Our Experience of Treating Wide Spectrum of External Ear Canal Atresia of Different Etiologies in Pediatric Patients

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Received: 8 March 2017 / Accepted: 9 May 2017 / Published online: 11 May 2017
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Abstract External auditory canal atresia (EACA) is a common otologic condition. Etiology can vary from congenital to acquired causes. It causes considerable difficulty to the patient. Bilateral ear canal atresia in children can lead to speech delays due to hearing impairment caused by this condition. Though easily diagnosed it is one of the most difficult conditions to treat. Acquired conditions can affect any age group. Restenosis following treatment is very common. This article focuses on the treatment of EACA due to different etiologies and emphasizes on special points of surgical treatment and follow up. Five cases of external auditory canal atresia was treated between 2014 and 2016. Two of them were congenital cases and three were acquired. One congenital atresia patient had pinna abnormalities in the form of one sided anotia and other side microtia. Another patient of congenital ear canal atresia had congenital cholesteatoma. Acquired atresia was due to osteoma, external trauma and surgical trauma following a condylectomy surgery. All the patients were treated surgically. A wide meatoplasty with split thickness skin graft lining the canal/cavity was done to avoid restenosis. Merocel wicks were used in all cases. Removal of localized granulations on follow up helped keep the canal patent especially in congenital EACA. All patients had significant hearing improvement following surgery. Child with bilateral atresia and pinna anomaly has now achieved normal hearing and speech milestones. She does not use any hearing aid. None of the patients developed restenosis of their reconstructed ear canal. One of the patient developed

granulations around the meatoplasty edges which was cauterized using silver nitrate. External auditory canal atresia surgery is a difficult surgery keeping in view the distorted anatomy and the propensity of restenosis of the newly constructed ear canal. Hearing restoration in the operated ear is all the more challenging. Meticulous planning with close discussion with patients and their caregivers regarding multiple surgeries has to be done. Using skin graft, doing a wide meatoplasty, using merocel wicks for ear canal dressing are few important aspects of this surgery which can give satisfactory results in long term.

Keywords Congenital external canal aural atresia · Acquired external canal aural atresia · Transmastoid approach · Canalplasty · Meatoplasty · Merocel ear wicks

Introduction

External auditory canal atresia both congenital and acquired is disabling to the patients. External auditory canal atresia is a rare condition commonly found in pediatric age groups. Acquired stenosis of the EAC has no age predilection and can affect all age groups. Treatment of external canal atresia is often challenging with varied results. Congenital EAC atresia is commonly associated with deformities of pinna and conductive hearing loss. Sensorineural hearing loss has also been reported [2]. Acquired EAC atresia often results in conductive hearing loss. Treatment of EAC atresia is challenging keeping in view the target of giving patients a hearing ear and to avoid restenosis of the ear canal. EACA was first described in Western medicine by Paulus of Aegina in the seventh century AD. Paulus recommended simple incision for

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opening the atresia. Later, physicians used a hot iron probe to maintain canal patency after incising the atresia. The first operation to correct an atresia of the external auditory canal (EAC) was performed in 1882 by Schwager [1]. In 1914, Page reported on a series of eight patients who underwent operation. Of these eight patients, five had a subjective improvement in their hearing after surgery. In the early twentieth century, techniques for correction of the atretic external canal have involved opening the antrum and aditus of the ear and lining the cavities with a skin graft. In these surgeries, the atretic bony plate was untouched, and the resulting hearing improvement was poor. In recent years improved radiologic and audiologic assessments along with improvements in surgical technology, such as the operating microscope and facial nerve monitoring, success rate after surgery has increased. We are describing four cases of external canal atresia due to four varied etiologies which were treated surgically in. All the patients had subjective and objective improvement in hearing. Age group of patients ranged from 4 to 35 years.

Case 1, 2 (Congenital Atresia)

A 4 year old girl came to us with complains of hearing loss and impairment of speech. She had left sided anotia and right sided microtia. Complete external ear canal atresia was present on both the sides. A persistent discharging sinus was present on the side of anotia just at the site of absent pinna. This was indicative of a preauricular sinus on the side of anotia. A final diagnosis of bilateral external auditory canal atresia with right sided microtia and left sided anotia with a “*preauricular*” sinus was made. Pure

tone audiogram could not be performed in this patient. Hearing evaluation was done using behavioral audiometry and brainstem evoked response audiometry (BERA). It revealed right sided moderate hearing loss and left sided moderate to severe hearing loss. HRCT revealed bilateral external ear canal atresia with. There was a bony atretic plate occluding the ear canal on the right side. Medially there was some soft tissue density indicative of a fibrous plug. Middle ear and mastoid were filled with soft tissue shadow with mild erosion of scutum and mastoid air cells. Ossicles on this side were malformed but oval window could be seen. Inner ear structures were normal. On the left side there was complete EAC atresia with bony atretic plate and the middle ear was grossly malformed. Inner ear was normal. Oval window was not clearly appreciated. Soft tissue shadow was filling the mastoid air cells. Radiological diagnosis of both sided congenital cholesteatoma was made keeping in view the soft tissue shadow and bony erosion of scutum (Fig. 1). MRI to confirm the presence of cholesteatoma could not be done due to high cost and unaffordability of the patient. Considering the radiological diagnosis of congenital cholesteatoma on the right side, mastoid exploration was planned. Surgery for excision of preauricular sinus on the left side was planned in the same sitting. The purpose of operation was two pronged. One was to ensure removal of cholesteatoma to make the ear safe and other was to restore some hearing on that side. Any form of amplification using osseointegrated temporal bone implants (BAHA) or bone conduction hearing aids was not possible due to cost factor. A transmastoid approach was planned to remove cholesteatoma and construct an external auditory canal on the right side. Skin markings were made using mastoid tip and the post

Fig. 1 CT scan temporal bone showing soft tissue shadow in the mastoid on both sides

auricular groove behind the pinna as a surgical landmark. Post auricular incision 1 cm posterior to the groove was given and flaps raised. The site of external auditory canal was represented by a small bony pit. Drilling was begun just posterior to this landmark. Antrum was opened. After having delineated the antrum, the bony atretic plate was drilled away just anterior to the antrum. A bony ear canal could be established which had a fibrotic plug medially. The fibrotic plug was carefully removed. A shiny whitish membrane like structure could be seen medially. No landmarks of the tympanic membrane could be visualized. Mastoid air cells was filled with congenital mesenchymal tissue. There were no cholesteatoma. Fused malleus and incus was present and a malformed stapes was seen. It was a mobile assembly. Facial nerve was lying outside its bony canal in its horizontal segment. Facial ridge (bone over the vertical segment) was intact and it was adequately lowered (Fig. 2). Temporalis fascia graft was placed over the mobile assembly of ossicles. It was then layered with split thickness skin graft. A wide meatoplasty was done and skin graft was stitched to its edges. Pre auricular sinus was excised completely on the opposite side. On progressive follow up the child developed some granulations at the junction of meatoplasty and skin graft. It was cauterized using chemical cautery with silver nitrate. A well epithelialized dry mastoid cavity and a patent external auditory canal could be achieved. In the third year of follow up the child is having near normal hearing on the right side and has achieved speech milestones as per her age.

Another case of unilateral ear canal atresia presented to our OPD with complains of foul smelling ear discharge from a small opening in the ear canal. The ear canal had only a pin hole opening since birth. He had moderately

Fig. 2 Transmastoid approach to treat external ear canal atresia. Figure shows a widely drilled mastoid cavity with deformed mobile ossicular assembly and a lowered canal

Fig. 3 Bony atretic plate of congenital aural atresia

severe conductive hearing loss on that side. HRCT temporal bones revealed soft tissue shadow filling whole of the ear canal, mastoid and middle ear with erosion of ossicles. Inner ear structures were normal. The stenotic external auditory canal was opened using multiple release incisions in the skin. External auditory canal was occluded with bony atretic plate with a very small opening on anterior aspect (Fig. 3). This bony plate was drilled. The EAC was seen to be filled with cholesteatoma. It was eroding the posterior meatal wall. Cholesteatoma was completely filling the mastoid cavity and middle ear. This was a case of congenital cholesteatoma which was causing extensive erosion of nearby structures. All the ossicles along with the stapes suprastructure was absent. A canal wall down mastoidectomy was done. Temporalis fascia graft was used to line the mastoid cavity. Wide meatoplasty was done. Patient now has a dry ear and mild conductive hearing loss. Period of follow up is one year. A second stage ossiculoplasty is planned for this patient to give adequate hearing benefits.

Case 3: (Acquired Atresia)

Another male child had presented to us with progressive hearing loss on the left side. He complained of progressive occlusion of his ear canal by a hard bony mass. Examination revealed a bony hard mass occluding the ear canal completely. It was lined by skin. HRCT temporal bones revealed a bony osteoma of the bony part of EAC which was completely occluding the ear canal. Middle and inner ear structures were normal. Pure tone audiogram revealed a moderate conductive hearing loss on that side. Surgical removal of the osteoma was planned. Post aural incision was given. The region of external auditory canal was completely occluded by compact hard bone. Drilling was

Fig. 4 Drilling of bony osteoma and making the ear canal patent

Fig. 5 Recanalization has been achieved following drilling of osteoma and a canalplasty. The tympanic membrane was perforated

commenced at this site. The bony osteoma was occluding the lateral bony ear canal. The bony osteoma was slowly drilled away (Fig. 4). A patent bony ear canal could be established. Medially the tympanic membrane was found to be perforated. Middle ear structures were normal (Fig. 5). Tympanic membrane was grafted using temporalis fascia and the raw bony ear canal was lined using skin graft. Along with bony canal plasty, meatoplasty was also performed to maintain the patency of the EAC and to avoid restenosis. Hearing was restored to normal level on that side. Three years since the surgery the patient is asymptomatic and hearing normally on the operated side.

Case 4, 5: (Iatrogenic Atresia: Secondary to Surgery and Trauma)

A girl aged 16 years had been operated for bilateral TM joint ankylosis in the Department of Maxillofacial surgery at our institute. After the surgery patient developed

Fig. 6 Cruciate release incision over the stenotic skin in a case of traumatic ear canal atresia

external ear canal stenosis on the left side. She also had conductive hearing loss. Incision for TM joint release was placed in the pre auricular region. During surgery there might have been some trauma to the external auditory canal skin following which there was fibrous stenosis of the ear canal. We approached this iatrogenic atresia from post aural route. Fibrous scar tissue was removed and canal patency was restored. Wide meatoplasty was done which was lined by skin graft. A patent canal with normal hearing was attained. Mouth opening which was achieved after TM joint ankylosis release was maintained.

Another case of external ear canal atresia secondary to trauma to ear after a road traffic accident had come to us with moderate conductive hearing loss. Only a small pin hole opening was present in the canal. HRCT temporal bones revealed a near normal bony ear canal, middle ear and inner ear. During surgery only a thin soft tissue/skin tag occluding the cartilaginous portion of ear canal was encountered. Release incision was placed around the pin hole opening and the skin tag occluding the canal was excised (Fig. 6). Part of conchal cartilage was also excised to do a wide meatoplasty. This was done to keep the EAC patent in the postoperative period.

Discussion

External auditory canal (EAC) atresia is not a common entity. Congenital atresia of the EAC is caused by a failure of canalization of the epithelial plug portion of the first branchial cleft. Persistence of the tympanic ring results in a bony atresia plate at the level of the tympanic membrane. Ossicular malformations may be seen as they arise from the first branchial cartilage (i.e., Meckel cartilage). Failure of the EAC to canalize results in conductive hearing loss. Concomitant ossicular malformations may result in additional conductive hearing loss. In addition, 11–47% of

patients also have a sensorineural hearing loss in the affected ear [2]. EACA may be seen with microtia. The auricle develops from ectodermal condensations and mesodermal condensations at 5 weeks' gestation. Often the mastoid system is filled with congenital mesenchymal tissue. In rare cases congenital cholesteatoma may be present.

Acquired stenosis or atresia is an uncommon disorder of the external auditory canal. Its incidence has been estimated at 0.6 cases per 100,000 [3]. It has different names in literature such as post-inflammatory medial meatal fibrosis [4, 5], post-inflammatory medial canal fibrosis [6], post-inflammatory acquired atresia [7], canal atresia [8], and stenosis of the external auditory canal [9]. Trauma is another possible cause of EAC atresia. Iatrogenic trauma from prior otologic surgery is a common inciting event [10], but direct trauma to EAC is a rare cause of atresia. Our series had patients having external canal stenosis due to etiologies ranging from congenital stenosis with pinna anomalies, congenital stenosis associated with cholesteatoma, acquired stenosis due to external trauma and surgical trauma and acquired stenosis due to osteoma of the canal. All these cases were managed using different surgical approaches and techniques.

Congenital external auditory canal atresia (EACA) requires a team approach and very close discussion with parents. Bilateral atresia usually requires surgery. Initial intervention is ideal at 4–5 years of age. Our patient with pinna anomaly and EACA was 4 years of age and decision to operate her was taken following a radiological diagnosis of congenital cholesteatoma. We also expected to restore her hearing so that her speech and learning milestones could be restored. Any form of amplification using osseointegrated implants was not possible due to cost factor. Congenital atresia have been treated traditionally by transmeatal route wherein drilling starts at the site of meatus just posterior to the glenoid fossa. A research group from Japan detailed their modified transmastoid approach. [19] They advocate visualization of the antrum, sinodural angle, and digastric ridge via the canal-open method, avoiding facial nerve injury, followed by preparation of a relatively large external ear canal with reconstruction of the posterior wall of the external ear canal. Our case was being operated for congenital cholesteatoma and hence we opted for the transmastoid approach. During surgery there was no cholesteatoma but only congenital mesenchymal tissue. The radiological diagnosis hence was not correct. For confirmation an MRI would be feasible. This again was not done in our case due to cost factor. Transmastoid approach for this patient was advantageous for minimizing postoperative complications of restenosis of the new ear canal and postoperative middle ear infection. It also gave us direct visibility of vital structures like the facial nerves and the ossicular assembly. On follow up her hearing on that side

improved considerably. The average PTA threshold on the operated side was 35 dB. At third year of follow up she has achieved near normal speech as per her age.

Another case of congenital EACA and cholesteatoma was operated via the transmastoid approach. Here the stenosis was fibrotic and cholesteatoma had completely eroded the posterior bony ear canal. The horizontal segment of facial nerve was eroded and no ossicles were present. Only footplate was intact. Modified radical mastoidectomy was performed. Wide meatoplasty was done to avoid any restenosis. This approach is a feasible approach to treat congenital external auditory canal atresia. Regular debridement of the cavity in both the cases avoided any restenosis of ear canal. In Battelino et al. [20] described a small case series of patients who had success in maintaining patency of the EACA repair with topical application of mitomycin-C. They reported success with both congenital and secondary fibrotic atresias of the EAC.

Acquired atresia due to osteoma is a very slowly progressive disease. Osteoma was arising from the inferior bony ear canal and had occluded complete canal. HRCT temporal bones revealed the presence of bony osteoma in the lateral aspect of the bony canal. Medially the ear canal, middle ear and inner ear ears were normal. This case was done through the transmeatal route. Osteoma was drilled and patent canal was established. Temporalis fascia graft and skin graft was used to line the canal and meatoplasty was done. This avoided any restenosis of the canal. Closure of air bone gap of 30 dB was achieved in the operated ear.

One of our patients developed ear canal stenosis secondary to trauma to ear canal following road traffic accident. This is a rare cause of EACA. Stenosis in these cases is usually fibrotic with normal bony canal. Stenosis was merely due to scar tissue formation in the ear canal. Scar tissue was released during which ear canal skin was partially sacrificed. A wide meatoplasty was done and split thickness skin graft was used to line the ear canal. Skin graft was stitched to meatoplasty edges. Merocel wicks were used to give adequate pressure to keep the graft in place. Merocel wicks improve healing rate. The House Ear Institute published its results in 2004 [11]. The new modifications included the use of the Argon laser, thinner split-thickness skin grafts, Silastic sheets, and Merocel wicks in the external auditory canal. With these modifications, hearing results were closure of the air-bone gap to 30 dB or less in 63% of patients, with a long-term postoperative air-bone gap of 30 dB or less in 50% of patients. Ossicular chain refixation occurred in only 4% of these patients. Doing meatoplasty and canal plasty avoid restenosis [11].

Another patient whom we have discussed was operated for temporomandibular joint ankylosis. Incision was placed in the preauricular region. Inadvertent entry in the ear canal had led to healing by scar tissue formation and eventual

complete stenosis of the ear canal. Conductive deafness was present on pure tone audiogram. Here also the fibrotic atretic plug was removed and lateral bony canal was lined using skin graft which was then sutured to edges of meatoplasty. Normal hearing levels were achieved in this patient.

Acquired ear canal stenosis is usually due to scarring from posterior canal skin. According to McDonald et al. [9], the essential step in canalplasty involves the generous widening of the posterior bony canal wall until mastoid cells are just encountered; the widening should be accomplished without creating any recesses that may later prevent proper cleaning of the ear canal. Cremers and Smeets [12] prefer to widen the auditory canal as much as possible without opening the mastoid, to increase the chance of achieving a sufficiently wide and dry auditory canal. They also claim that the amount of exposure that is achieved using the endaural approach is less than that achieved with the retroaural approach. The transmastoid approach is considered to be a safer route regarding the facial nerve, especially in trauma patients. McDonald et al. [9] emphasize the importance of using two pieces of split-thickness skin graft to cover the new ear canal and the de-epithelialized tympanic membrane, one being attached to the anterior margin, the other to the posterior margin of the new meatus. Adkins [13] covered the skin-deficient canal with a transposition flap in eight cases with no recurrence. Soliman et al. [14] removed a wedge of skin from the meatal floor and achieved a good result in 13 of 16 patients. Moore et al. [15] lined the canal with a full-thickness skin graft in one case with no recurrence. McCary et al. [16] used split-thickness grafts in 18 cases with one recurrence. Bell [17] used bilateral rotational skin flaps in nine cases with no recurrence.

Stucker and Shaw [18] advocated use of posteriorly based skin flap to reduce the chance of restenosis.

Main challenge in treating external ear canal atresia (EACA) of both congenital and acquired variety is its propensity towards restenosis. Restenosis is mainly due to granulation and secondary healing. A widely done canalplasty and meatoplasty along with careful placement of skin graft avoids granulation tissue formation and further scarring. This is essential to avoid restenosis.

All the reported cases are in continuous follow up and restenosis has not been operated. Hearing has improved in all the patients.

Conclusion

Surgical management of external canal atresia is a challenging surgery. Meticulous planning is required to achieve desired results. Long term follow up is essential and close

cooperation is required from both the surgeon and the patient's side. Our series emphasizes treatment of different varieties of external auditory canal atresia using different surgical approaches. We achieved good results following our surgery due to good planning and regular follow up. Wide canalplasty and meatoplasty are essential for good results. Merocele wicks used for ear packing not only helps to keep the skin graft in place but also accelerates better healing. These steps also avoids restenosis of the canal. Transmastoid approach is a good approach for congenital aural atresia. It provides better delineation of anatomical landmarks and helps make a wide canal which is not prone to restenosis.

Compliance with Ethical Standards

Conflict of interest The authors declare that they have no conflict of interests.

Ethical Approval All procedures performed in this series involving human participants were in accordance with the ethical standards of the institutional ethical committee.

Informed Consent Informed consent was obtained from all individual patients.

References

- Schwager K (2007) Reconstruction of middle ear malformations. *GMS Curr Top Otorhinolaryngol Head Neck Surg* 6:Doc01
- Shah RK, Shah UK (2015) External auditory canal atresia: background, pathophysiology, etiology. <http://emedicine.medscape.com/article/993857-overview#a5>. Nov 2015
- Becker BC, Tos M (1998) Postinflammatory acquired atresia of the external auditory canal: treatment and results of surgery over 27 years. *Laryngoscope* 108:903–907
- Katzke D, Pohl DV (1982) Postinflammatory medial meatal fibrosis. A neglected entity? *Arch Otolaryngol* 108:779–780
- Magliulo G, Ronzoni R, Cristofari P (1996) Medial meatal fibrosis: current approach. *J Laryngol Otol* 110:417–420
- el-Sayed Y (1998) Acquired medial canal fibrosis. *J Laryngol Otol* 112:45–49
- Bonding P, Tos M (1975) Postinflammatory acquired atresia of the external auditory canal. *Acta Otolaryngol* 79:115–123
- McKenna KX, Chole RA (1992) Traumatic external auditory canal atresia. *Am J Otol* 13:80–81
- McDonald TJ, Facer GW, Clark JL (1986) Surgical treatment of stenosis of the external auditory canal. *Laryngoscope* 96:830–833
- Lupin AJ (1976) External auditory canal stenosis. *Arch Otolaryngol* 102:458–460
- Teufert KB, De la Cruz A (2004) Advances in congenital aural atresia surgery: effects on outcome. *Otolaryngol Head Neck Surg* 131(3):263–270 (**Medline**)
- Cremers WR, Smeets JH (1993) Acquired atresia of the external auditory canal. Surgical treatment and results. *Arch Otolaryngol Head Neck Surg* 119:162–164
- Adkins WY, Osguthorpe JD (1981) Management of canal stenosis with a transposition flap. *Laryngoscope* 91:1267–1269
- Soliman T, Fatt-Hi A, Abdel Kadir M (1980) A simplified technique for the management of acquired stenosis of the external auditory canal. *J Laryngol Otol* 94:549–552

15. Moore GF, Moore IJ, Yonkers AJ, Nissen AJ (1984) Use of full thickness skin grafts in canalplasty. *Laryngoscope* 94:1117–1118
16. McCary WS, Kryzer TC, Lambert PR (1995) Application of split-thickness skin grafts for acquired diseases of the external auditory canal. *Am J Otol* 16:801–805
17. Bell DR (1988) External auditory canal stenosis and atresia: dual flap surgery. *J Otolaryngol* 17:19–21
18. Stucker FJ, Shaw GY (1991) Revision meatoplasty: management emphasizing de-epithelialized postauricular flaps. *Otolaryngol Head Neck Surg* 105:433–439
19. Yamane H, Sunami K, Takayama M et al (2004) Application of modified transmastoid approach methods to congenital atresia of the external ear canal with middle ear infection. *Acta Otolaryngol* 124:38–44 (**Medline**)
20. Battelino S, Hocevar-Boltezar I, Zargi M (2005) Intraoperative use of mitomycin C in fibrous atresia of the external auditory canal. *Ear Nose Throat J* 84(12):776–779 (**Medline**)