Original article:

Cholesteatoma….. revisited!!

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ABSTRACT

The stratified squamous epithelium of the tympanic membrane and external ear canal can migrate prior to being shed at the entrance to the external meatus.in this way the ear canal protects itself from filling with shed keratinocytes.

Present study of 50 cases was taken for study, 98% complained of otorrhea,72% of hearing loss, other complaints included post aural abcess, aural bleed, earache, facial palsy, vertigo and tinnitus.86%had foul smelling discharge, most common pathology seen was posterior superior retraction in followed by attic retraction. Hearing loss was mostly conductive type 82%.modified radical mastoidectomy and combined approach tympanoplasty was done in 42% and40%.middle ear reconstruction was done in 80% cases. Recurrence was about 20% in children; recurrence rate was 40% more in intact canal wall. No single treatment is sufficient for aural cholestetoma, and needs follow up.

INTRODUCTION

The stratified squamous epithelium of the tympanic membrane and external ear canal can migrate prior to being shed at the entrance to the external meatus.in this way the ear canal protects itself from filling with shed keratinocytes. Under some circumstances squamous epithelium accumulates within the temporal bone. If the squamous epithelium and accumulating keratinocytes are within the middle ear space, this condition is termed "Cholesteatoma". Failure of epithelium migration and accumulation of keratin within the ear canal is not generally termed Cholesteatoma. However if there is also focal erosion of external ear canal bone in association with keratin accumulation some authorities term this 'external canal cholesteatoma.

Cholesteatoma is a progressive destructive middle ear disease.mos cases occur in children and young adults, but it can affect any age. Skin builds up in layers and erodes the bone of the middle ear and mastoid .in its early stages cholesteatoma tends to attack the ossicles, the small bones conducting sound from the eardrum to inner ear, it can erode the inner ear causing partial deafness, sometimes with unpleasant smelling discharge and pain.it can erode the inner ear causing total or permanent deafness and tinnitus.

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The inner ear also contains the balance organs. If cholesteatoma erodes into the balance organ, vertigo, a severe form of dizziness results. Can also attack the facial nerve causing facial paralysis. In rare cases disease erodes upwards. The roof of the ear is the floor of the brain. If this thin plate of bone is breached, meningitis, brain abscess and death can result. The cholesteatoma is made of layers of dead skin like onion. Only the outer layer, known as matrix, contains live growing skin cells. Cholesteatoma represents the presence of a non-neoplastic accumulation of keratinizing stratified squamous epithelium along with desquamated keratin debris in the tympanic cavity and mastoid.

Surgical therapy is the mainstay of management. The primary surgical objective is eradication of all diseased tissue with a dry safe ear. Maintenance or restoration of hearing is a secondary goal. Management of cholestetoma requires prolonged, due to the significant rate of reciligent postoperative follow up due to the significant rate of recidivistic disease. This discussion will involve pathogenesis and management of cholesteatoma. Some underlying concepts regarding establishment of the pathologic process will be mentioned, as well as anatomic considerations pertinent to effective management of the condition.

Cholesteatoma has been classified as:

1. Congenital
2. Acquired

a) Primary acquired

b) Secondary acquired

Inactive or reasonably safe

Active cholesteatoma

Quiescent form

Uncertain.

Pathology of cholesteatoma-cholesteatoma appears as roundish or oval shaped friable mass which is whitish in color, pultaceous in consistency of a variable size ranging from a size of a walnut to even measuring over 5cm in diameter.

Microscopically the lesion is made up of three components i.e. the cystic content, the matrix and the perimatrix, the cystic content is composed of fully differentiated anucleate keratin squamous along with some purulent and necrotic matter in the cavity.

The matrix of cholestetoma consists of kertatinising squamous epithelium lining a cyst like structure.it comprises the following

1) The basal layer or stratum germinatum made up of columnar epithelium composed of small cuboidal cells displaying a basophile

2) The malphigian or spinal layer composed of larger cells cylindrical in shape, which becomes polyhedral in the superficial layer.

3) The granular layer in which cells become progressively flatter and contain hyper chromatic keratohyaline granular in their cytoplasm.

4) The stratum layer hyperkeratotic and desquamating the lamella of keratin from the cystic content.

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5) The lucid layer, often not detectable.

Pathogenesis

- Pressure necrosis were widely held prior to 1927, it was thought that expanding cholesteatomas exerted sufficient pressure upon underlying bone to impede circulation of blood, herby devitalizing.
- Abramson suggested that cholesteatoma may elaborate a collagenlytic enzymes in vivo which when released in the extracellular area near bone will lead to bone destruction.
- Cytokines have been proved to play an important role in the pathogenesis of cholesteatoma. Cytokines are naturally occurring substances which act as a messenger between cells. All cytokines are low molecular weight peptides. Among the cytokines network, tumour necrosis factor alpha (TNF-alpha) is an important member.it is responsible among other functions for the osteoclast activation and bone resorption. Together with neovascularisation.
- Enzymes seem to play an important role in bone distruction associated with cholestetoma. Lysosomal enzymes, cathepesin B has been observed in cholesteatoma tissues where it exhibited collengenolytic activity.
- Leucine amino peptidase and acid phosphatase found Bretlau et al (1982) concerned with bone resorption associated with cholesteatoma.

THEORIES

THEROIES OF CONGENITAL CHOLESTEATOMA

1. The first description of congenital of congenital cholesteatoma was published by Lucaae[1], Teed in 1936 and Michaels[2], fifty years later, identified an epithelial rest in the middle ear of the temporal bone of human fetuses. This epidermoid formation was shown to regress by 33 weeks of gestations. If it persists for unknown reasons, it can form a congenital cholesteatoma.
2. Tos[3] offered in 2000 a report on the acquired inclusion theory. He concluded that the place of origin of mesotympanic cholesteatoma does not fit with the location of epithelial formation described by Michaels. Based on the places of origin of mesotympanic (congenital)cholesteatoma being commonly connected to the malleus handle, the malleus neck or the long process of the incus, Tos proposed that keratinized squamous epithelium of the retracted eardrum might be implanted into the tympanic cavity. retraction can occur due to tubal dysfunction with retraction and adhesion of the eardrum, chronic secretary obits media and acute suppurative otitis media, all of which are very common during childhood.

THEORIES OF ACQUIRED CHOLESTEATOMA

1. METAPLASIA THEORY- Von trottsch was the first to consider an epidermal origin and supposed that differentiated respiratory and bony cells were capable, under the influence of chronic inflammation, to dedifferentiate and redifferentiate themselves into squamous epithelial cells[4].
2. MIGRATION THEOREY- Hbermann in 1888[5] and Bezold[6] in1890 described frequent concave but structural intact tympanic membrane without perforation characteristic for and occlusion of the Eustachian tube. Which supports the retraction pocket theory. Michaels[2] studied the squamous epithelial layer of adult tympanic membrane and described 3zones characterized by differences in their thickness.it was concluded that cholesteatoma might develop from the most active of these zones which is situated on the parts flaccid.

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3. RETRACTION POCKET THEORY- Wittmack in 1933[6] and is most widely accepted pathogenic mechanism of acquired cholesteatoma. Pathogenesis of cholesteatoma was the patient's predisposition to develop a hyperplastic mucosa, together with strongly reduced aeration of the mastoid cavity. The starting point was squamous epithelia in pars flaccid. Wittmack concluded that without the formation of retraction, the development of cholestetoma was impossible .in contrast; he frequently observed this formation of retraction pocket limited to Prussak's space, without formation of a manifest cholesteatoma. He further postulated that the accumulation of debris in the retraction pocket alone could not be responsible for further development into genuine cholesteatoma. He observed that large cholesteatoma may spread without the presence of infection, which he called dry cholesteatoma.

Aim and objectives:

- To study different types of presentations of cholesteatoma according to site –attic or posterosupeorior retraction.
- To study different modes of managemrnt of cholesteatoma, mainly different surgical techniques.
  To study exact etiopathogenesis in operated cases.
- To study the distribution in different age groups and various socioeconomic classes.
- To study the recurrence in operated cases.

Material and methods:

50 patients attending the ENT department were taken for study. These cases of cholesteatoma were taken for clinical evaluation, investigation and surgical treatment. After discharge patients advised to come for regular follow up.

1. Consent-informed written consent is obtained from all the patients undergoing surgery.
2. Source of data-patients coming to ENT outpatient department with complaints of ear discharge, earache, hearing loss.

Inclusion criteria:

- Ear discharge which may be blood stained, foul smelling(pungent/fishy odor)
- Cases with posterosuperior quadrant and attic pathology.
- Cases with evident intracranial complications.
- Congenital cholesteatoma with intact tympanic membrane.
- Patients giving informed and written consent.

Exclusion criteria:

- All cases with central perforation
- Safe dry ear

Observations and results:

- 50 pts taken out of which 27 males 23 females
- 54% male 46% FEMALES
- Ages were dispersed over a wide range of age group 5 to 55 years with mean age of 24 years
- Median age was 22

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TABLE 1

Symptoms	Total No. of Cases	Percentage (%)
Facial palsy	2	4
Vertigo or tinnitus	2	4
Aural bleeding or aural polyp	5	10
Postaural abscess or fistula	8	16
Earache	8	16
Hearing Loss	36	72
Otorhhea	49	98

GRAPH 1: SYMPTOM DISTRIBUTION

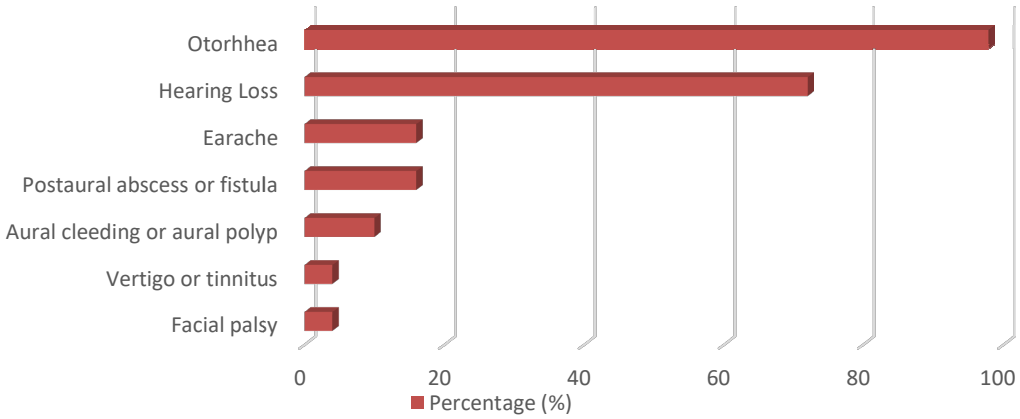

TABLE 2

	Thick and Scanty	Copious	Total
Foul Smelling	86% (42)	10% (5)	96%
Non-Foul Smelling	4% (2)	0% (0)	4%

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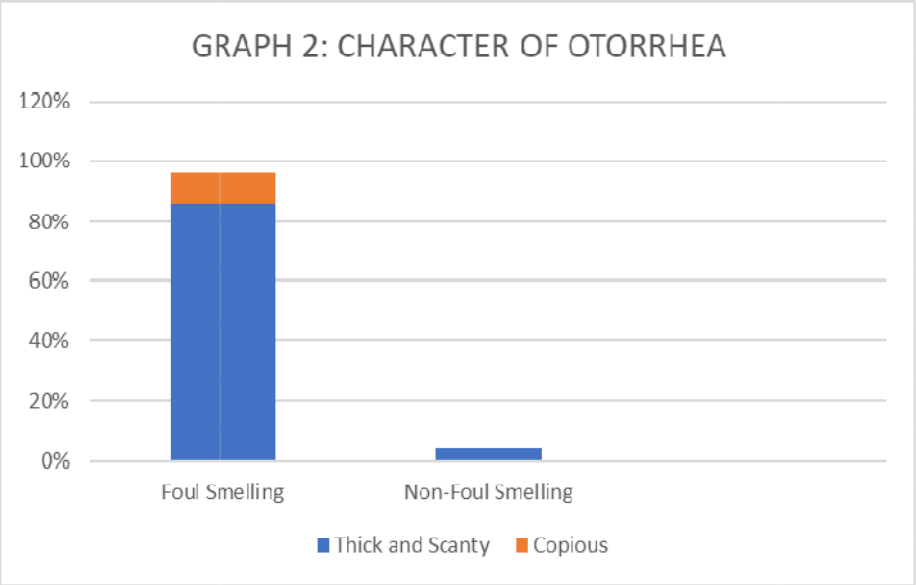

TABLE 3

Tympanic membrane findings	No. of patients	Percentage (%)
Postero superior retraction	28	56
Attic retration	20	40
Postero superior perforation (marginal)	1	2
Attic perforation	1	2
Total	50	100

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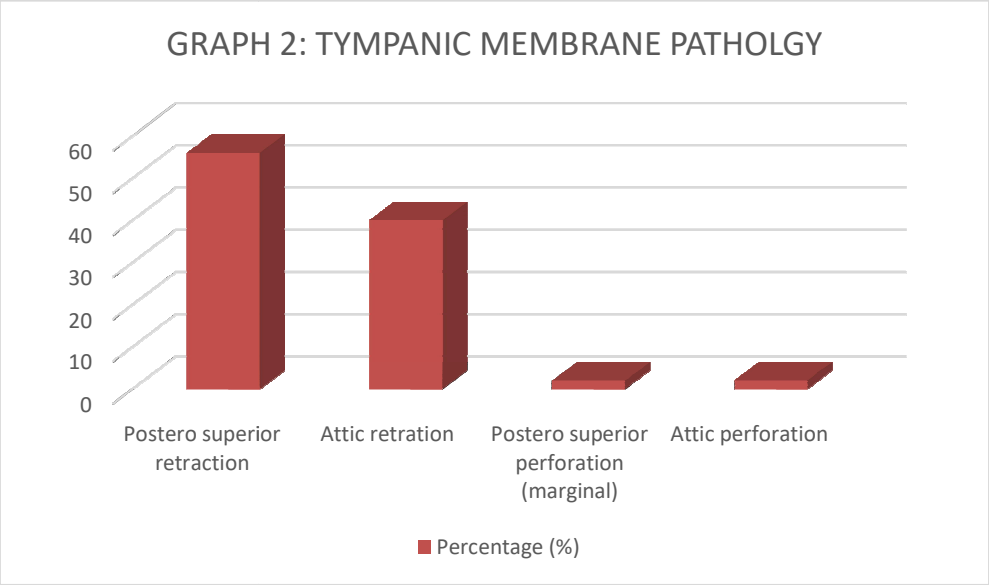

TABLE 4

Type	No. of Patients	Percentage (%)
Conductive	31	82
Mixed	5	13
Sensorineural	2	5
Total	38	100

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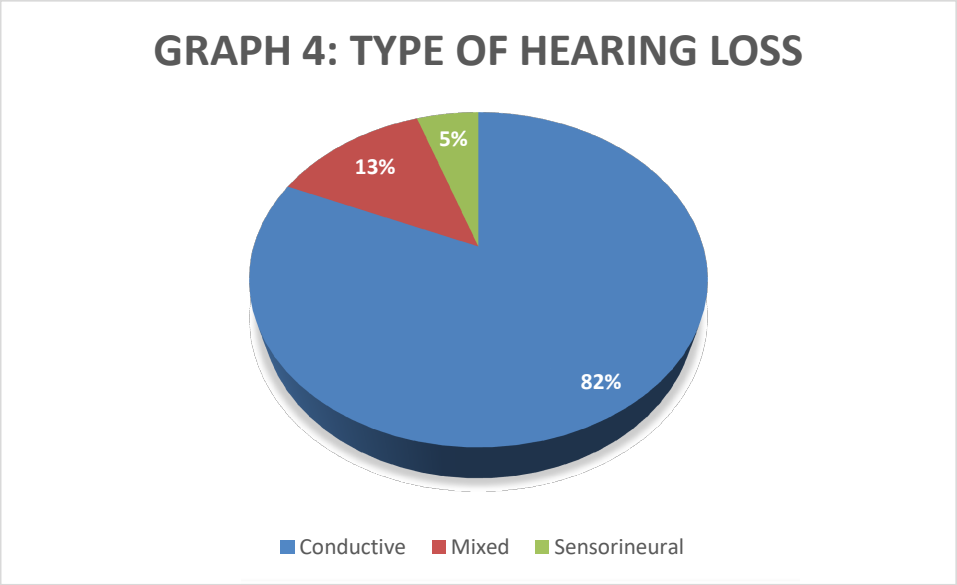

TABLE 5

Type of operation	No. of cases	Percentage (%)
Radical Mastoidectomy	4	8
Modified radical mastoidectomy	21	42
Combined approach tympanoplasty	20	40
Atticotomy	5	10

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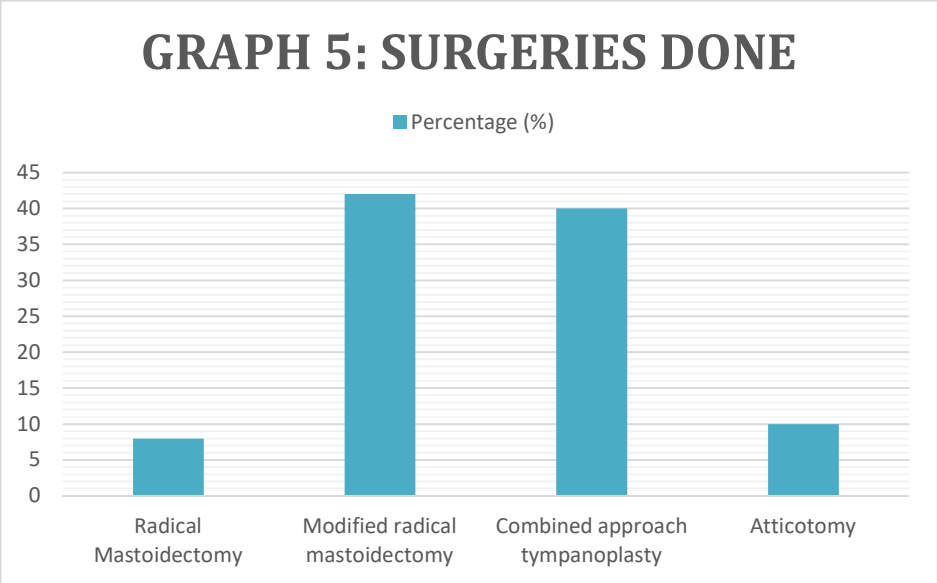

TABLE 6

Type of Surgery	Type of reconstruction	Total No. of cases	Percentage (%)
Radical Mastoidectomy	Type III tympanoplasty	1	2
	Not done	3	6
Modified radical mastoidectomy	Incus reposition	10	20
	Incus reposition Type I or III tympanoplasty	4	8
	Not Done	7	14
Combines approach tympanoplasty	Combined approach tympanoplasty (Type I or II)	20	40
Atticotomy	Type I Tympanoplasty	5	10

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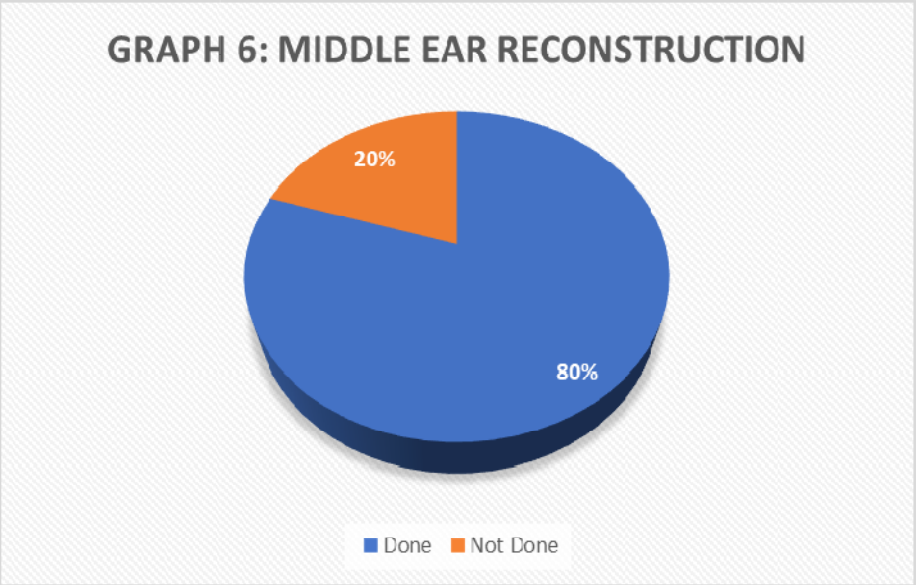

TABLE 7

	No. of Patients	No. of recurrences	% Recurrence
Adults	40	2	5
Children	10	2	20
Total	50		

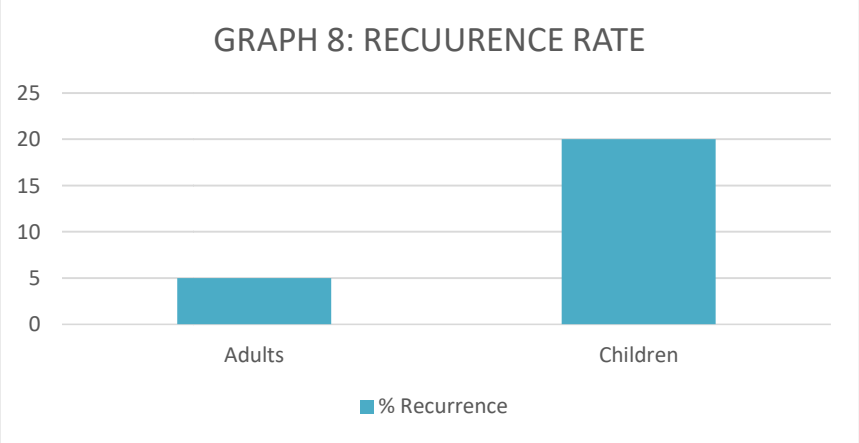

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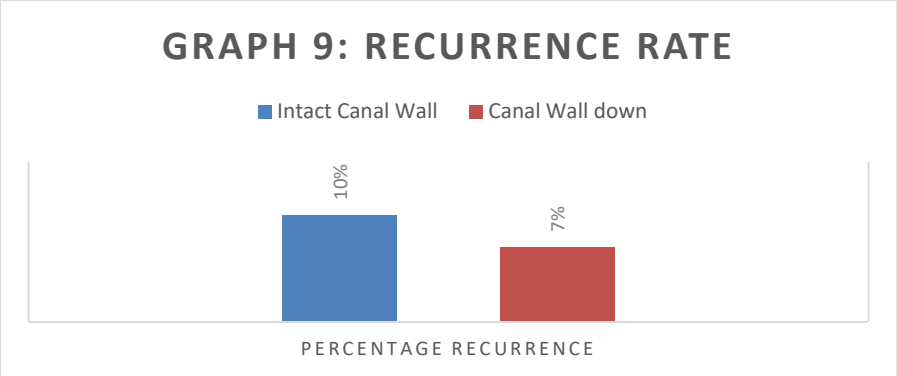
TABLE 8

	No. of Patients	No. of recurrences	Percentage Recurrence
Intact Canal Wall	20	2	10%
Canal Wall down	30	2	7%



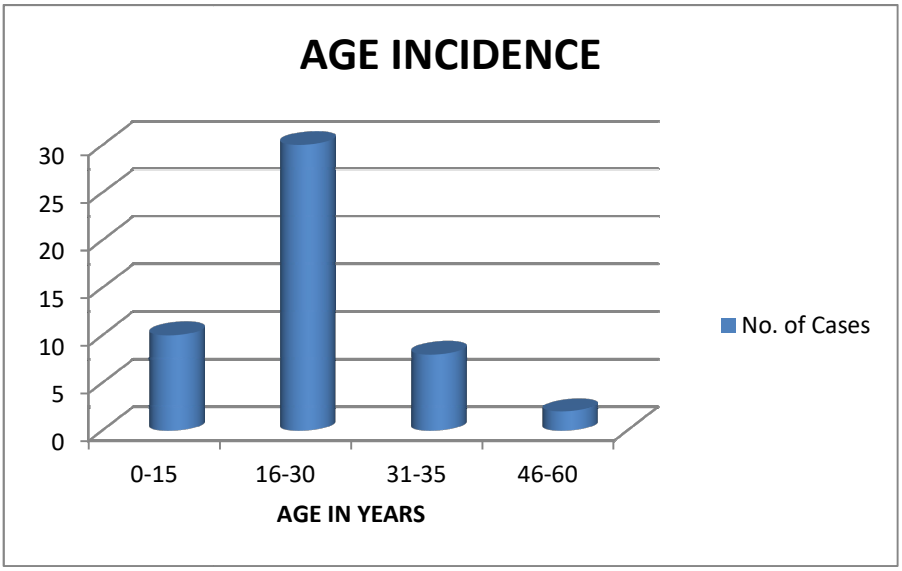


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TABLE 10

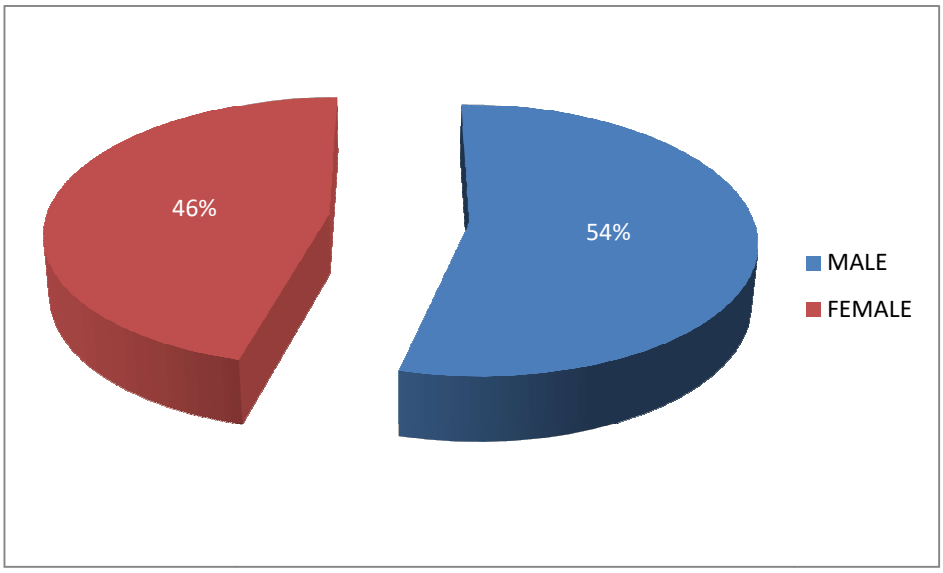

TABLE 11

Class	Number	Percentage (%)
Poor 68%	34	68
Middle 22%	11	22
Rich 10%	5	10
Total	50	100

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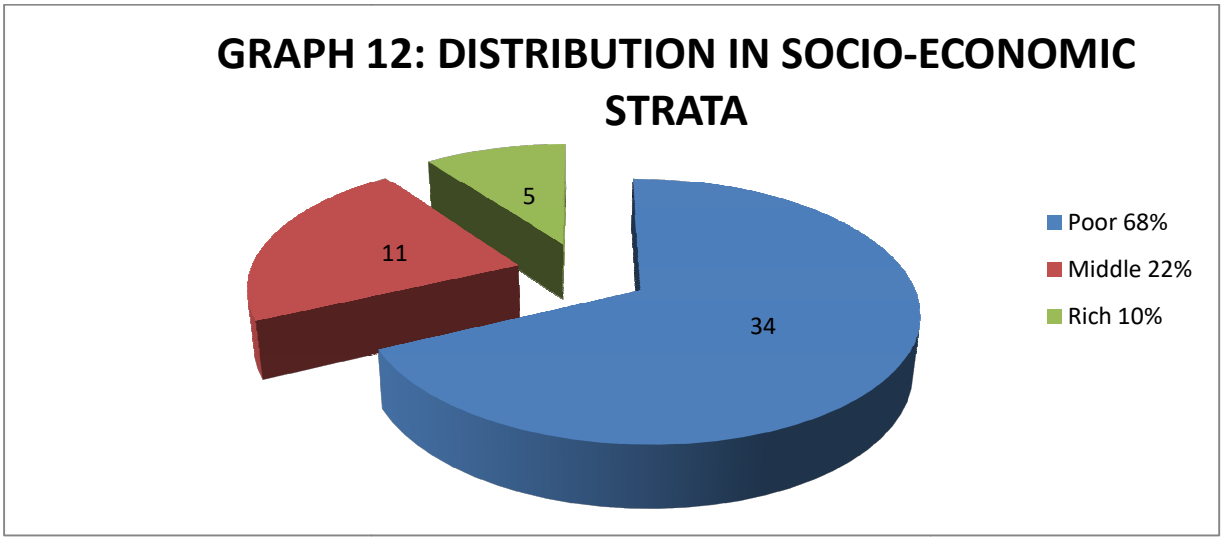

DISCUSSION

1. The diagnosis of cholesteatoma is usually made on otologic examination[7,8]. Since the signs and symptoms of ear disease may be lacking, cholesteatoma may go undetected.
2. Our study regarding the different modes of presentation of cholesteatoma revealed otorrhea as the most common presentation (98%)followed by hearing loss(72%).
3. In a similar study to identify the common presentations Garap JP and Dubey SP[9] found otorrhea to be the most common presentation in all age groups.post auricular abcesses and fistula were seen frequently. The results of our study are comparable with this study; the reason may be the poor socioeconomic background of the poor Papua New Guinean patients with majority of poor patients (60%) in our study.
4. In another study of clinical features by sheahan P.et al[10] in Ireland revealed hearing loss (78%) to be the most common presenting symptoms followed by otorrhea (60%).
5. Based on the observations of Bluestone et althea diagnosis of cholesteatoma is most effectively made with an atoscopic or, more accurately with otomicroscope[11]. Postereosuperior retraction pouch (56%)was the most commin finding followed by attic retraction(40%)in our study of 50 cases
6. Hirsch et al[12], favor the canal wall down approach for removal of cholesteatoma in children due to lower rate of recidivistic disease and a need for fewer revisions.
7. In our study of 50 cases, 60% underwent canal wall down and 40%canal wall up procedure. We adopted an individualized approach based on the extent of disease in our study. Sade J concluded that there is no single surgical treatment of choice for aural cholesteatoma.
8. Proponents of canal wall down surgery are Chang CC and Chen MK[13] who found a lower recurrence rate (3.8%) and high dry ear with this approach. Mill and Padgham[14] also reported a low residual disease (6%) and high dry ear rate (70%) with this approach in childhood cholesteatoma.

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9. In our study ,the recurrence rate (on 6months follow up) in children was found to be much higher (20%)than in adults (5%).this is comparable with the results obtained in Smyth's combined canal wall up series where the recurrence rate in children was 24% compared with 6%in adults.
10. The aggressive nature of cholesteatoma in children was reinforced by Rosenfeld et al (1992)[15] who recommended a diligent long term follow up including the necessity of a second look procedure in those children demonstrating ossicular erosion.
11. Our study also revealed a slight rate of recurrence in canal wall up procedures (10%) as compared to canal wall down procedures (7%) Austin's[16] study (1989) of 215 patients had revealed a recidivism rate of 4% for canal wall down and 39% for canal wall up procedures. The higher rate in Austin's study as compared to our study may be due to the fact that the follow up period in Austin's study was 17 years whereas in our study it was 6months.as the follow up period increases the recurrent rates will also tend to increase.
12. In our study ossicular erosion was found in 90% of patients with stapes erosion contributing 20%.Rosenfeld et al (1992) in their retrospective review of childhood cholesteatoma had found that recurrent residual cholesteatoma was associated with presence of ossicular erosion and not the approach used. Greistwood and Venables (1990) in their study found that the determining variables for the probability of residual disease were age and status of stapes. Young age and stapes erosion were strongly related to residual disease .hence we realize the importance for further follow ip for our cholesteatoma patients to detect residual recurrent disease.

CONCLUSION

1. There are important anatomic consideration in the management of cholesteatoma, and Eustachian tube function plays a vital role in its successful surgical treatment.
2. There is no single surgical treatment of choice for aural cholesteatoma .the extent of the cholesteatoma, the amount of preoperative distruction and size of the mastoid pnematization should guide the surgeon in choosing the type of operation for a particular ear. Which may range from a simple extraction of cholesteatoma (delivery)all the way to aradical mastoidectomy
3. Eradication of disease is the primary surgical goal followed by maintenance or restoration of hearing.
4. Cholrsteatoma is chronic disease with high rate of recidivism and requires diligent long term follow up.
5. Childhood cholesteatoma has an aggressive nature and has a high rate of recidivistic disease, hence the need for diligent long term follow ups.

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