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Clinical records

Ossicular chain interruption with present acoustic reflex

by

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It is claimed (Feldman, 1975; Wilber, 1976) that, theoretically, a conductive hearing loss with a normal tympanic membrane and a present acoustic reflex is highly characteristic of ossicular chain interruption. Usually the discontinuity is due either to a defect in the stapedial crura, or to fibrous adhesions between the stapes and the tympanic membrane, or to replacement of the long process of the incus by a fibrous band. On the other hand, only a few cases have been reported in the literature which have demonstrated this combination of anatomical and functional defects, as confirmed at surgery, following surgical (Hayes, 1977) or physical trauma (Shapiro *et al.*, 1981), congenital defects (Lidén *et al.*, 1974; Isenberg and Tubergen, 1980; Shapiro *et al.*, 1981) or diseases of the labyrinthine capsule (e.g. osteogenesis imperfecta) (Jenkins *et al.*, 1980) (Table I).

This paper reports the case of a stapes anomaly which caused a non-progressive, unilateral conductive hearing loss, with A type and W pattern tympanograms respectively, at 220 and 660 Hz. probe tone frequencies, and with an acoustic reflex elicited by contralateral stimulation. To our knowledge, this combination of findings has not been previously reported.

Case Report

On 15 March 1981, a 33-year-old woman was referred to our department with stable and non-fluctuant hearing loss in her left ear, first discovered 12 years previously. Tinnitus had also been present for the previous four months. The patient's history did not reveal any familial hearing loss, previous otitis, or history of head trauma.

The otologic examination revealed perfectly normal, semi-transparent tympanic membranes with normal light reflexes and an absence of any signs of previous infection. No abnormality of either auricle or external auditory canal was noted, and all the cranial nerves functioned normally.

Audiometry showed normal hearing in the right ear and a conductive hearing loss in the left ear, with a mean threshold of 68 db. HL. for the speech frequencies (0.5, 1 and 2 kHz.). The spondee dis-

TABLE I

OSSICULAR CHAIN INTERRUPTION WITH NORMAL ACOUSTIC REFLEX

Author	Case	Stapes	Incus	Etiology
Lidén *et al.* (1974)	2	Normal but with fibrous adhesion with tympanic membrane	Erosion of long process	Not mentioned
Hayes (1977)	1	Fractured crura; fixed footplate	Normal	Surgical trauma
Jenkins *et al.* (1980)	1	Fibrous crura; fixed footplate	Normal	Osteogenesis imperfecta
Isenberg and Tubergen (1980)	1	Absence of superstructure; mobile footplate	Shortened lenticular process	Congenital
Shapiro *et al.* (1981)	1	Fibrous crura	Normal	Congenital
	1	Normal	Anteriorly dislocated; fibrous union with the stapes	Physical trauma

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crimination score was 100 per cent in both ears and the SRT was reached a 80 db. HL. in the left ear.

In the right ear, tympanometry revealed A-type curves for both conductance and susceptance with 220 and 660 Hz. probe tone frequencies. In the left ear (Fig. 1, top), tympanograms were again of A-type at 220 Hz. probe tone, similar in shape and notch depth to the corresponding tympanograms of the contralateral ear. However, W-patterned curves were obtained with a 660 Hz. probe tone for both conductance and susceptance.

No acoustic reflex was elicited in the right ear by contralateral stimulation, but it was registered in the left ear with thresholds between 90 and 95 db. HL. for 0.5, 1, 2 and 4 kHz. stimulus frequencies. The ipsilateral acoustic reflexes were not investigated.

On 23 March 1981, an exploratory tympanotomy of the left ear was made, a posterior tympanomeatal flap was elevated, and the chorda tympani was cut. The stapedius tendon, a little longer than usual, was attached to the lenticular process of the incus. The stapes superstructure was completely missing but the footplate was of normal size and mobile. The malleus and incus were normal and mobile. No other abnormalities in the middle ear structure were found (Fig. 2). The stapedius tendon was sectioned and a 'wire-connective tissue' prosthesis 4.5 mm. length was inserted between the long process of the incus and the footplate.

The patient was discharged after five days. On 27 February 1982, examination revealed no trace of scarring in the tympanic membrane and almost normal hearing in the left ear, with closure of the air-bone gap to 10 db. HL. for the speech frequencies. The spondee discrimination score was again 100 per cent, but the SRT was lowered from the 80 db. HL. pre-operative value to 40 db. HL. Tympanometry then showed A-type curves for both conductance and susceptance at 220 and 660 Hz. probe tone frequencies (Fig. 1, bottom).

In table II are shown the values of the conductance (G) and the susceptance (B) at the surface of the tympanic membrane for the right ear (RE), for the left ear pre-operatively (LE_1) and for the left ear post-operatively (LE_2); the corresponding ranges in normal ears are shown for comparison. Note that, for A-type tympanograms, all the G and B values measured in the left ear (both pre- and post-operatively) fall within the normal range. They were calculated by averaging the two measurements of the tympanogram values with a pressure load on the tympanic membrane of 200 mm. H_2O (positive and negative) and by subtracting this averaged figure from the tympanogram's peak value. The data on normal ears were obtained from 50 normal subjects who were examined with the same equipment (Zito, 1976).

Discussion

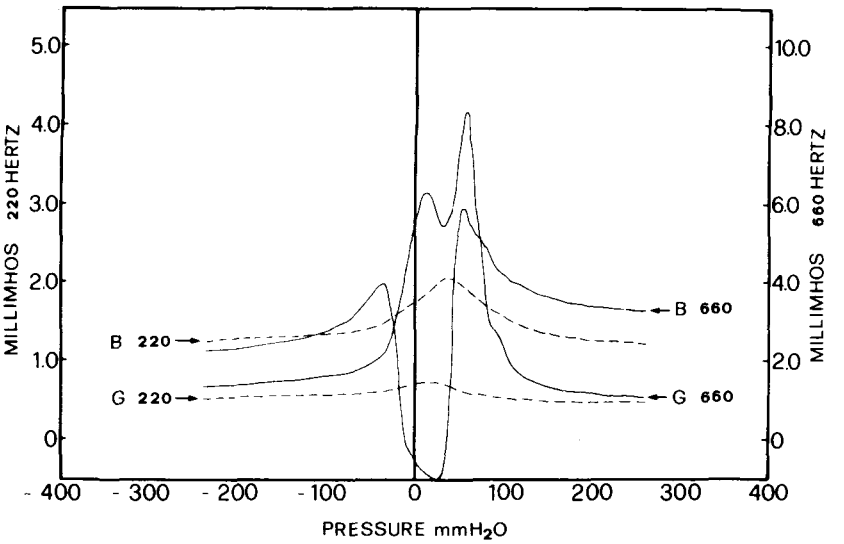
Analysis of this defect is directed specifically to the embryologic development of the auditory ossicles, and to the differential diagnostic tool offered by tympanometry.

The work of Bast and Anson (1949) on the development of the temporal bone in man has clearly shown that the stapes has a dual embryologic origin. The head, stapedial arch, and annulus are derived from Reichert's cartilage, while the 'lamina stapedialis' is a derivative of the otic capsule. Subsequent fusion of the annulus and 'lamina stapedialis', and secondary differentiation of the laminar tissue, provide the footplate, the stapedial rim and the annular ligament. The abnormal surgical findings in the present case could be explained by anomalous development or lack of further differentiation of only a part of Reichert's cartilage, since a normal annulus stapedialis was found; another possibility would be a preponderance of the normal erosive changes which occur in the stapes during later embryologic development.

Regarding the tympanometric data, the pre-operative findings showed completely normal tympanograms in terms of shape and notch-depth at a 220 Hz. probe tone frequency, while a W pattern was observed at 660 Hz. These findings are not in agreement with Alberti and Jerger (1974) who stated that 'there does not appear to be substantial clinical value in executing tympanograms at probe tone frequencies higher than 220 Hz.'. On the contrary, in human temporal bones, experimental interruptions in the ossicular chain always produce A-type tympanograms at 220 Hz. and W pattern curves at 660 Hz. (Roberto and Zito, 1983). In the case reported, the response to a 220 Hz. probe tone and the normal acoustic reflex would lead one to assume that there was a perfectly normal tympano-ossicular system. Only the 660 Hz. tympanometric findings provided correct information about the ossicular chain interruption observed at surgery.

The post-operative 220 Hz. tympanograms were similar to the pre-operative ones; only the notch depth was a little lower, though remaining within the normal range. On the contrary, the pre-operative W shape patterns observed with the 660 Hz. probe tone became A-type tympanograms after surgery, with a notch depth within the normal range. It must be concluded that a mass-controlled pre-operative tympano-ossicular system was converted surgically into a stiffness-controlled system.

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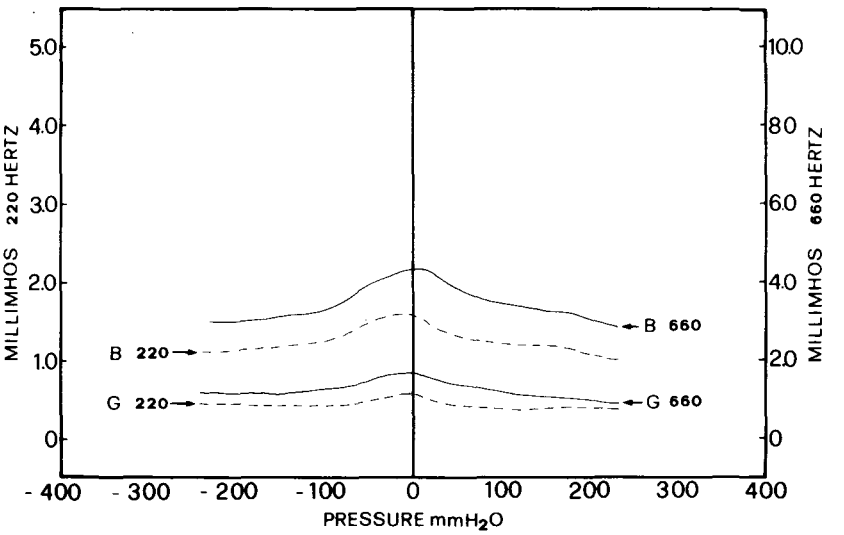


FIG. 1
Left pre-operative (top) and post-operative (bottom) tympanograms.

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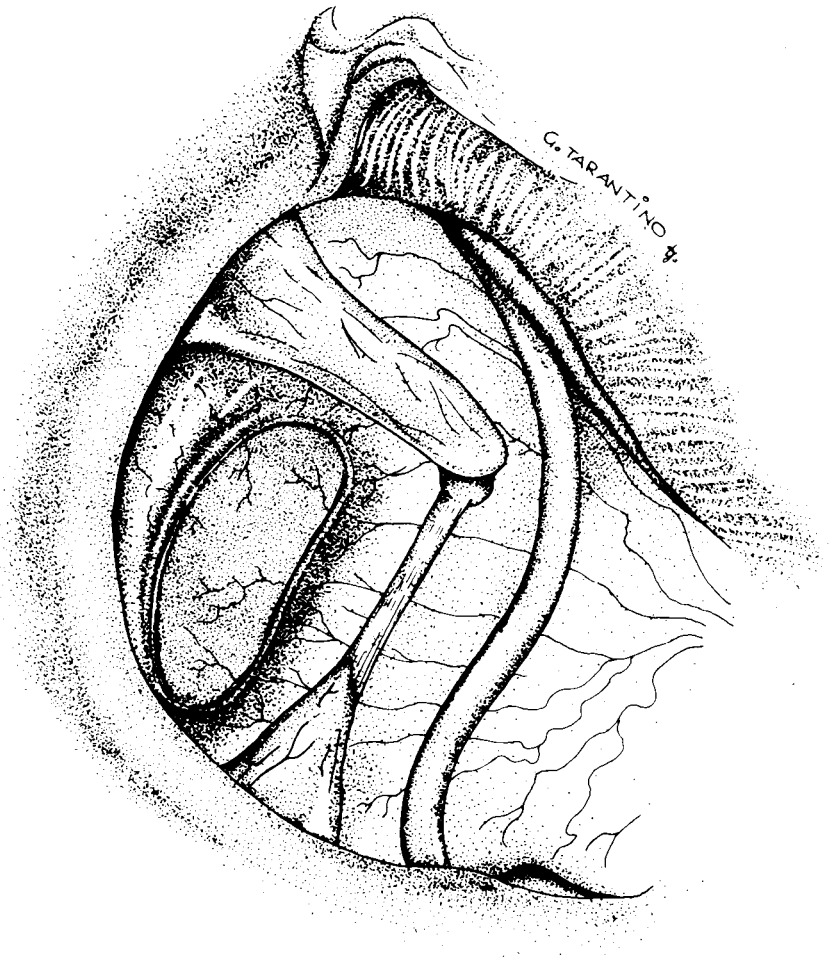


FIG. 2

Schematic drawing of ossicular defect. Note the absence of the stapes superstructure and the smooth, normal, mobile footplate; the stapedius tendon is attached to the lenticular process of the incus.

TABLE II

CONDUCTANCE (G) AND SUSCEPTANCE (B) AT 220 AND 660 HZ. PROBE TONE FREQUENCIES IN THE RIGHT EAR (RE), IN THE LEFT EAR PRE-OPERATIVELY (LE_1) AND IN THE LEFT EAR POST-OPERATIVELY (LE_2).

Probe tone	220 Hz.		660 Hz.	
Admittance component (mmhos)	G	B	G	B
RE	0.2	0.5	1.4	1.1
LE_1	0.25	0.7	not computed	not computed
LE_2	0.2	0.5	1.2	1.1
Normal range	0.1–0.5	0.2–1	0.5–4.5	0.5–3

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