CONGENITAL OSSICULAR ABNORMALITY
A Case Report

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INTRODUCTION

In children, congenital deafness presents not infrequently. The majority of cases are sensorineural in nature and congenital conductive deafness is far less common. The causes range from abnormalties and fixation of ossicles, ankylosed malleo-incudal joint and absence of one or more ossicles. We describe a case of congenital ossicular abnormality, presenting with conductive deafness.

CASE REPORT

A 9-year-old Chinese boy presented to our unit early in 1986 with bilateral deafness noted over the last few years. The deafness was moderately severe and non-progressive. The patient also had a severe speech handicap. There was no vertigo, tinnitus or previous history of ear disease. The ante-natal and post-natal history was normal as were his milestones. There was no history of familial deafness of consanguinity.

Examination revealed a normal child with normal ear canals and tympanic membranes. Tuning fork tests produced a negative Rinne response bilaterally and a centralised Weber's test. Audiometry test confirmed a bilateral conductive deafness of 70 dB and a tympanogram showed a type A curve for both ears. X-rays showed normal, well-pneumatised mastoids.

On 13 October 1986, an exploratory tympanotomy on the right side showed normal middle ear mucosa, a normal malleus and a thick long process of incus. The stapes was abnormal with a deformed posterior crus and a short anterior crus which was not in contact with the footplate (Fig.I).

A stapedectomy was performed and a Teflon wire-piston (0.6 mm X 4.50 mm) prosthesis was inserted. The outer ear was then packed with BIPP pack. Post-operatively, the patient recovered well with minimal vertigo and tinnitus. He was discharged on the 2nd post operative day.

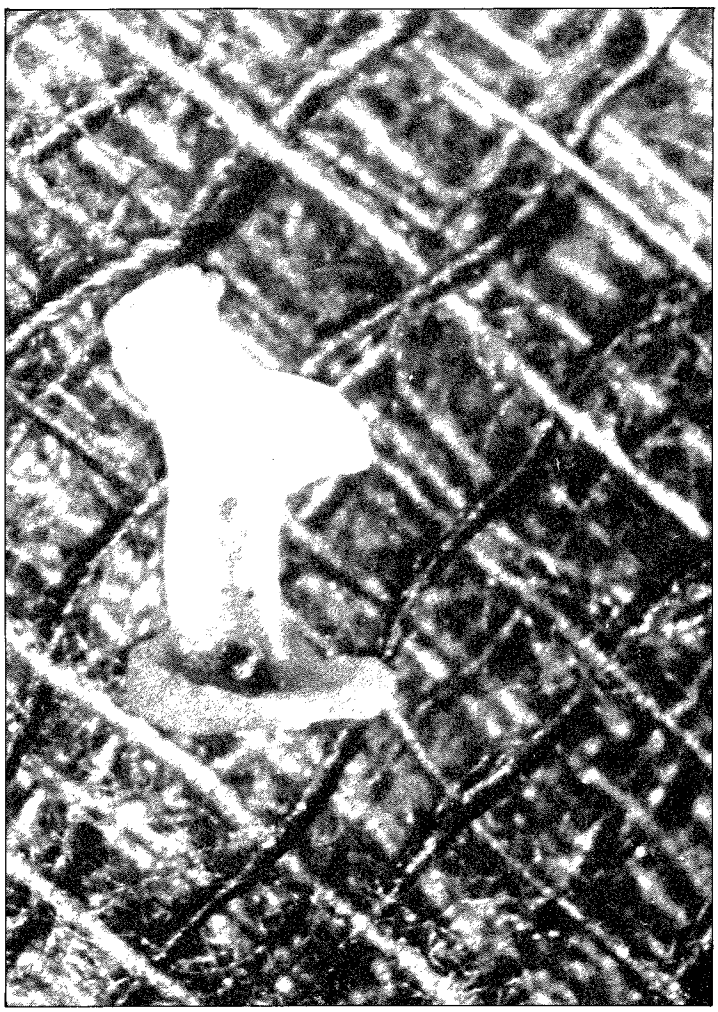

Fig. I Showing the abnormal stapes with short anterior crus.

DISCUSSION

Congenital conductive deafness occurs less often than congenital sensorineural deafness. Various abnormalties of the outer and middle ear structures, including the ossicles have been described.[1] The final appearance of the stapes is a result of remodelling that occurs during its development from a simple ring of bone perforated by the stapedial artery.[2] Various aetiological factors have been implicated that can lead to arrest of the remodelling, causing the stapes to be abnormal.

The diagnosis of congenital conductive deafness is made on clinical evidence. Exploratory tympanotomy reveals a marked variety of abnormalties that can occur in the ossicles.[3,4] Various ossiculoplastic procedures have been described to correct these congenital abnormalties. The results of these operations are generally satisfactory.

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