On syphilis and the ear—an otologist's view

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Although penicillin and other antitreponemal antibiotics have had a dramatic effect upon the prognosis of syphilitic disease, otological manifestations both early and late continue to be seen. Neonatal congenital syphilitic otolabyrinthitis and meningoneurolabyrinthitis are not encountered today, at least in Britain, presumably due to screening tests and appropriate therapy during pregnancy. These clinicopathological entities were described in detail by Rodger in 1940.¹

The otologist encounters *secondary or early syphilis* from time to time in one of two degrees of severity. The patients are likely to complain of malaise, slight fever, sore throat, enlarged lymph glands, vague headaches and classical skin rashes. These symptoms are spread over a month or two but at some stage the patient presents with tinnitus and deafness, usually bilateral, developing rapidly over a day or two. Vestibular symptoms are less likely though there may be dizziness on movement or on positional change. The pathology is a treponemal meningo-labyrinthitis. The hearing loss is high tone, not severe, partly sensory (end organ) and partly neural (central) as judged by auditory localisation tests and the finding of abnormal latencies on the auditory brainstem responses.²⁻⁴ It has been claimed that if a search is made for evidence of sensorineural deafness in early syphilis it may be found in as many as 17% of cases.⁵ The more severe variant, acute meningovascular syphilis, usually occurs within the first two years of infection. The general manifestations are similar but the headaches and fever are more prominent.

Ocular palsies, facial paralysis, deafness, tinnitus or vertigo of sudden onset may occur as primary symptoms.⁶ The multiplicity of possible presenting features may confuse the diagnosis. The serological tests are consistent with secondary syphilis and the CSF has an elevated protein and a moderate increase of mononuclear cells together with strongly positive tests including the FTA/ABS IgG and IgM. Antitreponemal therapy may reverse the hearing loss. Even untreated the infection runs a benign course though moderate deafness may be a permanent sequel.⁷ Not uncommonly the otologist sees the treated or untreated case many months later when the diagnosis can be overlooked. Treatment is best carried out with a venereologist and should be energetic. It is wise to check the CSF six to twelve months later.

Late syphilis of the temporal bone is a very different disease. Once established, its progress over long periods of time is relentless. Even with present day antitreponemal and anti-inflammatory agents, the prognosis is only somewhat better than it was in 1863 when Hutchinson, the London Hospital physician, wrote his treatise on inherited syphilis.⁸ To be treated it must be diagnosed. To be diagnosed it must be suspected. To be suspected it must be in the forefront of the clinician's mind when presented with any patient who complains of deafness, tinnitus, vertigo or ataxia.

Late syphilis affects the ear 10 to 40 years after the primary infection. The congenital variety is twice as common as the acquired; the former being commoner in younger females, the latter in older males. The pathological changes in the temporal bone are similar, beautifully described by Mayer and Fraser in 1936,⁹ by Goodhill in 1939¹⁰ and by others.¹¹⁻¹⁴ All three layers of the otic capsule are involved by osteitis. Gummatous inflammatory changes are seen including endarteritis and infiltration with lymphocytes, plasma cells and giant cells. The striking feature is the hydrops of the cochlear duct, saccule and utricle from obliteration of the endolymphatic duct in the region of its sinus, accounting for clinical features which mimic Menière's disease. Atrophic changes affect the organ of Corti, the cristae of all the semicircular canals and the cochlear and vestibular nerves. There may be ruptures of the basilar or of Reissner's membrane. In the late stages there is almost complete neuronal loss. The persistence of *Treponema pallidum* in several sites in late, treated human syphilis makes it likely that the spirochaetes demonstrated in the temporal bone by Mack *et al*¹⁵ were *T pallidum*.

The natural history has been described in detail.^{16,17} The deafness in late congenital syphilis is usually symmetrical while in late acquired disease it is often asymmetrical and may remain unilateral for months or years. The onset with tinnitus is sudden in 20% of cases and the symptoms fluctuate in 30%, especially the distortion and discrimination. Within the first two years of onset the pure tone hearing loss, as in Menière's disease, is low or low plus high-tone. As deafness progresses the loss becomes high tone and finally subtotal to total. In the earlier stages specialised hearing tests localise the lesion in the cochlea and transtympanic electrocochleography confirms the presence of hydrops by demonstrating an enhanced

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negative summing potential generated from the distorted basilar membrane.¹⁸ As neuronal loss progresses the auditory localisation becomes retrocochlear and speech discrimination deteriorates.

Vestibular disturbance can usher in the otological phase of the disease. Paroxysms of vertigo, reminiscent of Menière's disease, are seen in almost half these patients, while in the remainder there is a gradual loss of bilateral vestibular function leading to varying degrees of ataxia especially in the dark.

The rapidity of progression is variable. Within the first 5 years of otological manifestation the hearing loss may be as little as 50 dB or as great as 120 dB. After 15 years patients would be fortunate to have hearing thresholds as good as 80–90 dB. Most patients compensate to the slow loss of vestibular neurones and the severity of the damage may be evident only on caloric or rotational testing. The vertigo is rarely as troublesome as in Menière's disease.

In congenital disease there is likely to be or to have been evidence of syphilitic stigmata. Interstitial keratitis perhaps with choroidoretinitis, sometimes recurrent, is seen in ninety percent of cases. These acute episodes date from childhood, adolescence or early adult life. The otological symptoms and signs tend to present later and there may be no evidence of prior IK unless the tell-tale corneal vessels are detected on slit-lamp examination. An estimated one in three patients with congenital syphilis develops ear involvement,¹² but the figure could be higher—ear disease is undoubtedly the second commonest manifestation. Only one in five have Hutchinsonian teeth (not Hutchinson's!). The classical facies of saddle nose and frontal bossing is seen in 10%. Other features such as Clutton's joints or Dubois sign (short fifth metacarpal bones) are very uncommon.

Neurosyphilis is seen in late congenital disease but this is rarely florid since most patients have had several courses of anti-treponemal drugs over the years. The CSF is usually normal. In late acquired syphilis with deafness, there may be more evidence of neurological involvement such as lightning pains, loss of ankle jerk and of vibration sense, sensory changes, bladder symptoms, Argyll Robertson pupils or early optic atrophy. These patients may have elevated CSF protein and positive tests in the CSF.

In late otological disease the older screening tests for syphilis are often negative though the VDRL test has been positive in some 60%. The FTA ABS test is likely to be positive in almost 100%.¹⁶

Yaws due to *T. perpenue* is transmitted by direct contact usually in childhood. The weeping skin lesions leave the typical paper-thin scars particularly on the lower limbs. With immigration from parts of Africa, Central America, the West Indies and SE Asia yaws is no longer a rare disease in Britain. The late otological manifestations are identical to those of late congenital syphilis and the treatment is the same.

In the pre-penicillin era, Moore¹⁹ found that arsenicals had no effect in preventing advancing deafness in late congenital or acquired syphilis, though he did report temporary success in treating the sudden deafness due to early meningovascular disease. Though penicillin has become the treatment of choice for syphilis it has not solved the problems associated with late ear disease. There is now adequate clinical and pathological evidence to suggest that the established case will never be cured, though it may be arrested. Neonatal congenital syphilis treated adequately with penicillin does not prevent the onset of temporal bone gammatous disease in early adult life. Penicillin is usually given in high parenteral dosage with probenecid to block kidney excretion. Other antibiotics such as erythromycin, the tetracyclines, chloramphenicol, cephaloridine or the third generation cephalosporins are antitreponemal and may have to be considered if there is penicillin allergy, found in as many as 10% of patients with late syphilitic deafness¹⁶ due to prior therapy. The cephalosporins have the advantage of intramuscular administration and high levels of these antibiotics in bone. Upon diagnosing late syphilitic deafness a course of one of these agents is probably indicated though there is no proof or evidence of otological benefit.

Steroids are the treatment of choice for this disease. In 1952, Perlman and Leeks reported some success with ACTH.¹¹ Morton had indifferent results with cortisone.²⁰ By 1962 Hahn *et al* reported 50% of their patients improved with prednisone therapy.²¹ In 1968 Dawkins *et al*²² had some success. These were all small uncontrolled series.

In 1969 Morrison²³ reported that 50% of 26 patients treated with steroids had definite hearing gains but that relapses were being encountered; and in 1975¹⁶ his long-term results in over 100 patients were presented, analysing different regime results. The author's experience now runs to over 200 cases of late syphilis of the temporal bones including ten of late yaws. The rationale of initial treatment with both steroids and antibiotics is that steroids might encourage spirochaete multiplication at which time they would be more susceptible to antibiotic damage. It is equally possible that organisms in the temporal bones have lost their pathogenicity but not their antigenicity and that the treatment results reflect local anti-inflammatory properties. These concepts have been examined.^{16 17}

Steroid treatment consists of prednisone 10 mg thrice daily for the first week, 25 mg daily for the next two weeks, and thereafter a maintenance dose of 2.5 to 7.5 mg daily for months or years. More recently the alternative has been dexamethasone 4 mg qds for the first week, 4 mg tds for the second and maintenance therapy of 0.5 to 1.5 mg daily. If there is no hearing response in the first three weeks, some consider withdrawal advisable but maintenance therapy can prevent further hearing loss even if there has been no apparent benefit from initial therapy. The disadvantages of long-term steroids have to be weighed

against the benefits.

Hearing gains of 20–30 dB with improved discrimination can be expected provided the changes are not too advanced and the deafness history less than two years. Relapse rates following steroid withdrawal may be minimised by concomitant antibiotic therapy. Sudden deafness carries a poorer prognosis and a history of fluctuation a better one. Vestibular symptoms are dramatically controlled. Retreatment following relapse is rarely effective but patient control of maintenance therapy is desirable.

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