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Stapedectomy for far-advanced otosclerosis

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Abstract
Patients with far-advanced otosclerosis (FAO) may appear to be suffering from profound sensorineural hearing loss and are frequently directed to cochlear implantation programmes. In order to avoid such misdiagnosis, FAO should be considered in patients with non-measurable bone-conduction levels and air-conduction levels exceeding 85 dB. Specific clues can lead the clinician to suspect otosclerosis as the aetiology of hearing loss. A review of eight patients (nine ears) with FAO who underwent stapedectomy from 1985–1995 reveals that six of the eight (75 per cent) who had been unable to use a hearing aid pre-operatively obtained serviceable hearing with a hearing aid after surgery. This confirms that cochlear implantation is not the best treatment for all profoundly deaf patients; some are better off with stapedectomy.

Key words: Otosclerosis; Hearing loss, sensorineural; Stapedectomy; Cochlear implant

Introduction

Profound deafness has received increased attention in recent years, largely because of the availability of cochlear implants. Consequently, it is important to remember that a 'blank' audiogram with no response at any frequency does not necessarily mean absence of hearing. A blank audiogram may only mean that hearing is beyond the measurement limits of the audiometer (Frattali and Sataloff, 1993). For the speech hearing frequencies, this is around 110 dB for air conduction and 65 dB for bone conduction (Iurato *et al.*, 1992). Sheehy (1962) pointed out that patients with far-advanced otosclerosis (FAO) may present with that audiological profile. A patient with a 65 dB bone conduction level and a 60 dB air-bone gap would probably have a blank audiogram, suggesting total deafness. Such a patient would have great difficulty even with a powerful hearing aid. Failure to recognize the condition may result in a lack of treatment or unnecessary cochlear implant surgery. Stapedectomy could improve the air-conduction threshold of these patients to an easily aidable threshold (Glasscock *et al.*, 1996).

The term 'far-advanced otosclerosis' (FAO) was coined by House and Sheehy (1961) to describe clinical otosclerosis with an unmeasurable bone-conduction threshold and an air-conduction threshold of 85 dB or greater on a standard clinical audiometer. Iurato *et al.* (1992) proposed the term 'very far-advanced otosclerosis' (VFAO) to indicate otosclerotic patients with both bone- and air-conduction thresholds non-measurable on a standard clinical audiometer. The reason they gave a separate name for otosclerotic patients with a blank audiogram is to differentiate them from other post-lingually totally deaf patients who are candidates for cochlear implants.

Sheehy (1978) published specific diagnostic clues for FAO. The following clues can be obtained from the patient's history: (1) positive family history for otosclerosis, (2) progressive hearing loss beginning in early adult life, (3) paracusis during the early stage of the disease, (4) past use of a bone-conduction hearing aid, (5) previous audiograms showing an air-bone gap. In addition, the following criteria can be obtained from the physical examination: (1) normal voice, (2) positive Schwartze's sign (Frattali and Sataloff, 1993), (3) evidence of otosclerosis on high-resolution computed tomography (Von Glass and Philipp, 1988), (4) a Weber test lateralizing to the poor ear or a negative Rinne test by a 512-Hz tuning fork, (5) no other apparent cause for hearing impairment. However, in these patients with FAO and VFAO the diagnosis is presumptive and is confirmed only at surgery (Iurato *et al.*, 1992).

It is the aim of the present article to review the literature on FAO and VFAO, and to present the results of nine stapedectomies done for FAO and VFAO in Tanta University Hospitals, Egypt, over a 10-year period.

Patients and methods

A retrospective analysis was conducted of the charts of all patients with FAO or VFAO who underwent stapes surgery in Tanta University Hospitals from 1985 to 1995. All patients had air

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TABLE I
PATIENT SUMMARY

Case No.	Side	Pre-op PTA (dB HTL)	Post-op PTA (dB HTL)	Pre-op discrim (%)	Post-op discrim (%)	Patient satisfaction
1	L	85	65	18	70	Yes
2	L	95	70	12	65	Yes
3	R	100	70	10	68	Yes
4	L	NR	NR	CNT	CNT	No
5	R	NR	85	CNT	56	Yes
6	R	NR	95	CNT	16	No
	L	NR	75	CNT	52	Yes
7	R	NR	NR	CNT	CNT	No
8	L	NR	70	CNT	62	Yes

Pre-op PTA = pre-operative pure-tone average.
Post-op PTA = post-operative pure-tone average.
HTL = hearing threshold level.
Pre-op discrim = pre-operative discrimination.
Post-op discrim = post-operative discrimination.
NR = no response.
CNT = cannot test.
L = left.
R = right.

conduction levels exceeding 85 dB hearing threshold level (HTL) in the speech frequencies and non-measurable bone conduction. All of them were unable to benefit from amplification. For each case, demographic data, operative findings, surgical technique, prothesis type, graft type, pre-operative and post-operative pure-tone average (PTA) and speech discrimination, and incidence of complications were stored in a computer database. PTA was calculated by averaging the recorded hearing loss over 500, 1000 and 2000 Hz. The success of stapedectomy in treating FAO was determined by objective and subjective criteria (Frattai and Sataloff, 1993). Objective improvement may be noted in PTA and discrimination score, but subjective patient satisfaction with amplification is the real measure of success, because the operation was performed to restore hearing to an aidable level.

Results

Of the 11 operations reviewed, two were excluded from the study because no obvious conductive pathology was found on exploration. The remaining nine procedures were performed in eight patients; five women and three men. The age range was 52 to 67 years (mean age 61 years). All the patients were found to have otosclerosis of the oval window. All operations were stapedectomies with total footplate removal, a House wire was used as the prosthesis and perichondrium as the tissue graft. There were no complications.

The post-operative hearing results are summarized in Table I. Five patients could use a hearing aid successfully after stapedectomy. One patient required re-operation on the opposite ear due to inability to use a hearing aid post-operatively in the ipsilateral ear, and the other two patients refused re-operation. Thus, six patients could eventually use a hearing aid, making the overall success rate six out of nine operations (66.6 per cent) or six out of eight patients (75 per cent).

Discussion

Patients with FAO may appear to be suffering from profound sensorineural hearing impairment, and are frequently directed to cochlear implantation programs. In order to avoid such misdiagnosis, FAO should be considered in patients with air-conduction levels exceeding 85 dB and non-measurable bone-conduction levels. Certain clues may help, but the definitive diagnosis requires middle ear exploration.

The value of stapes surgery for this disease entity was first discussed in 1960 by House and Glorig. Sheehy (1964) reviewed 67 patients who underwent stapedectomy for FAO and concluded that there is no maximum bone-conduction threshold above

TABLE II
LITERATURE REVIEW

Authors	Total no. of cases	Hearing improved	Overall success (%)
House (1959)	4	3	75
Willis (1963)	28	21	75
Myers *et al.* (1963)	20	14	70
Sheehy (1964)	46	21	46
Sellars (1972)	10	8	80
Wiet *et al.* (1987)	2	2	100
Iurato *et al.* (1992)	3	3	100
Frattali and Sataloff (1993)	9	7	78
Glasscock *et al.* (1996)	11	9	82
This study	8	6	75

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which stapedectomy is contra-indicated. The most gratifying aspect of stapedectomy for FAO is converting the patient's hearing from non-serviceable to serviceable with conventional hearing aids. Our study proved that 66.6 per cent of ears (75 per cent of patients) were aidable after stapedectomy. These results compare favourably with those obtained in other series (Table II).

Since patient satisfaction with a hearing aid is the real measure of success, careful pre-operative counselling and informed consent are extremely important in patients with FAO (Frattali and Sataloff, 1993). The patients must be aware not only of the risks of the procedure, but also of the relatively limited goals. Normal hearing is not expected and the patient should be cautioned that amplification via a hearing aid will probably be needed after surgery to receive the optimum potential benefit (Wiet *et al.*, 1987). Sheehy (1978) found that in FAO the success rate was higher in those patients who had been wearing a hearing aid pre-operatively, ie. in ears which were 'trained' to hear by an aid.

The importance of considering the diagnosis of FAO when evaluating a patient with bilateral profound sensorineural hearing loss cannot be over-stressed. Compared to cochlear implantation, the stapedectomy is less-labour intensive, less expensive and involves less risk and less rehabilitation. Moreover, the hearing achieved with amplification following stapedectomy is generally better than that expected with cochlear implant. When a successful outcome is not achieved, the patient may still be suitable for cochlear implantation (Frattali and Sataloff, 1993).

A convincing histological explanation for increased bone-conduction threshold in FAO remains an issue for continued investigation. Schuknecht and Barber (1985) showed no correlation between bone-conduction thresholds and size of the lesion, activity of the lesion, involvement of endosteum or presence of a round window lesion. On the other hand, Wiet *et al.* (1987) reported moderate diffuse loss of hair cells and loss of cochlear neurons in the basal turn. Myers and Myers (1968) found atrophy of the spiral ligament with basilar membrane rupture.

Conclusion

Since FAO is one of the few conditions that permits treatment capable of converting the patient's hearing from nonserviceable to serviceable with conventional hearing aids, it is extremely important for otolaryngologists to be cognizant of this entity. Certain clues help, but definitive diagnosis needs middle ear exploration. When FAO is diagnosed, stapedectomy can provide aidable hearing in 75 per cent of patients.

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