CASE REPORTS

A FAMILIAL INCIDENCE OF OTOSCLEROSIS

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Carhart (1963) reports a family wherein two brothers were suffering from labyrinthine otosclerosis. Their mother had a long standing hearing loss and their own difficulties had been progressive since the ages of 11 and 15 respectively and the younger man exhibited a lower level of hearing loss. Their audiograms showed bilateral mixed hearing loss.

Although reports like the above are seen in large numbers where one or two members of a family suffer from otosclerosis, we do not find a report in the literature of a family of otosclerotics, where in all the siblings, their mother, grand father and a host of others are involved. This article presents one such family described. The members of the family all hail from Mysore city and experience progressive hearing loss, since the same age of about 15 years. The members reported to the Institute only after a considerable lapse of time since the onset of hearing loss, and hence we are at a loss to know about the beginnings of their hearing impairments.

Otosclerosis is defined as a localized disease of the bone in which the pathological changes are limited to the bony capsule of the labyrinth. This is more frequently seen in women than in men. Often otosclerosis starts in early adult life. Rarely it starts before the age of 10 years or after the age of 40 years. It is more common in whites (fair-haired people) than in negroes (dark-haired people). It runs in families. The chances of otosclerosis being passed on to a child is about 10 per cent, although where there is a strong family history it may be greater (100 per cent as in the family reported). Pregnancy is liable to initiate or accelerate the progress of deafness due to otosclerosis. The cordinal symptoms of otosclerosis is deafness—often insidious in its onset and only too frequently progressive in its course. The deafness may at first be limited to one ear, but sooner or later both are usually affected. At first the low frequencies are affected and gradually the higher frequencies will also be affected (Cawthorne 1968).

According to Carhart (1963), otosclerosis can cause four types of hearing impairments:

(i) traditional conductive impairment whose audiometric trade mark is an air-bone gap;

(ii) a sensory loss due to damage to elements within the organ of corti;

(iii) a neural loss resulting from harm to the fibres and cells of the auditory nerve; and

(iv) a condition which is an inner ear conductive disturbance due to mechanical interference with cochlear motion. This state can be classified

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as pseudo-sensorineural because its audiometric outcome is undistinguishable from a true sensory or a true neural loss.

The family to be reported in this article is an example which emphasises the need for careful examination of the family history, the age of onset of hearing impairment and the nature of the course of hearing impairment on the part of the audiologist to detect clinical otosclerosis.

The family reported here consisted of a 48 year old lady N, her daughter L, and her three sons M, D, and A. N's father E was suffering from diabetics and had hearing loss, the age of onset was not known. E's three brothers F, G, H had normal hearing, but each had a daughter who had hearing impairment. E was not alive and hence the nature of his hearing impairment was not known. According to N all these members' difficulties had been progressive since the age of about 15 years.

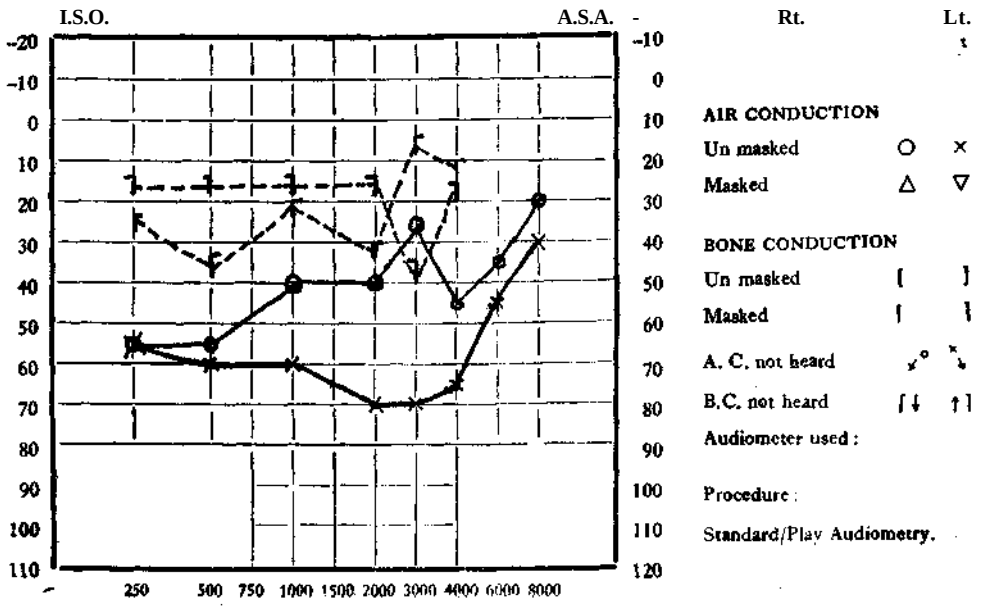
The details of the history and the relevant audiological findings of the members of the family are analysed briefly.

Case 1: The 48 year old lady N, complained that she suffered from bilateral hearing loss since the age of 14 years. She also reported that her hearing deteriorated further when she was pregnant for the third time. She had typhoid twice

AUDIOGRAM 1

Date: 17-1-69 Test No. 1 Tested by: T.G.

Audiogram of 48 year old lady N showing moderate mixed loss in right ear and moderate conductive loss in left ear



at the age of 13 and 16 years respectively. She is the second in order of birth. Her father E, had hearing loss. The case was examined at the All India Institute of Speech and Hearing, Mysore on 17-1-1969.

Her speech was normal and there were no significant psychological problems.

Her audiogram revealed that she had moderate mixed loss in right ear and moderate conductive loss in left ear.

She was again tested on 6-11-1969 and the results obtained showed essentially the same as those obtained on 17-1-1969, the maximum deviation at any one frequency being 10 d.B.

The ENT examination revealed that she had a deviated septum. Because of the family history and audiological evaluation the otologist rendered a diagnosis of otosclerosis. The case was advised to undergo stapes mobilization, as this case's prognosis was good and the surgery was bound to give her improvement in hearing.

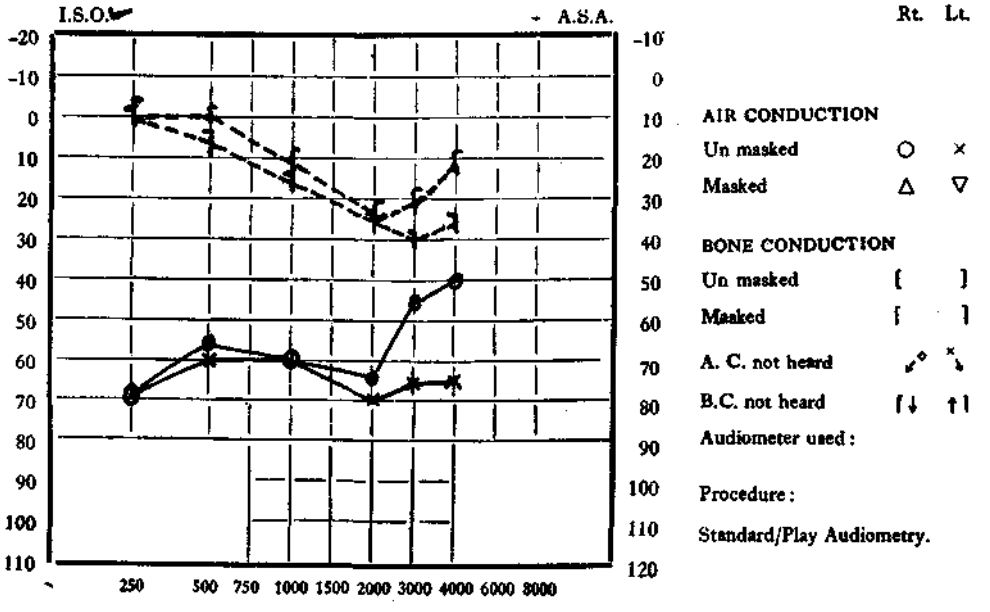
N married one of her blood relatives and hence L, M, D, A were children of consanguineous parents.

Case 2: L aged 28 years is the first daughter of N, and she had two children who were reported to have normal hearing. L had normal hearing till the age of 15 years and there upon her hearing acuity diminished gradually. She com-

AUDIOGRAM 2

Date: 17-1-69 Test No. 1 Tested by: T.C.

Audiogram of 28 year old lady L, showing bilateral mixed hearing loss



plained of bilateral hearing impairment and tinnitus in both the ears. She reported that she had typhoid at the age of 18 years.

Her speech was normal and there were no associated psychological problems.

Audiological testing indicated a bilateral moderate mixed hearing loss.

ENT examination at the Institute indicated negative aural physical factors. The otologist's diagnosis was otosclerosis based on the audiological findings and the familial incidence of hearing loss.

Case 3: M aged 25 years is the second child to N. He was reported to have had normal hearing till the age of 14 years and there upon his hearing acuity diminished. He is reported to have had head injury without bleeding at the age of 3 years; typhoid at the age of 13 years; and whooping cough at the age of 7 years.

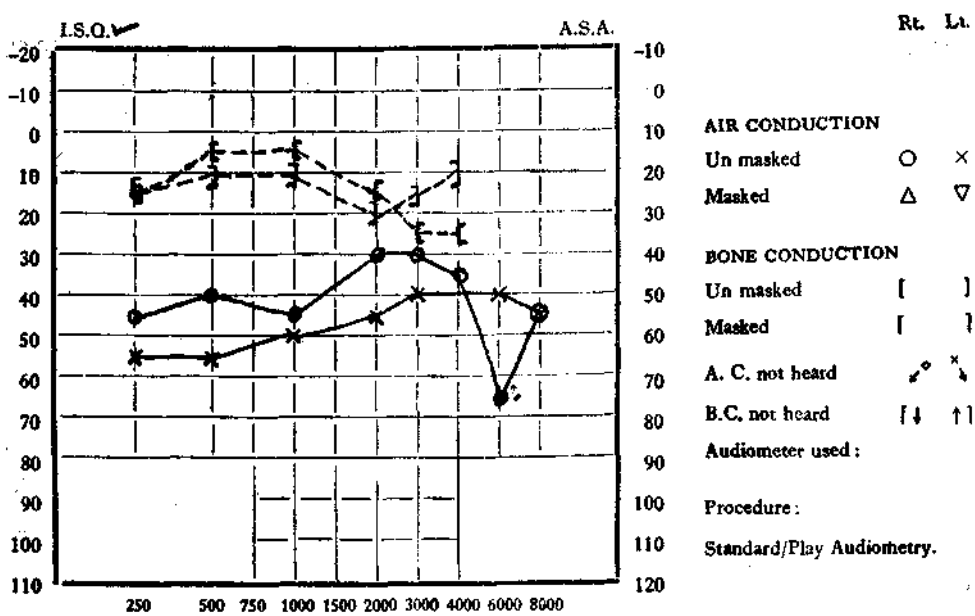
The speech examination revealed that his speech was babyish or infantile. The psychological examination revealed that the boy was mentally below average and got an IQ of 76 on the short version of Bhatia's battery.

The audiometric tests showed that he had bilateral conductive hearing loss.

AUDIOGRAM 3

Date: 17-1-69 Test No. 1 Tested by: T.C.

Audiogram of 25 year old M, showing bilateral conductive hearing loss



The absence of any aural physical factors, the presence of familial history of hearing loss and the bilateral conductive hearing impairment manifested enabled the otologist to diagnose the case as an otosclerotic.

Case 4: D aged 17 years is the third child to N, was reported to have had difficulties in hearing progressive since the age of 14 years. He had typhoid at the age of 3 years. He was reported to have hit a telephone pole resulting in head injury without bleeding or wound and to have been unconscious for few minutes at the age of 11 years. He also complained of occasional ear aches and tinnitus in both ears noticed during night times.

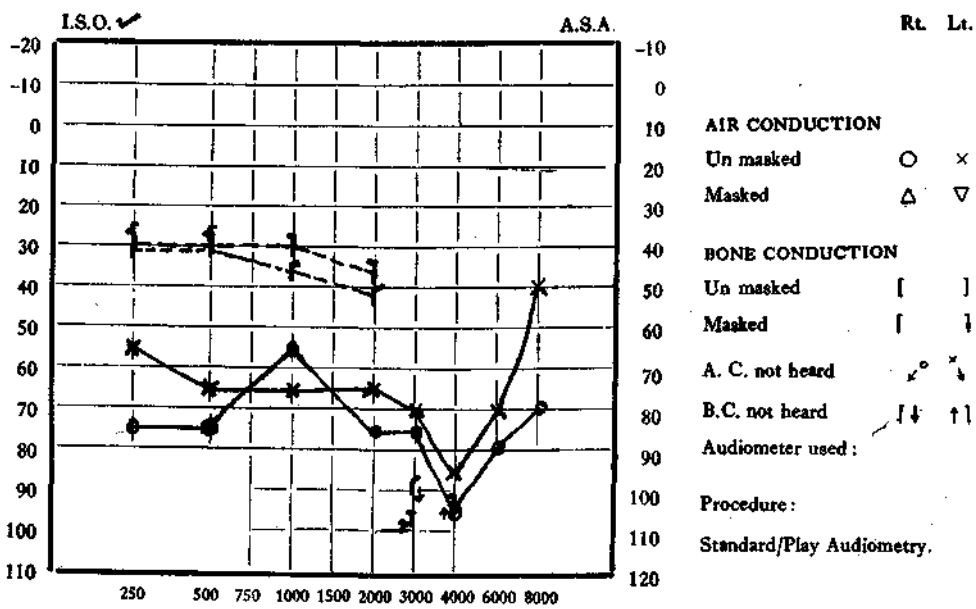
His speech was normal and the psychological examinations did not reveal anything abnormal.

The audiological examination revealed a severe bilateral mixed hearing loss.

AUDIOGRAM 4

Date: 30-12-69 Test No. 1 Tested by: T.C.

Audiogram of 17 year old boy D, showing severe bilateral mixed hearing loss



Repeat audiological testing on 18-7-1969 confirmed the results obtained on previous audiometric tests showing essentially the same levels and patterns as those obtained on 30-12-1968. The maximum deviation found at any one frequency was about 10 dB.

His right ear SRT was 75 dB and the discrimination score was 52 per cent. His left ear SRT was 70 dB and the discrimination score was 80 per cent. These were essentially the same during the retest.

Tone decay and SISI scores were negative for cochlear lesion.

ENT examination revealed that the auditory meatus Was narrow, and the neck glands palpable but not markedly enlarged. In view of the long standing family history the otologist's diagnosis was otosclerosis.

Since surgery was not thought to be of much use in this case, he was advised to buy and use a hearing aid.

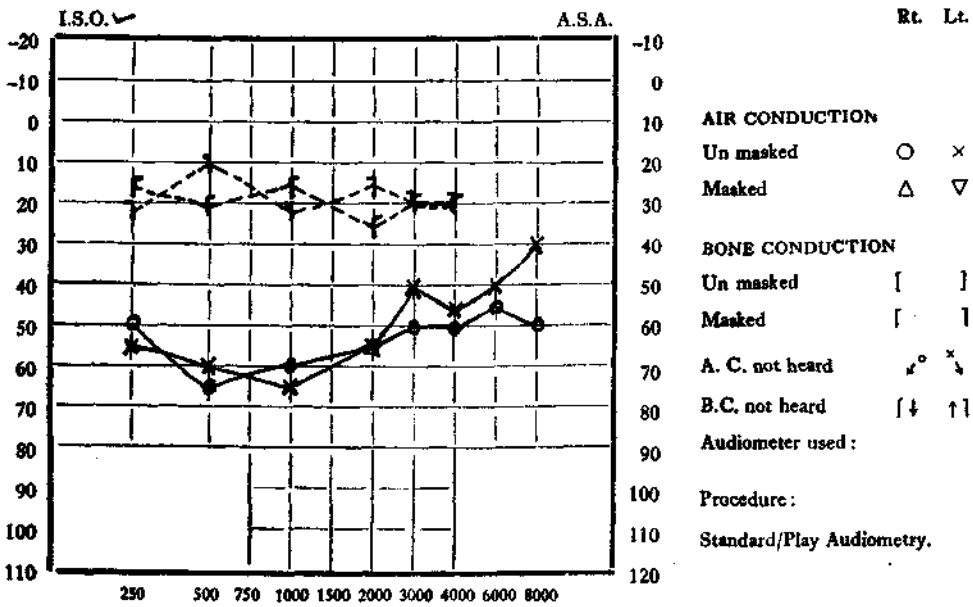
Case 5: A, aged 14 years is the fourth child to N. This boy was reported to have been experiencing difficulty in hearing since four or five months.

His speech was normal, and there were no associated psychological problems.

AUDIOGRAM 5

Date: 30-12-69 Test No. 1 Tested by: T.C.

Audiogram of 14 year old boy A, showing bilateral mixed hearing loss



The audiological testing indicated that he had bilateral mixed hearing loss.

Repeat tests also showed essentially the same values as obtained previously. The maximum deviation noticed at any one frequency was about 10 dB.

The SRT was 55 dB in both ears and the discrimination score was 100 per cent in both the ears.

The ENT examination revealed the following abnormalities: hypertrophied tonsils, septal obstruction, allergic rhinitis and small midline adenoids. He was advised to get his tonsils and adenoids removed. He was also advised to undergo stapes mobilization. Familial otosclerosis was the diagnosis made by the otologist.

Observations:

The tendency for otosclerosis to run in families is also seen in the family reported. Because of the strong family history all the siblings are affected. Some of the interesting facts revealed in the audiograms of these five members are mentioned briefly, although, an attempt is not made to explain as to why they are so.

None of the audiograms resemble each other. Each of them show their own unique audiometric configurations.

The three frequency puretone averages of all the five members are essentially the same in left ear (except M who shows 10 dB lesser loss) and they differ considerably in the right ear. Comparing the audiograms we find that all exhibit better hearing in the right ear through air conduction than in left ear except D who shows better hearing in the left ear than in his right ear.

The sex ratio of incidence in the case of these five members is 2:3. Showing that more males are affected than females contradicting the usual ratio of incidence in otosclerosis. This may be so, because we have considered only a single branch of the family and it may prove otherwise when the other branches of the family are investigated.

Even though all the members except D suffered from typhoid, apparently it has not made its impact on their hearing since the type of loss they exhibit has no relation to the type that would be expected as the outcome of typhoid.

The head injury reported in the case of M has not made its impact on his hearing since he acquired hearing loss only at the age of 14 or 15 years, several years after the head injury. In case of D, it is stated that the boy had hit a telephone pole and he lost his hearing soon after that. But the mother claims that he had normal hearing even after the head injury, and that he acquired hearing loss only at the age of 15 years. In this case we cannot precisely say whether head injury or the familial otosclerosis caused the hearing impairment, since there is no agreement between the mother and the son as to the time of onset of hearing impairment.

This report emphasises the importance of the careful examination of the family history, the age of onset and course of the hearing impairment in order that the audiologists can detect cases of clinical otosclerosis. Reports like this of similar families will throw more light on the nature of otosclerosis. The information these reports furnish will prove to be helpful for the audiologist to advice proper medical treatment.

This report is confined to only one branch of the family. However, attempts are being made to get in touch with the other members of the family and test them. The details of these investigations will be reported in a subsequent paper.

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Cawthorne, T. (1968) Otosclerosis. In Scott-Brown W. G., Ballantyne, J. and Groves, J. (Eds.) (1968) *Diseases of the Ear, Nose and Throat,* Vol. 2: London: Butterworth & Co. Ltd.