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# Assessment of stapes-pyramidal fixation by a bony bar

**Dr. Chandan Kumar[1*], Dr. Ratnesh Kumar[2], Dr. Umesh Kumar[3], Dr. Chandra Shekhar[4]**
[1, 3] Senior Resident, Department of (ENT), Nalanda Medical College & Hospital, Patna, Bihar, India
[2] Senior Resident, Department of (ENT), Patna Medical College & Hospital, Patna, Bihar, India
[4] Professor & HOD, Department of (ENT), Nalanda Medical College & Hospital, Patna, Bihar, India
[*] Corresponding Author: Dr. Chandan Kumar

**Abstract**
Symmetrical hearing loss averaging 30-60 dB (0-125-8 kHz) became apparent between 8 and 24 years of age. Tympanotomy showed a fixed stapes either through ossified stapedius tendon or through a bony bridge from the stapes to the pyramidal eminence in all patients. After surgical removal of the bony tendon hearing was normal. Both parents, four other sibs, and all grandparents had normal hearing. This family and a further published case suggest a possibly recessive inheritance of this form of conductive hearing loss.

**Keywords:** stapes fixation, conductive hearing loss, autosomal recessive, ossicular malformations

## Introduction

Conductive and sensorineural hearing loss is of genetic origin in between 20% and 60% of cases. Both conductive and sensorineural hearing loss is found as part of recognisable syndromes as well as without any associated abnormality. '2 Generally, genetic abnormalities are more often expressed as sensorineural hearing loss than as conductive hearing loss. Nevertheless, at least 50 different syndromes with conductive hearing loss caused by dysplasia or fixation of the ossicles of the middle ear or by malformations of the external auditory canal have been described [2].

The most common genetically transmitted form of isolated conductive hearing loss is otosclerosis. Its frequency ranges between 0-2 and 2% [3]. The basic defect is ossification of the stapes footplate causing fixation of the stapes and preventing transmission to the inner ear. The aetiology is unknown and is considered to be transmitted either autosomal dominantly with incomplete penetrance and variable expression or through the combined action of several additive genes (polygenic inheritance).'

Other isolated genetic malformations of middle ear ossicles include incudostapedial disconnection, which is autosomal dominantly inherited [5]. Recessive inheritance is observed in fixation of the malleus or incus or both [6]. Different degrees of malformation up to total absence of the stapes have been described although no familiality has been reported [8]. All these forms of malformation result in conductive hearing loss and can only be differentiated through tympanotomy.

We present here a family with a possibly new autosomal recessive form of hearing loss caused by stapes fixation owing to ossified stapes tendon. Four affected sibs show a similar clinical picture and pathological changes of the stapes tendon.

A 44 year old woman, the first daughter of a non-consanguineous family was first referred at the age of 24 years with progressive hearing loss in her right ear. No external ear anomalies were apparent. The tympanic membrane was intact. There were no findings or history of otitis media. Audiometry showed a symmetrical conductive hearing loss averaging 40 dB at 2 kHz in the right ear and 30 dB in the left ear (0 125-8 kHz).

## Research Study

Rinne test was bilaterally negative and stapedial reflex was also negative. Tympanotomy in the left ear showed a stapes fixed by a bony bar from the neck of the stapes to the pyramidal eminence parallel to the stapedial tendon. The stapes footplate was normal with no evidence of previous infection or otosclerosis. After removal of the bony bar, hearing was normalised. Subsequent tympanotomy in her right ear produced the same findings. Normalisation of hearing after surgery was documented through audiometry (5-10dB from 0-125-8kHz).

Audiometry showed conductive hearing loss of 30-50dB at 0-125-2kHz in her right and 30-40 dB at 0 125-8 kHz in her left ear. The stapedial reflex was negative. Left tympanotomy showed ossification of the stapedial tendon, which was cleaved. Postoperatively audiometry was normal. Tympanotomy in her right ear is still pending. Progression of hearing impairment in this ear over the years was documented by renewed audiometry.

A family with four sibs with bilateral stapes fixation is presented. Both parents and four other sibs have normal hearing which was verified through audiograms in all cases. No family history of hearing loss, except one case of probable presbycusis in the maternal grandfather, is documented. Age of onset was variable with progressive hearing loss starting between the ages of 8.

Patients reported progressive hearing loss which after three to five years reached 30-50 dB at 0-125-8 kHz. Hearing was completely normalised after surgical cleavage of the ossified stapedial tendon or the bony bridge, respectively. After 20 years of follow up the hearing in sib 1 is still normal.

Together with possible secondary changes caused by the surgery, a stapes plastic finally became necessary. This did then restore normal hearing, confirming the conductive nature of the hearing impairment in this sib also. Among the middle ear ossicle anomalies the stapes is the most often

affected ossicle. Some authors have discussed a relationship between malformations of the stapes and the persistence of the A stapedia, a ramification of the A carotis interna.

**Discussion**

In the last 10 years several cases with ossification of the stapedial tendon or a bony bridge parallel to the tendon similar to those described here have been published,"0-4 although only two were familial.13"4 Two sibs with similar findings were described by Grant and Grant,'3 a 9 year old girl and her older brother with ossified stapedial tendons. Although the authors did not mention whether the parents had normal hearing, this is probably the case as any hearing loss would probably have been noticed. Therefore this family is also compatible with a recessive pattem of inheritance.

Another case was published by Kinsella and Kerr,'4 describing a mother and her 5 year old daughter. Tympanotomy showed a bony bar extending to the stapes head from the pyramid, superior to a normal stapedius tendon. This family would suggest autosomal dominant inheritance. Several genetic factors, both dominant and recessive, could possibly lead to aberrant calcification of the stapes tendon itself or to formation of an adjacent bony bridge. As only tympanotomy can show the true nature of the underlying pathology, the full extent of this form among patients with conductive hearing loss remains unclear. In summary we present a family with a familial form of progressive conductive hearing loss owing to abnormal calcification of a tendon of the stapes, an inner ear ossicle. The inheritance pattern is suggestive of an autosomal recessive condition.

**Significance of the study**

During embryological development the stapes is formed out of the stapesblastema, which grows along the A stapedia. Normally the A stapedia obliterates during the third embryonic month. Persistence results in an increase of the calibre of the A stapedia and possibly leads to necroses in the blastema owing to compression. This can even lead to partial or total agenesis of the stapedial crura or to dysplasia of the stapes [9].

In the family presented the persistence of the A stapedia is probably not the reason for dysplasia, since ossification became apparent only later in life. Nevertheless, other similar vascular processes might have been responsible for the aberrant ossification of either the tendon itself or the formation of the adjacent bony bridge described. The most important differential diagnosis is otosclerosis.

In both diseases the stapes is involved in an abnormal ossification process, but different parts of the stapes are affected. In otosclerosis, ossification of the stapes footplate progressing to involvement of the entire oval window can be observed. The otosclerotic change may spread from the oval window to the round window and the basal turn of the cochlea and into the stapes [3]. In the patients described here, tympanotomy showed an ossified stapedial tendon and a bony bar parallel to the tendon, respectively, which excludes otosclerosis.

**Conclusion**

The similarity of the stapedial changes observed in all four sibs, together with the normal hearing in both parents and grandparents, suggests autosomal recessive inheritance. The fact that both parents as well as the grandparents of each line are not affected argues against the possibility of autosomal dominant inheritance. Although we cannot completely rule out mitochondrial inheritance with heteroplasmy in the mother, the specific nature of the changes observed as well as the similar degree of severity in the sibs argues against such a possibility.

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