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Congenital syphilitic deafness*

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MANY will feel that this subject is purely academic, but this is not the case. Congenital syphilis is still with us and it still affects the labyrinth. Indeed, inner ear symptoms can develop years after the congenital condition appears to have been cured. In the past five years we have seen twenty-five cases and there is a fair chance that every now and then every otolaryngologist gets the opportunity of making the diagnosis. Recognition and treatment in the early stages of congenital syphilitic labyrinthitis may lead to the preservation of hearing in those who otherwise would be condemned to irreversible bilateral profound deafness.

Our interest in this subject was first stimulated about five years ago by Dr. Jerome Goldman, an ophthalmologist in the Massachusetts Eye and Ear Infirmary in Boston (Goldman and Girard, 1967); he found treponema pallidum in the anterior chamber of the eyes of patients who had received what was regarded as adequate anti-syphilitic treatment. Following massive doses of ampicillin the treponema pallidum disappeared from the aqueous humor.

It seemed reasonable to postulate that spirochaetes were not only in the eye fluids but also in the inner ear fluids, and that the failure of penicillin to stop the progressive inner ear damage in congenital syphilis lay in its inability to reach adequate concentrations in perilymph or endolymph. We decided, therefore, to use massive doses of ampicillin in congenital syphilitic labyrinthitis.

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Diagnosis

Of course, one must diagnose this condition before treating it. Dr. Lawton Smith (1969), an American Ophthalmologist interested in late syphilis, made a particularly apt statement about its diagnosis. Up to 1906, he points out, syphilis was difficult to diagnose and difficult to treat. In 1906, with the discovery of the Wassermann reaction it became easy to diagnose but still difficult to treat. When penicillin was introduced it was thought to be easy to diagnose and easy to treat. One is now seeing patients treated in the past with penicillin with the development of negative Wassermann reactions, developing late ocular, neurological and otological complications, so completing the cycle to make it once more difficult to diagnose and difficult to treat.

Two main prerequisites are essential if these difficult cases are to be diagnosed. Firstly, one must be aware of the condition. Secondly, one must not use the Wassermann reaction as a test to exclude it, as many have been treated with penicillin in the past and developed a negative W.R. Acquired syphilis rarely affects the ear but congenital syphilis can do so at almost any age. It does so most often in the forties and fifties and the presentation is often almost indistinguishable from Meniere's disease. The possibility of congenital syphilitic labyrinthitis should be considered in any patient with a fluctuating sensorineural hearing loss (Kerr, 1969). In otological syphilis, both ears become involved eventually and the disease is often rapidly progressive.

There may be features in the past history of the patient which are suggestive of this condition, the most common manifestation being inflammatory eye disease, treated by injections and sometimes leaving behind scarring of the cornea.

The ultimate tests are serological, although as already stated, not the Wassermann; probably the most reliable is the fluorescent treponemal antibody absorption test (FTA-ABS), which tends to remain positive, long after the Wassermann has become negative.

Treatment

While these people also may have tinnitus and vertigo, their main problem soon becomes one of deafness—a deafness that, while fluctuating, is relentlessly progressive in most cases. Our main objective in treatment, therefore, has been to reverse, or at least halt the progression of the deafness.

In this paper we will give a more or less chronological account of our efforts to preserve hearing and do not intend to give a detailed analysis of the twenty-five cases we have seen.

The first patient whom we treated with ampicillin had a three-year history of bilateral fluctuating deafness. Figure 1 shows her audiogram in October 1967 before she started treatment. Our routine speech dis-

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crimination test is based on a number of lists of twenty-five monosyllables presented to the test ear at 40–45 decibels above the average air conduction thresholds for 500, 1,000 and 2,000 Hertz, the other ear being masked. The number of words repeated correctly is expressed as a percentage (Kerr and Smyth, 1972).

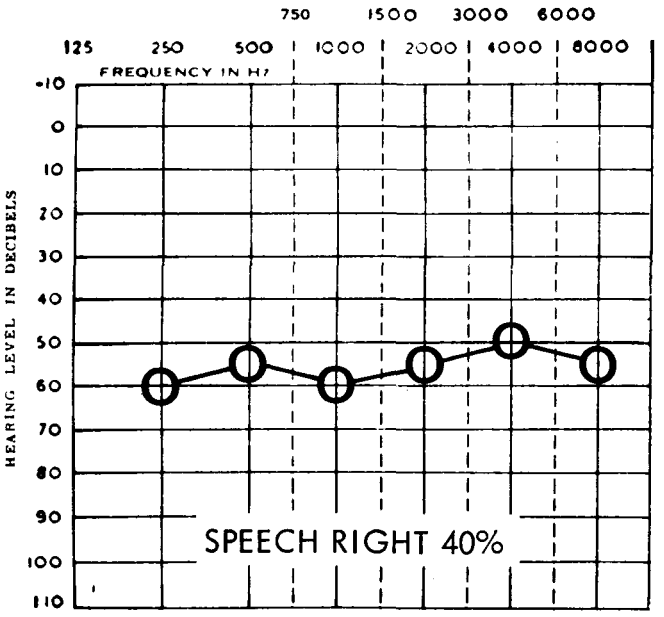


FIG. 1.

The right audiogram in October 1967. The hearing on the left side was similar to that on the right and responded similarly to treatment.

This first patient in this series had been seen on previous occasions without the diagnosis having been made. But, clinical suspicions having been aroused, she was asked some questions, which, since then, have become fairly routine. As a child she had had inflammation of the eyes with a temporary visual loss treated successfully by injections. Her Wassermann reaction had been tested in June 1966, over a year earlier, and was negative, but her FTA-ABS test was done in September 1967 and was positive. She was started on ampicillin 1·5 grams six-hourly for four weeks.

Figure 2 shows the hearing levels over the two to three years before starting on treatment. As the speech discrimination in congenital syphilitic deafness tends to fluctuate much more widely than the pure tones, both aspects of hearing must be represented. In fact, to report changes in pure tone hearing, without reference to speech discrimination can be quite misleading (Kerr and Smyth, 1972). Therefore, we have expressed, on

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one graph, the changes in both pure tone hearing and speech discrimination. The response to treatment is seen in Figure 3.

There has not been much change in the pure tone threshold, but the speech discrimination score has improved dramatically and is no longer fluctuating in the former severe manner. She has no doubts about the improvement in her hearing, despite the minor change in the pure tone levels.

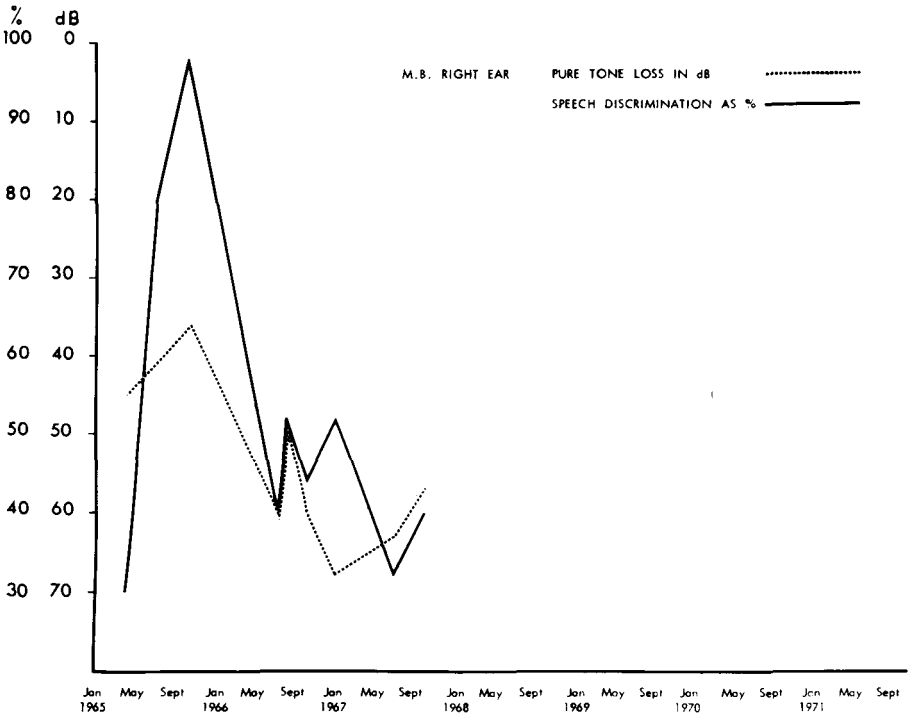


FIG. 2.

The fluctuating hearing (pure tone hearing and speech discrimination) on the right side before treatment are well demonstrated on this graph. The hearing on the left side was similar.

The next patient that we saw with this condition had been profoundly deaf for over ten years and had a speech discrimination score of nil in both ears. Filled with enthusiasm from our first case, he was given ampicillin in the same massive dose, but unfortunately did not have any response whatsoever. This is not surprising when one looks at the pathology illustrated in this temporal bone section in an advanced case of congenital syphilitic labyrinthitis (Fig. 4). Miracle drugs alone would be of no value—to restore the hearing in such a case would require a miracle itself. We had another similar case shortly afterwards, when, optimism overcoming

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realism, we tried again, but again got the same absence of any response to treatment.

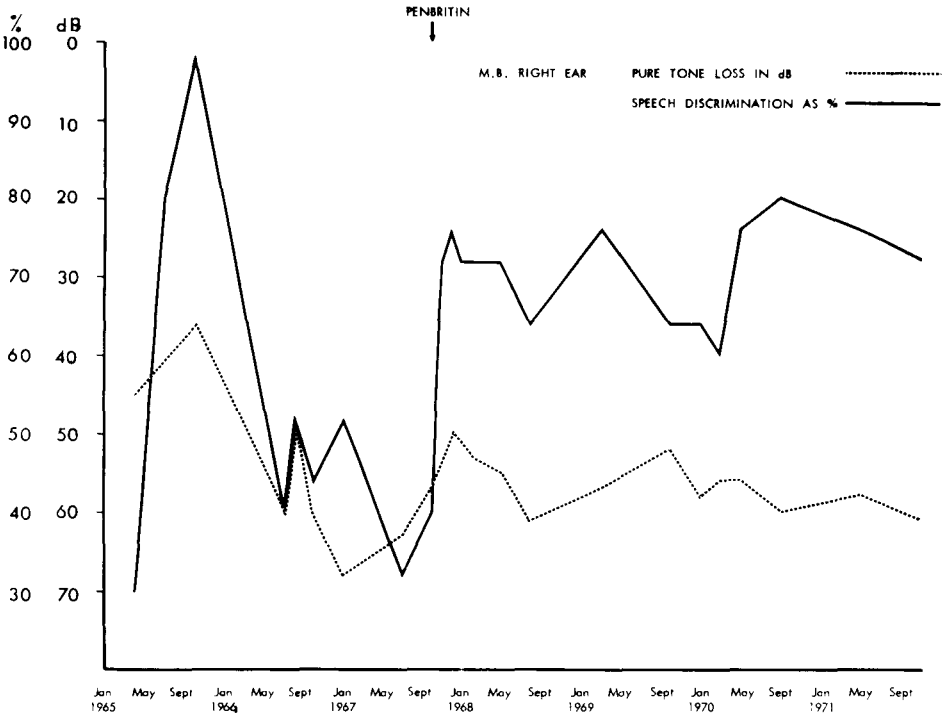


FIG. 3.

The hearing on the right side after treatment. Note the marked improvement in speech discrimination without much change in pure tone hearing.

However, we soon had some more encouragement with a patient who presented with this audiogram (Fig. 5). He was given ampicillin in a dosage of 1·5 grams six-hourly for four weeks and had a marked improvement in his speech discrimination (Fig. 6). However, unlike the first case, this was not maintained, and eight months later his hearing was even worse thar before treatment. Re-treatment was considered, but this time Prednisone, 10 mg. three times daily for ten days, tailing off over the following ten days, was given along with ampicillin, given again in the dosage of 1·5 grams six-hourly for four weeks. The response is illustrated in Figure 7.

We began, therefore, to include prednisone in our standard treatment and in the next two cases had encouraging results.

We then ran into trouble again in a patient with useful, although poor, hearing in an only hearing ear (Fig. 8). She got a minimal response to penbritin and prednisone with speech discrimination rising only from

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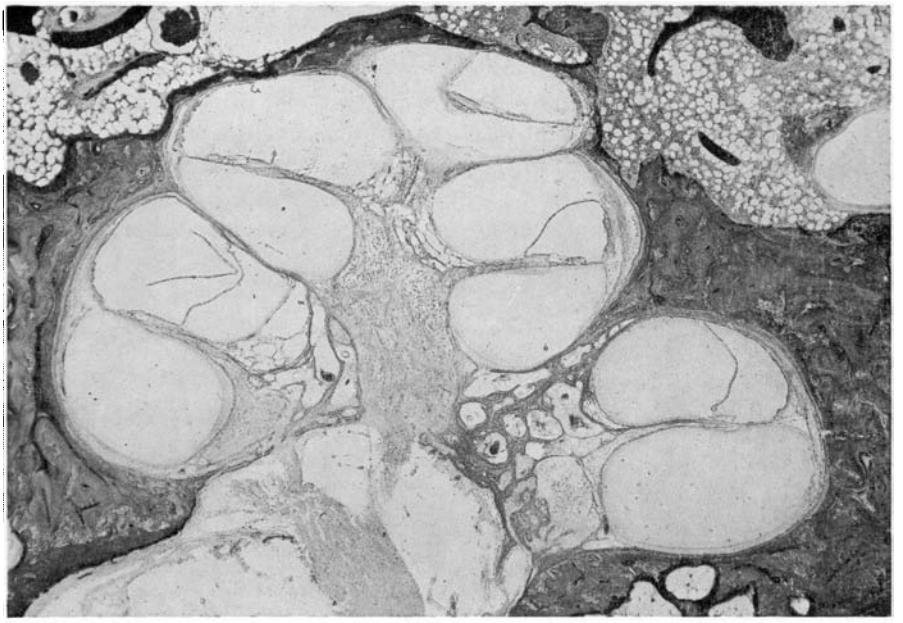


FIG. 4.

The cochlea in a case of advanced congenital syphilitic labyrinthitis, with profound deafness. There is marked erosion of the cochlear capsule, severe damage to the Organ of Corti, hydrops, rupture of Reissner's membrane and almost complete absence of spiral ganglion cells. (Courtesy of Professor Ruedi and Professor Schuknecht.)

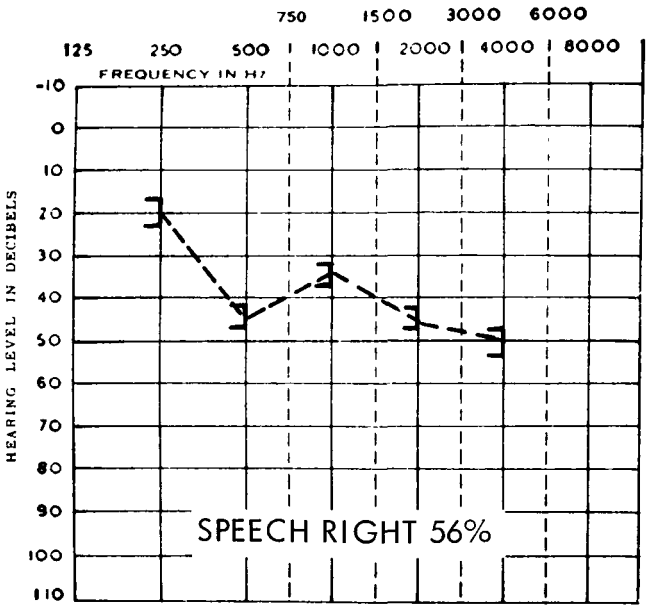


FIG. 5.

The bone conduction levels are recorded as this patient also had a mild degree of conductive deafness. As in Figures 1, 2, and 3, the left ear was similar to the right before treatment and responded similarly.

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40 to 60 per cent, but after three months this began to fall off again (Fig. 9). As she had no hearing whatsoever in the other ear we felt that something further should be done. She was started therefore on intramuscular ACTH, 40 units weekly, with the addition of oral prednisone for the first ten days. Figure 9 shows the dramatic response in her speech discrimination as a result of this treatment but again the pure tones did not alter significantly. This patient also had no doubt about improvement in her hearing.

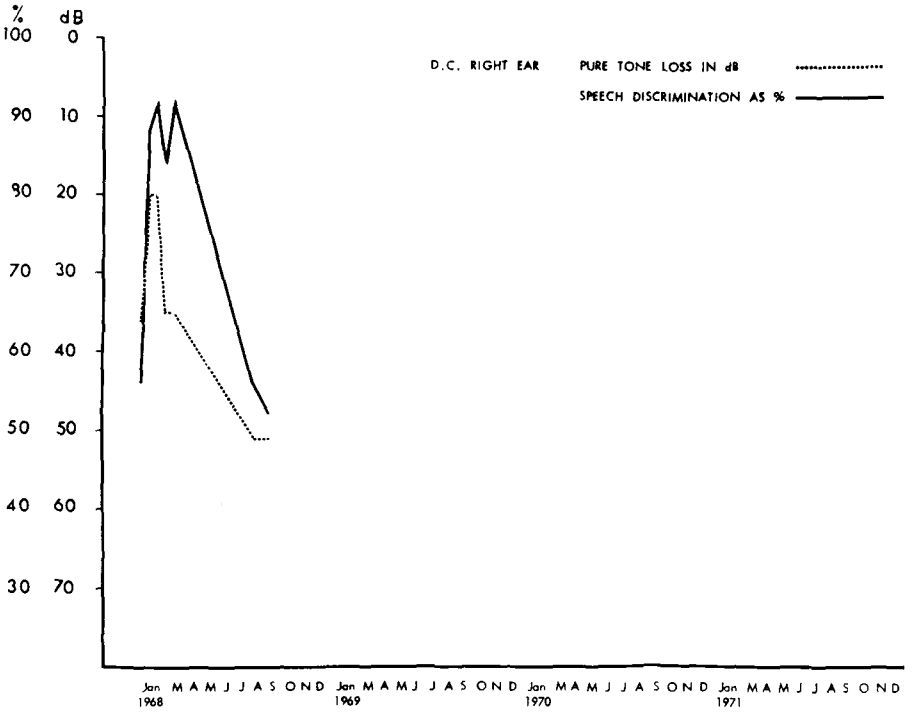


FIG. 6.
The response to ampicillin, given in January 1968.

Discussion

The cases described above illustrate the types of response we have had. Our policy now is to leave untreated any who have had profound deafness with complete loss of speech discrimination for a period of over six months. We have never got any response from these people. This is not surprising as complete destruction of sensory end organs and neural tissue in the inner ear probably had occurred and there is no medical treatment that could be expected to alter that.

In the other cases our standard procedure is to start with ampicillin 1·5 grams six-hourly for four weeks, accompanied by prednisone 10 mg.

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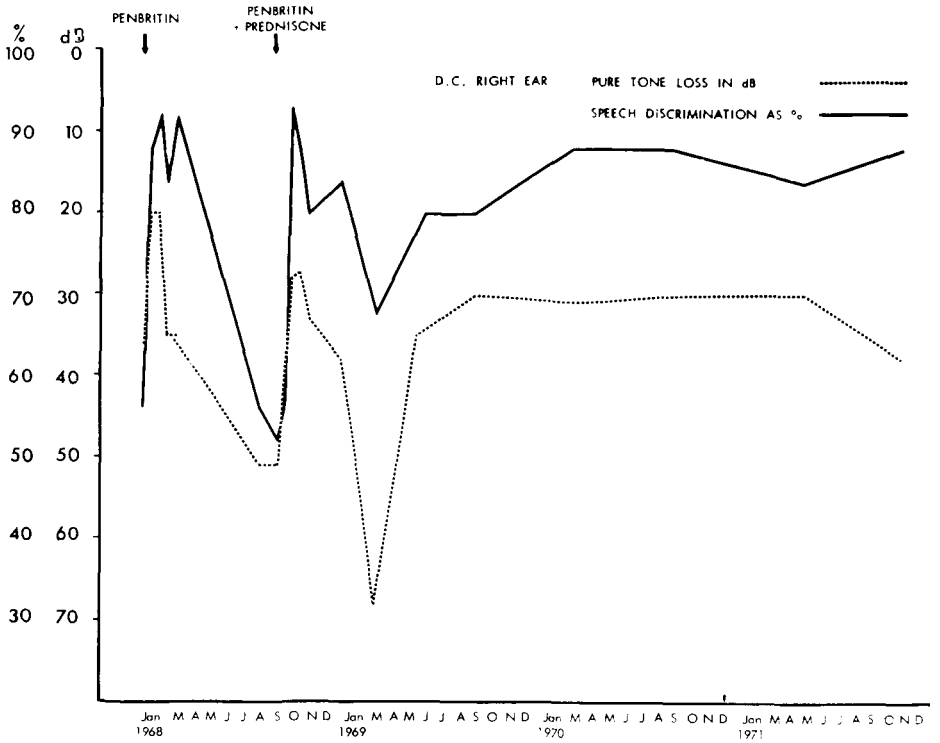


FIG. 7.

The response to ampicillin and prednisone in September 1968.

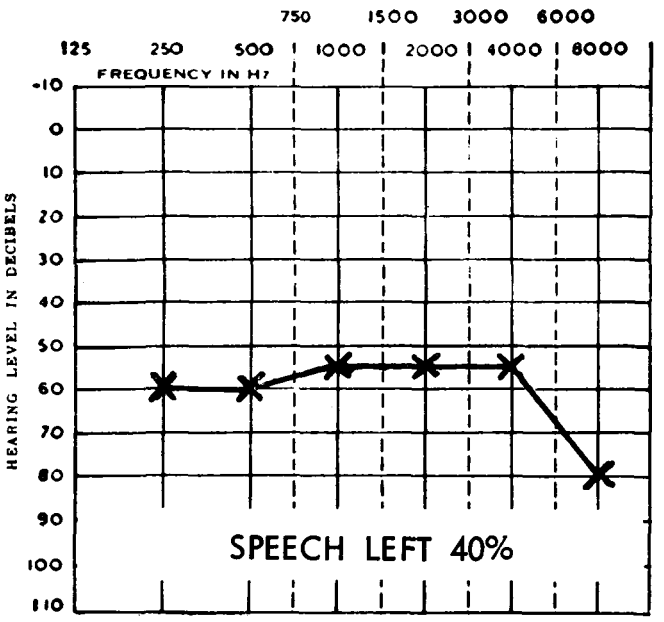


FIG. 8.

The hearing on the left side. The right ear was profoundly deaf.

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three times daily for ten days, tailing off over the succeeding ten days. Usually, there is a response to this combination.

If the hearing does not improve, or the initial improvement is not maintained, we then proceed on an *ad hoc* basis, sometimes to regular injections of ACTH but being influenced by such factors as the rate of progression, the level of deafness and the hearing in the other ear.

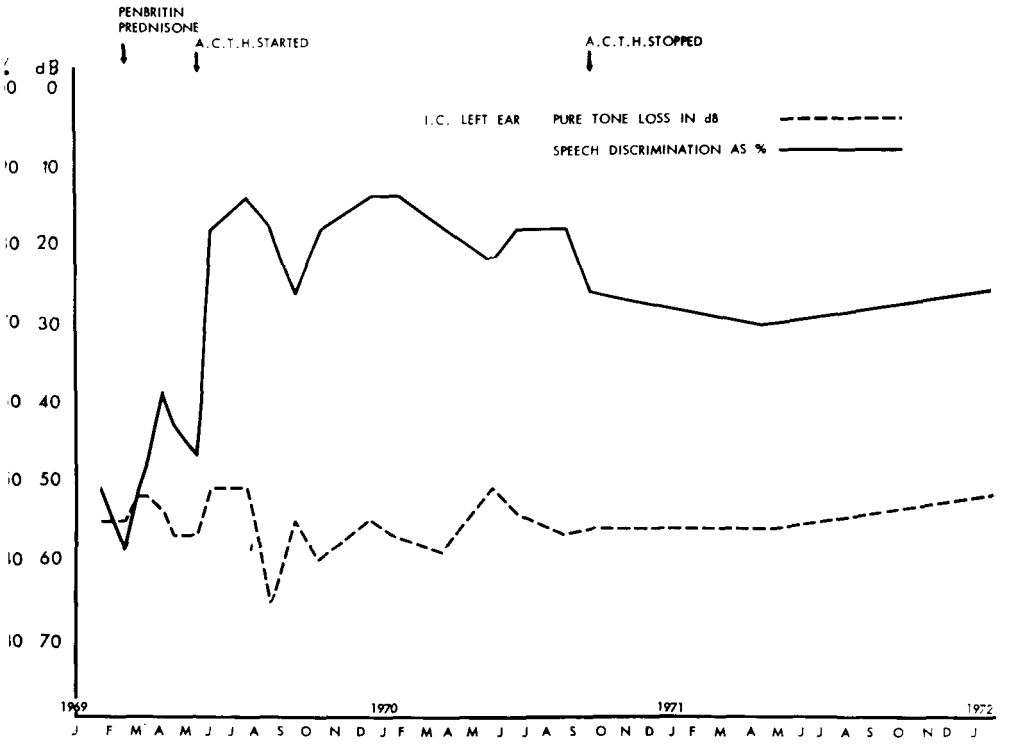


FIG. 9.
The hearing over a three-year period.

The most critical situation arises when one ear is already profoundly deaf with progressive deafness in the one remaining hearing ear. We are prepared to give indefinitely intramuscular ACTH in such patients. At present we have three patients on weekly or twice-weekly injections of ACTH 40 units intramuscularly, with maintenance of the hearing. We have tried the effect of reducing the dose in two of these, but had to increase it again because the hearing deteriorated. The fourth patient to get ACTH, has been off it for one year, and her hearing has been maintained (Fig. 9).

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By energetic treatment none of the 12 patients who, when first seen at the clinic had serviceable hearing, has lost this. Most are hearing better now than at the outset of the treatment and only in one patient has there been any progression of the hearing loss. In this case, however, he still has only a mild degree of deafness despite the progression and, therefore, we feel, does not yet need ACTH.

As mentioned earlier, our main concern has been with the deafness, rather than tinnitus and vertigo. Fortunately, improvement in hearing usually is accompanied by improvement in the other symptoms, and where necessary, the addition of antivertiginous drugs usually eliminates the problem.

What is happening in these people is difficult to say. We demonstrated that ampicillin reached levels in human and animal perilymph, sufficient to kill the spirochaete (Kerr *et al.*, 1970).

However, treponema pallidum has not yet been found in perilymph or in endolymph. But, since we became interested in this condition, it has been found (Mack *et al.*, 1969) in the enchondral bone of the labyrinthine capsule (Fig. 10). This situation explains the difficulty in treating congenital syphilitic labyrinthitis. Enchondral bone is dense and almost avascular, and antibiotics, therefore, have difficulty in getting at any spirochaetes in it. It is probably an even worse situation than would occur with spirochaetes in the perilymph or endolymph.

It is likely, therefore, that there is nothing specific about the effects of ampicillin, the significant factor being only the massive dose. We have used very large doses of intravenous penicillin in two cases, with equivocal results, but in the future plan to use it again in any cases with a poor response to ampicillin.

The use of steroids is not new and has been reported many times in the past (Perlman and Leek, 1952; Hahn *et al.*, 1962; Karmody and Schuknecht, 1966; Dawkins *et al.*, 1968); presumably the steroids are suppressing an immune reaction. How or where this is occurring is uncertain, but because of the wide fluctuations of the speech discrimination, without much change in pure tone hearing, we suspect that the action is in relation to the auditory nerve, in either the spiral or Rosenthal's canal, rather than in the organ of Corti.

Prognosis

And what is the prognosis? In those cases where a single course of ampicillin and prednisone stabilized the hearing, we feel that there is a good chance that the disease process has been arrested. We are not so confident about those on regular ACTH injections but think that untreated at least three out of the four would be profoundly deaf by now. They may still become profoundly deaf but at least that day is being postponed.

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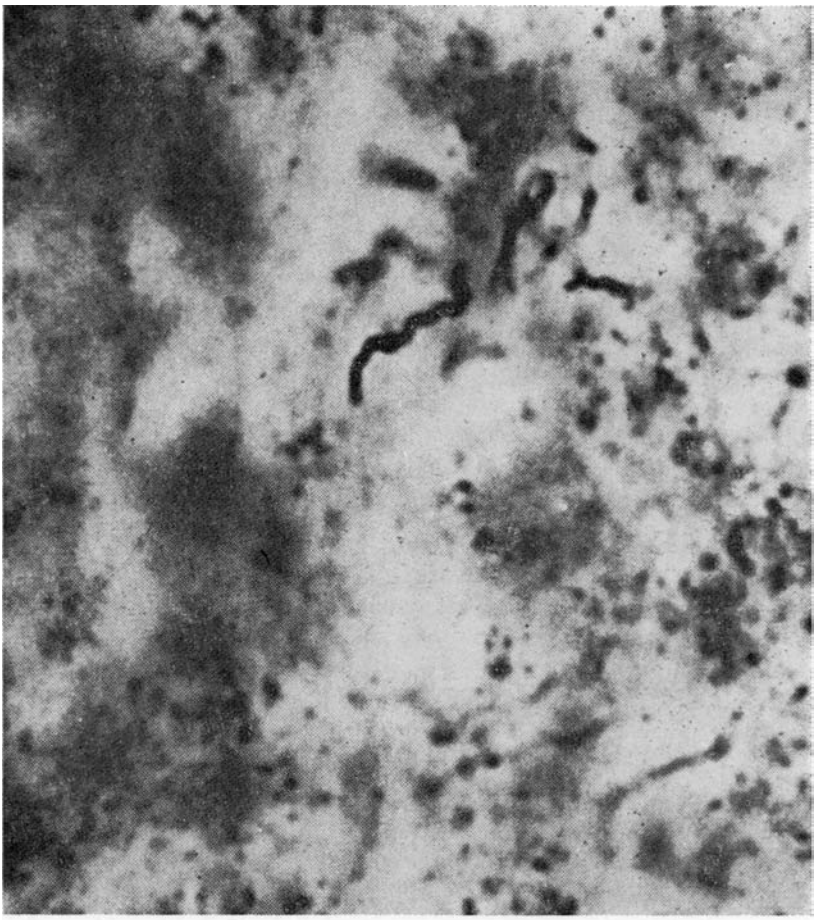


FIG. 10.

Treponema pallidum in the enchondral bone of the cochlear capsule. (Courtesy of Dr. J. Lawton Smith, *Archives of Otolaryngology*, 1969, **90**, 11–14. 'Copyright 1969, American Medical Association'.)

Summary

Congenital syphilitic labyrinthitis is uncommon but not rare. While the hearing in this condition fluctuates, the disease tends to be relentlessly progressive leading to bilateral profound deafness; consequently, early and effective treatment is important.

In advanced cases, where there is severe destruction of the inner ear structures, it is obvious that medical treatment cannot be expected to restore hearing.

In less advanced situations, where the hearing is still fluctuating, an adequate level of antibiotic at the site of the organism may result in an improvement. Ampicillin and penicillin have been given in massive doses

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with encouraging results. As the disease process probably also involves an immune reaction, an improvement in symptoms would be expected, and has been obtained, following the use of steroids.

In twelve cases the symptomatic progress of the disease has been reversed or halted, for up to four years, using various combinations of antibiotic and steroids; in four of these cases, weekly injections of ACTH have been required to achieve this.

Whether profound deafness is being prevented or merely postponed will be known ultimately only with the passage of time.

Acknowledgements

We are indebted in this work to so many colleagues that they could not all be mentioned by name. We wish, however, to acknowledge the co-operation we have had from those in the Ear, Nose and Throat, Venereology, Metabolic and Photographic Departments of the Royal Victoria Hospital, Belfast, and the Nephrology Department of the Belfast City Hospital.

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