Management of Congenital Aural Atresia

Robert F. Yellon and Françoise Denoyelle

Introduction

Management of microtia and congenital aural atresia (CAA) are challenging yet rewarding entities in reconstructive surgery and otology. Preservation of hearing and the seventh cranial nerve to correct hearing loss. High-resolution computed tomography (CT) and seventh nerve monitoring increase the safety of atresiaplasty.

Demographics and Associated Deformities

The incidence of microtia/CAA is 0.83-17.4 in 10,000. In a report on 1200 patients with microtia by Brent, 58% of cases were right-sided, 32% were left-sided, and 9% bilateral. Associated deformities included facial asymmetry (36.5%), seventh nerve weakness (15.2%), cleft lip or palate or both (4.3%), urogenital defects (4%), cardiovascular malformations (2.5%), and macrostomia (2.5%). Haploinsufficiency for the HOXA2 gene has been associated with autosomal dominant bilateral microtia and hearing loss.

Microtia may occur due to in utero obliteration of the stapedial artery or hemorrhage into local tissues. Known teratogens for microtia include thalidomide, isotretinoin, vincristine, colchicine, and cadmium. Genetic syndromes associated with microtia/CAA such as hemifacial microsomia (also known as Oculoauriculovertebral Spectrum or Goldenhar syndrome), Treacher-Collins syndrome, and Branchio-Oto-Renal syndrome should be identified. Hemifacial microsomia may include cardiac, cervical spine, auricular, ocular, and renal anomalies. Genetics consultation is advised.

Initial Evaluation

Parents are advised that the two important factors are first to maximize hearing and then to achieve optimal cosmesis. Otoacoustic emissions and/or auditory brain stem response testing should be performed within the first few months, to document hearing function in the normal ear and the degree and type of hearing loss in the atretic ear. The bone conduction is usually, but not always, normal in CAA. Conductive hearing loss is usually maximal at 60dB. With congenital external auditory canal stenosis (CEACS), there may be less conductive hearing loss.

In the past, for unilateral cases, no intervention was considered necessary other than preferential seating and monitoring of hearing. Recently, evidence
is mounting that unilateral hearing loss has adverse outcomes and that early treatment may be considered.\textsuperscript{9-11} For unilateral cases, acquisition of binaural hearing, improved sound localization, and optimizing hearing in noise are desirable. The need for surgical reconstruction for unilateral CAA is controversial.

Computed tomography of the temporal bones, may be obtained at one year to evaluate the anatomy for possible atresiaplasty reconstruction and to rule out possible cholesteatoma.\textsuperscript{12-14} Some would postpone the CT scan until the age of 5-7 to avoid radiation exposure.

Parents should be counseled about realistic expectations for both hearing and cosmesis with the various surgical and non-surgical options. Parents should be educated about the options of prosthetic auricles, rib graft auricles, Medpor\textsuperscript{®} (Stryker, Kalamazoo, MI) auricles, prosthetic auricles clipped to bone anchors, BAHA\textsuperscript{®} (Cochlear Corp., Lane Cove, Australia), Ponto\textsuperscript{®} (Ponto, Oticon Medical, Askim, Sweden), Sophono\textsuperscript{®} (Sophono Inc., Boulder, CO), Vibrant Soundbridge\textsuperscript{®} Middle Ear Implants (Med-El, Innsbruck, Austria), Sound Bite\textsuperscript{®} (Sonitus Medical, San Mateo, CA) devices and Softbands\textsuperscript{®}. The child should be seen at yearly intervals.

**Timing and Sequence of Surgeries for Microtia and Congenital Aural Atresia**

Historically, most authors advocate CAA repair following microtia reconstruction. Previous operations may create scarring and compromise blood supply that may decrease the chances of successful cartilage graft implantation. However, Roberson et al.\textsuperscript{15} reported a small series of early atresiaplasties followed by later microtia reconstruction with Medpor implants and temporoparietal flaps. Zhao et al.\textsuperscript{16} reported a series of 1460 ears in 1300 children with microtia/CAA who underwent canal wall down atresiaplasty at the same time as microtia reconstruction with rib graft, as the first stage of reconstruction.

Currently, the optimal age to begin auricular reconstruction is generally considered age 7 or more years, although others have recommended reconstruction from 2-9 years. For the occasional case of CAA without microtia and for CEACS, the atresiaplasty may be undertaken after the age of 4 years. The appropriate age for atresia repair is debated. Proponents of early surgery believe that acquisition of binaural hearing, sound localization, and improving the ability to hear sound in noise at as early age is beneficial. Proponents of late atresiaplasty believe that the decision for atresiaplasty should be delayed until the patient is old enough to take part in the decision and is willing to accept the risks of facial nerve injury and SNHL.

**Congenital Aural Atresia – Grading Systems of Favorability for Atresiaplasty**

The critical factor in the decision for reconstruction of CAA and CEACS is patient selection based on anatomy. Yeakley and Jahrsdoerfer\textsuperscript{17} devised a grading system based on CT of the temporal bone and auricle to determine which patients are candidates for atresiaplasty (Table 1). Each favorable factor is awarded 1 point. A score of less than 6 indicates that the patient is not a favorable surgical candidate, while a score of 6 or greater indicates that the patient is a candidate for atresiaplasty. The higher the total number on the grading scale, the more likely is a good hearing outcome after surgery. The presence of a normal stapes is the only factor that is given 2 points (Figure 1).
Table 1. Jahrsdoerfer Grading System for Computed Tomography of Congenital Aural Atresia

<table>
<thead>
<tr>
<th>Anatomic Structure</th>
<th>Points Awarded</th>
</tr>
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<tbody>
<tr>
<td>Stapes favorable</td>
<td>2</td>
</tr>
<tr>
<td>Oval window open</td>
<td>1</td>
</tr>
<tr>
<td>Middle ear well pneumatized</td>
<td>1</td>
</tr>
<tr>
<td>Facial nerve favorable</td>
<td>1</td>
</tr>
<tr>
<td>Incus/malleus favorable</td>
<td>1</td>
</tr>
<tr>
<td>Incus/stapes connected</td>
<td>1</td>
</tr>
<tr>
<td>Mastoid well pneumatized</td>
<td>1</td>
</tr>
<tr>
<td>Round window open</td>
<td>1</td>
</tr>
<tr>
<td>Auricle normal</td>
<td>1</td>
</tr>
<tr>
<td>Maximum Total</td>
<td>10</td>
</tr>
</tbody>
</table>

If total >6 – favorable for atresiaplasty  
If total <6 – unfavorable for atresiaplasty  
(From: Yeakley J, Jahrsdoerfer RA. CT evaluation of congenital aural atresia: what the radiologist and surgeon need to know. J Comput Assist Tomogr 20:724, 1996.)

Figure 1. Coronal computed tomography scan of right temporal bone of child with microtia/congenital aural atresia. This ear is favorable for atresiaplasty. Note the well-pneumatized middle ear and mastoid cavity, well-formed malleus-incus complex, and the stapes present in the oval window.  

Figure 2. Axial CT scan of temporal bone in congenital aural atresia showing the malleus-incus complex directly lateral to stapes rather than in the normal antero-lateral position. This anomaly makes surgical visualization and exposure to the stapes, incudostapedial joint and oval window more difficult or not possible.  

Yellon and Branstetter and Dedhia, et al. recently reported modification of the Jahrsdoerfer grading system for CAA candidacy for atresiaplasty (Table 2). The new and/or modified anatomic considerations with their respective incidences included the presence or absence of:
1 - low-lying tegmen mastoideum (13%);
2 - malleus/incus complex (MIC) large and positioned directly lateral to stapes rather than the usual anterolateral position (24%) (Figure 2);
3 - facial nerve obstructing oval window (41%); and 4 - facial nerve turning anterolateral and obstructing lateral surgical approach (21%)\(^{18,19}\)

<table>
<thead>
<tr>
<th>Anatomic Structure</th>
<th>Points Awarded</th>
</tr>
</thead>
<tbody>
<tr>
<td>Stapes favorable</td>
<td>1 or 2</td>
</tr>
<tr>
<td>Oval window open</td>
<td>1</td>
</tr>
<tr>
<td>Middle ear well pneumatized</td>
<td>1</td>
</tr>
<tr>
<td>Incus/malleus favorable</td>
<td>1</td>
</tr>
<tr>
<td>Incus/stapes connected</td>
<td>1</td>
</tr>
<tr>
<td>Mastoid well pneumatized</td>
<td>1</td>
</tr>
<tr>
<td>Round window open</td>
<td>1</td>
</tr>
<tr>
<td>Auricle normal</td>
<td>1</td>
</tr>
<tr>
<td>Tegmen mastoideum normal, mildly low, severely low</td>
<td>1 or 2</td>
</tr>
<tr>
<td>Malleus-incus vs. stapes position</td>
<td>1</td>
</tr>
<tr>
<td>Facial N. at oval window normal</td>
<td>1</td>
</tr>
<tr>
<td>Facial N. posterior or lateral to middle ear</td>
<td>1</td>
</tr>
<tr>
<td><strong>Maximum Total</strong></td>
<td><strong>14</strong></td>
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Any anomaly of the inner ear excludes the child from atresioplasty because of an increased incidence of sensorineural hearing loss (SNHL) following surgery.\(^{20}\)

**SURGICAL APPROACHES**

Atresioplasty via a canal wall-down mastoidectomy was advocated by earlier surgeons such as Schuknecht\(^{14}\) but fell out of favor due to poorer hearing and mastoid cavity problems. More recently, good hearing results with canal wall down atresioplasties have been reported.

Most surgeons favor the anterior approach described by Jahrsdoerfer. An incision is made and the landmarks of the zygomatic root and the temporomandibular joint are identified. The more anterior (towards the temporomandibular joint) and superior (towards the dura) one drills, the safer it is because this area is furthest from the facial nerve. Caversaccio et al.\(^{21}\) and Seigert\(^{22}\) reported CT-based image guidance navigation system for atresioplasty. Drilling continues to the bony atretic plate. The MIC is usually fixed to the bony atretic plate, and excessive drill energy may be transmitted by drilling on the plate. The safe technique is to use diamond
burs to thin the atretic plate to eggshell thickness and then remove it gently with picks and curettage. Angled telescopes and lasers may be useful (Figure 3). Ossiculoplasty techniques may be needed, including partial ossicular reconstruction prostheses or stapedectomy. If stapes surgery is required, it is preferable as a planned second stage procedure.

When the MIC is large and directly lateral to the stapes, it is difficult to assess the mobility of the stapes and the incudostapedial joint. The new EAC and bony annulus should be widened up to 15 mm in diameter; however, the EAC diameter may be limited by dura, temporomandibular joint, facial nerve, or mastoid air cells.

A fascia graft is placed to reconstruct the tympanic membrane. A split-thickness skin graft is used to line the external auditory canal. A small disc of thick silastic sheeting is placed over the temporalis fascia graft and the partially overlapping skin graft to anchor these grafts and prevent lateralization. Pieces of Merocel® (Medtronic Xomed, Jacksonville, FL) packing coated with bacitracin and expanded with ofloxin drops are placed in the EAC. Teufert and de la Cruz reported improved hearing outcomes using Merocel®, argon laser, thin split-thickness skin graft (0.008 in.), and intraoperative placement of the silastic disc.

An external auditory meatus is created by excising tissue from the appropriate location on the reconstructed auricle after a pedicled skin flap has been elevated. The reconstructed auricle is placed in its original position. The split-thickness skin graft is brought out through the external auditory meatus, and sutured to the edges of the skin and flap from the meatus. More packing is placed. The packing and silastic disc are removed after 8-12 days.

**Complications of Atresiaplasty**

Facial nerve injury has been reported in 0-7% of cases. SNHL has been reported in 0-7.5% of atresiaplasties. Stenosis is the most common complication and most common reason for revision surgery, occurring in 0-25% of cases. Injection of triamcinolone into the meatus at initial surgery or subsequently may decrease stenosis.

Lining the meatus with a pedicled full-thickness skin graft can decrease stenosis. Chang et al. reported a 15% incidence of meatal stenosis using anterior and inferior periosteal flaps and a superiorly based skin flap to line the meatus versus a 25% rate without. Wicks, stents, and molds can decrease or prevent stenosis. Lateralization of the tympanic membrane may require revision surgery in 0-18% of cases.
Combined Microtia and Atresioplasty

Several surgeons have reported combining stages of microtia with atresioplasty. Seigert\textsuperscript{22} developed a three-stage method for complete reconstruction of microtia and CAA. Cho and Lee\textsuperscript{33} developed a two-stage complete microtia reconstruction with atresioplasty. Yellon\textsuperscript{29} combined atresioplasty with tragal reconstruction following the cartilage implantation, lobule rotation and elevation stages of microtia reconstruction.

Hearing Results for Atresioplasty

With the anterior approach to atresioplasty, Jahrdoerfer\textsuperscript{24} and Lambert\textsuperscript{38} reported a 25 dB or smaller hearing threshold for 70\% of patients. Shonka et al.\textsuperscript{39} reported that a Jahrdoerfer grade of >7 predicted an 89\% chance of <30 dB SRT. Dobratz et al.\textsuperscript{26} and Lambert\textsuperscript{13} noted better hearing and decreased need for revision surgery when the native ossicular chain was preserved versus ossiculoplasty. Results for revisions are generally worse than results for primary surgeries.\textsuperscript{28,29,40}

An air-bone gap <30 dB may be achieved in most patients, ranging from 55\%-94\% in various series.\textsuperscript{28,29,30,34,40} Patel and Shelton\textsuperscript{30} reported that atresioplasty has a learning curve and that hearing results improve with surgeon experience. Canal wall-down atresioplasty was historically associated with poorer hearing outcomes.\textsuperscript{14} More recently, fairly good hearing results with canal wall down atresioplasties have been reported,\textsuperscript{16,41,42} including a large series (n=1480) performed simultaneously with rib cartilage grafting for microtia reconstruction. Lambert\textsuperscript{13} and others\textsuperscript{17,39,46} reported that atresioplasty hearing results may undergo deterioration over time.

Sound localization has been shown to improve after unilateral atresioplasty.\textsuperscript{43}

Congenital External Auditory Canal Stenosis (CEACS)

Schuchtenicht developed a four-level grading system for CEACS and CAA.\textsuperscript{14} The Type A anomaly is isolated meatal stenosis, which is the mildest form. Canal cholesteatoma may develop medial to the stenosis. Isolated meatal stenosis may be repaired via canaloplasty with drilling of the bony ear canal and thinning of the soft tissues with skin flap preservation.

The type B anomaly involves narrowing of the entire canal. Middle-ear anomalies are common. Canaloplasty is performed if narrowing is severe, associated with hearing loss, or canal cholesteatoma. Yellon\textsuperscript{44} observed that about 50\% of cases of CEACS have a partial atretic plate, small tympanic membrane, and/or ossicular anomalies.

Type C anomalies have complete CAA but relatively normal middle ear structures, favorable pneumatization, and good facial nerve position. These ears may be reconstructed by atresioplasty. Type D anomalies are the most severe, with complete CAA, poor pneumatization, major anomalies, and are not candidates for surgery.

Jahrdoerfer and Chole\textsuperscript{45} reported a 50\% incidence of canal cholesteatoma by the age of 20 years with EAC diameter < 4 mm. It is likely that some of these cases were untreated as children. A neonate with an EAC diameter of >2.5 mm who is managed in the office or operating room every six months for examination and cerumenectomy may outgrow the problem by age 4-5 years. Canal cholesteatoma
is more likely if the diameter of the EAC is <2mm and the EAC is not surgically widened. Canal cholesteatoma may be present with postauricular sinuses. When surgery is indicated for CEACS, the endaural approach described by Lempert is preferred. If a partial atretic plate is attached to the MIC, the plate is thinned and removed gently. The removal of the partial atretic plate and enlargement of the bony annulus may necessitate placing a fascia graft to enlarge the tympanic membrane. If exposed bone in the EAC is extensive, then a split thickness skin graft is required; a full thickness skin graft will be too bulky and obstruct the EAC. Wicks or stents may be placed if stenosis occurs. Triamcinolone injection may decrease stenosis.

Alternatives for Congenital Aural Atresia—Implantable Hearing Aids in Children

Several types of implantable hearing devices may be used for CAA in children as an alternative to atresiaplasty: percutaneous (crosses skin) bone conduction devices (BAHA®, Ponto®), transcutaneous (closed skin) bone conduction devices (Sophono®, and BAHA Attract®), and active middle ear implants (Vibrant Soundbridge®). In the United States, the minimum age for the BAHA®, Ponto® and Sophono® is 5 years, and the minimum age for the Vibrant Soundbridge® is 18 years. In other countries, the BAHA®/Ponto®/Sophono® devices have been implanted in children younger than 5 years. The Soundbite® is a nonsurgical device that is attached to the teeth and uses an external processor. The minimum age is 18 years in the United States. The internal vibrator of the Bonebridge® (MedEl, Innsbrück, Austria) is usually too large to be inserted in a small child’s atretic mastoid bone.

Children with bilateral CAA or in selected cases of unilateral CAA (highly motivated parents or children with hearing loss in the opposite ear) are typically fitted with a bone conduction device/sound processor strapped to the head with a Velcro band during the first months of life. Devices using headbands include the Bone Anchored Hearing Aid (BAHA), the Ponto®, and the Sophono®. The hearing device on a headband may also be used as a trial to see if the patient accepts the device prior to implantation. The traditional conduction hearing aid with metal headband is also a less expensive option. Compliance can be an issue.

Evidence is accumulating that that unilateral CAA has an impact on learning skills. While developmental outcomes of children with unilateral sensorineural deafness have already been studied, the etiology of the deafness may also cause developmental delay (such as congenital infection with cytomegalovirus). Kesser, et al. showed that 65% of school children with unilateral CAA had need for extra services including hearing aids (13%), individualized educational programs (48%), FM systems (33%), and speech therapy (48%). Hearing rehabilitation for unilateral CAA or CEACS should be approached on an individualized basis.

Early rehabilitation (5-8 years of age) should be offered to children with language impairment or school difficulties, but otherwise it may be appropriate to wait for a decision by the child as he/she gets older. Atresiaplasty is a complex, lengthy, potentially risky surgery. Hearing results with the implants are usually better than results of atresiaplasty, and BAHA®/Ponto®/Sophono® surgery is
relatively fast and has less risk. On the average, BAHA® results in better hearing and lower cost per decibel of hearing improvement than atresiaplasty. BAHA® fixtures and abutments may also be used as anchors for prosthetic auricles.

**Bone Conduction Devices Clipped to a Percutaneous Abutment**

The BAHA® device with percutaneous implantation (open skin), was described by Tjellström more than 30 years ago. In one recent study of 15 patients with bilateral CAA (children and adults), the average hearing improvement was 33 ± 7 dB after a mean follow-up of 6.5 years.

The BAHA®/Ponto® is useful for children with CAA who have unfavorable anatomy for reconstruction, those who are poor candidates for a lengthy atresiaplasty, or for families that desire good hearing with less risk. The BAHA® and Ponto® have a visible metal abutment and sound processor and are less favorable from a cosmetic standpoint than atresiaplasty.

The standard surgical procedure requires general anesthesia in children and can be performed in one or two stages, according to the preference of the surgical team. Two-stage procedures are usually performed for younger and syndromic children with thinner bone (<2.5 mm). A simplified procedure is now used with new types of abutments (longer and/or Dermalock®- hydroxyapatite composite), without reduction of the soft tissue or removal of hair follicles.

Cutaneous complications and implant extrusion are the main complications after BAHA® implantation in children. The rate of cutaneous severe complications was estimated to range from 9.4 to 37%, and the loss of the implant ranged from 14 to 25.9%.

The planned site for BAHA® placement is 5.5 mm posterior superior to the external auditory canal or planned site of the external auditory canal. For children who have microtia reconstruction planned, Bajaj and colleagues advise placement of the BAHA® 6.5-7.0 cm from the planned site of the external auditory canal to avoid interference with microtia reconstruction.

Hearing results for children with bilateral CAA who underwent BAHA® placement have been impressive. Children with unilateral CAA who had BAHA® placement had improved quality of life reported but only mild functional improvement. A recent report by van der Pouw et al. described improved sound localization and speech perception in a small series of children with bilateral aural atresia who underwent bilateral BAHA® placement. Nadaraja, et al., did a systematic review of hearing results comparing osseointegrated bone conduction devices versus atresiaplasty. They found a mean hearing improvement of 38 dB with the osseointegrated bone conduction devices (100 ears) and 24 dB with atresiaplasty (516 ears).

**Transcutaneous (closed skin) Bone Conduction Device- Sophono® Alpha 1® and 2® and BAHA® Attract**

The Alpha 1®and then Alpha 2® bone-anchored implant were introduced in 2006 (Sophono Inc., Boulder, Colorado, USA). The external processor is held on by a transcutaneous (closed-skin) magnet. The vibrations are transmitted through the skin to the bone by surgically implanted magnets. An intermediary plate located between the external device and the skin allows modification of the strength of the
magnet. A retrospective study reported an average hearing gain in free field pure tone average of $31.2 \pm 8.1$ dB, without extrusion or severe cutaneous reactions in over 100 patients.\(^6^6\) The Sophono® is better cosmetically than percutaneous devices, but has slightly poorer amplification and speech discrimination.\(^6^9\) The skin incision is 7.5 cm in length and located 6 cm behind the pinna. Implant magnets are secured by 5 screws (Figure 4a and 4b). The implantation may be bilateral and/or combined with another stage of microtia reconstruction without an additional incision. The BAHA® Attract (Cochlear Corp., Lane Cove, Australia) is a similar new promising device but little data are available.

![Figure 4A. Sophono alpha, incision.](image)

![Figure 4B. Sophono alpha, implant fixation.](image)

**Closed Skin Active Middle Ear Implants**

In young children outside the United States, the Vibrant Soundbridge® middle ear implant may be used, because the placement of the floating mass transducer to the ossicular chain is independent of skull growth. First indicated only for SNHL in adults, the indications for this device were extended to conductive deafness to children in Europe.\(^7^0\) In 2009, implantation of the Vibrant Soundbridge® was reported in 7 cases (in 3 children aged 10, 11 and 15 years and 4 adults) with unilateral hearing loss from CAA.\(^7^1\) The incision is performed in the hair, in an area away from the vestigial auricle, in order to allow future ear reconstruction (Figure 5a and 5b).

![Figure 5A. Middle ear implant Vibrant Soundbridge, incision.](image)

![Figure 5B. Middle ear implant Vibrant Soundbridge, postoperative aspect.](image)
The floating mass transducer can be crimped onto the long process of the incus, stapes or into the round window. In a French collaborative study in children (12 cases of CAA), the floating mass transducer was crimped onto the long process of the incus (8 ears) and to the stapes suprastructure (4 ears) after removing the fused MIC. The mean functional gain with the Vibrant Soundbridge® was 36.9 ± 11 (mean aided PTA of 28.3 ± 10 dB). An option when the MIC is directly lateral to the stapes (Figure 2) is to crimp the floating mass transducer onto the short process of the incus, since the location of the MIC precludes crimping on the stapes.

Conclusion

Surgical management of microtia and CAA continue to be challenging areas of reconstructive and otologic surgery. Attention to detail, careful patient selection and planning, adequate training, and excellent judgment are required for good atresiaplasty results. Several implantable hearing devices are available. Selection depends on the child’s age, thickness of cortical bone, severity of middle ear malformation, surgeon’s experience, and finally, the patient’s preference.

References


67. Siegert R. [Magnetic coupling of partially implantable bone conduction hearing aids without open implants]. Laryngorhinootol 2010; 89: 1–6. (German)


