

# UNSAFE CSOM WITH POST-AURICULAR ABSCESS IN PATIENTS WITH MICROTIA: OUR EXPERIENCE

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### Abstract:

Microtia is a developmental anomaly of ear due to failure of fusion of hillocks developing from 1st & 2nd pharyngeal arches. Microtia occurs once in every 6000 births. In 10% of cases, the problem is bilateral. The right side is more commomly affected than the left and males are more often affected than females.

Incidence of atresia of external auditory canal is 0.5 per 1,00,000 population, incidence of membranous atresia being 20 times less than the solid form. Congenital aural stenosis as compared to aural atresia, carries a much greater risk of cholesteatoma. Jahrsdoerfer & Cole reviewed 600 cases of major congenital ear malformation. Fifty patients (54 ears) were found to have aural stenosis. The most significant finding was that in children of 12 years or older with a meatus narrower than 2mm, 91% develop cholesteatoma.

Here, we will describe 2 cases with congenital anomaly of external ear, presenting with recurrent abscess in post aural region. Case one was having unilateral microtia with atresia of external auditory canal whereas case two was having bilateral microtia with stenosis of bilateral external auditory canal. Both of cases were operated and reconstructive procedures were done for better audiological as well as better cosmetic outcome and disease free ear.

Key words- Congenital Microtia, Ear Canal, Constriction, Pathologic, Cholesteatoma, Abscess

# Introduction:

A study by Becker & Tos<sup>1</sup> showed the incidence of atresia of external auditory canal to be 0.5 per 1,00,000 population, incidence of membranous atresia being 20 times less than the solid form.

Congenital aural stenosis as compared to aural atresia, carries a much greater risk of cholesteatoma.

Jahrsdoerfer & Cole<sup>2</sup> reviewed 600 cases of major congenital ear malformation. Fifty patients (54 ears) were found to have aural stenosis.

The most significant finding was that in children of 12 years or older with a meatus narrower than 2mm, 91% develop cholesteatoma.

Microtia is the developmental anomaly of ear due to failure of fusion of hillocks developing from 1st & 2nd pharyngeal arches. Microtia occurs once in every 6000 births. In 10% of cases, the problem is bilateral. The right side is more commomly affected than the left and males are more often affected than females.

The normal size of the auricle at birth is 66% of the length & 76% of the width of an adult ear. By the age of six, the auricle has attained 90% of adult proportion<sup>3</sup>.

# Case 1:

A 7 year old male child was brought to the out patient department with complain of right sided post auricular swelling and pain since last 2 days.

On Examination - There was a globular post-auricular swelling on the right side of size 2x2 cm2, fluctuant in nature, tender to touch with tense, inflammed and excoriated overlying skin having pus points.

Local examination further revealed: the child had right sided microtia with atresia of external auditory canal. External ear consisted of lobule, tragus and a small bulge of cartilagenous tissue behind an atretic canal.

However, lobule and tragus of the diseased ear were present alomost at the same level compared to the other side. His left ear was completely normal.



Figure 1 : Photograph at initial presentation

On further enquiry, the parents gave a history of similar post-auricular abscess of same ear, which ruptured and healed spontaneously without any treatment.

Blood investigations revealed raised wbc count (18.2x103/ml), predominantly polymorphs. Other parameters were within normal limit

Microscopic examination of the pus from post-auricular abscess showed gram positive cocci in cluster with no growth of pathogenic organism on culture. BERA: bc-abr suggestive of cochlear pathology in right ear. Ac-abr could not be done due to external ear pathology. Ac-abr of left ear suggestive of minimal hearing loss.

Pure tone audiometry (PTA) & tuning fork tests gave inconsistent results.

High resoluion CT (HRCT) temporal bone showed right sided post-auricular thickening with soft tissue & fluid, along with thickening of pinna. Eac showed fluid & granulation with circumscribed cystic lesion in middle ear. Ossicles were clear, Internal acoustic canal (IAC) was normal.

Operative findings: in order to locate the antrum of the diseased ear the mastoid tip of that side is palpated and its position is compared with that of the opposite side. Bilaterally the mastoid tip & the tragus were lying at the same level. Lobule was almost 2cm above & anterior to the mastoid tip. Eac was probed and found to be blind measuring 3mm

A curvillinear incision was given passing over the post-auricular abscess upto 1cm above the mastoid tip running behind the lobule. After initial dissection spine of henle was identified which was rudimentary. Mac ewan's triangle was not well defined. Mastoid antrum & attic were hypoplastic. Cholesteatoma was seen in antrum, sinodural angle, attic and going further anteriorly into the eustachian tube opening. Eustachian tube orifice provided an important landmark to identify the otherwise malformed middle ear. Cholesteatoma sac was found lying on the vertical segment of the facial nerve. Dura was low lying. Incus and malleus were malformed & fused together. Canaloplasty done. Meatoplasty was done by pulling and suturing the tragus more anteriorly & lobule further inferiorly. A stent was kept to maintain the patency of the EAC.



Figure 2: Post operative photograph

# Case 2:

A 17 year old female was brought to ent opd with complain of left post auricular swelling with pain and left ear discharge since last 5 days. She also complains of left sided facial asymmetry and decreased hearing from the same side since childhood.

On physical examination, the patient had bilateral microtia with a diffuse swelling over left post-auricular area extending upto the angle of mandible and lateral aspect of upper third of neck. The swelling was fluctuant and tender to touch.

On left side, there were two cartilagenous spicules in place of the lobule lying inferiorly to the meatus and an isolated tag of soft tissue almost 2 cm posterior to the eac. Tragus was rudimentary. Left external auditory canal was stenosed & filled with foul smelling whitish discharge. Bilateral eac were placed more posteriorly & at a lower level, almost 3 cm below the level of the horizontal line drawn from the lateral canthus of eye on the temporal bone.



Figure 3: Pre operative photo of case 2

Past history of 2 similar episodes were present 4 yrs & 8 yrs ago for which the patient underwent incision and drainage.

Blood investigations revealed raised wbc count (14.1x103/ml), with other parameters being within normal physiological range.

Microscopic examination of pus from the abscess showed gram positive cocci but without growth of any organism.

Audiological evaluation: Pure tone audiometry showed 55db conductive hearing loss in left ear. Bone conduction masking could not be done as the patient is having postauricular pain.

Right ear- Normal hearing, bone conduction threshold within normal limit.

Tympanometry & Otoacoustic Emission (OAE) could not be done due to active discharge.

Tuning fork test:

	Pre op	perative	
	256hz	512hz	1024hz
Rinne:	+/NP	+/NP	+/NP
Weber:	>		
ABC:		Eq/NP	

[ NP: Not perceived, Eq: Equivocal, + : Positive]

HRCT temporal bone: homogenously mildly enhancing soft tissue density seen infiltrating the left external and middle ear cavities with erosion of posterior & inferior bony wall of eac, scutum & long process of incus. It is seen completely filling epi, meso & hypo tympanum. It is also extending in prussack's space, aditus & mastoid air cells with their erosion & opacification. It is seen eroding the lateral semicircular canal with patchy dehiscence of facial canal. Evidence of heterogenously enhancing thick walled collection is noted in left posteroinferior auricular region.

Operative findings: probing revealed a stenosed eac running downward & posteriorly. An initial small incision of 2cm was given just below and almost 1.5cm posterior to the cartilagenous remnant away from the mastoid tip to drain the abscess. The incision was extended upward and anteriorly running between the soft tissue tag and the eac, upto 1.5cm above the meatus. Spine of henle was rudimentary. Drilling revealed sclerosed mastoid antrum. Granulation tissue present in aditus, attic & middle ear. Malleus & incus were malformed & fused; stapes head & stapedius tendon were visualised. Round window found to be placed anteriorly. Lateral semicircular canal was eroded. Abscess found to extend from eac inferiorly. Canaloplasty was done. The soft tissue tag was pulled & sutured superior to the meatus. Meatoplasty was maintained by keeping a stent.



Figure 4: Post operative photo of case 2

Tuning fork test:

	Post operative			
	256hz	512hz	1024hz	
Rinne:	+/+	+/+	+/+	
Weber:		>		
ABC:		Eq/Eq		
[ NP: No	t perceived, Ed	η: Equivocal, -	+ : Positive]	

# Discussion:

The auricle or pinna develops from a series of small cartilaginous tubercles or Hillocks. Hillock 1-3 comes from 1st or mandibular arch & 4-6 comes from 2nd or hyoid arch. According to park<sup>4</sup>, hillock 1 produces the anterior portion of the ear lobule, hillock 2 tragus & hillock 3 the ascending helix. Of the 2nd arch hillocks, 4 & 5 produces antihelix & helix, with 6 contributing to the posterior lobule.

By the end of fifth week, five branchial arches are discernable. In a 38 day old embryo, six hillocks have developed in the mesenchymal tissue of the first (mandibular) and second (hyoid) arch and a process of fusion produces a primitive ear in the 50 day old embryo.

Both case 1 & 2 presented with microtia, case 1 unilateral where as case 2 bilateral. [malformation, such as anotia and microtia, are likely to be caused by the disturbance of development at seven or eight weeks gestational age.] In both the cases tragus & lobule (1st arch structures) though present, 2nd arch anomaly was more pronounced.

The ear initially forms in the neck region and moves upward onto the head by week 10. In case 2 this migration was affected & bilateral ears were low set.

Case1 had atresia of right external auditory canal where as case 2 presented with stenosis of bilateral canal.

At 28 weeks, a core of ectoderm canalizes from medial to lateral and eventually breaks through to communicate with the conchal depression. Failure of canalization or more likely lack of ectodermal migration can lead to atresia of external auditory meatus and partial canalization leads to meatal stenosis (diameter of the canal less than 4mm).

In both the cases malleus & incus were malformed and fused. The head of the malleus and body and short process of the incus develops from meckel's cartilage (first arch derivative), where as, the manubrium of the malleus, long process of the incus and stapes suprastructure arise from reichert's cartilage (second arch derivative). The process begins at 4 weeks & adult shape, size & ossification is present by 25 weeks.

The full-sized outline of membranous labyrinth is formed by 25 weeks of gestation.

The scc starts to develop at 35 days. Cochlea is also formed by 25 weeks.

The organ of corti starts developing as a single block of heaped up ectodermal cells at about 11 weeks. Within this mass develop inner & outer hair cells & then specialized supporting cells.

Recent studies using high resolution computed tomography suggest a higher rate of inner ear congenital anomalies affecting between 10 and 47 percent of patient with atresia.

BERA of case 1 is suggestive of cochlear pathology in right ear.

# Conclusion:

Abscess are the presenting feature of congenital ear atresia. The age varies depending on deformity. So prevention is to perform reconstruction as soon as guidelines indicate inorder to prevent any complication. We should be alert for bony meatal, middle and inner ear abnormality during surgery. The diseased ear are the ear with altered anatomy. Hence we will have to anticipate them. As far as hearing is concerned, restoration should be done. Staged cosmetic correction is preferred.

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