

**TITLE: Congenital Aural Atresia**

**SOURCE: Grand Rounds Presentation, UTMB, Dept. of Otolaryngology**

**DATE: October 17, 2007**

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Congenital aural atresia (CAA) refers to the absence of external auditory canal. It occurs in 1/10,000-20,000 live births. It is more common in male and occurs more frequently on the right side. It is more commonly unilateral and bony as opposed to cartilaginous.

### **Embryology:**

Embryology explains many associations that occur with CAA. At around 6-8 gestational week, epithelial core from the first branchial groove forms meatal plate. This recanalizes around week 21 and forms external auditory canal. During a similar time period, auricle and middle ear structures form; the first and second branchial arches give rise to auricle between week 8-12 and to middle ear structures around week 5-12. Inner ear structures, on the other hand, develop solely from otic capsule and starts at an earlier date, around week 3. Hence the most commonly associated anomalies to CAA are malleus/incus fusion and microtia. It is notable that the severity of microtia dictates the degree of middle ear malformation. Less commonly associated are the malformations of the footplate of the stapes or inner ear structure. Unlike other ossicles, the footplate of the stapes has dual origin from the second branchial arch as well as otic capsule.

### **Incidence:**

CAA is mostly sporadic. Syndromic associations such as Treacher Collins, Goldenhar's, De Grouchy, branchio-oto-renal, and Crouzon syndrome occur in about 11-47%. It is familial in about 4.9-10.3%.

Other non-syndromic associations include facial asymmetry, facial nerve abnormality, cleft lip/palate, urogenital defects, cardiovascular malformation, macrostomia, and congenital cholesteatoma.

### **Presentation:**

Patients usually present as newborns with microtia or having failed hearing screen. A careful history should be taken including exposure to teratogens such as thalidomide, isotretinoin,

vincristine, cholechicine or cadmium as these could cause microtia. Family history of CAA or other ear anomalies should be elicited. Hearing status, language development, history of any ear surgeries as well as frequent ear infections should be asked.

On physical exam, one should look for the location of condyle, size of mastoid, craniofacial anomaly, and severity of microtia. Severity of microtia is graded from I-IV. Grade I is when the auricle is in normal shape but slightly smaller. Grade II still has normal cartilaginous framework, but the size is about half of normal. Grade III is severe microtia without normal shape of the auricle. Grade IV is anotia.

## **Grading:**

One can also grade severity of atresia based on more extensive anatomical findings. One such classification is by Schucknecht. Grade A is stenotic ear canal with normal middle ear and inner ear anatomy. Grade B has stenotic ear canal with some degree of middle ear abnormality such as fusion of malleus and incus. Grade C has complete atresia but relatively preserved anatomy of middle ear and inner ear. Finally, grade D has both atresia of the canal as well as abnormal middle ear, inner ear and neurovascular structures.

## **Hearing:**

Audiological assessment should be done when the patient presents at age 2-3 months with auditory brainstem response (ABR). It will typically show a normal bone conduction level but maximal conductive hearing loss with air conduction level near 60 dB. This is both due to the absence of canal and impaired mobility of the ossicles. If ABR shows unilateral hearing loss with normal contralateral hearing, then no intervention is necessary. Behavioral audiogram should be done at age 6 months to confirm ABR. If ABR shows bilateral hearing loss, then the child should be fitted for bone conduction hearing aid ideally before the age of 6 months. They should be enrolled in speech therapy early on.

## **Imaging and Prognosis:**

High resolution CT scan (HRCT) should be obtained at several months. Things to look for include the status of the inner ear, the extent of temporal bone pneumatization, the course of the facial nerve and the presence of the oval window/stapes footplate. Additionally, one should also assess the thickness of the atretic bone, soft tissue contribution to the atresia, size/status of the middle ear cavity, and the presence/absence of congenital cholesteatoma.

Jahrsdoerfer et al. came up with grading system of CAA based on CT findings. It gives each anatomical structure such as presence of stapes, oval window, middle ear space, and normal facial nerve either 1 or 2 points, with the presence of stapes footplate being the only one with 2 points. Out of 10 points, if the patient achieves 8 or 9, that correlates with excellent prognosis when undergoing a reconstructive surgery, likely to achieve post-operative air bone gap of 20-30dB 80% of the time, according to their study. On the other hand, if their score is equal to or less than 6, then the patient is a poor candidate for the surgery.

## **Counselling:**

Based on this scheme, one should counsel parents of the patient with CAA about the prognosis of the reconstructive surgery. Regardless, if the patient has bilateral conductive hearing loss, he or she should be started on bone conduction hearing aid (BCHA) right away. Bone conduction hearing aid uses a headband or glasses and has a sound transducer which vibrates mastoid bone and stimulates cochlea directly. About 25 years ago, another kind of bone conduction hearing aid called BAHA or bone anchored hearing aid was developed in Sweden. It has an implantable titanium screw with abutment which osseointegrates into the patient's temporal bone. It overcomes many of conventional BCHA's weaknesses such as contact site discomfort, lack of cosmetic appeal and attenuation of sound through soft tissue. It requires a two-staged surgery under general anesthesia for younger children, the first one for the implantation of the titanium fixture and the second one for skin-penetrating abutment, since young children's skull continue to grow. Despite the need for surgery, BAHA does not preclude a later reconstructive surgery owing to its posterior/superior location

## **Hearing Improvement:**

There are many studies attesting to the hearing results in patients using BAHA. Grandstrom et. al in 1993 showed that 39 patients fitted with BAHA for poor Jardoerfer score (6 or less) showed 100% satisfaction with speech threshold. Hakansson et al published BAHA results with 147 patients in Sweden in 1995 which showed that if pre-implantation pure tone average is less than 45, then 89% patients stated improvement in hearing. This went down to 61% for pre-implantation PTA between 46 and 60 and 22% for those with pre-implantation PTA of >60. Lustig et al looked at 40 patients in US using BAHA and showed 80% of those exhibited air bone gap (ABG) < 10 whereas 60% had ABG < 5. The BAHA consensus statement from 2004 suggested then that BAHA to be used when there is conductive hearing loss with ABG > 30 and also with conditions that preclude use of air conduction system such as in CAA or with persistent otorrhea. It also stated the need for bilateral fitting of hearing aids in children with bilateral conductive hearing loss. In children who are waiting for reconstructive surgery, BAHA can be used in patients younger than 2 with use of elastic band such as Softband BAHA.

## **Treatment:**

The goal of reconstructive surgery is to create a patent, skin-lined external auditory canal with post operative ABG between 20-30. The hearing result must hold in long term; in pre-lingual and school-aged children, the hearing result must be adequate for language development and give maximal education benefit. It should also create an esthetically pleasing auricle.

Atresiaplasty is a difficult surgery for many reasons. There are various altered anatomical sites, so that usual surgical landmarks are absent, particularly when anomaly of facial nerve might be present. There are other technical difficulties such as placing skin graft and separating ossicles from atretic bone. One needs to be wary of such complications as meatal stenosis and sensorineural hearing loss.

Because CAA is usually present along with microtia, one must approach this as a multi-disciplinary and multi-staged procedure. One approach adopted by Jahrsdoerfer et al is to have

auriculoplasty involving cartilage graft harvest, implantation, lobule reshaping and raising of the framework by a reconstructive surgeon in 3 different stages and then proceeding with atresiaplasty. One can combine formation of tragus with atresiaplasty. Each stage is separated by minimum of 3 months. The timing of surgery is case dependent, but usually the earliest time one would consider surgery is at age 5-6 for bilateral CAA. At any earlier age, the rib cartilages from which microtia repair will be harvested from are not mature. Also, the patient has to be old enough to be able to participate in post-op care such as dressing changes. Reconstruction of the auricle must precede atresiaplasty since one would need an undisturbed vascular bed for the best possible reconstruction.

Contrary to other ear surgeries, the better hearing ear is operated on first. Atresiaplasty itself has multiple parts, including canaloplasty, tympanoplasty and meatoplasty. One must employ intra-operative facial nerve monitoring because there are facial nerve anomalies such as when its vertical portion makes a more acute angle than usual, thereby re-crossing the middle ear space.

## **Follow-up Care**

Follow-up care is extensive, with meatal sutures coming out at 6 days, packing coming out in 2-3 wks and the first audiogram at 8 wks. Antibiotic drops should be used for 8-12 wks. The patient must come back for audiogram and periodic debridement of the new canal every 6-12 months.

## **Results of Treatment::**

The hearing results of the surgery were looked at by several authors. Jardoerfer et al published hearing results in 90 patients in 1992 they operated on. Out of these, 37 patients had Jahrdoerfer's grade of 8 or 9 out of 10. 78% of these patients had speech reception threshold of 10-25 dB and SRT <10 in 11%. Chandrasekhar in 1995 looked at 95 patients. 60% of those going through primary surgery achieved air bone gap closure of <30 and whereas 54% of revision surgery patients did. In 1997, Murphy et al published results showing 33% of those with partial atresia achieving SRT<20 and 15% of those with total atresia. De La Cruz et al published results of 116 operated ears, of whom 58% showed ABG<30 for primary surgery. For long term hearing results, De La Cruz et al looked at 90 ears with post-operation follow-up of greater than 6 months. He showed that on average, hearing improved with both pure tone average and air bone gap, but in long-term follow-up, hearing did deteriorate a bit, although not statistically significant. Ossicular chain re-fixation was the most common reason for hearing deterioration after the surgery. In summary, the success rate of surgery is dependent on the experience and expertise of the surgeon. In addition, what is termed a success in each study is different. The most commonly used measure is post-op ABG <30, and even that is not good enough for pre-school or school-aged children who need SRT <15 for maximal educational benefit.

## **Complications:**

There are many complications of the surgery. Lateralization of the tympanic membrane occurs in about 22-28% of the time and up to 12 months after the procedure. Meatal restenosis occurs in 8-12%. Fixation and discontinuity of ossicles happen at 11.5% of the time.

Sensorineural hearing loss and facial nerve injury is less common, occurring at 2 and 1% respectively.

Evans et al in 2006 revisited the question of “surgery or implantable hearing aids?” in a paper that compared costs, rate of complication, and hearing results. The paper showed that with atresiaplasty, the hearing gain on average is 17.3dB per ear whereas it is 31.8dB for BAHA. The cost for reconstructive surgery was \$51,506 whereas the cost for 1-stage BAHA and 2-stage HAHA respectively were \$42,449 and \$28,341. This comes out to cost/dB gain of hearing of \$2909 for reconstructive surgery, \$1238 for 1-stage BAHA and \$826 for 2-stage BAHA. In addition, they found out that 93% of patients who had the reconstructive surgery needed some form of sound amplification whereas BAHA patients achieved hearing level <15dB.

Complications-wise, there were far more serious and frequent complications such as



TM for the surgery compared to BAHA in which local inflammation/infection of skin (7.5%) and failure to osseointegrate (2.5%) were the main risks.

that binaural neurons develop early on only when there are binaural inputs present. There are reports of children with minimal or unilateral hearing loss having social and educational impacts without intervention. Wazen et al. looked at the implication of using BAHA in patients with unilateral CAA. He showed that they not only had the expected improvement in the thresholds for tonal and spondee, but also saw a significant improvement in handicap scores, reported by patients. This is from gaining binaural advantage in sound localization and speech perception in noise.

## **Conclusion:**

In conclusion, early identification of CAA is important for hearing amplification and special education. Patient classification with audiologic testing and CT images using such system as Jahrdorefer's is important for predicting results of the reconstructive surgery. BCHA should be fitted before age 6M for bilateral CAA. BAHA affords better hearing results and satisfaction than BCHA for non-surgical candidates. Reconstructive surgery needs a careful planning and timing with multidisciplinary effort. Before offering surgery, parents must understand that that the surgery produces a questionably adequate hearing result for children. BAHA on the other hand produces good hearing results and is more cost effective for hearing gain than the reconstructive surgery. Finally, binaural hearing amplification with BAHA is recommended for further gain in speech in noise and localization.

## **Case Studies:**

### **Case #1**

AL is a 53 year old Hispanic male with no significant past medical history who presented to our clinic as a referral from audiology. He was wearing a bone conductive hearing aid on the right side for left CAA and wished to be refitted for another one. On exam, he had grade 3 microtia with complete atresia on left side and normal auricle with normal canal and tympanic membrane on the right. He was wearing a BCHA in a hairband. His audiogram showed mixed severe to profound hearing loss bilaterally with conductive hearing loss on the right mostly. When asked about the possibility of reconstructive surgery or BAHA, patient stated that he was pretty happy with BCHA and wished no changes in his life.

### **Case #2**

RP is a 10 month old boy returning to our pediatric ENT clinic for a follow-up after failing initial newborn OAE. He was born with right partial atresia. On physical exam, he had microtia and right side with small, stenotic canal. Left side had a narrowed EAC at the junction of bony-cartilaginous junction. CT showed bony atresia on right side, making contact with middle ear ossicles with otherwise normal inner ear and middle ear. Left ear also had stenotic, tortuous canal with additional, second tympanic membrane medially. Patient was developing speech normally. He was referred to audiology for fitting with either BCHA or BAHA for binaural benefit.

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