CONGENITAL ABSENCE OF INCUS & STAPES SUPERSTRUCTURE

L E Loh

SYNOPSIS

A case of congenital absence of incus and stapes superstructure is presented. The audiological investigative findings and technique of repair are reported. The various aspects of congenital ossicular defects; classification, embrology, diagnostic clues are discussed.

SING MED J. 1988; 29:182–183

INTRODUCTION

Ossicular defects (discontinuity) may result from various causes such as congenital malformation, infection of middle ear, cholesteatoma and trauma.

Congenital malformation of middle ear ossicles takes several forms depending upon the extent of maldevelopment during the embryonic stage of fetal development. This case highlights the involvement of the incus and stapes.

CASE REPORT

N.K.H., a 28 year old Chinese lady, has impaired hearing in the left ear since the memory of her childhood, the impairment is non-progressive and there is no history of head injury nor family history of deafness.

Examination of the ears reveals that the tympanic membranes, auditory canals and auricles are normal. Rinne's test reveals a conductive deafness in the left ear, Weber's test reveals localisation to the left.

Preoperative audiometry reveals a moderate degree of conductive deafness in the left ear; average air-bone gap for the speech frequencies being 55 db. There is some degree of sensorineural impairment in the high frequencies bilaterally. The right audiogram is otherwise normal (Fig. 1). Tympanometry reveals type A tympanogram bilaterally at both 275 & 660 Hz. However, the left tympanogram has a high compliance. Acoustic reflex threshold is abscent bilaterally. Xray of the mastoids is normal.

A left tympanometry was carried out on 28.6.84 to explore the middle ear. It was found that the long process of the incus and the superstructure of the stapes were absent. The malleus, the body of the incus and the footplate of the stapes were normal. The handle of a homograft malleus was utilised to bridge the patient's handle of malleus and stapes footplate. This was achieved by amputating the head of the homograft malleus and grooving the surface of amputation to receive the patient's handle of malleus.

Department of ENT
Tan Tock Seng Hospital
Moulmein Road
Singapore 1130

L E Loh, MBBS, FRCS, AM
Registrar

Post-operatively, the patient's hearing improves. The left audiogram carried out one month post-operatively reveals an average air-bone gap of 20 dB for the speech frequencies; a closure of 35 dB (Fig. 2). A repeat audiometry carried out eight months post-operatively shows that her hearing has remained stable.

DISCUSSION

A number of ossicular defects can exist either in solo or in combination. Generally, such defects have been classified into four types: 1. Ossicular defect involving only the incus. 2. Ossicular defect involving the incus and the stapes. 3. Ossicular defect involving the incus and the malleus. 4. Ossicular defect involving the incus, malleus and stapes.

Austin (2) in a study of 1151 patients with non-congenital chronic ear disease found 11.6% had type 2 defect i.e. loss of long process of incus and stapes superstructure. This was found to be usually associated with cholesteatoma. No study has quoted any figure on type 2 defect due to congenital malformation as in this case report.

In order to understand the various combination of ossicular defects due to congenital malformation, one needs to understand the embrology of the development of ossicles. Hough (1) in a study of congenital malformation of the middle ear states that the first pharyngeal arch participates in the development of the head of malleus and the body of incus. The second pharyngeal arch participates in the development of the handle of malleus, long process of incus, stapes superstructure and footplate. The latter additionally receives contribution from the labyrinthine capsule by a process of fusion. Thus a maldevelopment of the second pharyngeal arch may produce defects involving the handle of malleus, long process of incus and stapes superstructure in various combination, as occured in this case report.

The following clues point to possible congenital middle ear malformations: 1. a history of deafness since birth or since the memory of early childhood. 2. a family history of malformation of the middle ear. 3. malformation of external ear. 4. unilateral, non-progressive conductive deafness.

Numerous articles have been published on the techniques and materials used in the repair of ossicular defects; autograft or homograft incus or malleus and synthetic prosthesis(3)(4). With respect to homograft, it should not be harvested from donor who has a positive serology, a past history of jaundice, evidence of carcinoma or infection of the middle ear. Homograft

should be stored in 70% ethyl alcohol at room temperature.

At the time of implantation, it should be washed with normal saline to remove the alcohol and should be denuded of all tissues.

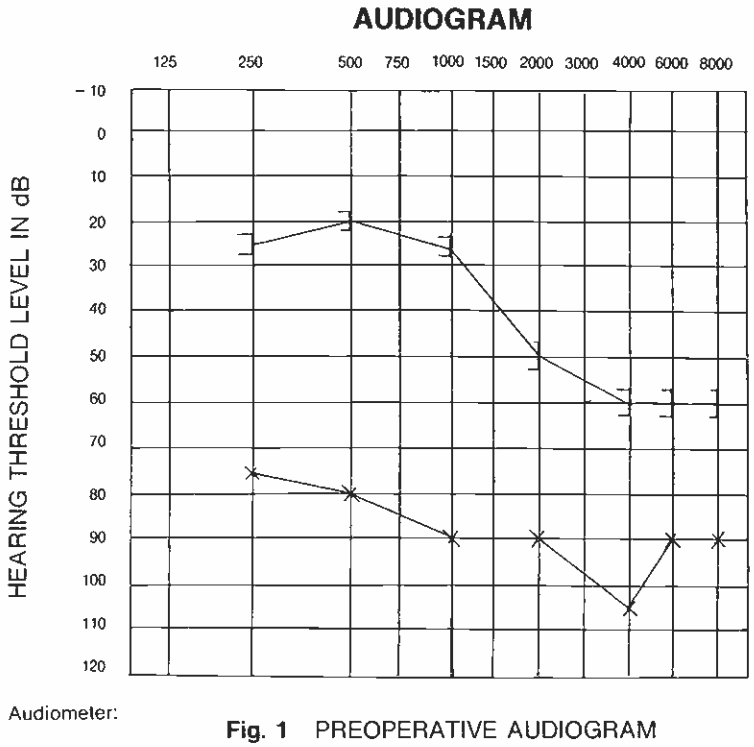

Fig. 1 PREOPERATIVE AUDIOGRAM

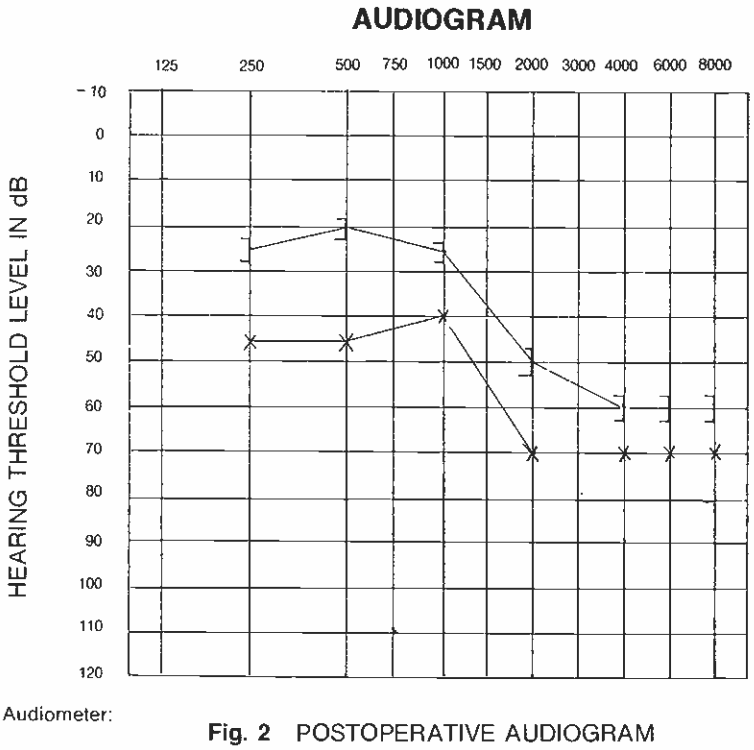

Fig. 2 POSTOPERATIVE AUDIOGRAM

REFERENCES

1. Hough J V D: Congenital Malformations of the Middle Ear. Arch Otol 1963; 78:335–43.
2. Austin D F: Ossicular Reconstruction Arch Otol 1971; 94:525–35.
3. House W F etal: Incus Homografts in Chronic Ear Surgery. Archives of Otolaryngology 1966; 84:148–53.
4. Pennington C L: Incus Interposition Techniques. Ann Otol Rhinol & Laryngol 1973; 82:518–29.