

Letters to the Editor

Autoimmune Inner Ear Disease and Ulcerative Colitis

Dear Sir,

We read with interest the paper by Jacob *et al.* (1990) on a case of sensorineural hearing loss which occurred in a patient with ulcerative colitis, episcleritis and giant cell arteritis.

We would like to report upon two more patients, both of whom had ulcerative colitis without any previous systemic complications, who developed total deafness and were in fact seen in our cochlear implant assessment clinics.

Case 1: A 30-year-old woman developed acute ulcerative colitis in 1982 and was managed with topical steroids and salazopyrine. In 1985 she developed vertigo and left sided hearing loss of sudden onset. Four weeks later she again developed vertigo and this time right sided hearing loss. On both occasions she was treated with histamine, dextran and steroids to no avail and when assessed in the cochlear implant clinic in 1988 she was totally deaf. She has since been implanted with a Nucleus 22 channel intracochlear device and has good open set speech with little recourse to lip reading.

Case 2: A 33-year-old man had sudden onset of total right sided vestibulocochlear failure at the age of 15 at about the same time as being diagnosed as having ulcerative colitis. Three months later the hearing on the left side started to diminish and deteriorated over the space of a year until he developed total hearing loss. He is still under consideration for implantation.

Neither patient had been treated with any commonly recognized ototoxic drug.

The two main proposals for the aetiology of ulcerative colitis are i) infective and ii) immunological.

Strong evidence was provided for the immunological theory when Takahasi and Das (1985) isolated an auto-antigen in normal colon to which much of the tissue-bound immunoglobulin in ulcerative colitis was specific. They later found the antigen in skin and biliary tract which would offer an explanation for the systemic effects of pyoderma gangrenosum and pericholangitis (Das *et al.*, 1987).

There have been several reports in the literature of SNHL associated with ulcerative colitis. Of note is that fact that the hearing loss often recovers, at least in part, with aggressive treatment with steroids and cyclophosphamide or azathioprine (Summers and Harker, 1982, Weber *et al.*, 1984, David and Rees, 1987). Plasmapheresis, although of value in some other cases of autoimmune hearing loss, proved disappointing in the treatment of a patient with sensorineural hearing loss and ulcerative colitis (Luetje, 1989).

Autoimmune inner ear disease may occur due to

1. Loss of immunological tolerance to self antigens, either organ specific or as part of a generalized autoimmune state at T or B cell level.
2. Autoimmunity due to hidden self antigens which are exposed due to tissue damage.
3. Cross reacting antibodies or self antigen modification by infection or drugs.

The damage, whether T or B cell mediated, may be directed at inner ear structures themselves, or at a retrocochlear level, or at the vascular supply of the inner ear.

That autoimmune inner ear disease exists is no longer doubted, but it is proving extremely difficult to find a reliable test to diagnose it. Numerous techniques have been described for looking for T cell or B cell autoimmunity in the ear, but although encouraging results are reported, they seem to bear no relation to whether the patient will respond to immunosuppressive therapy. Indeed McCabe, who originally described autoimmune hearing loss in 1979 has now adopted a policy of treating all patients that he suspects clinically of having autoimmune inner ear disease with cyclophosphamide and steroids, such is his disillusionment with the predictive value of laboratory testing (McCabe, 1989).

Until a satisfactory test is found we would recommend that all patients developing sensorineural hearing loss in association with ulcerative colitis should be treated with systemic steroids and if there is no improvement, azathioprine or cyclophosphamide should be added to the regimen.

Yours faithfully,

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Reply

Dear Sir,

We would agree that all patients with ulcerative colitis who develop progressive or sudden sensorineural hearing loss be treated with steroids followed by azathioprine if unresponsive. Any patient with an 'autoimmune disorder' who develops significant sensorineural hearing loss in the presence of indicators of immunological disturbance should be assumed to have an auto-immune hearing loss and treated actively.

There are no reliable specific tests yet for this form of hearing loss and treatment should be instituted on what clinical and immunological information is available.

Yours faithfully,

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Tympanometry in ossicular discontinuity/otosclerosis

Dear Sir,

In their interesting case report (JLO, Vol. 104 pp 560–561) of an unusual case of bilateral conductive deafness due to discontinuity of the stapes crura from the footplate, Dr Hoare and his colleagues state that their pre-operative diagnosis was otosclerosis.

It would be interesting to know how such a diagnosis was reached and if a pre-operative tympanometric evaluation was performed on the patient. I would expect the tympanogram to show an abnormally high peak since the discontinuity of the ossicular chain would markedly increase middle ear admittance, whereas if otosclerosis was the only middle ear pathology admittance would be low, normal or at most—if there was a coincident hyper-

mobility of the tympanic membrane—a little higher than normal.

If either of the patient's ears had hearing close to normal, the contralateral acoustic reflex would provide additional information since in otosclerosis it would be absent whereas in this case of discontinuity of the ossicular chain medial to the insertion of the stapedius muscle there would be some form of reflex since the muscle would exert its action on the ossicular chain. Testing the patient's right ear some time after surgical correction of hearing of the left ear was achieved may have provided this interesting information.

I would like to point out that tympanometry should be performed in all cases of hearing loss (certainly in all cases of conductive hearing loss) since it is a valuable diagnostic aid to therapeutic planning.

P. N. Eliopoulos,
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Reply:

Dear Sir,

We would disagree with Dr Eliopoulos's assertion that 'tympanometry is a valuable diagnostic aid to therapeutic planning' in cases of conductive deafness. It can confirm a mobile drum and so exclude a middle ear effusion, but this is usually clear from clinical examination including pneumatic otoscopy. Distinguishing otosclerosis from ossicular discontinuity is not essential preoperatively, provided that the patient's consent has been obtained for the higher risks of surgery nearer to the footplate. Tympanotomy is required for definitive diagnosis as well as treatment if there is a conductive hearing loss with a normal eardrum. Our patient had an unremarkable tympanogram. Stapedial reflexes were not tested.

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