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Otosclerosis- A Clinical Study

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Hearing loss is the most prevalent chronic condition which can impact the quality of life not only of the individual who has hearing loss, but also their frequent communication partners, employers, family members, friends, and acquaintances. Otosclerosis is a localized hereditary disorder affecting endochondral bone of the otic capsule that is characterized by disordered resorption and deposition of bone. An otosclerotic lesion consists of areas of bone resorption, new bone formation, vascular proliferation and a connective tissue stroma.
This present work has been undertaken to evaluate the patient of clinically suspected otosclerosis on the basis of patients history, clinical examination and audiological test (pure tone audiometry and impedence audiometry) who visited in the department of ENT, Gauhati Medical College and Hospital, Guwahati.

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I. Aims And Objectives:

1. To access the incidence of otosclerosis in this region, mainly in relation to age, sex, unilateral or bilateral ear and special community of patient.
2. To diagnose early so that curative treatment can be provided.
3. To evaluate all conductive type of deafness presenting in our OPD with available investigative facilities like audiometry, impedence and tuning fork test.
4. To provide effective treatment to the patients may be surgery, hearing aid or follow up.

II. Methodology

- Patients presenting with conductive hearing loss were taken up for the present clinical study for a period of one year. In all cases, a detailed history was taken, a thorough clinical examination was then performed.
- A fixed scheme of investigations was followed in each case. It included pre- and post-operative pure tone audiometry and impedence audiometry.
- Total number of studied cases were 35 for a period of one year.

Inclusion Criteria

- Intact tympanic membrane.
- Conductive hearing loss.
- As type of tympanogram in impedence audiometry.
- Negative Rinne's test and webers lateralized to the affected ear in tunning fork test.

EXCLUSION CRITERIA

- History of trauma or any surgical intervention of the study ear with intact tympanic membrane.
- Tympanosclerotic patch or with healed tympanic membrane.
- Mixed hearing loss.

REVIEW OF LITERATURE

Otosclerosis was first described in the early 18th century by Valsalva, who noticed a stapes ankylosed by ossification of its ligament in the course of dissecting a temporal bone of a deaf patient. In 1869, von Troltsch named the final inactive sclerotic stage of the disease, "otosclerosis." Siebenmann designated the active, hyperemic stage as "otospongiosis" in 1912. Toynbee a century later accumulated post mortem evidence of stapes ankylosis as a common cause of deafness.Adam Politzer in 1894 coined the term otosclerosis.

III. Results And Observations

Table 1: Showing distributions of age, sex, ear involvement and family history:

AGE(in yrs)			SEX		EAR INVOLVEMENT		FAMILY HISTORY		TOTAL NO OF CASES
20-30	31-40	41-50	M	F	Unilateral	Bilateral	+ve	-ve	
20	11	4	12	23	16	19	20	15	35

Of the total number of suspected cases of otosclerosis, 23 no of cases (65.71%) were female and 12 no cases (34.29%) were male which showed a female: male ratio of almost 2:1. Majority of patients 57.14% belongs to age group between 20-30 years. 57.14%(20 out of 35) cases gave positive family and 15 cases(42.86 %) gave negative family history. We found unilateral ear involvement in 16 no of cases while bilateral disease was seen in 19 cases.

Table 2: Distribution of clinical symptoms:

Sl no	Symptoms	No of cases	Percentage
1.	Hearing loss	35	100
2.	Tinnitus	30	85.71
3.	Ear blockage	15	42.86
4.	Autophonia	7	20.00
5.	Vertigo	0	0

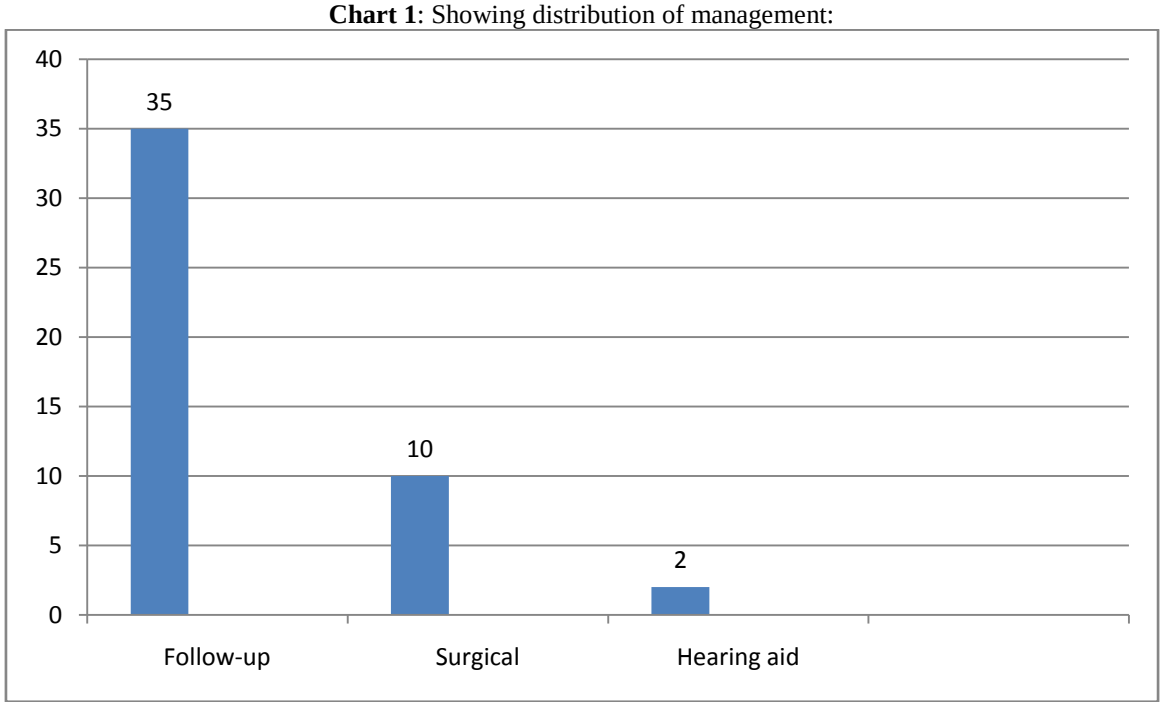
All the patients complained of reduced hearing (100%), 30 out of 35 patients complained of tinnitus(85.71%), ear blockage was complained by 15 cases(20%) and vertigo by none of the patients.

Table 3: Showing pure tone audiometric finding:

Degree of hearing loss (in dB)	No of cases	Percentage	CARHARTS NOTCH		
25-40	6	17.14	Present	Absent	Total
40-55	26	74.29			
55-70	3	8.57	14	21	35
	35	100			

The above table shows 26 of 35 cases came with 40-50 degree hearing loss(74.29%), 6 of 35 cases with 25-40 degree hearing loss(17.14%) and 3 of 35 cases with 55-70 degree hearing loss (8.57%). Of the total cases, 60% of cases (21 out of 35) with absent carhart's notch in audiogram and 40% cases with positive notch in audiogram.

Chart 1: Showing distribution of management:



Of the total patients of clinically suspected otosclerosis, all cases were undergoing follow-up and the patients included were unilateral hearing loss with normal hearing contralateral ear or were cases who refused

surgical treatment and using hearing aids, and also the post-operative patients. 10 cases were treated surgically(stapedotomy) and 2 case opted for hearing aids.

Table 4:Distributions of operative findings during stapedotomy:

Sl no	Intra-op findings	No of cases
1.	Fixed stapes footplate	8
2.	Floating stapes footplate	1
3.	Adhesive otitis media	1
	Total	10

Of the total cases opted for surgery, intra-operative findings of 8 out of 10 cases was with fixed footplate, 1case resulted in floating of footplate and 1case were diagnosed as adhesive otitis media.

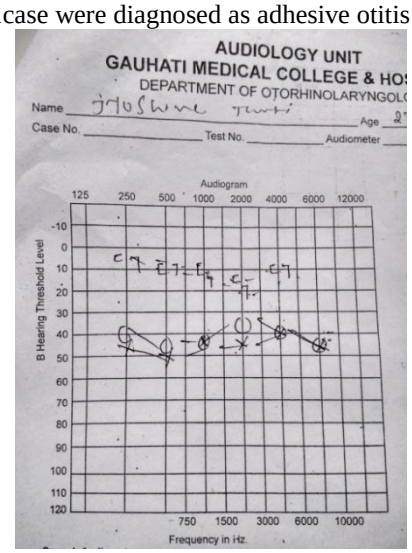


Figure 1: A pure tone audiogram in a patient of otosclerosis with carhart's notch.

(a) (b)

.**Figure 2: (a) and (b) showing image of TEFLON PISTON**

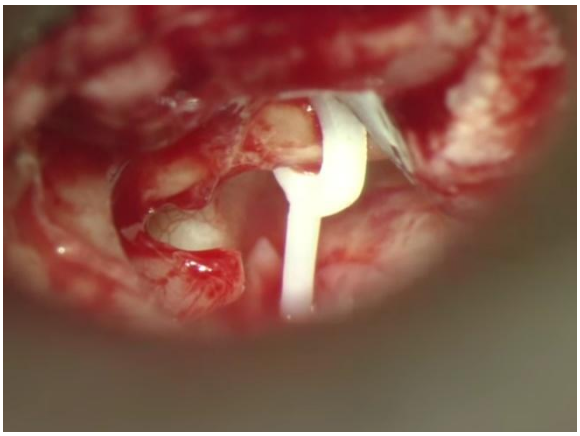


Figure 3: TEFLON PISTON PLACED AFTER STAPEDOTOMY

IV. Discussion

Hearing loss is the most prevalent chronic condition faced by adults, especially older adults; however, it is often not routinely screened. Schuknecht and Kirchner (1974) defined clinical otosclerosis as stapes fixation due to an otosclerotic lesion. The prevalence of clinical otosclerosis in epidemiological studies has been significantly less.

When the sex distribution of the total patients was studied, of the total 35 cases of suspected clinical otosclerosis ,two-third of the cases were female and one-third cases were male; which showed a female:male ratio of 2:1. It has been debated whether pregnancies and oral contraceptives have any effect on the development of otosclerosis. The incidence of the disease is more common in women, and some studies have shown that pregnancy can aggravate symptoms (Precechtel 1967, Gristwood and Venables 1983). Possible effects of oral contraceptives have also been discussed, however a study including 17032 women with oral contraceptives did not demonstrate any increased risk of ear disease including otosclerosis (Vessey *M,* Painter *R in 2001).*One clinical study showed that there is a strong evidence that otosclerosis can worsen with the pregnancy (F.Gentil et al.2016) where 53 cases were studied, out of which 35 reported worsening of hearing and 18 cases did not relate auditory worsening with pregnancy.

The age distributions of the patients of suspected otosclerosis, it was found that majority of the patients, 57.14% presented between 20-30 years, between 31-40 years accounted for 31.43% and 11.43% of cases between 41-50years. According to Batson L, Rizzolo D.2017 the most common age group presenting with hearing loss from otosclerosis is 15-45 years. The average age of onset is 30.

The most common symptoms of otosclerosis with which the patients presented in the out-patients department was with progressive reduced hearing(100%). In clinical otosclerosis, there is conductive hearing loss due to stapes fixation (Schuknecht and Kirchner 1974). Tinnitus was complained by 85.71% of the patients which was followed by ear blockage in 42.86% of cases. The prevalence of tinnitus in patients with otosclerosis is reported to be 65-92% (Gristwood and Venables 2003, Oliviera 2007).The mechanism of tinnitus according to the neurophysiological model of tinnitus by Jastreboff (1990) is compensatory activity of the neuronal network induced by hearing loss. Around 20% of patients presented with autophonia while none of the patient presented with vertigo.

When ear involvement of all the cases was studied, bilateral involvement was seen in 19 cases and 16 no of cases with unilateral involvement. Examination of both pairs of temporal bones from individuals have shown that otosclerosis is usually bilateral, with involvement of both ears in 70–90 percent of cases (Schuknecht HF et al.1985).

On coming to the familial history of otosclerosis, in the present study 57.14% of cases gave positive family history and 42.86% with negative family history. The hereditary pattern recognized so far is autosomal dominant with reduced penetrance (40%)((Moumoulidis et al. 2007, Markou and Goudakos 2008). Although a familial link is well established, 40-50% of clinical cases are sporadic (Sabitha et al. 1997, Moumoulidis et al. 2007, Markou and Goudakos 2008).

When audiometric findings of the cases of otosclerosis were studied, 74.29% of cases had hearing loss between 40-55 decibels, 17.14% of cases with 25-40 db while 8.57% with 55-70 db hearing loss. At early stages, only low-frequency hearing loss is noticeable. But if the disease remains active and the footplate becomes increasingly immobile, hearing loss will eventually occur across all frequencies. Also of all the cases of otosclerosis studied, 40% had positive carhart notch and 60% with negative carhart's notch. A depression of bone conduction thresholds at 2000 Hz (Carhart's notch) is often seen in otosclerosis, but it is not considered pathognomonic (Kaz J.2002).

And amongst the total patients of clinically suspected otosclerosis, 35 cases were undergoing follow-up as the patients had unilateral hearing loss with normal hearing contralateral ear and also patient included in this group were who refused surgery or were not fit for surgery or were followed up post operative and patients with hearing aid.10 cases were treated surgically(stapedotomy) and 2 case opted for hearing aids.

Stapedotomy has been shown to give better high-frequency gain and postoperative speech discrimination scores (Fisch 1982, Bailey et al. 1983, Cremers et al. 1991, Spandow et al. 2000, Kos et al. 2001). Stapedectomy in turn, may give better gain at the lower frequencies (Persson et al. 1997, Kos et al. 2001).

Of the total cases stapedotomy, intra-operative findings of 3 out of 5 cases was with fixed footplate, 1case resulted in floating footplate and 1case were diagnosed as adhesive otitis media.

V. Conclusion

- Otosclerosis is not only a middle-ear disease, but also an inner-ear disease which is a progressive yet treatable form of hearing loss.
- Almost all surgically treated subjects will eventually need hearing aid rehabilitation.

- Lastly, to conclude I must say that proper history taking, thorough clinical examination are essential for early diagnosis and management along with audiological investigations.

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