

Congenital Aural Atresia - A clinical study and review of literature



Medical Science

KEYWORDS : Congenital; External Auditory Meatus; Atresia

Dr. Chirag Prafulchandra Vaghela

10, Shivkunj Society, Nr. Radhaswami Satsang, B/h, R-T-O, Ranip, Ahmedabad, Gujarat – 382480

Dr. Priyank Vinodbhai Rathod

1, Krushna park, Nr, Ambalal Park, Opp Tulsi Appartment, Karelibaug Vadodara, Gujarat – 390018

ABSTRACT

A female patient of pubertal age group presented to our institute, Smt. Shardaben General Hospital, Saraspur, Ahmedabad in June, 2007 with external aural deformity and bilateral deafness since birth. Patient was previously operated 7 years back by a plastic surgeon to rectify the external aural deformity. Patient was not relieved on otologic aspect and still had hearing problem due to Congenital Aural Atresia. Patient was operated for restoration of the hearing mechanism through tympanoplasty using temporalis fascia and for reconstruction of the external auditory canal by split thickness skin graft.

Introduction:

Congenital Aural Atresia is a term used for a rare spectrum of ear deformities present at birth that involves the variable degree of failure of development of external auditory canal (EAC). Often this deformity may involve the tympanic membrane, the ossicles and middle ear spaces to varying degrees. The inner ear development is mostly normal. The main aim of the otologist is to restore the hearing of the patient with minimal cosmetic derangement.

The incidence of congenital aural atresia is 1 in 10,000 to 1 in 20,000 live births.^{1,2} Unilateral aural atresia is three to five times more common than the bilateral variety and right side is most commonly involved in unilateral cases.¹ It occurs sporadically but its autosomal involvement is also seen. Aural atresia has been reported to occur in association with hydrocephalus, posterior cranial hypoplasia, hemifacial microsomia, cleft palate and genitourinary abnormalities. It has also been described as part of various syndromal abnormalities including Treacher-Collins, Goldenhar's, Crouzon's, Mobius', Klippel-Feil, Fanconi's, DiGeorge, VATER, CHARGE and Pierre Robin.¹

Case Report :

A 12-year-old female child presented to our hospital with features of bilaterally absent external ear canal opening and deafness since childhood. Patient was operated by plastic surgeons at the age of 5 years with a view to correct the external deformity. Patient presented to our institute with the persistent complain of deafness and absent canal opening.

On general physical examination the child was moderately built, moderately nourished. There were no signs of anemia, icterus, cyanosis, clubbing or lymphadenopathy. Vitals were within normal limits. The medical history was insignificant.

Patient had Type B atresia of external canal and was having bilateral conductive hearing loss in pure tone audiometry. Patient was accessed by the grading system proposed by Jahrsdoerfer and Type II Atresioplasty (Canaloplasty) was done through postauricular approach, epitympanic part of temporal bone was removed and Type I tympanoplasty done. Flap was created on the anterior aspect of the external canal wall and then it was covered by split skin thickness graft taken from the right thigh region.



Fig. I : Pre operative photo of patient



Fig. II : Atresia of Right Ear Canal



Fig. III : Atresia of Left Ear Canal



Fig. IV : Intraoperative Right Atresioplasty

Discussion :

Old literature supported that only bilateral cases should be operated on because of the risk involving facial nerve injury and sensorineural hearing loss post surgery.

The condition of middle ear can be mostly judged by the appearance of the external ear.^{2, 3} To reap the benefit of surgery the middle ear should be well aerated. For selecting the ideal candidate of surgery for congenital aural atresia apart from detailed physical assessment there should be normal cochlear function which should be supported by audiometric or evoked response evidence of hearing tests and normal inner ear anatomy which should be supported by radiometric evaluation.^{2,3}

In case of bilateral aural atresia early amplification is essential and bone conduction hearing aid is advised after medical and

audiological evaluations in first few months of life. The chief goal in such patients is to provide sufficient hearing so that amplification is no longer needed. Unilateral cases are delayed till the 5 to 6 years of age so that patient is compliant and aware of the surgical procedures.²

The external ear canal is developed from the first brachial cleft and initially is represented by a solid core of epithelial cells that extends medially to the area of the tympanic ring and first pharyngeal pouch. The medial portion of the external ear canal is formed by the tympanic bone. In the third embryonic month, this structure begins to ossify eventually forming the tympanic ring and osseous ear canal. The osseous ear canal continues its lateral growth during the first and second postnatal years. Malformation of the tympanic bone produces atretic bone at the level of the tympanic membrane and results in atresia of ear canal.² The ossicles except the vestibular portion of the stapes footplate (otic capsule derivative) are formed from the first and second brachial arches, hence stapes footplate is never fixed.²

Congenital aural Stenosis predisposes the canal to cholesteatoma formation. Destructive changes from cholesteatoma secondary to congenital aural stenosis begin to appear during adolescence. Cholesteatoma is directly proportional to the stenosis of canal. Cholesteatoma is not seen in stenosis greater than four millimeters and surgery is advisable in stenosis less than two millimeters.

Congenital aural atresia is classified variously on temporal bone studies, clinical evaluations, surgical findings and type of repair or a combination of these modalities. The distribution of the cases faced among general population is chiefly classified into following four types.⁴

Type A (Meatal) Atresia

This type is limited to the fibrocartilaginous part of the external auditory canal. The atretic area has an opening that is too small to permit the spontaneous egress of desquamated keratin. If untreated it can lead to cholesteatoma formation. Meatoplasty is the preferred surgery.

Type B (partial) Atresia

In this type there is narrowing and tortuosity of both fibrocartilaginous and bony part of external auditory canal. The manubrium is frequently short or curved and the malleus may be fixed to the tympanic annulus or walls of the epitympanum. A partial bony shelf may incompletely separate the middle ear into a lateral compartment that contains a deformed malleus and incus and a medial compartment that has a normal stapes and normal windows. Hearing loss is mild to severe.

Type C (Total) Atresia

This type of atresia includes all cases that have totally atretic canal but well developed pneumatized tympanic cavity. A fibrous tract or a miniscule dermal tract may exist, but more commonly, the canal is a shallow pit or is totally missing. There is a partial or complete bony atresia plate. Characteristically the tympanic membrane may be missing, but occasionally remnants of the pars propria are present. The manubrium is usually absent and when present, it is deformed and angled towards the promontory. Characteristically the head of the malleus is slightly deformed and has a fibrous fixation to the atresia plate or to the incus. Incus may be hypoplastic and the long process may fail to contact the head of stapes. Facial nerve may take anterior course.

Type D (Hypopneumatic Total) Atresia

This type has all the dysmorphic features of type C Atresia and has a reduced pneumatization of the temporal bone. Radiological features reveal abnormal course of facial nerve and abnormalities of bony labyrinth.

The feasibility of surgery on benefit analysis is made on the grading system in which 5 out of 10 is the minimum score to perform the surgery, below which there is no significant operating advantage.³

Grading system of candidacy for surgery of congenital aural atresia 3

Sr No.	Parameter	Points
1.	Stapes present	2
2.	Oval window open	1
3.	Middle ear space	1
4.	Facial Nerve	1
5.	Malleus – Incus complex	1
6.	Mastoid pneumatized	1
7.	Incus – stapes connection	1
8.	Round window	1
9.	Appearance external ear	1
Total available points		10

Operation is divided in two parts : cosmetic and functional. Cosmetic results are generally achieved by plastic surgeons and it is usually delayed until the child is about 7 years of age.

The classification of surgery is principally based on the type of mechanical system that is created to accomplish transmission of the sound and also partly on the surgical approach.⁴

Type I Atresioplasty (Meatoplasty ^{4,6})

This is used for type A atresia. Endaural incisions are made, and the skin of the posterior wall of the canal and adjacent concha is elevated as a laterally based (koerner) flap. Conchal cartilage and soft tissue are removed to create an adequate canal. Ossicles if eroded by cholesteatoma are repaired. All unepithelialized surfaces are covered by split thickness skin.

Type II Atresioplasty (Canaloplasty ^{4,6})

Canaloplasty is a term for an eclectic series of surgical procedures designed to restore patency to the canal. Although the underlying pathologic condition may vary, canaloplasty is indicated in cases in which the canal is stenotic or atretic to either 1) correct a conductive hearing loss, 2) prevent recurrent infection related to trapped cerumen and keratin debris, or 3) control a canal cholesteatoma, which is inherently destructive.⁶

This procedure is done to construct an external auditory canal and for mechanism of sound transmission through the middle ear without entering the mastoid.

Postauricular incision is kept and dissection is done till the atretic bony canal. Mastoid cortex is exposed and mandibular fossa, mastoid tip, temporal line and area of the mastoid fossa are identified.

Drill work is started at the antero superior aspect of mastoid fossa and carried along the epitympanic region. Facial nerve is taken care of and malleus and body of incus are widely exposed without causing much acoustic trauma.

A conventional meatoplasty is performed and postauricular flap is repositioned to form an external auditory meatus that is in alignment with the bony canal and held in place with appropriate sutures. Graft is placed to cover the ossicles and extend 2mm on the canal wall.

Type III Atresioplasty (Canaloplasty with strut ⁴)

This is done in discontinuity of ossicles or fixation of the malleus or incus to the epitympanic walls. Meatoplasty and canaloplasty are done alike the type I and II atresioplasty. This procedure involves removal of malleus and incus and insertion of strut to bridge the gap from the head of the stapes to the fascial graft.

Type IV Atresioplasty (Mastoidectomy with Stapedioplasty ⁴)

A wide tympanomastoidectomy is performed using postauricular approach and classical Wullstein type III tympanoplasty is done placing the tympanic graft over head of stapes. Atresia plate is removed and the posterior canal wall is taken down to the facial canal. Malleus and incus are removed and postauricular fibrous tissue is used to obliterate the mastoid cavity and

form the posterior meatal wall. Facial graft is used to bridge the middle ear space and is used for meatoplasty. Canal alignment, skin grafting and packing is done as previous types. Fenestration is kept reserved for oval window abnormalities.

Facial nerve injury, Labyrinthine injury, canal stenosis, Chronic infection and conductive hearing loss are the complications linked with Congenital Aural Atresia surgery.^{1,2} Facial nerve follows aberrant course in 25-30% of cases of aural atresia. It is typically displaced antero-laterally in comparison to normal. The bend at second genu tends to be more acute and the nerve crossed the middle ear in a medial to lateral direction so that at the level of round window, the nerve may be lateral to the middle ear space and encased in atretic bone. Five most susceptible situations for facial nerve injury are as follows 1) making the skin incision; 2) dissecting in the glenoid fossa; 3) during the

canalplasty; 4) transposing the facial nerve; and 5) dissecting soft tissue in the preauricular area. Injury to facial nerve can be avoided by improved imaging techniques and intraoperative monitoring.¹

Conclusion:

Congenital aural atresia surgery is among the most difficult and challenging surgeries for the otologic surgeon. If performed by experienced otologists, repair of this deformity can be performed safely and with predictable results. The goals of atresia surgery are to restore functional hearing, preferably without the requirement of a hearing aid, and to reconstruct a patent, infection-free external auditory canal. Successful accomplishment of these goals, in the face of such an operative challenge, can make atresia repair one of the most rewarding surgeries for the otologic surgeon.

REFERENCE

- 1 Rosen EJ, Gadre AK, Quinn FB, Ryan MW. 2003 Jan; Congenital Aural Atresia. UTMB, Department of Otolaryngology | 2 Lambert PR, Dodson EE. Congenital Malformations of the External Auditory Canal. Otolaryngologic clinics of North America 1996 Oct; 29(5):741-760 | 3 Jahrsdoerfer RA. Surgery for Congenital Aural Atresia. Surgery of the Ear; 5th Edition :389-99 | 4 Schuknecht HF. Congenital aural atresia. Laryngoscope 1989;99:908-917. | 5 Jahrsdoerfer RA, Cole RR. The Risk of Cholesteatoma in Congenital Aural Stenosis. Laryngoscope 1990 Jun; 100(6):576-8 | 6 Parisier SC, Levenson MJ, Hanson MB. Canalplasty. Otolaryngologic clinics of North America 1996 Oct; 29(5):867-886