Atresiaplasty in Congenital Aural Atresia

What the Facial Plastic Surgeon Needs to Know

Douglas S. Ruhl, MD, MSPH, Bradley W. Kesser, MD*

KEYWORDS

• Atresia • Microtia • Otologist • Congenital aural atresia • Atresiaplasty • Canalplasty • Atresia surgery • Conductive hearing loss

KEY POINTS

- Evaluation of hearing early in life is essential in patients with congenital aural atresia so options for early auditory habilitation can be presented to the family.
- Bone conduction hearing devices are a necessity in children with bilateral microtia/atresia to support normal speech and language development.
- Microtia surgery with rib grafting should precede canalplasty; however, canalplasty should precede microtia repair when the Medpor or SuPor implant is used.
- Surgeons must consider the possibility of an aberrant or superficial facial nerve in these patients.
- It is imperative that the reconstructive surgeon and the otologist work together to coordinate care.

INTRODUCTION

Patients with congenital aural atresia (CAA) present with a spectrum of severity that ranges from ear canal stenosis to complete absence of the external auditory canal. This rarely occurs in isolation, but is more commonly associated with microtia or other craniofacial dysplasias (see later). The middle ear and ossicles are affected to varying degrees.[1] It has been estimated that CAA occurs in 1 out of 10,000 to 20,000 births.[2] Males are more commonly affected than females, and atresia affects the right ear more frequently than the left ear. Unilateral atresia is much more common than bilateral atresia (3- to 7-fold difference).[1,3] Approximately 10% of patients with CAA have an associated syndrome including Treacher Collins, Goldenhar, hemifacial microsomia, branchio-otorenal syndrome, de Grouchy, and Crouzon, so a careful physical examination is important during the initial visit.[3]

Patients with microtia and atresia may face multiple surgeries to correct the malformation. Ideally, children should be evaluated at a multidisciplinary center shortly after birth. These children require the services of an audiologist, otologist, reconstructive surgeon, speech therapist, and possibly a developmental pediatrician. The parents and child face many difficult decisions. How will they address the hearing problem? Should they correct the microtia? Should the child have canalplasty surgery? When should one order the computed tomography (CT) scan to evaluate for canal surgery? Who should do these surgeries, and at what age?

Disclosure Statement: The authors have nothing to disclose.
Department of Otolaryngology–Head and Neck Surgery, University of Virginia School of Medicine, PO Box 800713, Charlottesville, VA 22908, USA
* Corresponding author.
E-mail address: bwk2n@virginia.edu

Facial Plast Surg Clin N Am 26 (2018) 87–96
https://doi.org/10.1016/j.fsc.2017.09.005
1064-7406/18/© 2017 Elsevier Inc. All rights reserved.

This article discusses several important aspects of atresia surgery that the reconstructive surgeon should be aware of to ensure that these patients have comprehensive care with optimal outcomes.

ASSESSING HEARING

When a child is diagnosed with microtia/atresia, usually in the newborn nursery, the hearing should be assessed as early as possible with air and bone conduction auditory brainstem response testing. Auditory brainstem response testing is mandatory in the postnatal period to determine hearing thresholds in both ears.[4] Typically, the atretic ear has a moderate to severe conductive hearing loss with air conduction thresholds in the 50- to 65-dB hearing level range and normal bone conduction thresholds (0- to 10-dB hearing level). In patients with unilateral microtia/atresia, the hearing status of the contralateral (normal) ear is equally, if not more important. It should not be assumed that the hearing is normal in the nonatretic ear just because the external ear looks normal. Air conduction auditory brainstem response testing should be performed in the contralateral, normal ear to verify normal hearing.

When the child is older, behavioral audiometry with air and bone conduction testing should be done to continue to monitor hearing status and cochlear integrity. Children between ages 1 and 3 can be tested behaviorally with visual reinforced audiometry and those ages 3 to 6 with conditioned play audiometry.[3] Testing is generally recommended every 3 to 6 months for the first 2 years of life, and if the hearing is stable, every 6 to 12 months thereafter.

HEARING HABILITATION

Based on audiometric testing results, options are presented to the family for hearing habilitation. Use of a soft or hardband bone conducting hearing device is a necessity for normal speech and language development in children with bilateral CAA. In children with unilateral CAA, the data are not as convincing about the benefits of a bone conducting device. Although normal hearing in one ear is sufficient for normal speech and language development, the long-term effects of aiding versus not aiding the atretic ear (with a bone conducting device) are unknown. Parents should never be discouraged from having their child with unilateral CAA use a bone conductor, but the necessity or the benefits of a bone conductor in a child with unilateral CAA remains under investigation.[5]

In children with bilateral CAA, the bone conducting device should be fit as early as possible and worn until the child is at an age where additional options (eg, canal surgery; osseointegrated bone conducting device, such as the BAHA system [Cochlear Americas, Englewood, CO] or Ponto [Oticon Corp, Somerset, NJ]) are available. During the interim, however, it is important to ensure that adequate auditory input is presented during the child's critical language development period between the ages of 0 and approximately 8.

Older children have several options regarding hearing habilitation. For children with unilateral CAA, parents may elect to do nothing and simply monitor the hearing and speech and language development, providing benefits in the classroom, such as preferential seating, a frequency modulated (FM) system, or other resources through an individualized educational plan (IEP).[5] Implantable, osseointegrated bone conducting devices are an excellent option for patients with bilateral CAA, if the family does not wish to pursue canalplasty, or for patients who are poor candidates for surgery.[6] The hearing outcomes of these devices are consistently excellent, often equivalent or better than the audiometric results gained from canalplasty.[6–9] However, these devices have some drawbacks. The Food and Drug Administration prohibits implanting patients less than 5 years old. These devices do not support good sound localization. Some patients find the device cumbersome, and they can have wound care issues, such as inflammation, granulation, moisture at the site, or skin overgrowth. These skin issues could be a significant problem for the reconstructive surgeon if the device is placed too close to the surgical field for the auricular reconstruction. Communication between otologist and reconstructive surgeon is critical so that the osseointegrated bone conducting device is placed in a favorable position for auricular reconstruction.

Atresia surgery, when performed properly and in the appropriate patient, can achieve outstanding hearing results. Even if hearing results decline over time, having an external auditory canal can allow the patient to wear a traditional in-the-ear hearing aid, which may bring the hearing into the normal range.[8]

Parents hunger for solid, reliable information about microtia/atresia, and early consultation with an experienced reconstructive surgeon and otologist starts their journey off on the right track even if nothing, other than monitoring the hearing, needs to be done in the 0- to 4-year-old child's life. After the early auditory needs have been met, the child's candidacy for auricular and/or atresia surgery is undertaken around the age of 4 to 5.

Download English Version:

https://daneshyari.com/en/article/8806067

Download Persian Version:

https://daneshyari.com/article/8806067

Daneshyari.com