Otologic and Audiology Aspects of Microtia Repair

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ABSTRACT

Congenital abnormalities of the outer ear pose a reconstructive challenge for plastic surgeons and otologists. Many patients with microtia of the auricle have concurrent atresia of the external auditory canal. The hearing loss associated with canal atresia can have long-lasting effects if not treated promptly and appropriately. The diagnosis and workup for canal atresia requires an otologic evaluation. Audiologic and radiologic evaluations direct treatment, which varies depending on unilateral or bilateral presence of atresia. Rehabilitation of hearing loss can be performed via hearing aids, bone-anchored conductive devices or canalplasty. Due to the complexity of treatments, communication between the reconstructive plastic surgeon and the otologist is necessary to detect hearing loss and determine the best method of restoring hearing in conjunction with microtia repair.

KEYWORDS: Canal atresia, audiology, canalplasty, BAHA

Congenital developmental abnormalities of the external ear are divided into external ear canal stenosis/atresia and external auricular microtia/anotia. Children born with these problems face difficulties in hearing and communication, as well as the social stigma of auricular deformity. Here we focus on otologic and audiologic considerations for these patients because they are often encountered together.

CONGENITAL CANAL ATRESIA

Canal atresia is a broad term referring to a lack of the external ear canal. It describes a spectrum that also includes a stenotic ear canal. Associated with this malformation is improper development of the middle and inner ear. Therefore, a discussion of embryologic development is important to understand the need for a thorough diagnostic workup.

Embryology of the Inner, Middle, and External Ear

Otologic structures are largely derived from the first and second branchial apparatus and the otic placode. Each branchial apparatus is made up of the branchial arches, pharyngeal pouches, branchial clefts, and branchial membranes. The placode is first visible as an ectodermal thickening on the lateral aspect of the head. The placode invaginates, forms a pit, and then detaches from the surface as a vesicle. This otocyst goes on to form the inner ear membranous structures. These structures are subsequently innervated by the cochlear and vestibular nerves. As they enter the otocyst, they exert an inductive influence to produce neuroepithelium. The neuroepithelium becomes the sensory cells of the semicircular canals, saccule, utricle, and cochlea of the inner ear. These structures are enveloped by a cartilaginous capsule that develops into the petrous portion of the temporal bone.

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In the fourth week, the first pharyngeal pouch forms the tubotympanic recess. This develops into the tympanic cavity, mastoid air cells, and the Eustachian tube one week later. The Eustachian tube is critical in ventilation of the middle ear space by connecting with the nasopharynx. The first branchial cleft is the only cleft to remain as a postnatal structure. It develops into the external auditory canal and its epithelium in the fourth week.¹ As the cleft and pouch come closer, a membrane containing mesoderm is created. This tissue becomes the tympanic membrane and its middle fibrous layer is derived from the mesodermal layer. The ossicles span from the tympanic membrane to the oval window of the inner ear. The first arch cartilage ossifies to form the upper portions of the malleus and incus in the sixth week. The second arch cartilage ossifies to form the manubrium of the malleus, incus long process, and the stapes suprastructure (head, neck, and crura). It also forms the styloid process. The stapes footplate is derived from the otic capsule.

Subsequently, at 8 weeks, the primordial external canal becomes occluded by an ectodermal plug. This plug gradually resorbs as its cells degenerate. By the 28th week, the canal is fully canalized. Any perturbations to this process can lead to canal tortuosity, stenosis, or atresia. In cases of atresia, an atretic plate remains.

The external pinna is developed from a cluster of mesodermal thickenings referred to as the hillocks of His. The first branchial arch contributes to the anterior cluster and the second branchial arch contributes to the posterior cluster. The first branchial cleft separates them. Eventually, they develop into distinct parts of the auricle.

The structures of the inner ear, middle ear, external auditory canal, and auricle derive from different embryologic structures at various times of development. Therefore, it is accepted that one part may develop abnormally while adjacent structures are normal. However, it is easy to see that development of one structure can have a direct impact on the development of its neighbor. This is why a patient with an obvious microtia deformity must be thoroughly evaluated for deformities of the remainder of the external, middle, and inner ear.

Epidemiology of Canal Atresia

Canal atresia occurs in ~1 in 20,000 live births. It has been associated with many congenital syndromes, but is more often seen in isolation. It occurs more commonly in males and when unilateral, in the right ear.² Bilateral atresia occurs in around 30% of all cases. Inner ear abnormalities coexist in 12 to 50% of cases.

Diagnosis and Workup

At initial evaluation, patients can be easily divided into unilateral or bilateral atresia. A thorough history and

physical is necessary to determine contributing factors and to determine if the atresia is part of an associated syndrome or concomitant abnormalities. The external auditory canal can be graded as normal, stenotic, blindly ending, or atretic. A stenotic or blindly ending canal may elude diagnosis as it may only be appreciated on otoscopy.

An important pathologic finding in stenosis and atresia is canal cholesteatoma. Cholesteatoma is a term for trapped keratinizing squamous epithelium occurring in the temporal bone. Cholesteatoma can be recurrently infected and cause local bone destruction.

In general, the occurrence is highest in meatal stenosis (100%), moderate in partial atresia (27%), and lowest in complete atresia (4%).³

UNILATERAL ATRESIA

Patients with unilateral atresia with normal hearing on the contralateral side typically go on to develop normal speech. Because it is not regarded as a significant functional deficit, some authors advocate proposing surgery once the patient becomes an adult. Others have demonstrated consistent and successful results to restore hearing and have advocated the surgery to restore binaural hearing.

BILATERAL ATRESIA

Bilateral canal atresia is approached differently secondary to the inherent deficit of hearing of the child. Hearing is vital within the first few years of life for development of normal language. Drawing correlates from patients with congenital deafness, aural rehabilitation is initiated within the first few years of life, with many otologists performing cochlear implantation before one year of age. Delay of treatment beyond 5 years of life results in poor development of speech and language. Additionally, cognitive and social-emotional development can be affected.

Audiologic Testing

Ability to hear sound is divided into a conductive component and a sensory neural component. The conductive component relies on the external and middle ear; the sensory neural component is dependent on the inner ear cochlea and the cochlear nerve. Together, the mechanical energy of sound is converted to neural signals that are delivered to higher-level auditory processing centers of the brain.

Atresia or stenosis of the canal can present with a 45 to 60 dB hearing loss secondary to conductive impairment. Audiologic testing is therefore important to determine the presence of sensorineural hearing. The goals of a surgical intervention would be to either bypass or correct the conductive deficit. Therefore, any intervention is contraindicated if there is any sensorineural hearing loss. Alternatively, aural rehabilitation with

nonsurgical methods could be initiated quickly if a hearing loss is detected.

Auditory Brainstem Response

Audiologic testing in pediatric patients is highly dependent on the patient's ability to participate in responding to stimuli.⁴ The first line of testing used in the youngest patients is with a measurement of auditory brainstem response (ABR). A well-defined series of electric potentials can be recorded from the scalp when acoustic stimuli are presented to the ear. Waves can be observed that correlate with neurologic function beginning at the cochlear nerve and through auditory processing centers of the brainstem. These responses are not affected by sleep or sedation. Alternatively, poor patient cooperation can impede accurate results. Therefore, the testing is usually done while the patient is sedated.

Multichannel air and bone conduction ABR is used in canal atresia, especially when it is bilateral. It is particularly useful when conditions of hearing loss may make it difficult to determine the side of origin of a response. One scenario is when the patient has a large conductive deficit in one ear and a large sensorineural deficit in the contralateral ear. When measuring bone conduction, an oscillator is placed on the temporal bone and vibratory stimuli are passed through the bone. In adults, an intensity of less than 10 dB can be transmitted to the contralateral ear, eliciting a response. Fortunately, the levels are as high as 15 to 25 dB in 1-year-old infants and as high as 25 to 35 dB in neonates, making it easier to determine the side of a response.⁵

Visual Reinforcement Audiometry

Visual reinforcement audiometry (VRA) is a mode of audiologic testing used on patients between 5 to 6 months of age. At this age, patients begin to respond to sounds by turning their head laterally. VRA relies on operant conditioning techniques where head turning toward sound stimuli is reinforced with a pleasant visual event, such as seeing a toy. The conditioning delays habituation to the sounds and enough stimuli can be presented to determine a threshold of sound intensity. This technique can determine ear-specific responses if earphones are used and can also determine bone conduction. Infants who are 12 months old generally have more accurate and reliable results.

Conditioned Play Audiometry

As children get older, conditioned play audiometry (CPA) becomes the preferred testing modality. With CPA, the child is taught a motor skill in response to detecting a sound stimulus. Tasks include dropping a building block into a bucket or placing a peg into a board.

By varying the motor tasks, audiologists can present enough stimuli to determine both air and bone thresholds for a child beginning between 2 to 3 years of age.

Pure Tone Audiometry

Conventional measures of hearing involve multiple tests, but the most commonly used is the pure tone average. This test involves presenting sound stimuli to each ear via air conduction or bone conduction at various frequencies and intensities. The patient then responds to a detected stimulus by pressing a button or raising their hand. The intensity level where half of the sounds are heard determines a threshold. Generally, patients as young as 5 years old can cooperate to successfully undergo this testing. Other aspects of conventional testing that are useful are tympanometry and acoustic reflexes.

Radiographic Imaging

Imaging studies are important for all children with canal atresia. It provides valuable information regarding the development of the middle and inner ear. Noncontrast temporal bone computed tomography (CT) scanning is the preferred study. The most important prognostic factors for a good outcome are the size of the tympanic cavity, pneumatization of the mastoid, the status of the ossicles, and the course of the facial nerve (Fig. 1). Generally, a CT is performed around age 4, once the middle ear and mastoid are fairly aerated and the patient is close to potential surgery. It could be performed earlier to rule out a canal cholesteatoma.

Various grading systems have been developed to categorize the severity of atresia, but the system proposed by Jahrsdoerfer has given surgeons the ability to determine the likely success of performing an atresia reconstruction (Table 1).⁵ When considering a canal-plasty for unilateral atresia, surgery is reserved for patients who are highly motivated and have a good to excellent prognosis based on radiographic findings. The threshold for offering a surgical correction is much lower for patients with bilateral canal atresia.

TREATMENT

It is important to understand that not all treatment for congenital canal atresia involves a surgical intervention. Additionally, when surgeries are considered, there are usually multiple stages required to restore hearing successfully, which is in conjunction with the multiple stages usually required for microtia repair.

Nonsurgical Treatment

Hearing aids should be considered for any child who demonstrates a permanent bilateral hearing loss exceeding

Figure 1 Coronal computed tomography scan of the temporal bone in a patient with canal atresia. Note the thick atretic plate where the external auditory canal is usually found.

20 dB Hz between 1000 and 4000 Hz. Bone conduction hearing aids are used since the canal atresia precludes the use of standard hearing aids. Ideally, they should be initiated within the first few months of life. Bone conduction hearing aids are similar to conventional air hearing aids, except they have an oscillator that replaces the speaker. The oscillator is pressed firmly to the skin over the mastoid via a metal or elastic headband. Vibrations are then passed through the bone where they are detected by the cochlea and processed as sounds. If the normal-appearing ear is found to have a hearing defect, then behind the ear hearing aids are usually recommended. As the patient and their external ear canal grows, the molds needed to fit into the canal can be replaced quickly and at a reasonable cost.

Table 1 Jahrsdoerfer Grading System for Atresiaplasty Candidacy

Parameter	Points
Stapes present	2
Oval window open	1
Middle ear space	1
Facial nerve normal	1
Malleus-incus complex present	1
Mastoid well pneumatized	1
Incus-stapes connection	1
Round window normal	1
Appearance of external ear	1
Total available points	10
Rating	Type of candidate
10	Excellent
9	Very Good
8	Good
7	Fair
6	Marginal
≤5	Poor

Surgical Treatment

CANALPLASTY

Canalplasty is a surgical procedure aimed at restoring the conduction of mechanical sound energy. Normal transmission of sound requires an intact tympanic membrane, mobile and intact ossicular chain, and transmission of energy to the stapes footplate that overlies the oval window. The oval window is in communication with the cochlea where mechanical energy is converted to neural impulses. Therefore, the surgery requires creating bony canal, repairing any ossicular malformations, repairing the tympanic membrane, and creating a communication through the skin to the canal and lining it with squamous epithelium.

Historically, these surgeries were complex with variable hearing results. Therefore, canalplasty was offered at an early age for bilateral canal atresia patients and reserved until adulthood for unilateral atresia patients. Over time, audiologic and diagnostic testing proved beneficial in selecting patients with favorable anatomy and advances in surgical techniques led to more consistent and improved results. Therefore, patients with unilateral atresia were offered canalplasty during childhood.

Unilateral atresia is usually repaired at around 6 years of age. The timing of the intervention has been linked to the timing of any planned microtia repair. The main factors affecting the timing are sufficient time for rib cartilage development to create a new auricular framework balanced by the initiation of schooling where the patient may be subjected to teasing because of the deformity. Bilateral atresia repair may be initiated earlier. However, repair of the microtia before the age of 5½ years is technically more difficult⁶ and some authors recommend waiting until age 5 for proper pneumatization of the middle ear and mastoid.³

Surgical planning involves good communication between the plastic surgeon and otologist. The otologist focuses on the reconstruction of the external ear canal and the middle ear. Traditionally, reconstruction of a microtia/atresia involves a five-stage approach. The canalplasty is delayed until the third stage. Therefore, the cartilaginous framework is implanted and the lobule is created before the canalplasty is attempted. The rationale for this order is to preserve an unoperated field for the microtia and to avoid needing to center the framework over the drilled-out canal.

Two Approaches

ANTERIOR APPROACH

A postauricular incision is created behind the auricle or the cartilage framework if microtia reconstruction had been initiated. Then, the tympanomandibular joint and the floor of the middle cranial fossa (referred to as the tegmen plate in this region) serve as anterior and superior boundaries for a bony circular canal directed medially. The facial nerve generally courses more anterolaterally than typically encountered. Thin bone can be left over these structures and will guide drilling through the atretic plate. Care must be taken to avoid transmitting vibratory energy from the drill to the ossicles as this may result in sensorineural hearing loss. Once the middle ear is opened, visualization and palpation of the ossicular chain can help identify sites of fixation or discontinuity.

An ossiculoplasty is then performed, where a prosthetic device is used to replace one or more bones or restore continuity to the chain. Next, fascia from the temporalis muscle can be used to reconstruct the tympanic membrane. Finally, the bony canal needs to be lined with epithelium and made to communicate with the outside world through the meatus. A meatoplasty is performed by incising the skin of the concha and debulking the soft tissue underneath to allow an anteriorly based flap to line the anterior canal wall. The remaining bone is then lined with a split thickness skin graft. Packing material is then placed to hold the grafts and flaps in appropriate position. The material is kept in place until epithelialization is complete.

TRADITIONAL MASTOID APPROACH

In the mastoid approach, the middle ear is approached from posterior dissection through the aerated mastoid bone. The surgeon is directed by the sigmoid sinus posteriorly and the tegmen plate superiorly until the antrum is reached. This cavity is a stable landmark; it is where the mastoid air cells and the tympanic cavity communicate. From this point, the stapes and incus can be disarticulated to avoid vibratory trauma to the cochlea, and the facial nerve can be identified. These

landmarks allow the atretic bony plate to be removed; the surgery mirrors a canal wall down mastoidectomy. A fascia graft can be placed over the stapes and a small middle ear space is thus created. A meatoplasty will then allow the new mastoid bowl to communicate with the outside and epithelization can occur from the skin flaps. Because this cavity cannot clear keratin debris, routine cleaning by an otolaryngologist will be necessary throughout life.

COMPLICATIONS AND SUCCESSFUL OUTCOMES

The most dreaded complication of canalplasty is injury to the facial nerve. This is particularly high if the nerve has a greatly abnormal course and if the aeration of the middle ear and mastoid is poor. Most otologists have adopted the use of intraoperative neurophysiologic monitoring of the facial nerve to assist in its identification. Audiologic success is commonly measured by achieving a speech reception threshold of 25 dB or lower. Generally, this could be achieved in ~75% of patients if stringent radiographic selection criteria are used to determine favorable patients. Delayed complications of canal stenosis or tympanic membrane graft lateralization can lead to worsening hearing months to years after an initially successful surgery.

Bone-Anchored Hearing Aids

Bone-anchored hearing aids (BAHA) consist of a small sound processor clipped to a percutaneous abutment, which is attached to an osseointegrated titanium implant. Sounds are detected by a microphone, processed, and then transmitted by an oscillator to the abutment. The vibration is transmitted directly to the cochlea through the bone of the skull base. The initial Food and Drug Administration (FDA) indication was for conductive and mixed hearing loss and the technique is used where traditional surgery is unlikely to be successful or feasible. For patients that are considered poor candidates for canalplasty surgery, BAHA has become a reasonable alternative.

Prior to surgery, the expected subjective and objective benefit from the BAHA can be demonstrated using a bone conduction hearing aid. If the patient does not appreciate any gain in hearing quality, the surgery could be deferred. The titanium abutment is placed into the temporal bone, generally posterior and superior to the anticipated location of the external auditory canal (Fig. 2). The surgery entails raising a split-thickness skin flap and removing all subcutaneous tissue while preserving the periosteal layer of the skull. Next, the periosteum is incised, a hole is drilled in the bone, and an abutment is screwed into the hole. The skin graft is replaced over the surgical defect and it is punctured to allow the

Figure 2 Microtia and canal atresia patient demonstrating the typical location of a BAHA abutment. Generally, the abutment and sound processor are well hidden if the patient has long hair.

abutment through. Many variations have been described in the procedure, including the need for a second stage. Several weeks are needed before it can be used in adults and a slightly longer time is needed in children.

Despite FDA approval of use beginning at 5 years of age, many articles report its successful use in younger patients. Advantages over the bone conduction hearing aid include avoidance of skin problems associated with chronic pressure, better sound quality, and improved high-frequency hearing because the vibratory signal is more directly transmitted to the bone. Complications with this technique generally involve skin-healing issues. Because of the need for a skin graft, issues such as graft loss or hypertrophic scarring and abutment overgrowth can sometimes delay use of the hearing aid device.

CONCLUSIONS

Congenital canal atresia is a serious malformation that is commonly associated with microtia of the auricle. These

patients need a multidisciplinary approach to achieve a functional and cosmetically acceptable reconstruction. Patients with a noticeable deformity of external auditory canal development require prompt evaluation by an otologist. The patient can then be evaluated both audiotically and radiologically for conditions that may need prompt intervention. If a patient is found to have a bilateral hearing deficit, immediate aural rehabilitation is needed to avoid sequelae such as delayed language development. Conservative measures such as bone conduction hearing aids and traditional hearing aids can help dramatically. When the patient is of proper age, surgical options such as a BAHA or canalplasty can permanently rehabilitate a conductive hearing loss. The decision-making algorithm is highly variable and depends on the surgeons, the family, and ultimately the patient. Overall, despite continued controversies, there are many options to restore functional hearing to these patients.

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