MANAGEMENT OF CONGENITAL ATRESIA EAR

Lt Col SB MAHAJAN*, Air Cmde LK KOCHAR^{vsm,†},
AK GUPTA[#]

Abstract

A total of 3 out of 5 cases of major congenital ear malformation seen in out patient department in two years in 1997 and 1998 were operated for functional reconstruction of external auditory canal, middle ear and ossicular chain. 2 cases of bilateral malformation of auricle grade 2 were advised otoplastic repair prior to tympanoplasty. Out of the 3 cases operated by anterior transatretic bone approach, 2 cases had good hearing improvement in air conduction threshold of up to 30 db hearing level (averages of 0.5, 1.2 and 3 khz). One case developed external canal stenosis by six months post operatively and drop in hearing. There was no facial nerve injury in any of the cases. This paper analyses the importance of facial nerve delineation preoperatively based on its embryological development and its relationship with temporal bone and membranous labyrinth. It also focuses on the various parameters like timing of surgery, importance of (CT) scanning, use of facial nerve monitor and surgical expertise and experience which govern the surgical intervention and its results in congenital ear atresia.

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Introduction

Congenital atresia of the ear, a relatively rare entity has an incidence varying from 1 in 8000 births with not more than 30% of these bilateral, to 1 in 10,000 to 1 in 20,000 live births with 11% to 47% associated inner ear abnormalities [1,2]. Congenital malformation of the ear has both aesthetic and functional implications depending upon the severity, extent and bilaterality of the disability. The complexity of the disease requires a systematic approach of management, right timing of initial evaluation and recommendation for surgical correction of the ear malformation.

Congenital malformations are amenable to surgery for functional reconstruction of the conductive pathway of sound from external ear to inner ear. The aim of surgery is to create a patent skin lined external auditory canal and to achieve closure of air bone gap to 20 to 30 db. The much dreaded hazards of surgery like injury to facial nerve, membranous labyrinth, post operative canal stenosis and infections have been obviated by regular use of high resolution computerised tomography in preoperative evaluation [3,4], recent introduction of facial nerve monitoring during surgery [5] and better understanding of surgical application of embryological anatomy of 7th nerve and related structures of middle ear.

To prevent injury to facial nerve it is important to assess and delineate the nerve route pre-operatively

which is very likely to be abnormal. Sataloff [6] has rationalized through his studies that if the 7th nerve were abnormal in its path, the arrest of nerve development in the temporal bone would have generally occurred at the same time as developmental arrest of the other structures in a malformed ear. Visual examination and high resolution radio imaging would help in pin pointing the developmental arrest in gestational growth of fetus and thus predict and correlate it to development and localization of facial nerve route. Thorough knowledge of temporal bone anatomy and embryological development supplemented by radio imaging and surgical expertise will go a long way in achieving desired long term results in surgery of congenital atresia.

Material & Methods

A total of 5 cases of congenital atresia were seen in 2 years from 1997 to 1998 in ENT OPD. 2 cases had bilateral congenital malformation with microtia grade 2 as per Jahrsdoerfer classification [7]. Both cases were below 5 years of age. These cases were advised to use hearing aid and referred to plastic surgeon for auriplasty prior to atresia repair. 3 cases aged between 9 and 18 years had unilateral atresia grade 1. They were followed up for 1 to 2 years prior to surgery. The brief description of 3 cases operated is as follows :

Case No. 1

10 year old male child presented with a small, low and forward placed left pinna of normal shape. External auditory canal was fully absent with poorly developed mastoid. Right ear was normal in shape, size and location. Tuning fork test revealed absent air conduction with normal bone conduction in left ear. Pure tone audiometry showed no response on air conduction (AC) but bone

*Classified Specialist (ENT), 150 GH C/o 56 APO, †Consultant (ENT), Army Hospital (R&R), New Delhi, #Medical Suptd, Post Graduate Institute of Medical Education and Research, Chandigarh

Fig. 1: CT scan of left ear showing complete bony atresia with thick atresia plate, body of Malleus and Incus fused, crura of Stapes well formed, middle ear space moderately formed with partially pneumatized mastoid, Cochlear coils and semi circular canals well delineated.

conduction (BC) was at 20 db in left ear. Right ear had AC and BC at 15 db and 10 db, respectively. CT Scan showed complete absence of external canal. The atretic plate was present and adherent with fused mass of malleus and incus. Middle ear space was present but smaller than opposite side. Vestibule and semicircular canals were well developed. Facial nerve was not discernible.

The likely time of developmental arrest was analyzed at around early 8th week due to (a) the failure of resorption of epithelial core to form external canal suggesting malformation earlier than 21 weeks gestation (b) the appearance of auricle indicating the arrest at beginning of 8th week (c) the small and partial middle ear space with fused mass of malleus and incus suggesting the arrest late in 7th or early 8th week. Accordingly, the facial nerve location in its horizontal position was predicted to be above the oval window and vertical portion to be more anteriorly and superficial than in fully developed ear.

Case No. 2

A 9 year old male child was first examined at 5 years age and subsequently reviewed for aesthetic reason and poorly developed speech. He had near normal left pinna. External canal was completely absent but meatal pit was present. Tuning fork tests suggested conductive hearing loss with good cochlear reserve. CT scan revealed absent external canal with partial middle ear space. Atretic plate was fused with malleus incus mass. Stapes was present. Fallopian canal was not delineated. Developmental arrest was suggested at around 8th week like in first case. Other salient features were hypoplasia of left mandible and slurring of voice.

Case No. 3

An 18 year old male patient presented with complaints of mal-developed left ear and absent ear canal with defective speech. Clinically the left pinna was found to be well formed but low and forward placed with absent external auditory canal. Tuning fork tests and pure tone audiometry suggested conductive hearing loss

with good cochlear reserve in the affected ear. CT scan showed absent external canal, well formed middle ear space, malleus and incus. Fallopian canal was delineated in both vertical and horizontal portions in normal location. Developmental arrest of 7th nerve is suggested in 20th week of gestation in view of (a) well formed middle ear space with ossicle and well formed auricle with adult shape suggest arrest after 8 to 20 weeks, whereas failure of resorption of inner core of epithelium suggests arrest before 21 weeks gestation. Hence, facial nerve location was likely to be normal at 20 to 21 weeks i.e. horizontal portion to be above oval window and vertical segment to be normal or little more anterior and superficial in view of failure of epithelization of core epithelium.

Surgical Procedure

All the cases were operated under general anaesthesia. The anterior-posterior approach as practised by Jahrsdoerfer [8] was used in all cases. Through a post auricular incision soft tissue was lifted and mastoid cortex exposed. A careful drilling was confined to an area bounded by temporomandibular joint (Glenoid fossa) anteriorly and middle fossa dura (Tegmen tympani) superiorly. This expectedly overlay the epitympanum. Drilling was done till atretic plate was reached. The atretic plate was invariably encountered fused with malleus and incus in attic. A diamond burr was used to thin out the atretic plate to paper thin and then removed by fine hook with minimal manipulation of ossicular mass. Ossicular mass was not dislodged to view the stapes. In most cases the ossicular chain though deformed, was mobile. Facial nerve was less prone to injury as it lay medial to the fused ossicular mass in anterior superior part of atretic plate [9] compared to its posterior inferior part wherein the nerve lay lateral and anterior. The cavity was widened keeping the fallopian canal when seen as its posterior limit.

Temporal fascia grafting was done by placing it over the ossicular mass and tucking in the graft in an artificial sulcus made all around at the junction of external canal and middle ear. This was followed by wide double than normal size meatoplasty. A split thickness skin graft was taken from abdominal wall or inner thigh to line the wide external canal. One edge of the skin graft was cut in zig-zag fashion such that 3 to 5 points were created. The new canal was circumferentially lined with skin, taking care that zig-zag tips partially overlap the fascia without burying skin under each other. The skin graft was maintained by antibiotic soaked gauze packing which was kept for 2 weeks. A disc of silastic sheeting 0.020 inch was kept over the neotympanum.

Result

All 3 cases had grade 1 microtia, case no 1 & 2 had small contracted middle ear cavity. Malleus and incus head were found to be fused. Malleus was fused with atretic plate as well. Ossicular chain was found to be mobile in all 3 cases. No attempt was made to visualize the stapes deliberately.

The tympanic segment of facial nerve was normal in all except a small dehiscence part anterosuperiorly in case no 1. The vertical segment was more anterior and lateral in case no 1 & 2 leading to small and narrow middle ear cavity. The vertical segment was normal in case no 3 ensuring near normal reformed cavity. In case no 2, thin band of chorda tympani nerve was seen lateral to ossicular mass flushed with atretic plate.

All 3 cases have shown improvement in air conduction threshold upto 30 to 35 db HL. Case no 2 developed postoperative canal stenosis by nearly 50%, leading to deterioration in hearing level by 6 month follow up. He was advised revision surgery at an interval of 1 year. Case 3 developed discharge postoperatively which was managed conservatively and a tiny perforation healed through

chemical cautery.

Discussion

Congenital atresia of ear apart from aesthetic disability is responsible for functional impairment of hearing and speech development, depending upon extent and bilaterality of disease. Even when the malformation is unilateral, surgery is warranted to create a safe ear canal and to establish access to the middle ear to restore binaural hearing. There is a growing evidence that the development of language and early learning are enhanced with binaural hearing [8]. Major concerns and criteria before undertaking any reconstruction, pertain to timing of surgery, preoperative evaluation, surgical technique and avoidance of any intraoperative or postoperative complication.

In case of bilateral atresia optimum age at which surgery should be undertaken in one ear is around 5 years, whereas the standard teaching to operate on unilateral atresia is not before adolescence. Lember [10] believes that in order to provide binaural hearing and proper speech development, corrective surgery in properly selected children with unilateral atresia can be undertaken as early as 4 years of age. An ideal case would be one without associated facial deformity, without a gross difference in middle ear space (not less than 50% of the opposite side) and presence of ossicular mass as seen in CT scan. Any otoplastic repair by the plastic surgeon needs to be taken up first. In cases with bilateral atresia, bone conduction hearing aid should be used till such time surgical repair is done.

The preoperative evaluation of the patient should include audiometric analysis, consisting of behavioural, pure tone audiometry and brain stem evoked response audiometry. The latter will be of great help in cases with bilateral atresia, wherein, it will help to assess the cochlear function. Surgery on non functioning ear or the only hearing ear, needs cautious approach and expertise of skilled otologic surgeon.

High resolution CT is mandatory in evaluating and analyzing the shape and size of middle ear, any abnormality of tympanic bone, type of atresia plate, presence and state of ossicular chain, inner ear status, any soft tissue opacity (cholesteatoma) or infection of tympanum and possibly the course of facial nerve canal. The presence of very small middle ear (i.e space less than 50% size of normal side) and absence of ossicles in a unilateral atresia case is a contraindication for surgical correction [10].

Major consideration during surgery is prevention of injury to facial nerve. The course of facial nerve canal

usually can be delineated by CT scan, however, if the facial nerve canal is not developed, it might be impossible to obtain an accurate localization radiographically. In such cases or otherwise, judgement based on understanding of embryology could be the surgeons only guide to the position of facial nerve. Sataloff has studied and rationalized the development of facial nerve and its position at a specific point in its gestational life. He emphasizes that if the facial nerve were abnormal the arrest of facial nerve development in temporal bone would generally occur at the same time as developmental arrest of other structures in malformed ear. Hence if the malformation of external or middle ear structures can be timed in gestational age by visual or radio imaging it will be possible to predict the location of facial nerve. It will be a boon to have a fair idea of location of facial nerve during surgery. This supplemented by intraoperative facial nerve monitoring will make atresia surgery more safe and rewarding.

Jahrsdoerfer has popularised the anterior approach as described in surgical procedure. The advantage of this approach is that there is less manipulation in the vertical segment of facial nerve and horizontal part is protected by the ossicular mass. There are no chances of mastoid cavity problems. The external canal needs to be widened as much as possible keeping the vertical part of fallopian canal as its posterior boundary. This will facilitate placement of skin graft lining in external canal as well.

In the posterior transmastoid approach as advocated by Glasscock [11], mastoid cavity is created by drilling the bone from sinudural angle anteriorly into antrum and facial recess. The atretic plate is then lifted after separating incudo-stapedial joint. In this approach there are more chances of injury to vertical portion of 7th nerve and also mastoid cavity problem. Modified anterior approach is useful in patients with thick bony atresia plate and poorly pneumatized mastoid wherein maintaining medial orientation is difficult and dissection too far posteriorly or inferiorly endangers facial nerve or lateral semicircular canal (LSCC). In such cases, initial posterior dissection with limited exposure of sinodural angle enables facial nerve and LSCC identification. This is followed by anterior approach to create new ear canal. This combination of anterior and posterior approach acts as intact canal wall for mastoidectomy.

All the 3 cases in this study have been operated by anterior-posterior approach.

To conclude, the surgery of congenital ear atresia

can be made safe and rewarding (a) by good case selection (b) proper preoperative evaluation and analysis using radio imaging and knowledge of embryological anatomy (c) use of intraoperative facial nerve monitoring. Anterior trans-atretic approach is relatively safe with minimal manipulation of ossicular mass and avoids mastoid cavity problems. The facial nerve is expected to be abnormal in most cases as is stapes expected to be mobile in majority of the cases. And finally the postoperative office care is as important as the surgical technique. Early detection of narrowing of meatus may be managed by dilatation and restenting over a period of several months without reoperation successfully.

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