

TITLE: Congenital Aural Atresia

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Introduction

Congenital aural atresia is a term used to refer to a spectrum of ear deformities present at birth that involve some degree of failure of the development of the external auditory canal (EAC). Often, the malformation will also involve the tympanic membrane, ossicles and middle ear space to varying degrees. While associated abnormalities of the auricle are common, the inner ear development of these patients is most often normal. The challenge to the otologist is to restore the sound conduction pathway through the atretic EAC and malformed middle ear to the normal cochlea.

The incidence of congenital aural atresia is approximately 1 in 10,000 to 20,000 live births. Unilateral atresia occurs three to five times more commonly than bilateral atresia. Males are more often affected than females and in unilateral cases the right ear is more commonly involved. This anomaly most often occurs sporadically although cases of autosomal dominant or recessive inheritance have been reported. 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 (1,2,3).

Embryology

Congenital aural atresia occurs as a result of abnormal development of the first and second branchial arches and the first branchial groove. Development of the external ear begins during the fourth week of gestation as six mesenchymal proliferations enlarge to form ridges known as the hillocks of His. These hillocks, which surround the first branchial groove or primitive meatus, fuse to form the primitive auricle by the third month of gestation.

The EAC develops from the first branchial groove beginning in the eighth week of gestation. Epithelial cells from the meatus proliferate forming a solid core of cells, known as the

meatal plug. This core of cells migrates medially toward the outgrowth of the first branchial pouch, which will eventually form the middle ear cleft. The meatal plug contacts the middle ear cleft by the ninth week of gestation. This solid core will then recanalize to form the epithelial lined EAC, but not until the sixth or seventh month of gestation.

Ossicular development begins in the fourth week and, at this time, the malleus and incus appear as a fused mass. Separation into two distinct ossicles typically occurs by the eighth week of gestation. The first branchial arch, Meckel's cartilage, contributes to the development of the head and neck of the malleus and the body and short process of the incus. The second branchial arch, Reichert's cartilage, leads to the development of the manubrium of the malleus, the long process of the incus and the stapes suprastructure. By the sixteenth week of gestation, the ossicles are of adult size.

Inner ear development from the otic placode begins during the third week of gestation. Invagination of the otic placode to form the otic vesicle is apparent by week four, and by the sixth week the semicircular canals have taken shape. The utricle and saccule have formed by the eighth week. Development of the cochlea begins during the seventh week, and by week twelve the complete two and a half turns have formed. The membranous labyrinth is entirely developed by fifteen weeks gestation and ossification of the surrounding otic capsule is complete by twenty-three weeks gestation.

The nerve of the second branchial arch is the facial nerve. Its development begins with the differentiation of neuroblasts from the acoustico-facial primordium between four and five weeks gestation. The course of the nerve is completely formed by seventeen weeks. However at this time, the nerve is located in a more anterosuperior position. The eventual migration of the facial nerve to its normal adult position is dependent upon the normal development of the tympanic ring and mastoid.

From this summary of otologic embryology, we can see that completion of development of the external ear occurs rather early in gestation while the recanalization of the EAC occurs later. Therefore, a severely deformed auricle is likely an indicator of associated EAC, middle ear, facial nerve and possibly inner ear anomalies. In contrast, aural atresia in the presence of a normal auricle most likely represents a later arrest in development and has a higher likelihood of normal middle and inner ear structures (1,4,5).

Classification

Several different classification or grading systems for congenital aural atresia are present in the otolaryngology literature. Altmann's classification, first reported in 1955 but still widely utilized today, divides atresia into three groups based on the clinical assessment of the severity of the malformation. Group I atresia is characterized by a small EAC, hypoplastic temporal bone and tympanic membrane (TM), a normal or small middle ear cleft and normal or mildly deformed ossicles. Group II includes those cases with an absent EAC, an atretic plate, a small middle ear space and fixed and malformed ossicles. Group III is characterized by an absent EAC, a severely contracted or absent middle ear space, and absent or severely malformed ossicles.

De la Cruz made modifications to Altmann's classification system so that cases are categorized into major and minor malformations. The minor category is characterized by normal mastoid pneumatization, normal oval window, reasonable oval window-facial nerve relationship and a normal inner ear. The major category is comprised of cases with poor pneumatization, abnormal or absent oval window, abnormal course of the horizontal facial nerve and inner ear anomalies (1).

Yet another classification system was introduced by Schuknecht in 1989. This system divides atresia cases into four types based primarily on intraoperative findings and the type of surgical repair required. Type A atresia is limited to the cartilaginous EAC and is addressed with meatoplasty. Type B atresia is characterized by narrowing of both the cartilaginous and bony EAC along with a small TM and mild deformity of the malleus and incus. This type of atresia most often requires canalplasty, possibly with ossicular chain reconstruction (OCR) as well. Type C atresia cases have complete EAC atresia but a well-pneumatized middle ear and mastoid. The TM and ossicular malformations are more severe than in type B and there is a higher likelihood of facial nerve anomalies. Canalplasty and OCR will be necessary to correct type C atresia. Type D atresia involves complete EAC atresia and poor middle ear pneumatization. In these cases, associated facial nerve or inner ear anomalies often preclude surgical intervention (3).

Probably the most clinically useful classification system was introduced by Jahrsdoerfer in 1992. This system establishes a score (up to 10) based upon findings of high resolution CT scans of the temporal bone. The parameters of an open oval window, width of the middle ear cleft, facial nerve course, malleus-incus complex, mastoid pneumatization, incudostapedial continuity, round window patency and auricle appearance are assigned a value of one point. The presence of a stapes is given a higher priority and assigned two points. The final score has been used to predict the likelihood of successful atresia surgery. A score of 8 out of 10 correlates to an 80% chance for restoration of hearing to normal or near-normal levels defined as speech reception thresholds (SRT) between 15 and 25 dB. Cases with a score of less than or equal to 5 are generally not considered for surgical intervention (6,7).

Patient Evaluation

The first step in the evaluation of a patient with congenital aural atresia is to obtain a complete history and perform a thorough physical exam. Given that the majority of these cases will be discovered in the newborn, the history is focused on the details of pregnancy. It is important to ask about prenatal care and to determine if the mother was exposed to infections, drugs or alcohol during pregnancy. Parents should also be questioned about any family history of ear deformities or syndromal anomalies. Physical examination will, of course, involve a complete head and neck exam. Specifically, the degree of microtia is assessed and the severity of EAC atresia noted. In cases of EAC stenosis, attempts should be made to visualize the TM and ossicles and their presence and position documented. Additionally, examination of overall craniofacial development is necessary to assess for the presence of associated branchial arch anomalies.

Next, accurate audiologic evaluation in the newborn period is mandatory. Infants with any degree of ear anomaly should be marked as high-risk for associated hearing loss and have auditory brainstem response (ABR) testing before leaving the hospital. The initial priority in cases of unilateral atresia is to evaluate the auditory function of the unaffected ear. Normal hearing in one ear will allow for essentially normal speech and language development. However, the incidence of both conductive and sensorineural hearing loss in the nonatretic ear is greater in patients with unilateral atresia than in the general population. Therefore, it is essential that any auditory dysfunction in the “normal” ear be diagnosed early on so that appropriate amplification can be implemented if necessary. If surgical repair of unilateral atresia is considered later on, audiologic testing to confirm normal cochlear function in the involved ear will be necessary.

In cases of bilateral aural atresia, early evaluation with both air and bone conduction ABR testing is necessary. The bilateral conductive component makes testing somewhat more difficult by creating a masking dilemma. Evaluation of ear specific cochlear function is possible by measuring the Wave I response ipsilateral to the stimulation. Patients with bilateral atresia should be fitted with bone conduction hearing aids as early as possible to optimize speech and language development.

Once auditory function has been established, either via the unaffected ear in unilateral atresia cases or with bone conduction aids in bilateral cases, further evaluation is not necessary until the child has reached the age of 5 or 6 years. At this time, a high resolution CT scan of the temporal bones in both the axial and coronal planes is indicated. This study will allow evaluation of middle ear pneumatization, ossicular anatomy, inner ear morphology and the course of the facial nerve. This is the most useful study to decide if a patient is a candidate for surgery and to predict the likelihood of successfully reestablishing normal hearing as discussed previously. An indication for obtaining a CT scan earlier would be those patients with congenital aural atresia that present with a draining ear or acute facial palsy, which could indicate an underlying cholesteatoma (1,2,4,5).

Surgical Repair

There are two absolute requirements for a patient with congenital aural atresia to be eligible for surgery: 1) normal inner ear morphology demonstrated on CT scan and 2) normal cochlear function demonstrated by audiologic testing. A score of 5 out of 10 or less by the CT scan grading system may be considered a contraindication to atresia surgery because these patients are not likely to have an appreciable hearing improvement and are at higher risk for surgical complications. Patients with a score of 6/10 are considered “marginal” candidates, 7/10 “fair”, 8/10 “good”, 9/10 “very good” and 10/10 “excellent.”

Once the decision to operate has been made, the timing of repair must be planned. Patients with auricular malformation should undergo microtia repair first to avoid scar tissue formation compromising the local blood supply. Most authors agree that this process should begin around age 5 to 6 years at which time costal cartilage is sufficiently developed for harvest and optimal development of the mastoid process has occurred. Opinions vary, however, as to

whether atresia repair should be performed between Stages 2 and 3 of microtia repair or 2 months after the final stage of microtia repair.

There are basically two techniques for atresia repair—the transmastoid approach and the anterior approach. According to most authors, the transmastoid approach is not used as frequently but may be a preferable option in some cases. This approach begins with drilling the mastoid to allow identification of the sinodural angle, which is then followed anteriorly to the antrum. The lateral semicircular canal is identified and used as a landmark. The facial recess is opened and the incudostapedial joint (if present) may be separated. The atretic plate is then carefully removed. Ossiculoplasty and tympanoplasty proceed in the usual fashion and the newly created EAC is lined with a split thickness skin graft. The main disadvantages of the transmastoid approach are the creation of a larger defect that must be skin grafted, prolonged healing of the cavity and the presence of a mastoid bowl that requires lifelong maintenance.

The anterior approach, popularized by Jahrsdoerfer, is the most common method of atresia repair utilized today. In this technique, drilling begins at the atretic plate just posterior to the temporomandibular joint and inferior to middle fossa dura. Dissection proceeds medially following these two landmarks to the epitympanum where the fused malleus and incus can be identified. The safest area to drill is anterosuperiorly because the facial nerve is consistently located medial to the ossicles in the epitympanum. The most likely area to encounter an aberrant facial nerve is while drilling posteroinferiorly so this should be performed only after identification of other landmarks. Care must be taken not to drill directly on the ossicular mass to avoid trauma to the inner ear. The atresia plate is thinned and removed and any fixation of the ossicles to the atretic bone is lysed, either sharply, or, perhaps more safely, with the carbon dioxide laser.

Next, continuity of the ossicular chain must be assessed and if found to be intact, no manipulation is indicated. Cases in which the incudostapedial joint consists of just a fibrous attachment or is altogether absent warrant ossiculoplasty with a PORP. Unstable or absent suprastructure of the stapes requires reconstruction with a TORP. Both of these statements assume a mobile stapes footplate, which is most often the case. In those rare patients with a fixed footplate, some type of fenestration procedure will be necessary along with ossiculoplasty.

A temporalis fascia graft that had been harvested earlier and allowed to dry is used to recreate the TM. Optimally, the new TM will be centered on the ossicular mass to maximize hearing results. If a PORP or TORP had been used for ossiculoplasty, placing a small piece of cartilage between the table of the prosthesis and the fascial graft will help to minimize chances of extrusion. Finally, a split thickness skin graft, .012-.015 inches in thickness and approximately 6x6cm, is harvested for lining the new EAC. Most authors report using a donor site of the upper inner arm, but alternatives include the upper thigh or buttock. The skin graft is placed into the EAC, overlapping the TM facial graft medially. Care must be taken to ensure that all bone is covered with skin and that the skin graft edges are not folded over on itself. The ear canal is then packed with Nu-gauze or Merocel sponges impregnated with antibiotic ointment.

The native or reconstructed auricle is often located anteroinferior in relation to the newly created EAC. The auricle can be repositioned by undermining soft tissue and possibly excising

redundant postauricular skin. An external meatus is then created by excising skin, subcutaneous tissue and cartilage from the auricle. Alternatively, an anteriorly based flap of conchal skin and cartilage can be incised and folded into the new EAC to line the anterior portion of the canal. The lateral edge of the skin graft is brought through the meatus and sutured to the skin edges and this area is packed similarly to the medial EAC. The postauricular incision is closed in the usual fashion, possibly with the addition of some tacking sutures to the periosteum to maintain the posterior position of the auricle and to keep the meatus widely open. A mastoid dressing is applied.

The mastoid dressing is removed on postoperative day 1. The timing for removal of the EAC packing varies between authors, some recommend removing it altogether at 10 days to 2 weeks. Other recommend removing and replacing the meatal packing at 2 weeks, starting antibiotic ear drops at that time then removing the entire pack at 3 weeks. Frequent visits are required after packing removal to address any granulation tissue formation and to remove any desquamated skin (1,2,4).

Results

The hearing results reported in the literature after surgery for congenital aural atresia are somewhat difficult to interpret because of different classification systems used to describe the atresia preoperatively, different criteria for selecting surgical candidates, different definitions of a “successful” outcome and different time periods of patient follow-up after surgery. However, the majority of authors consider a successful hearing result to be 25-30dB or less after surgery. Using this definition, the percentage of cases with a successful outcome reported in the literature varies from 12-71% (8). Unfortunately, the majority of these papers are citing the hearing results obtained in the early postoperative period. An important concept to be familiar with when counseling patients about this surgery preoperatively is the stability of the hearing results over time. This issue was addressed by Lambert in 1998 by comparing early postoperative hearing levels (<1yr postop) to levels after longer follow-up (1-7.5yrs, avg 2.8yrs). He found that 60% of cases had hearing levels of 25dB or better and 70% were at 30dB or better in the early postoperative period. This diminished to 46% and 50% with longer follow-up. Additionally, he found that nearly one third of cases required revision surgery, most often for restenosis of the EAC or lateralization of the TM. After revision surgery, hearing levels of 25dB or less were achieved in 50% of cases and levels of 30dB or less in nearly two thirds of cases. He also commented that of those patients with an exceptional result after primary surgery (hearing level 10-20dB) 83% maintained this outcome over longer periods of follow-up (9).

Complications

The most frightening complications of atresia surgery are facial nerve injury and iatrogenic hearing loss, however canal restenosis, TM lateralization and chronic infection are much more common. The rate of EAC restenosis is also variable in the literature, ranging from 8-50% of cases. These figures have been found to correlate to the initial severity of the atresia with more severe cases having a higher likelihood of restenosis. If the restenosis is mild a recurrence of the conductive hearing loss is unlikely and the patient may be observed. More significant narrowing will cause epithelial trapping and predispose the patient to infection; these

cases typically will require dilation or revision canalplasty. If restenosis is limited to the lateral soft tissue portion of the EAC and is caught early it may be responsive to injection with steroid solution.

A lateralized TM presents with a gradual worsening of the hearing level and can occur up to 12 months after surgery. This problem has been reported to occur in 5-26% of cases and is best prevented by meticulous technique at the primary surgery. Taking care to create a good bony annulus upon which to anchor the graft, anchoring the graft medial to the malleus as well as using a silastic button to hold the fascial graft in place will all help to prevent TM lateralization.

Chronic infection of the newly constructed EAC may occur as a result of the lack of normal keratin migration in the skin-grafted canal and the lack of production of protective cerumen. Buildup of keratin debris and trapping of water can lead to epithelialitis and chronic otorrhea. This problem can be minimized by creating a widely patent meatus and addressing any canal restenosis. Patients should be counseled on aural hygiene and recommended to return for microscopic debridement once or twice a year (1,3,4,9).

The facial nerve follows an aberrant course in 25-30% of cases of congenital aural atresia. It is typically anterolaterally displaced in comparison to its normal course. The bend at the 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 the round window, the nerve may be lateral to the middle ear space and encased in atretic bone. Most reports in the literature say that in the hands of experienced surgeons the incidence of facial nerve injury is 1.0-1.5%. The majority of injuries reported were temporary palsies that had return to normal function over several months. Facial nerve transection and permanent dysfunction is exceedingly rare. In a review of over 1,000 surgeries for congenital aural atresia, Jahrsdoerfer and Lambert report only 10 cases of facial nerve injury. In their review, they noted five situations in which the facial nerve was most susceptible to injury: 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. Careful study of the preoperative temporal bone CT with mapping of the facial nerve course is of paramount importance to avoid this devastating complication. Intraoperative facial nerve monitoring and improved imaging techniques both help to avoid nerve injury (10).

High frequency sensorineural hearing loss has been reported to occur in up to 15% of patients undergoing atresia surgery. This occurs either as a result of transmission of drill energy to the inner ear while removing atretic bone, direct drill injury to the ossicles or traumatic manipulation of the ossicles. Although the first mechanism is largely unavoidable, the others can be prevented by using meticulous surgical technique around the ossicles. Although the hearing loss generally occurs in the 4,000-8,000 Hz range and does not affect the speech frequencies, cases that have experienced a loss in speech discrimination have been described (1,3,4,9).

Controversies

The management of the patient with unilateral congenital aural atresia remains controversial in today's literature. Historically, the teaching has been that a patient with normal hearing in one ear is capable of normal speech and language development. This idea, along with

the potential complications of atresia surgery and the unpredictability of hearing results after surgery, have lead many authors in the past to recommend against surgery in unilateral atresia cases. However, recent research has shown that children with unilateral hearing losses do suffer from auditory, linguistic and cognitive impairments that can have a negative effect on their education. Additionally, a unilateral hearing loss causes difficulty with sound localizaton and speech recognition in the presence of background noise. These impairments, along with improved methods of patient selection using high resolution CT and improved surgical techniques with more predictable hearing results after surgery, have lead some authors to begin recommending surgical repair of unilateral atresia.

If surgery is elected for the patient with unilateral atresia, the second issue of controversy is the timing of repair. Many authors would recommend postponing any surgery until the patient is old enough to participate in the decision making process and understand the potential complications and limitations of repair. More recently, several authors are recommending unilateral atresia repair at the age of 5-6 years after microtia repair, as in bilateral cases. These authors emphasize the importance of appropriate patient selection for unilateral repair, primarily reserving surgery for those patients that have favorable anatomical findings on CT scan and whose parents have realistic expectations for surgery and understand the demands of postoperative follow-up (8,11).

Conclusion

Nearly every report on congenital aural atresia begins by saying that it is among the most difficult and challenging surgeries for the otologic surgeon. That being said, in the hands of 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.

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