterine life. The new-born infants show all the signs of congenital deafness, and the static labyrinth is not excitable. The abstractor has examined both ears from a syphilitic foetus of seven months, and found otitis media on both sides; abnormal marrow spaces between the cartilage and periosteal bone; small cell infiltration of the eighth nerve. Changes were present in the membranous labyrinth, but these appeared to be of a "post-mortem" nature.

2. In Young Children.—M'Bride (Diseases of the Throat, Nose, and Ear) states that, in children suffering from congenital syphilis, what appears to be a case of simple Eustachian catarrh, runs an unfavourable course. Treatment does no good, and the drumheads remain thickened and indrawn while the deafness is severe and persists. M'Bride further says that in children hereditary syphilis often gives rise to a combination of middle-ear catarrh and labyrinthine deafness. From this it would appear that, if cases of middle-ear catarrh do not rapidly clear up—especially if they are accompanied by severe deafness—it is advisable to have the Wassermann or Noguchi reaction tested.

Alexander (Ohrenheilkunde des Kindesalters) says that middle-ear catarrh is common in syphilitic children and appears to be a syphilitic manifestation. There is only slight or no improvement after the absorption of the exudate. The middle-ear symptoms are thus replaced by those of an inner-ear lesion. These catarrhal affections develop into conditions which clinically correspond to otosclerosis. The prognosis is not good on account of the poor nourishment of the child. According to Cheatle, Eustachian obstruction occurs in the "snuffling" stage of congenital syphilis. Cheatle holds that syphilis alone will not cause suppurative otitis media; pyogenic organisms are necessary.
Urban Pritchard calls attention to those cases of deafness in children where removal of adenoids does no good. He holds that these are due to syphilis. Madden (quoted by Cheatle) describes a case of gumma of the middle ear in which there was a deep opening behind the ear discharging thick caseous material. At the operation carious bone was removed, but the child died after repeated bleedings from the sinus. The autopsy revealed a large gumma involving the petrous pyramid. Gummata were also present in the dura mater and in the parietal and occipital bones.

3. Late Type of Congenital Syphilis, in which the deafness occurs usually between the seventh and thirteenth years.

(a) Deafness due to a Neuro-Labyrinthitis spreading from the Meninges.
—Siebenmann and Mayer believe that the condition is due to a syphilitic meningitis which extends to the labyrinth. Mayer holds that this is a recurrence or an exacerbation of the condition which he has found in the internal auditory meatus of still-born syphilitic infants (vide supra). It is claimed in support of this view that Kretschner and Tobler have found lymphocytosis in the cerebro-spinal fluid in older cases of congenital syphilis.

(β) Vascular Changes.—Baratoux believes that the late form of congenital syphilitic deafness is due to changes in the vessels (endarteritis).

(γ) Otitis Media followed by Paralabyrinthitis or Invasion of the Labyrinth.—As will be seen later, many otologists agree that the tympanic membranes are seldom normal in the late form of congenital syphilitic deafness. This points to a past attack (or attacks) of otitis media. Politzer states that congenital syphilitic disease of the labyrinth is often associated with middle-ear catarrh or suppuration. The abstractor holds that, at least in some cases, the late form of deafness is due to syphilitic otitis media—possibly with mixed infection—which either invades the bone of the labyrinth capsule, giving rise to a chronic form of osteomyelitis, or breaks through into the hollow spaces of the inner ear, causing labyrinthitis. In support of this view of the pathology the abstractor puts forward the following case, for permission to record which he is indebted to Dr. Logan Turner:—D. W., male, aged 20 years, had measles at the age of 4 years, followed by otorrhoea. The father died of aneurysm at the age of 35; the mother stated that she had had four still-born children before the birth of the patient; afterwards she had three more still-born children. At the age of 9 years the patient had interstitial keratitis and 6 months later syphilitic ulceration of the pharynx, followed by contraction. At this period also the boy became markedly deaf, and within a year the deafness was complete. At the age of ten years enlarged glands (syphilitic or tubercular?) were removed from the neck. The patient's speech was very indistinct, but his mother
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could understand it. For some time before his admission to the Infirmary the patient had been very drowsy. Examination showed that he was emaciated. The incisor teeth were notched and the corneas very cloudy. Syphilitic scars were noted round the knees. Wassermann reaction negative. The breathing was so noisy as to keep other patients awake at night. Only fluid food could be swallowed, on account of stenosis of the hypopharynx. The nasopharynx was entirely shut off from the mouth and oesopharynx by scar tissue. Suspension laryngoscopy revealed marked cicatricial narrowing of the upper aperture of the larynx. The patient died suddenly. Autopsy showed syphilitic changes in the skull-cap, brain membranes, lungs, heart, aorta, and liver. The brain was markedly oedematous. As an example of congenital syphilitic disease of the ear the case is not free from objection, e.g. Wassermann reaction negative (this is not uncommon in congenital syphilis in patients who reach 20 years), and presence of suppurative otitis media (to the abstractor this appears quite natural). Enlarged glands were removed from the neck (these also may have been syphilitic).

Microscopical examination of the middle and inner ears on both sides showed the same conditions—purulent otitis media, with great thickening of the submucous tissues; ankylosis of the head of the malleus to the outer attic wall; invasion of the marrow spaces surrounding the bony capsule of the labyrinth by a chronic form of osteomyelitis, with numerous giant cells but no caseation; marked erosion of the bony labyrinth capsule by the osteoclastic marrow; invasion of the semicircular canals by erosion of their bony walls and formation of granulation tissue in the perilymph spaces in such a way as to occlude the perilymph and endolymph spaces; marked dilatation of other parts of the membranous labyrinth, utricle, sacculi, and membranous cochlea. The two divisions of the eighth nerve, along with the facial nerve, were normal.

The abstractor also claims that Walker Downie's classical case supports this last view (γ) of the pathology of congenital syphilitic deafness. Downie's patient became deaf at the age of eleven years. The deafness was accompanied by severe pain in both ears, though there was never any discharge. (All cases of otitis media do not suppurate and discharge.—Abs.) The deafness became complete in six months. Otoscopy showed the drumheads to be indrawn and opaque (as if from old otitis media.—Abs.).

The patient died from meningo-encephalitis resulting from a fungating gumma of the right parietal bone. Macroscopic examination of the ears showed that the seventh and eighth nerves were healthy. The middle ears were normal (at the time of death). The semicircular canals were obliterated by new bone formation and the vestibule largely filled up. The modiolus of the cochlea was thickened.
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These findings appear to correspond to a stage of sclerosis and ossification following an attack of labyrinthitis.

B. Clinical Aspect of the "Late" Form of Congenital Syphilitic Deafness.

This is not the place to enter in detail into the symptoms and signs of congenital syphilis, but attention may be called to the wizened appearance of the child at birth, the presence of skin lesions, enlargement of the liver, spleen, and lymphatic glands, nasal catarrh (snuffles) albuminuria, and disease of the long bones, as also of those of the face and head. Headache is common, and it is stated that the presence of severe nocturnal headaches in a child should suggest congenital syphilis.

Hutchinson, in 1861, first described the three cardinal symptoms and signs of the late form of congenital syphilis—notched, peg-shaped incisor teeth, cloudy cornea, and deafness. Wanner states that the deafness usually comes on between the ages of 7 and 9 years, or between 11 and 13 years, though later cases are not uncommon. (Cheatle reports a case aged 54.) Females are affected much more frequently than males (15 to 6), whereas in other forms of ear disease the proportion is 6 males to 4 females. As a rule the eye trouble (interstitial keratitis) comes on three or four years before the deafness. Cheatle states that choroiditis or iritis may be present though the cornea are clear. Sometimes there is a recurrence of the eye trouble with the onset of deafness. "Hutchinson" teeth are present in 50 per cent. of cases.

The deafness may come on gradually or suddenly (apoplectiform type). In some cases it is stated that the deafness comes on in a single night. It may or may not be accompanied by giddiness and disturbance of balancing. Stuempke states that in hereditary cases the deafness is usually gradual and bilateral, while in secondary acquired syphilis it is sudden and unilateral.

Otoscopy.—Politzer, Bezold, Wanner, and Hopmann all agree that the drumheads are very seldom normal. Of 33 cases examined by the abstractor, the tympanic membranes were only normal in 4, and of 31 cases observed by Nager only 9 were normal.

Functional Examination (Cochlear Apparatus).—As a rule the bone-conduction is greatly shortened and may be altogether absent. The watch is not heard by bone- or air-conduction. The upper tone-limit is much reduced, while the lower tone-limit is normal or raised. Bezold states that both ends of the scale of hearing are contracted in most cases until, finally, only an island remains; even this may eventually disappear. Rinne's test is usually positive, i.e. air-conduction is better than bone-conduction, and the tuning-fork, when placed on the middle of the vertex, is heard in the better ear. In a few cases, on the other hand, the deafness belongs to the middle-ear type.
Vestibular Apparatus.—Of 24 cases examined by the abstractor, the vestibular reaction to rotation and cold syringing was absent in 14, reduced in 7, and only normal in 4. Bárány states that the vestibular apparatus fails to react in almost all cases of congenital syphilitic deafness. Hennebert (La Presse Oto-laryngologique Belge, 1909, No. 5) was the first to call attention to the presence of “compression” nystagmus in some cases of congenital syphilitic disease of the ear. Increase of the air-pressure in the external meatus by means of a valveless Politzer bag causes a slow movement of the eyes to the same side, while aspiration brings about a slow movement to the opposite side. Hennebert finds that in these cases the rotation reaction is always absent while the caloric reaction is reduced. Buys and Alexander have also reported cases. Alexander thinks that Hennebert’s sign is due to a change in the nerve endings, while Bárány holds that it is due to excessive mobility of the stapes. The abstractor believes that his findings (vide supra) in the case of congenital syphilitic disease of the ear point to another explanation—the bony wall of one of the canals is eroded by inflammatory changes in the marrow, and the hollow spaces of the canal are partly filled up by syphilitic granulation tissue, so that the rotation and caloric reactions cannot be produced. The more powerful pneumatic test is, however, able to bring about an abnormal or reversed vestibular reaction.

Diagnosis.—According to Alexander, congenital syphilitic deafness must be diagnosed from the following conditions:—(1) Idiopathic atrophy of the auditory nerves—a rare condition. Many supposed cases are really syphilitic. (2) Hereditary non-syphilitic nerve deafness. (3) Atypical cases of otosclerosis. Bezold advises that suspected cases should be examined for the presence of ozæna; saddle-back nose; perforation of the nasal septum; scars on the palate, pharynx, and angles of the mouth; deformity of the skull. Many cases show defective mental development and infantilism. The abstractor has found that a clinical history somewhat like the following can often be obtained from the patient’s mother:—The patient is the eldest living child, but the mother had numerous miscarriages and still-births before the patient was born. These miscarriages, etc., occurred at gradually lengthened periods of pregnancy. Sometimes two or more members of the family show the typical Hutchinson triad of symptoms. Glover reports a case in which the eye and ear symptoms of congenital syphilis were found in the second generation, but as this is the only case on record it appears to be open to considerable doubt.

Fowler advises the application of Noguchi’s skin test in the diagnosis of doubtful ear cases. The Wassermann reaction is, of course, an alternative. (See Treatment, section IV.)
III. ACQUIRED SYPHILIS.

A. External Ear.

1. *Primary syphilis* of the outer ear is rare. Sohier Bryant has collected 32 cases of this condition. The disease is inoculated by kissing, biting, the use of infected towels, or by piercing the ear with infected instruments. Barbier records a case of phagedenic chancre due to piercing the ear. He states that Chinese barbers use the same instrument for cleaning the external meatus that they employ for removing condylomata.

2. *Secondary syphilitic* affections of the external ear and meatus appear to be more common. Desprès has observed 5 cases with papules in the external meatus among 1200 cases of syphilis. Fowler, Pogany, Guettich, Klauss, and Beck record cases of secondary affection of the external ear at periods of from 4 to 7 months after the primary sore. Knapp states that condylomata of the external meatus begin as red efflorescences, which gradually enlarge, and are followed by diffuse swelling of the meatal walls combined with secretion. Panse (*Pathologische Anatomic des Ohres, 1912*) says that pigmented scars and contraction may follow healing. In most instances the condition appears to be *secondary to syphilitic middle-ear suppuration*. Klauss says that condylomata may cause severe pain, while Pritchard states that they may be mistaken for epithelioma.

*Treatment.*—In Fowler's case an injection of salvarsan was given, and 5 days later it was found that the auricle had improved, while in a week all manifestations had gone except the scar in the tympanic membrane. Beck advises the local application of calomel, while Pogany uses white precipitate ointment or corrosive sublimate drops.

3. *Tertiary Syphilis.*—Cheatle states that the auricle and external meatus may be the seat of gummatous ulceration. M'Bride (*Diseases of the Throat, Nose, and Ear*, p. 490) reports a case of deep ulceration of the auricle.

B. Middle Ear.

1. *Primary Sore.*—West states that over 100 cases are on record of chancre at the orifice of the Eustachian tube. (These cases belonged mostly to the pre-antiseptic days, in which unsterilised vulcanite Eustachian catheters were transferred from an infected nose and naso-pharynx to a healthy one. Modern methods have rendered primary syphilis of the nose and naso-pharynx of very rare occurrence.—Abs.)

2. *Secondary Syphilis.*—This is probably of much more common occurrence than is usually supposed. In the abstractor's opinion investigation of this subject would well repay the time and trouble
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expended. Cheatle states that syphilitic naso-pharyngitis may spread up the Eustachian tube to the tympanum and give rise to the symptoms and signs of catarrhal otitis media. Bezold says that if the labyrinth is affected in the secondary stage of acquired syphilis one usually finds otitis media as well. As a rule only one ear is involved. The onset of deafness in these cases is usually rapid. Bourgeois reports a case of middle-ear suppuration cured by anti-syphilitic treatment. The onset of otitis was not painful, but the condition lasted 3 months. The case was cured within 10 days under specific treatment. M'Bride (p. 663) finds that in acquired syphilis the patient may become suddenly deaf at a varying period after infection. Tinnitus is marked, and giddiness may be present. The immediate cause may be sea-bathing (several cases). The drumhead is thickened, but inflation produces no improvement in hearing.

3. Tertiary Syphilis of the Middle Ear.—Politzer states that syphilitic otitis media is usually due to an affection of the naso-pharynx. The otitis may be a simple catarrh, a suppurative inflammation, or an adhesive process. The deafness is usually marked. Ulceration of the mucous may occur, and be followed by caries and necrosis of the tympanic walls associated with facial paralysis. The diagnosis is difficult as the appearances are not characteristic, but rapid destruction of the drumhead and loss of bone-conduction are in favour of syphilis. Politzer states that syphilitic disease of the labyrinth may appear along with the otitis media.

Otitis media in the tertiary stage of syphilis appears to be of common occurrence. The affection may not be entirely due to the spirochete of syphilis, but at any rate it appears that the presence of syphilis alters the clinical course of otitis media. Syphilitic disease (gumma or ulceration) of the mucous membrane of the naso-pharynx is usually present. In most cases disease of the labyrinth is combined with the otitis media (syphilitic pan-otitis). It appears that in these cases the middle ear has little power of repair, and the labyrinth but small capacity for resistance.

Gruenberg records a case of a male, aged 21, who had had syphilis for 2 years. The naso-pharynx was ulcerated. Within 6 months of infection there was nerve deafness with tinnitus. Death occurred from syphilitic cachexia. Post mortem the left ear showed serous exudate in the tympanum, with thickening of the mucosa; the drumhead was intact. The incus was embedded in inflammatory tissue and the bony wall of the promonotory was thickened. The bony capsule of the cochlea showed wide marrow spaces. In the membranous labyrinth there were only slight changes—atrophy of the spiral ganglion in the basal coil. There were no syphilitic changes in the vessels.

M'Bride (Diseases of Throat, Nose, and Ear, p. 548) reports the case
of a middle-aged man, suffering from acute otitis media, with severe pain, headache, and a copious discharge of pus. A mastoid operation was proposed. A history of syphilitic infection was obtained and iodide given. Within 3 days the pain had gone, the discharge rapidly stopped, and the membrane healed.

Ritter records a case in which there were tertiary syphilitic ulcers at the ostium of the Eustachian tube on both sides. Bilateral catarrhal otitis media was also present. Bruhl reports a case of a male, aged 35, who had contracted syphilis 16 years before. The patient suffered from severe headaches, tinnitus, and deafness, followed later by facial paralysis, giddiness, and nystagmus. The right ear showed catarrhal otitis. A tender swelling formed on the mastoid, but under treatment with mercury and iodide the swelling disappeared in 14 days. The giddiness and tinnitus improved, but the deafness remained.

Dan M'Kenzie records a case of sudden bilateral deafness in a tertiary syphilitic while the patient was under treatment by mercury and iodide. The patient was suffering from gumma of the cervical glands, which were excised on a diagnosis of tuberculosis. The wound healed well but the gland trouble recurred. Brunk has demonstrated a case of gumma of the mastoid accompanied by syphilitic affection of the floor of the nose. Braun had a case of syphilitic infection in a male, aged 31, who had undergone mercurial treatment at the time of the chancre 6 years before. Braun found a gumma of the right inferior turbinal and right tonsil. The drumhead on this side showed evidence of catarrhal otitis media. Deafness and tinnitus came on rapidly and the bone-conduction was shortened. Vestibular reaction present. Salvarsan caused rapid improvement.

Lueders reports 3 cases of tertiary syphilis in which the middle ears were affected. As usual the labyrinth was involved in all, while in one the facial nerve was paralysed. Specific treatment resulted in the cure of the middle ear condition, but the deafness remained. Beck has shown a case of gumma of the middle ear in a patient who had syphilis 20 years before. Cheatle describes 3 cases of gumma of the naso-pharynx with deafness due to Eustachian obstruction.

Pogány comes to the conclusion that, in diseases of the middle ear, if the bone-conduction is greatly shortened, we must suspect syphilis. He records 2 illustrative cases.

Bárany has recorded a case of Hennebert's sign (vide supra) in a case of acquired syphilis. For 10 years the patient had suffered from deafness and tinnitus in the right ear with nystagmus. Movements of the head, however, caused no giddiness. The rotation reaction was normal but the caloric reaction was diminished. Compression brought about slight movements of the eyes along with giddiness.

It will thus be seen that in tertiary syphilis of the middle ear the labyrinth is almost always involved.
C. Labyrinth and Eighth Nerve—“Neuro-Labyrinthitis.”

In the present state of our knowledge it appears to the abstractor to be impossible to distinguish clinically between syphilitic diseases of the labyrinth and those of the eighth nerve. It is true that in affections of the labyrinth alone the galvanic reaction is retained, while in those of the eighth nerve it is lost; but data on this point are given in so few instances that, for the present, cases must be grouped under the title of “neuro-labyrinthitis.”

Beck (Münch. med. Wochenschr., 1913, p. 2778) finds that in 80 per cent. of syphilitic patients the bone-conduction is more or less shortened although the hearing is normal. Beck attributes this to raised intracranial pressure, as he finds the same condition in cases of brain tumour and hydrocephalus. Beck performed lumbar puncture on some of his syphilitic cases and found that after this the shortening of the bone-conduction disappeared for a time, though it returned in two or three days. Cases in which the primary sore is still present and the Wassermann reaction is negative do not show this shortened bone-conduction. The latter only appears with the constitutional symptoms. The sign may be of value in the diagnosis of syphilis.

1. Secondary Syphilis.—Frey states that cases of neuro-labyrinthitis in secondary syphilis are much more common than is usually supposed, and claims that the work of Moriac in 1879 proves this. Habermann, in 1896, reported 66 cases of disease of the acoustic nerve, of which 34 occurred in the secondary stage. Mayer has collected 65 cases occurring since 1896, and of these 13 belonged to the secondary stage. Labyrinth symptoms, i.e. tinnitus, deafness, giddiness, nausea, vomiting, disturbance of balancing, and vestibular nystagmus have occurred as early as the seventh day after the appearance of the primary sore (Politzer, quoted by Pogány, M. f. O., 1913, p. 1333). Mayer states that the trouble may come on three weeks after the appearance of the primary sore. At the time of onset of the first skin rash neuro-labyrinthitis is by no means uncommon. The pathology of the condition is unknown, but Stuempke holds that the membranous labyrinth is involved by a syphilitic infiltration.

Beck has examined 600 cases of secondary syphilis and finds that complaints of giddiness and disturbance of balancing are by no means rare. In some of these nothing abnormal is to be found in the ears or nervous system. In others there are changes in the posterior cranial fossae, as evidenced by nystagmus of central origin and cerebellar disturbance of balancing. The third group consists of vestibular cases which show alteration in the galvanic irritability of the vestibular nerve. At the onset tinnitus is present in most cases, while giddiness is noted in 50 per cent. These symptoms may be observed before the skin rash appears. In the later stages of
secondary syphilis the ear trouble almost always comes on along with a recurrence of the rash (Mayer). Disturbance of hearing is gradual and may vary in intensity; it is usually bilateral. Functional examination shows an affection of the eighth nerve. Didsbury reports the case of a man, aged 28, who became deaf (within a few days) only four months after infection. Paracusis lontainé was present, i.e. the tuning-fork was not heard on the mastoid but was heard on the knee or foot. According to Roosa, diplacusis is a symptom of syphilitic disease of the eighth nerve, and in this opinion Freytag agrees, as the paracusis is accompanied by giddiness and loss of certain tones.

Vestibular symptoms vary from slight giddiness up to vomiting and disturbance of balancing. Mayer has not observed in secondary syphilis a case of isolated disturbance of the vestibular apparatus such as has been reported in several cases after the injection of salvarsan. According to Beck, the cochlear branch of the eighth nerve is more sensitive to toxins than the vestibular, e.g. quinine, salicylates, arsenic. Bárány, Bondy, Neumann, and Beck have, however, observed cases of isolated loss of conduction of the vestibular apparatus in the secondary stage of syphilis without "606." Neuro-labyrinthitis may occur whilst the patient is undergoing mercurial treatment, as is shown by the case of Neumann, whose patient woke up one night after his fifteenth mercurial injection with marked tinnitus in the right ear, soon followed by giddiness, vomiting, and cold sweats; the vestibular reaction was diminished. After the seventeenth injection there was sudden abducens paralysis on the same side. Salvarsan was given, but proved useless.

The prognosis appears to be better in the neuro-labyrinthitis of secondary syphilis than the form which occurs in the tertiary stage.

Treatment.—Alexander and others hold that it is dangerous to give salvarsan alone if there be any affection of the auditory nerve apparatus. Mercurial treatment should always be combined with “606.” Citelli reports three cases of neuro-labyrinthitis in recent syphilis in which salvarsan acted promptly after other remedies had failed. Lowinski has observed a patient who suffered from sudden occipital headache, followed in two days by facial paralysis (primary sore still present). Wassermann-reaction+. Five days after an injection of salvarsan the paralysis had disappeared. Möller, on the other hand, had a case in which salvarsan failed but strenuous mercurial treatment resulted in slight improvement. Stein’s patient was greatly improved by injections of salicylate of mercury.

2. Tertiary Syphilitic "Neuro-Labyrinthitis."—In the tertiary stage of acquired syphilis cases of neuro-labyrinthitis are not infrequently met with. In several of the recorded cases paralysis of other cranial nerves was also present, e.g. facial, oculomotor, glosso-pharyngeal, etc. It would appear therefore that these cases approximate closely to those
in which the eighth and other nerves are involved by gummatous infiltrations of the brain membranes at the cranial base (see later). Cheatle records the case of a man of middle age who contracted syphilis in 1894 and was treated with mercurial pills till 1900. In 1909 he suffered from pain and tingling in the right arm and right side of the face, and in 1910 suffered from tinnitus, deafness, and pain in the left ear, along with unsteady gait. Cheatle saw the case three weeks later and found the drumheads normal. Nerve deafness was present. Inunction of mercury improved the condition. Cheatle records two other similar cases. In one of these there was a gummatous mass to be felt in the naso-pharynx. West records the case of a female who suffered from an (acquired) tertiary affection of the seventh and eighth nerves. There was total deafness in one ear, with complete facial paralysis. Wassermann reaction+. The facial paralysis cleared up under mercury and iodide but the deafness remained. Harmon Smith (New York Med. Journ., 17th March 1906) records a case of syphilitic necrosis of the temporal bones. Double optic neuritis was present, along with paralysis of the facial and glosso-pharyngeal nerve, deafness, and disturbance of balancing. Operation revealed necrosis reaching through the vestibule and cochlea to the apex of the petrous pyramid and backwards to the foramen magnum. Death occurred in spite of energetic anti-syphilitic treatment.

Wassermann Reaction in Nerve Deafness of Obscure Origin.—All otologists know how often they have to be content with a diagnosis of "nerve deafness of obscure origin" after they have excluded such causes as senile or arteriosclerotic changes, to the patient's occupation (boiler-makers, rivetters), injury, the various infectious diseases, including influenza and cerebro-spinal epidemic meningitis, labyrinthitis following middle-ear suppuration, anaemia and other blood diseases, toxic neuritis as from quinine or salicylates, etc. Such unexplained cases have been investigated as regards the Wassermann reaction by Pogany, Zange, Beck, Busch, and Arzt. In all, these observers have tested 209 cases and found the Wassermann reaction positive in 77, i.e. 36 per cent.

3. Acquired Syphilitic (Gummatous) Affections of the Eighth Nerve.—Rosenstein (C. f. O., 1905, p. 165) holds that syphilitic affections of the eighth nerve are rare as compared with similar diseases of the labyrinth. The central portion of the nerve may be involved by gum mata, aneurysm, or scars, while the peripheral part may be affected, along with other cranial nerves, by gummatous infiltration of the meninges and nerve sheaths. These conditions are often preceded by severe headache—sometimes of long duration. Rothmann (quoted by Rosenstein) describes the case of a male who suffered from headache, nausea, vomiting, diplopia, dysphagia, with ptosis, and loss of smell on the right side. There was also hoarseness, and pain in the left arm and leg. Examination revealed optic atrophy and paralysis.
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of the third, fourth, fifth, sixth, seventh (paresis), and eighth nerves. The other cranial nerves were also involved, as shown by paresis of the palate, disturbance of taste, mastication and swallowing. The recurrent laryngeal nerve was paralysed, but the pulse and respiration were not disturbed. All three cranial fossae were evidently affected. The patient recovered under mercurial inunction. It is noteworthy that both ears showed purulent otitis media, and Rosenstein states that it is by no means uncommon to find this combination of otitis media with multiple paralysis of the cranial nerves. Similar cases are recorded by Mendel and Rosenstein. Kobner's case (quoted by Rosenstein) was that of a man, aged 36, who had had severe headache and depression for two years. There was hyperaesthesia of the face and of hearing. Later, sudden paralysis of the swallowing apparatus occurred. The autopsy revealed exostoses of various forms and sizes at the base of the skull; the brain membranes were thick and cicatricial.

4. Affections of the Eighth Nerve in Tabes.—Ramsay Hunt describes five cases of otalgia due to tabes. In one case degenerative changes were found post-mortem in the pars intermedia of Wrisberg. Cheatle has observed one case of this kind. Haug (Die Krankheiten des Ohres, etc., 1893) reports a tabetic case in which the acoustic nerve and ganglion were affected. Beck states that an affection of the cochlear nerve may be an early symptom of tabes. He reports the following case: Male, aged 34, had syphilis at the age of 22, and was treated by inunction of mercury. Severe headaches at times.—Wassermann reaction +. In the last five years the patient had suffered from deafness and tinnitus in the left ear, and of late the right ear had also been affected. The middle ears were normal. The left ear was quite deaf and the right could only hear a loud noise. Rombergism present. Vestibular reactions normal. Mercurial inunctions were of no benefit.

IV. Treatment of Syphilitic Affections of the Ear.

Treatment.—Such questions as that of making syphilis a notifiable disease and of the careful regulation of the marriage of syphilitic persons do not come within the scope of the present résumé. Nevertheless it remains true that the prevention of syphilitic deafness would be much more satisfactory than the more or less unsuccessful attempts to cure syphilitic affections of the ear once they have occurred. Much was hoped for at one time from salvarsan, but this remedy has proved very disappointing in the treatment of ear syphilis—indeed many, as will be seen later, regard it as a frequent source of ear troubles. Mott (Brit. Med. Journ., 1915, vol. i. p. 192) points out that, as the normal cerebro-spinal fluid contains no protein or leucocytes, spirochætes find in it a safe retreat, and are with great difficulty eliminated, because mercury and arsenic do not pass through the choroid plexus.
With regard to congenital syphilis, Findlay and Robertson (Glasgow Med. Journ., 1914, vol. ii. p. 401) hold that antenatal treatment gives the best results. They have treated seven pregnant women suffering from syphilis with salvarsan combined with mercurial inunction. None of the babies showed evidence of syphilis and in no case was pregnancy interrupted. Boardman (Journ. Cutan. Dis., 1914, p. 545) states that congenital syphilitic infants should be treated for three or four years by means of mercurial inunction combined with neo-salvarsan. The latter is injected into the external jugular or scalp veins at intervals of two or three weeks (dose 0.015 gr. per kilo of body weight).

One of the great difficulties in treating cases of congenital syphilitic ear disease is the fact that children are seldom brought when the deafness first comes on. As a rule a period of months or even years elapses—at least in hospital work—before the cases are seen. If the condition were at once diagnosed by the patient's doctor the prognosis might be somewhat better. Ordinary anti-syphilitic treatment appears to be of little value, for several cases are on record in which the deafness appeared while the child was undergoing mercurial treatment on account of the eye trouble. Wanner, however, advises inunction of mercury combined with the administration of potassium iodide. Cheatle states that if thyroid extract be given along with mercury and iodide a better result may be obtained. Alexander favours the use of mercurial injections followed by salvarsan. Belam reports a case benefited by mercurial inunction, but states that the deafness is apt to recur. Bezold holds that iodide is of no value. Dench has found that in some cases injections of pilocarpine are of great benefit. Pritchard believes in local blistering. Politzer says that we should try pilocarpine first of all, while iodide and mercurial inunctions should be used later. McBrice (p. 694) records a case of deafness due to congenital syphilis which cleared up, apparently, under treatment with pilocarpine, so that a stranger could not tell that the patient was deaf. The only trouble that remained was that melodies seemed discordant or painful. Siebenmann has seen great improvement after the administration of salvarsan, but Scheibe does not agree with this statement. So far as the abstractor knows, no cases have been reported in which the new remedies "intramine" and "ferrivine" have been used in syphilitic disease of the ear. On the whole, it must be admitted that treatment appears to be of little value. It is very important, however, that the speech should be retained, and, for this reason, the child should, as soon as possible, be admitted to a deaf-mute school. Unless this be done the speech will rapidly deteriorate, so that within a few months in the case of younger children—up to a year in the case of older children—the speech cannot be understood or may be altogether lost.

In the treatment of acquired syphilitic ear diseases, Stuempke (C. f. O.,
1909, p. 450) advises the use of mercury as soon as possible. He records two cases of secondary syphilis in which this drug caused a rapid improvement, not only in the vestibular but also in the cochlear condition. Collins (Brooklyn Med. Journ., November 1905) reports six cases of labyrinth syphilis which rapidly improved under mercury and pilocarpine. Beck (C. f. O., 1911, passim) states that mercury is the best remedy in early syphilitic lesions of the eighth nerve; he uses inunctions or injections. Vály (M. f. O., 1914, p. 260) holds that mercury and potassium iodide are indicated, not only in the secondary but also in the tertiary stages. Guastoni (C. f. O., 1908, p. 531) has obtained good results from injections of corrosive sublimate into the veins according to the method of von Baccelli. Braun (C. f. O., 1911, p. 297) records a case of syphilitic infection of the inner ear cured by salvarsan (vide supra). The methods of Swift and Ellis and of Ogilvie of introducing salvarsanised serum into the spinal canal were recently described by Eason in an abstract in the Journal. So far as the present abstractor knows, these methods have not hitherto been employed in dealing with syphilitic neuro-labyrinthitis.

V. SALVARSAN AND THE EIGHTH NERVE.

Since the introduction of salvarsan attention has been called to the frequency of cases of "nerve relapse" after the injection of the remedy. In many of these the eighth nerve is involved—alone, or along with others. As we have already seen in an earlier part of this résumé, cases of neuro-labyrinthitis are by no means rare in the secondary stage of syphilis, quite apart from injections of salvarsan. The question then is—Have such cases become more frequent than they used to be in the pre-salvarsan era? It is by no means easy to give a confident answer. Some authorities, such as Alexander and Bruehl, hold that syphilitic neuritis of the eighth nerve was rare in the early stages of syphilis before the introduction of "606." They believe that the salvarsan is to blame. Ehrlich, Benario and Frey, on the other hand, maintain that the nerve trouble is due to syphilis and not to the remedy. Ehrlich contends that salvarsan produces a sterilisation through the blood, except in some of the osseous canals through which the cerebral nerves pass. Here the circulation being sluggish, a few spirochetæ escape the action of the remedy, and proliferate, causing an inflammation and consequent swelling, with pressure upon the nerve, thus interfering with its function. Nichols (Journ. Amer. Med. Assoc., 2nd March 1912) suggests that after salvarsan, which kills most of the spirochetæ at once, no natural resistance is established so that a focus in the nervous system, which has escaped the salvarsan, can develop unopposed. The best way to prevent nerve involvement is to administer mercury and potassium iodide along with salvarsan, and to repeat the injections as often as necessary.
Frequency of Nerve Relapse after Salvarsan.—Statistics on this point vary very greatly. Finger, Kren, Biehl, and Frey have (together) observed only 6 cases of nerve relapse affecting the eighth nerve in 732 cases injected with salvarsan. Benario has collected 62 cases of eighth nerve affection out of 14,000 treated with salvarsan. On the other hand, Zeisl and Botella (together) have noted 28 cases out of 266 patients injected. Botella thinks that these lesions of the auditory nerve apparatus may be due to—(1) Herxheimer reaction; (2) nerve relapse; (3) increase of an existing defect; or (4) arsenical neuritis.

From the clinical point of view the cases may be divided into two groups—(A) early, and (B) late.

A. Early Cases.—The trouble may come on within a few hours—up to a day—after injection. This form is usually regarded as an example of Herxheimer reaction due to swelling of the nerve in the narrow bony canal of the internal auditory meatus. (The Herxheimer reaction is said to occur where spirochaæte are present, or where syphilitic processes are going on.) The isolated affection of the vestibular apparatus is characteristic of this form of the affection. Rapid and complete recovery appears to be the rule.

Beck records the following illustrative case:—A girl of ten years was infected extragenitally by her sister who had syphilis. Salvarsan was given intravenously, and six hours afterwards there was marked giddiness and nausea, with loss of vestibular reaction on one side. By next day the condition had cleared up. Other similar cases are reported by Beck, Finger, and others.

B. The late form occurs from four weeks to six months after the injection. In several of the reported cases otitis media was present. Beck (Muench. med. Wochenschr., 1911, p. 2217) records the case of a male, aged 28, who got an intravenous injection of salvarsan at the time of the first skin rash. Six weeks later there was marked tinnitus and deafness in both ears. The vestibular apparatus showed loss of reaction. Beck states that arsenic was found in the urine four months after the injection of salvarsan. The case was treated with salicylate of mercury. The vestibular apparatus recovered, but the deafness remained. Somewhat similar cases are recorded by Kinck (M. f. O., 1911), Neumann (C. f. O., 1912, p. 124), and Rimini (Deut. med. Wochenschr., 1913, No. 2, p. 71). Neumann’s case is of special interest:—Male, aged 20, was injected with salvarsan. Four weeks later tinnitus was present and the gait was uncertain; high tones lost, but other tests normal. Three months later the symptoms recurred. Rotation reaction lost; caloric reaction diminished; galvanic reaction normal. Neumann draws the conclusion that the retrolabyrinthine portion of the nerve was normal because the galvanic reaction was present. The endolymph apparatus was also normal, because the caloric reaction was present. The loss of the rotation reaction Neumann
explains by a lesion of the first neurone, sufficient to do away with the physiological stimulus (rotation), but not enough to abolish the irritability for artificial stimuli, e.g. cold syringing and galvanism.

It seems quite possible that both factors—syphilis and salvarsan—may be to blame, as both are nerve poisons.

To summarise the arguments against the view that salvarsan is the cause of the ear trouble—(1) Cases of neuro-labyrinthitis were common during the stage of secondary syphilis before the days of salvarsan (Mauriac, Paris, 1879), and are not infrequently met with at the present time in cases in which “606” has not been employed. (2) Similar attacks are recorded during treatment by mercury and iodides. (3) Examination of the cerebro-spinal fluid in cases of “nerve relapse” appears to show that the condition is due to syphilitic meningitis (Voss, C. f. O., 1913, p. 291). When the optic nerve is affected the changes appear to be inflammatory (Schanz and Tobias). (4) Cases in which salvarsan is vigorously pushed suffer less than those which receive a single dose (Ehrlich). (5) Syphilitic affections of the inner ear may be improved by the injection of salvarsan (Lange, C. f. O., 1912, p. 277). (6) In the early stages of syphilis an endotoxin is formed which is liable to injure the acoustic, optic, and facial nerves or their terminations (Willeutt, Journ. Amer. Med. Assoc., 1915, vol. ii. p. 602). (7) Long interval between the injection of “606” and the appearance of the labyrinth symptom, at least in some cases.

In Favour of the View that Salvarsan is the Cause of the Ear Trouble.

(1) Deafness has followed the use of salvarsan in a non-syphilitic case (lichen ruber) quoted by Lange (C. f. O., 1912, p. 277).
(2) Meningitic changes occur in the cerebro-spinal fluid after the injection of salvarsan, though before the injection the fluid was normal (Kinek, C. f. O., 1913, p. 292).
(3) Arsenie in large doses is a nerve poison (Evely, Pogány, M. f. O., 1913, p. 1333), and the eighth nerve is a “locus minoris resistentiae,” especially in cases in which it is already affected by syphilis.
(4) Japanese waltzing mice suffer from a congenital lesion of the vestibular nerve. Ordinary white mice, when injected with arsacetin, become waltzing mice. Subsequent microscopic examination shows degeneration of the vestibular nerves (Rothig).
(5) Alexander states that he saw fewer cases of labyrinth disease in syphilis during the seven years prior to the introduction of salvarsan than in four months after its introduction (C. f. O., 1911, p. 291).
(6) Failure of further doses of salvarsan to effect a cure of the condition.
(7) Analogy between the effects of atoxyl on the optic nerve and salvarsan on the auditory.

Literature.—[C. f. O. = Centralbl. f. Ohrenheilk.; Z. f. O. = Zeitschrift f. Ohrenheilk.; M. f. O. = Monatschr. f. Ohrenheilk.; A. f. O. = Archiv f. Ohrenheilk.]
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