

**Case Report**

## **CONGENITAL AURAL ATRESIA- OUR EXPERIENCE WITH THREE DISTINCT CASES**

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### **ABSTRACT**

Congenital aural atresia, a rare entity, may be accompanied with variations in course of facial nerve and congenital cholesteatoma. In the present study, we describe three distinct cases of aural atresia, with successful management, according to the extent of atresia. Management of atresia is essentially a team work, comprising of otologist, plastic surgeon, radiologist and paediatrician, thus making this rare anomaly a special case.

**Keywords:** *Meatal Atresia, Congenital, Cholesteatoma*

### **INTRODUCTION**

Congenital aural atresia is a birth defect that is characterized by hypoplasia of the External auditory canal, often in association with dysmorphic features of the auricle, middle ear and occasionally the inner ear structures (Schuknecht, 1989).

Aural atresia is characterized by incomplete development of the external ear with variations. The incidence is approximately 1.5 in 10,000 to 15,000 births, with a 25% bilaterality and a male predominance (Mastroiacovo *et al.*, 1995).

Atresia occurs as a result of maldevelopment of first and second branchial arches and first branchial cleft, though it may be a part of syndromes such as Treacher Collins syndrome, Crouzon's syndrome, Klippel Feil syndrome, Pierre Robin syndrome and Goldenhar syndrome (Rosen *et al.*, 2003; Jahrsdoerfer *et al.*, 1990).

In 92% of the cases, there are associated malformations of the pinna. It may vary from Grade I in which the ear is smaller in size with majority of structures present to a deficiency of pinna sub structure (Grade II) to a classic peanut deformity (Grade III) (Marx *et al.*, 1926). The tympanic bone can be present with variations that range from mild hypoplasia to complete absence. When the tympanic bone ring is absent there is a bony wall termed atretic plate that constitutes the lateral wall of the middle ear. In patients with atresia, the middle ear cavity may be smaller than normal, with ossicular deformities along with it. Congenital cholesteatoma has been reported in about 4-7% of cases of congenital external auditory canal (EAC) atresia (Jahrsdoerfer *et al.*, 1990).

Abnormalities of the facial nerve are also common in cases of aural atresia, especially the dehiscence of the fallopian canal, and abnormal course namely alterations in the second genu, which provides a more anterior position in the mastoid portion and also more lateral in its exit at the level of the stylomastoid foramen. The inner ear development is mostly normal, as it develops early in embryogenesis.

The usual presenting symptom is deformity followed by hearing loss, unilateral or bilateral. Hearing loss is conductive and ranges from 45 to 60 dB in complete atresia and from 30 to 40 dB in partial atresia.

The main aim of the otologist remains restoration of the hearing of the patient, with otoplastic procedures either at the same sitting or afterwards. Management of atresia is essentially a team work, comprising of otologist, plastic surgeon, radiologist and paediatrician, thus making this rare anomaly a special case.

### **CASES**

#### **Case Report 1**

A 20 year male patient presented with absent left external ear canal opening and hearing diminution, since childhood. Patient was operated by an otologic surgeon one year back, but had persistent

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complaints. There was no significant medical history. There were no congenital or hereditary factors involved. Examination showed grade I auricle atresia (the auricle is of a smaller size than normal, but has all the structures and these are well defined), with complete occlusion of EAC. There were no syndromic anomalies. Hearing loss was mild conductive (36.66 dB) in left ear. Right ear had normal hearing. HRCT scan showed presence of left EAC occlusion, which appeared to be non bony. The ossicles, middle ear and inner ear were normal. Under general anaesthesia, patient was operated.



**Figure 1: CT scan showing atresia with cholesteatoma debris**



**Figure 2: Thumb print concha**

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**Figure 3: Bony bar representing ossicular assembly**

A post auricular approach was used. After exposing the mastoid cortex, the soft tissue was elevated anteriorly. The spine of Henle was absent. Bony walls of EAC were traced. A probe was inserted through the rudimentary external EAC opening, with the tip pointing towards the exposed bony canal. Excess intervening soft tissue was removed, creating a neocanal followed by split skin grafting. The tympanic membrane with ossicles was seen. An adequately sized Endotracheal tube was cut and inserted to act as a stent/scaffolding in the neocanal to maintain the contour. Postoperative period was uneventful. On follow up post 6 months, patient showed improved hearing with well developed EAC.

#### **Case Report 2**

A 4 year old female patient came with complaints of decreased hearing in right ear since birth. Examination showed grade 1 auricle atresia with “thumb print” concha (figure 1). Partly developed cartilaginous EAC was present.

Hearing loss was mild conductive type (33.33 dB). HRCT showed absent bony EAC, with presence of approximately 3mm long blind pouch representing rudimentary EAC. The tympanic membrane was not commented upon by radiologist. Middle ear cavity and ossicular chain were unremarkable.

Exploration was done via postaural approach. Spine of Henle was absent. Bony EAC was absent. Mastoid process appeared larger than normal. Tracing anteriorly, a depression anterior to a cribriform like area was seen in the bone. Drilling under magnification was initiated in the region. Tympanosquamous and tympanomastoid sutures were subsequently identified and traced. Removing adequate bony atretic plate, middle ear space was identified. Ossicular assembly was present in the form of a single ossicular bar (figure 2) across the middle ear cavity which appeared mobile. Neo bony annulus was created for supporting the temporalis fascia graft which was placed over the ossicular complex and split skin graft which was placed over neocanal. Stenting was done. Patient on follow up showed well formed EAC with graft uptake.

#### **Case Report 3**

A 16 year old female patient presented with hearing loss and deformity of the ear. A rudimentary EAC with stenosis and Grade 3 auricle atresia was present on clinical examination. Hearing loss was moderate conductive type (53.33 dB) on audiometry and hearing loss was her primary concern.

HRCT scan showed cartilaginous EAC atresia with bony EAC slightly superior to the position of rudimentary external opening, with soft tissue debris probably cholesteatoma in left canal (figure 3).

Under GA, exploration was done. Incision was given posterior to the auricular tags after palpation of posterior canal wall. Dissecting anteriorly, the bony EAC was traced. There was cholesteatoma debris in the EAC which was removed. The bony EAC was found deficient inferiorly. There were two tips present

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in mastoid area. One being sharp, probably malformed and rotated styloid process and the other being normal mastoid tip. The tympanic membrane was identified normal in appearance after separation of soft tissue and elevation of flap, with identification of lateral process and handle of malleus. Neocanal thus formed was secured with Tiersch skin graft.

Postoperatively the patient had grade I facial paresis. On follow up, patient had an improved hearing with well developed EAC. The facial paresis showed recovery within 4-5 hours. Patient is planned for a second stage otoplasty procedure as per patient preference.

### **DISCUSSION**

As described by Schuknecht, congenital aural atresia is of four types. In Type A, atresia is limited to fibrocartilaginous part of EAC. Type B shows atresia of fibrocartilaginous and bony part of EAC. In type C, while EAC is totally atretic, tympanic cavity is well pneumatised. Type D is described as having total atresia with poor pneumatisation of the temporal bone and these patients are poor candidates for surgery (Rosen *et al.*, 2003).

In C type of atresia, facial nerve anomalies are more common and there can even be abnormalities in the course of internal carotid artery and jugular bulb, which can be hazardous during surgery (Schuknecht, 1989).

EAC atresia could be fibrous, osseous or mixed type and middle ear anomalies may accompany the atresia. Congenital aural stenosis predisposes to canal cholesteatoma formation. Cholesteatoma is directly proportional to the stenosis of canal. Cholesteatoma is not seen in stenoses greater than four millimeters and surgery is advisable only in cases showing stenosis less than two millimetres (Jahrsdoerfer *et al.*, 1990). Review of literature says that only bilateral cases of meatal atresia should be operated on because of the risk involving facial nerve injury and sensorineural hearing loss post surgery. But as per recent trends even unilateral cases are operated to give a better functional and cosmetic outcome.

The first ever case of atresia operated had complication of facial nerve paralysis. This represents a risk when a new canal is surgically created in the posteroinferior portion.

To have better hearing outcome, the middle ear should be well aerated (Mastroiacovo *et al.*, 1995), (Rosen *et al.*, 2003) Selection of the case for surgery is of paramount importance for good results.

For selecting the ideal candidate of surgery for congenital aural atresia, apart from detailed physical assessment there should be normal cochlear function supported by audiometric or evoked response evidence of hearing. Normal inner ear anatomy should be documented by radiometric evaluation by HRCT or MRI temporal bone (Rosen *et al.*, 2003). Jahrsdoerfer suggested a grading scale to determine success of aural atresia repair using various parameters determined from a preoperative CT scan and examination. A score of <5/10 indicates an unfavourable outcome of atresia repair (Rosen *et al.*, 2003).

Surgery for congenital aural atresia has two aspects cosmetic as well as functional. Cosmetic part of the surgery is generally the domain of plastic surgeons and it is usually delayed until the child is about 6-7 years of age.

The main aim of the otologist is to restore hearing with a patent EAC. The surgical procedure ranges from meatoplasty to canaloplasty to mastoidectomy with stapedioplasty, depending on the type of atresia present and based on clinical, audiological and radiological findings. (Schuknecht, 1989)

Congenital aural atresia surgery is among the most difficult and challenging surgery for the otologic surgeon. When performed by an experienced otologist, repair of this deformity can be accomplished safely with predictable results. The goals of this surgery are to restore serviceable hearing and to reconstruct a permanent, patent, infection-free external auditory canal; keeping in mind the probable aberrations in facial nerve course, anatomic variations and hence increase in incidence of complications. A thorough knowledge of embryogenesis and anatomy of ear with extensive preoperative workup is essential to enable successful and complication free surgery. Moreover a regular followup and packing is essential to prevent restenosis and infection. Successful achievement of these goals in the face of such an operative challenge makes atresia repair one of the most rewarding and challenging surgery for an otologist professionally.

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