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# Relief of severe hyperacusis and diplacusis in a deafened ear by cochlear labyrinthectomy

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**Abstract**
A professional musician with intolerable hyperacusis and dysharmonic diplacusis in a severely deafened ear was successfully relieved of his symptoms by deliberate destruction of the cochlea.

**Key words: Tinnitus; Labyrinthectomy**

## Introduction

Hyperacusis with loudness intolerance is well recognized in association with cochlear hearing losses (Dix, Hallpike and Hood, 1948; Groves, 1979). We report a case in which very severe hyperacusis, with dysharmonic diplacusis binauralis, both arising in a severely deafened ear, were relieved by cochlear labyrinthectomy.

## Case report

A 58-year-old professional musician was first seen in 1991. Almost 40 years previously, during military service, he had sustained acute acoustic trauma to his left ear, when a rifle had been discharged adjacent to the left external auditory meatus. This produced immediate left deafness (to which extent no previous records were available), associated with a bitonal tinnitus in the left ear. Over a few years preceding the current consultation, the left ear had deteriorated progressively, and there had been episodes of rotatory vertigo, typically lasting twenty-four hours with vomiting. Five months prior to consultation, the patient had developed severe distortion in the left ear, with dysharmonic diplacusis. Musical notes were perceived in the left ear at a pitch one semitone different from that in the right. This was associated with very severe loudness intolerance.

Professionally, the patient played and taught the piano, as well as conducting practical piano examinations, and tuning instruments. Work was becoming intolerable, because the louder percussive sounds of the instrument were distressingly unpleasant and distorted. Neurotological examination was normal apart from left-sided deafness and a tendency to stagger to the left on Unterberger's test.

Initial investigations disclosed normal hearing for pure tones on the right, with a severe sensorineural loss on the left, characterized by no measurable hearing above 3 kHz on that side (Figure 1). High definition, contrast-enhanced CT scans of the posterior cranial fossa were within normal limits. Thyroid function was normal and serology for *Treponema pallidum* was negative. In view of the excellent hearing in the right ear and the normal audiogram on that side, together with the severity of the loss on the left, no further audiometric studies were performed, but warm and cool air calorics were undertaken. These produced no electronystagmographic response on the left, and an ice-water caloric on that side in the clinic likewise produced no subjective response and no visible nystagmus.

In view of the distressing symptoms, combined with poor hearing, a left labyrinthectomy was advised, with a view to destroying residual cochlear function. Through a permeatal approach, the stapes was removed, the contents of the vestibule aspirated, and, to ensure cochlear destruction, the round and oval windows were drilled into continuity (Graham and Goldsmith, 1994) and sealed with a vein graft, before replacement of the stapes. Post-operatively, there was vertigo with vomiting and destructive nystagmus, rather unexpected in view of the absent caloric function prior to operation. The symptoms subsided sufficiently to allow his discharge five days post-operatively. At review, four months after surgery, all loudness intolerance had been abolished together with the diplacusis, and there was no residual hearing in the ear (Figure 2). Although experiencing occasional unsteadiness, he was able to ride a bicycle. His left tinnitus (louder prior to surgery than at its onset in the 1950s) continued unchanged. Fortunately, it did not unduly disturb him. He was able to pursue his profession free of distressing auditory phenomena.

## Discussion

Labyrinthectomy has traditionally been used in the relief of intractable peripheral vertigo in severe cases of unilateral Ménière's disease, where residual speech perception on the affected side is poor. The alternative option of selective vestibular neurectomy has been strongly supported in recent years, because of its potential to conserve residual hearing (Fisch, 1976). Selective vestibular labyrinthectomy by ultrasound (Stahle, 1976), cryotherapy or perilymph perfusion of ototoxics (Shea, 1994) has also been advocated. Deliberate cochlear destruction has few, if any, recognized indications at present; it has long been recognized as of little value in controlling distressing tinnitus in a deaf ear.

In the present case, a musician suffered incapacity because of intolerable loudness discomfort and dysharmonic diplacusis, both apparently engendered by

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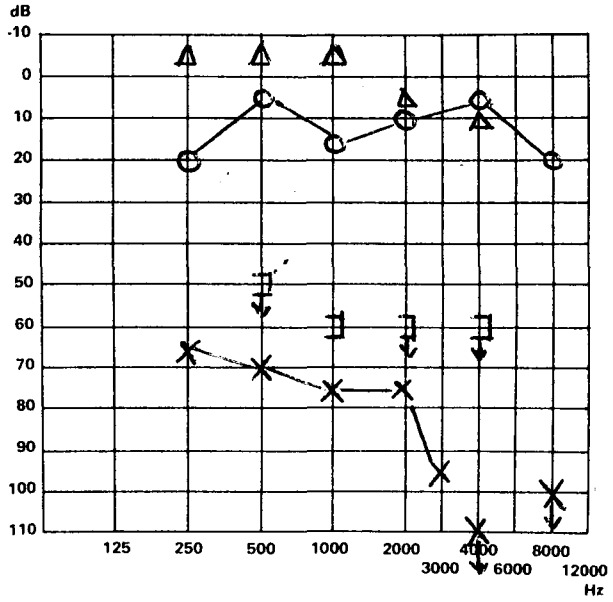


FIG. 1
Pre-operative audiogram.

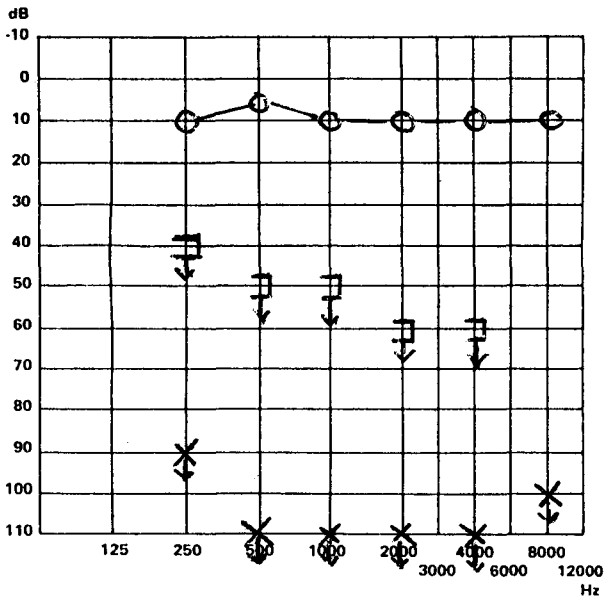


FIG. 2
Post-operative audiogram.

progressive cochlear damage following acute unilateral acoustic trauma (itself an interesting sequence of events). His hearing on the affected side was of no value to him, and it was therefore decided to destroy the cochlea in the hope of abolishing both residual hearing and its unwanted consequences. Alternatives designed to produce selective vestibular destruction would, of course, have been of no value in this instance. No suggestion was made to the patient that his tinnitus would be relieved, and indeed it was not, although presumably originally of peripheral origin. The procedure succeeded completely in its primary objective.

Under these rare circumstances, deliberate cochlear destruction finds a place in the armamentarium of the otologist.

## Conclusion

We report an unusual case of loudness intolerance and harmonic distortion in a severely deafened ear, relieved by cochlear labyrinthectomy, which we would not hesitate to recommend for similar cases.

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