Otologic and Audiology Concerns of Microtia Repair

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Semin Plast Surg 2017;31:127–133.

Abstract

Keywords

- ▶ microtia
- ▶ aural atresia
- ▶ ear reconstruction
- ▶ atresioplasty
- ▶ hearing rehabilitation

Microtia is a congenital auricular deformity that commonly presents with associated congenital aural atresia. The most acute concern in these patients is concomitant hearing loss at birth. A team-based approach by plastic surgeons and otologists is necessary to address both the otologic and audiology concerns of microtia and atresia. Hearing rehabilitation is imperative; yet it should not compromise the aesthetic components of reconstruction and vice versa. Here, the authors propose a framework to evaluate and manage patients with microtia and atresia with the goal of optimizing functional and cosmetic outcomes.

Microtia is a congenital deformity of the pinna that frequently occurs in conjunction with a spectrum of other abnormalities. Congenital aural atresia is the most commonly associated condition that involves an underdeveloped auditory canal and malformed middle ear ossicles. Given the irregular external and middle ear anatomy, audiology impairments plague children with microtia and atresia. Children who lack serviceable hearing encounter significant developmental delays in speech, language, cognition, and socialization. The social stigma associated with visible ear deformities also generates significant psychoemotional concerns. To avoid these long-lasting consequences, the restoration of hearing is the primary focus of microtia repair and atresioplasty. However, attention should also be given to the aesthetic component of repair without compromising hearing rehabilitative options. For these reasons, microtia with atresia remains one of the most complex otologic problems to address as it involves appropriately timed integration of hearing rehabilitation with auricular reconstruction.

Diagnostic evaluation is crucial in determining the severity, laterality, and associated abnormalities of microtia. Through audiology testing and imaging of temporal bone morphology, a surgeon can determine surgical candidacy and devise a cohesive plan for repair. Children with microtia and their parents must be counseled in advance regarding

the optimal timing of reconstruction and the possible need for adjuvant conductive hearing aids.

In a team-based approach driven by plastic surgeons and otologists, children can achieve both hearing restoration and successful aesthetic results. Here we outline the evaluation process of microtia and atresia with specific considerations of concomitant hearing loss. Our purpose is to provide a framework for managing microtia and atresia to achieve desirable audiology and cosmetic outcomes.

Ear Development

The ear consists of three distinct parts: the outer, middle, and inner ear. Otologic structures from the external and middle ear originate from the first and second branchial arches in embryonic development. The external ear forms in the sixth week of gestation just prior to the formation of the middle ear.¹ The ossicular chain consisting of the malleus, incus, and stapes, develop at approximately 6 to 8 weeks gestation and are adult-sized by the time of birth.² The tympanic membrane has also matured by full-term gestation.² Pneumatization of the mastoid bone and growth of the external ear continue after birth until completion at approximately 5 to 6 years of age.³ The inner ear structures, the cochlea and vestibular system, are derived from separate embryonic

Issue Theme Ear Reconstruction; Guest Editors: Tuan A. Truong, MD, and Renata S. Maricevich, MD

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Tel: +1(212) 584-4662.

DOI <https://doi.org/10.1055/s-0037-1603957>.
ISSN 1535-2188.

derivatives than the middle and external ear and send sensorineural projections to the brain for interpretation of auditory signals.¹

A disruption at any point in ear development can result in otologic malformations with earlier arrests in development resulting in more severe defects. Because the external ear forms before the middle ear, the presence of microtia indicates likely subsequent malformation in the middle ear ossicular chain and dysplasia of the tympanic membrane.³ In fact, the severity of microtia has been shown to inversely correlate with how well-developed the middle ear is.^{4,5} In severe cases, the ossicles may even be fused together and to the atretic bony plate, preventing sound vibration. Ultimately, auricular and external auditory canal development must be tightly coordinated with middle ear formation to obtain normal hearing functionality.

Because the inner ear is derived from separate embryonic structures than other ear components, most patients with microtia have normal inner ear anatomy. Hence, sensorineural transmission of auditory signals through the cochlea is intact. In fact, 85% of hearing deficits in microtia and atresia are conductive in nature as a result of bony middle ear abnormalities and a stenotic ear canal, whereas the rest are of the sensorineural or mixed type.⁶

Epidemiology and Associated Anomalies

The prevalence of microtia is approximately 1 to 20 in 10,000 births worldwide, occurring almost twice as frequently in males and with incidence higher in the Hispanic, Native American, and Asian populations.⁷ Microtia is unilateral in 60 to 70% of cases, most commonly affecting the right side.³ Microtia rarely occurs in isolation because of subsequent malformations of the middle ear in later stages, whereas isolated aural atresia has been known to occur in otherwise normal-appearing ears.³

All but the mildest forms of microtia have some degree of conductive hearing loss secondary to aural atresia and ossicular chain disruptions. Bilateral microtia cases are also associated with a higher rate of other abnormalities, such as facial clefts, microphthalmia, mandibular hypoplasia, holoprosencephaly, vertebral anomalies, and polydactyly.⁸ Isolated microtia is considered the mildest condition in the spectrum of hemifacial microsomia, which results from malformations in the first and second branchial arch structures.² When microtia does appear to have a genetic component, it is most frequently in the form of Treacher Collins syndrome, where it presents as bilateral microtia and is an autosomal dominant trait.^{7,8}

Environmental and genetic factors play an intertwining role in causality of microtia, including teratogenic medications (i.e., thalidomide, retinoic acid), in utero vascular disruption, and inherited syndromes.^{7,8} Anomalous development of the first and second pharyngeal arches in Goldenhar's syndrome and Treacher Collins syndrome result in anotia and microtia.² Single gene disorders, such as branchio-otic syndrome and branchio-oto renal syndrome, are also associated conditions.^{7,8} Microtia can also be a clinical

feature of autosomal trisomies due to chromosomal aberrations disrupting normal organogenesis.^{7,8}

Diagnostic Workup

Malformed ears are diagnosed at birth and should undergo global assessment of both structural and functional abnormalities. Identifying hearing deficits are especially important as this can alter the management approach. To address these functional concerns, evaluation involves three essential components: a comprehensive examination of the ears, bilateral audiologic testing, and radiographic imaging.

Initial and Preoperative Examinations

A thorough history identifies predisposing factors that contribute to the child's ear deformity, especially familial syndromes or teratogen exposure.⁹ On examination, ear vestige size, shape, rotation, canal position, and facial symmetry are noted. Otoscopic visualization of the ear canal and tympanic membrane is required in microtia patients due to the strong correlation in abnormalities of these structures.⁴ The morphology of the mandible, eyes, oral cavity, and spine are also evaluated to rule out associated anomalies such as hemifacial microsomia.¹⁰ In syndromic cases, a genetic evaluation may be indicated, especially for congenital sensorineural hearing loss that cannot be repaired by microtia repair and atresioplasty.

Based on the examination, a microtia grade is assigned using any of the historical staging systems (►Fig. 1). The Marx¹¹ classification system is a popular method describing the severity of deformity (►Table 1). High-grade microtia is considered grades III–IV and is useful when determining surgical candidacy for atresioplasty. The Nagata¹² system uniquely classifies microtia based on the surgical correction of the deformity (►Table 2).

During preoperative evaluation, the normal ear is traced onto a clear X-ray film as a template for reconstruction of the malformed ear. The distance between the helical rim and mastoid in the normal ear helps approximate the projection of the reconstructed ear. The relationships of the ear to the nose, lateral canthus, and oral commissure are useful landmarks to symmetrically align the reconstructed ear.

Audiologic Assessment

A child with any ear deformity should undergo comprehensive hearing diagnostics within the first few months of life. Auditory brainstem response testing is the most accurate tool as it delineates the degree and type of hearing loss specific to each ear.¹³ Most patients with microtia and atresia are diagnosed with a conductive hearing deficit of 40 to 60 dB.¹⁴

Even if microtia and atresia appear unilateral, an audiogram of the contralateral ear is necessary to confirm normal sound conduction. Particularly in cases of unilateral microtia and atresia, protection of the better hearing ear is crucial to prevent hearing impairment and assist in the development of appropriate speech. Identifying any degree of hearing loss allows early intervention with conductive hearing aids until corrective surgery can be performed. After initial examination, patients with atresia are followed every 6 to 12 months

Fig. 1 Right microtia classified as grade III type (Marx system) or lobular type (Nagata system).

to re-evaluate acoustic function and ensure preservation of hearing in the normal ear.¹⁵ Based on the laterality and severity of the hearing loss, surgeons can better tailor their management approach to incorporate hearing amplification devices with planned future ear reconstruction.

Radiographic Imaging

Preoperative imaging of temporal bone morphology is performed at approximately 5 years of age to determine surgical candidacy.¹⁶ Earlier imaging may not be accurate as pneumatization of the mastoid bone and maturation of the external ear is still ongoing, thus unnecessarily providing radiation exposure to the child's brain.

A high-resolution computed tomography (CT) scan in coronal and axial views is the preferred method for imaging. The important structures to visualize include the outer ear canal, ossicles, middle ear aeration, facial nerve, and mastoid pneumatization.^{9,17} The general direction of drilling through the temporal bone can be plotted for atresioplasty.¹⁷ CT also rules out a congenital cholesteatoma, which can cause local destruction of bone and complicate atresia repair.¹³

The Jahrsdoerfer's¹⁸ atresia classification system based on CT findings is popularly used to predict the likelihood of significant hearing improvement after atresioplasty (→ **Table 3**). A score greater than 7 suggests an 85 to 90% chance for significant postoperative hearing restoration, whereas a score less than 5 essentially disqualifies patients from surgery.^{18,19} For example,

Table 1 Marx staging system for microtia

Grade	Description
Grade I	Abnormal auricle with all landmarks identifiable
Grade II	Abnormal auricle with only some landmarks identifiable
Grade III	Very small auricular tag
Grade IV	Anotia, or complete absence of external ear

patients with Treacher Collins syndrome or hemifacial microsomia rarely obtain a score above 6; as such they are poor surgical candidates because they gain minimal hearing post-operatively.¹⁹ This predictive scoring system highlights that successful microtia repair and atresioplasty is measured by not only cosmetic results, but also the restoration of hearing to near normal range.

Management of Microtia and Atresia

Microtia repair fundamentally encompasses two components: cosmesis and acoustic functionality. Improvement of the aesthetic appearance of the ear, even in minor cases of microtia, can tremendously uplift a child's self-esteem. In patients with associated hearing loss, ear reconstructive approaches become even more comprehensive by incorporating techniques that address functional hearing issues. Ultimately, auricular construction for aesthetic improvement goes hand-in-hand with hearing rehabilitation. Treating functional deficits should not compromise aesthetic reconstructive options and vice versa. Through extensive collaboration between plastic surgeons and otologists, children with congenital microtia have the opportunity for both hearing restoration and good aesthetic

Table 2 Nagata classification system for microtia

Type	Description
Anotia	Lack of auricular tissue or minute ear remnant
Lobular	Ear vestige with lobule and helix; no concha, auditory meatus, or tragus
Conchal	Ear vestige and lobule with concha, auditory meatus, and tragus
Small conchal	Ear vestige and lobule with small indentation instead of concha
Atypical	Cases that cannot be characterized by other types

Table 3 Jahrsdoerfer's grading system of candidacy for atresiaplasty

Parameter	Points
Stapes bone present	2
Oval window open	1
Middle ear space	1
Facial nerve	1
Malleus/incus complex	1
Mastoid pneumatization	1
Incus-stapes connection	1
Round window	1
External ear appearance	1
Total possible score	10

Note: 10: Excellent; 9: Very good; 8: Good; 7: Fair; 6: Marginal; ≤ 5 : Poor

outcomes. Appropriate family counseling is crucial to manage expectations and explain the need to delay treatment until the optimal age for successful results.

Goals and Indications of Repair

The primary goals of auricular repair and atresiaplasty are to rehabilitate acoustic function to near normal status and improve cosmetic appearance.^{9,16} Hearing restoration and ear reconstruction can then secondarily improved speech, school performance, social acceptance, and psychoemotional development.^{9,16}

Indications for surgical microtia repair depend on the severity of the deformity and associated anomalies. Patients with high-grade microtia (grades 3–4), a Jahrsdoerfer¹⁸ score less than 5, abnormal cochlear function, and no apparent middle ear space are not good candidates for atresiaplasty as corrective surgery would unlikely benefit hearing.¹⁹ Significant inner ear abnormalities, best seen on preoperative magnetic resonance images, are also contraindications to functional surgery.^{9,19} Hence, these patients are managed nonoperatively prior to age 5 with bone conductive hearing devices on the soft band. After age 5, there is sufficient skull thickness to support osseointegrated implants like the bone-anchored hearing aid (BAHA) or Ponto.

Options for Microtia Repair

There are several options for microtia reconstruction, with the main three consisting of autologous reconstruction with autologous rib, composite reconstruction using an alloplastic framework, and finally prosthetic reconstruction. As the initial alloplastic material used was frequently silastic which demonstrated a high rate of extrusion, autologous reconstruction has been traditionally favored. Recently, however, the use of porous polyethylene frameworks such as Medpor have demonstrated integration with good aesthetic results without donor-site morbidity, and are thus being explored as a possible alternative to the traditional autologous reconstruction with rib graft.²⁰

Stages of Microtia Repair in Autologous Reconstruction

Historically, the staged approach to autologous ear reconstruction was first described by Tanzer, then modified by Brent²⁰ and Nagata, and further refined by Firmin.^{12,21,22} Although a detailed description of surgical techniques is beyond the scope of this review, recognition of each stage is helpful in clarifying the best time to incorporate atresiaplasty during auricular reconstruction.

Brent's²² reconstructive approach is considered at age 6, performed in four stages that are spaced approximately 3 months apart. Stage I includes fabrication and placement of a costochondral graft in the subcutaneous pocket adjacent to the ear vestige. Stage II involves lobule transposition. In stage III, the cartilage framework is elevated, and a split-thickness skin graft is used to cover posterior defects. Symmetry adjustments, tragus reconstruction, and conchal excavation complete ear reconstruction in stage IV.²²

The Nagata¹² modification condenses the number of operations into two stages: The first stage combines cartilage graft placement with lobule transposition and tragus reconstruction; the second stage elevates the framework and closes posterior defects with a temporoparietal fascia flap.

Technical Overview of Atresiaplasty

Atresiaplasty is a technically challenging procedure generally requiring the skills of the most-experienced otologic surgeons. Atresia repair involves reconstruction of the external auditory canal and the tympanic membrane, as well as the middle ear ossicular chain if necessary, all for the purpose of gaining audiological function.^{17,23} First, a postauricular incision is made to dissect down to the mastoid periosteum, taking care to avoid exposure of the previously implanted rib cartilage. A canal is drilled through the temporal bone with care to preserve the facial nerve. The ossicular chain of the middle ear is exposed and repaired if possible. The tympanic membrane is then reconstructed using fascia harvested from the temporalis muscle. A split-thickness graft from the scalp or arm is then used to line the external canal. Finally, the auditory canal is aligned with the conchal bowl of the reconstructed ear, suturing the skin lining each portion to exteriorize the auditory meatus. The canal is packed with antibiotic-soaked sponge and silicone sheeting and replaced several weeks postoperatively.^{17,23}

Timing of Auricular Reconstruction and Atresiaplasty

Optimal timing of microtia repair is primarily influenced by the maturity of the external ear and adequacy of donor rib cartilage. The outer ear continues to grow after birth, reaching approximately 85% of its maximal size by age 5.² Additionally, donor rib cartilage is typically not adequate in volume or stability until ages 5 to 6.¹⁶ For these reasons, autologous costochondral reconstruction was traditionally considered at ages 6 to 7 as proposed by the Brent technique.²² The best age for good reconstructive potential must also be balanced with early intervention to reduce psychological anguish from a craniofacial deformity. The Brent²⁰ method, which can be performed at ages 5 to 6, maintains this balance, whereas the Nagata¹² technique does not as it is performed later at age

10 due to the need for a more rigid construct and thus sturdier donor cartilage. By this time, significant psychological morbidity may have already occurred.

In unilateral conditions, the normal ear is used as a template to design the dimensions of the reconstructed ear. If repair is performed prior to 6 to 7 years of age, the reconstructed ear may eventually appear smaller, rotated, and abnormally positioned once the normal ear has finished growing. Psychological concerns occasionally resurface in these children as they age, often requiring a revision procedure to rectify asymmetry.

In bilateral cases of microtia/atresia, reconstruction should be staggered, with the first ear repaired at approximately 6 years of age. If a reconstructed ear is not given enough time to heal, the shape of the graft framework is jeopardized when the child's head is manipulated for the contralateral repair.¹⁶

If auricular reconstruction is planned, atresioplasty is usually incorporated into this timeline for better aesthetic results. From a surgical perspective, atresioplasty is delayed until after the initial two stages of microtia reconstruction (in Brent technique, or first stage in Nagata technique) are complete.²³ This is due to the fact that the cartilage graft has to be placed subcutaneously under a pocket of mastoid skin. If the overlying skin has been manipulated by prior atresia repair, the scarred tissue may not be pliable or perfused well enough to contour over the grafted cartilage. Thus, atresia repair is performed approximately 3 months after framework placement and lobule transposition in conjunction with the latter stages of auricular reconstruction.^{17,23}

Atresioplasty performed during the final stages of microtia repair involves the harmonization of surgical techniques; the plastic surgeon elevates the cartilage framework while the otologic surgeon drills the bony canal and repairs the middle ear.¹⁴ The skin flap over the concha is then raised to create an exterior opening for the auditory canal and sutured to the skin graft lining the canal.¹⁴ A disadvantage of this coordinated

sequence is that the reconstructed canal may have suboptimal fitting with the concha of the already placed ear framework.

To achieve desirable results from auricular reconstruction and atresioplasty, the plastic surgeon and otologic surgeon must plan several aspects of the procedure in advance. This includes the timing of procedures, the positioning of the ear canal in relation to the auricular construct, the vascular supply of flaps, surgical incision sites, and the positioning of conductive hearing devices. For instance, postauricular incisions used to place bone-conducting implants can inadvertently compromise possible skin grafts for later reconstructive stages.²⁴ Surgeons must also plan the best positioning of implants, which Bajaj et al²⁵ found to be approximately 7 cm posterior to the external auditory meatus to produce good acoustic functionality, while preserving tissue needed for reconstruction.

For these reasons, surgeons must reassure patients and their families of the benefits of waiting until the optimal age of 6 to 7 years of age for auricular reconstruction and atresioplasty. Extensive discussion regarding postoperative wound care, head bandages, drains, and activity restrictions increase compliance to prevent devastating complications and revision procedures. When children reach this age for microtia repair, they are usually aware of their condition and mature enough to cooperate with the substantial postoperative care required.

Hearing Rehabilitation Options

Autologous ear reconstruction improves aesthetic outcomes, but does not address the functional hearing issues in microtia and atresia. There are various options available for hearing rehabilitation in microtia and atresia, such as conventional bone conducting aids, osseointegrated implants, and atresioplasty. The decision to use a particular method depends on the timing, the severity of the deformity, and the laterality of hearing loss (—Fig. 2).

In unilateral microtia and atresia, hearing loss is isolated to the affected ear. The contralateral ear typically has a good

Fig. 2 Proposed hearing rehabilitation for cases of microtia and atresia based on severity and laterality. This is a simplified approach and is not an exhaustive list of options available to patients. Osseointegrated implant includes bone-anchored hearing and Ponto devices.

range of hearing, so children usually develop normal speech patterns.⁶ Nevertheless, children with unilateral ear conditions can have loss of directional sound and impaired speech perception in noisy environments.²⁴ Efforts to preserve hearing in the normal ear, such as prompt treatment of otitis media, are necessary to avoid further exacerbation of hearing loss. Recent studies also recommend fitting a bone conductor on an elastic band (also known as a soft band) in patients with unilateral microtia and atresia starting as young as 6 months old.^{6,24} Hearing amplification promotes superior binaural hearing, thus enabling better school performance.¹⁰ Once patients reach approximately 6 years old, they can undergo auricular reconstructive surgery with atresioplasty and/or implantation of BAHA for excellent cosmetic and hearing results.^{16,26} Corrective surgery in unilateral cases can be delayed until adolescence or adulthood, but the benefits of enhanced binaural hearing versus the risk of postoperative stenosis and degradation of hearing should be weighed.

Patients with bilateral microtia and/or atresia are usually functionally deaf bilaterally. Hearing amplification early in life is imperative to prevent poor speech and social development. As the critical plasticity period for binaural brain development has already passed once children reach the age of surgical ear reconstruction (6–7 years old), if treatment of hearing loss is delayed until this point, hearing rehabilitation is unlikely to occur despite restorative efforts.¹⁰ Therefore, until the child has reached the optimal age for ear reconstruction, the primary focus of management in bilateral cases should be to augment hearing with conductive devices. The soft band and other band-retained bone conductors are popular choices.^{6,24} As the child grows and the skull bone is thick enough to implant an osseointegrated device, the soft band can be converted to a more permanent bone-anchored solution like a BAHA or Ponto.^{27,28} The BAHA, approved for placement after the age of 5, bypasses any obstruction caused by atresia and helps conduct vibration across bone to preserve hearing.^{27,28} This osseointegrated implant is long-lasting and does not jeopardize the integrity of a reconstructed auricle. Likewise, a middle ear implant called the Vibrant Soundbridge (MED-EL Corp.) is functionally comparable to the BAHA, but does not have the cosmetic concerns of a visible hearing aid.⁶

In about half of microtia/atresia cases, the middle ear anatomy can be repaired in atresioplasty with immediate postoperative hearing gain. However, long-term studies suggest that beyond the first postoperative year, hearing deteriorates secondary to canal restenosis, bony regrowth, and lateralization of the tympanic membrane.^{28,29} In fact, Li et al²⁸ suggest that 30% of patients still require assistance for mid-range hearing loss after atresioplasty. Atresioplasty alone has an average gain of 17 dB, whereas combined atresioplasty with BAHA has an average hearing gain of 44 dB.^{30,31} This suggests that combining atresioplasty and BAHA offers more reliable results. It is important to note that in severe cases of high-grade deformity, atresioplasty may not even be an option; patients may only undergo auricular reconstruction for aesthetic benefits with fixation of an osseointegrated implant to rehabilitate hearing function.

Complications and Follow-Up

Microtia reconstruction and atresioplasty are generally well-tolerated, producing improvements in cosmetic appearance, hearing, and psychosocial attitudes of children.³² The reconstructed auricle, ear canal, and tympanic membrane are regularly assessed to evaluate for skin necrosis, redundant skin, early canal stenosis, and infection. Postoperative audiograms at 4 weeks are necessary to determine the amount of hearing restoration and provide a baseline for long-term follow-up.¹⁷ Repair is considered to be successful if hearing in the reconstructed ear is brought to within 15 to 25 dB of the normal ear; greater than a 35 dB difference is considered the threshold for failed corrective surgery.¹⁴ Annual clinical and audiometric surveillance is crucial to detect any delayed worsening of hearing due to canal restenosis, bony overgrowth, or tympanic membrane lateralization.^{22,33}

The use of intraoperative electromyographic monitoring is also useful in avoiding facial nerve damage, but it is not a guarantee, which may result in postoperative facial palsy, either temporary or permanent.³⁴ Chronic skin and implant-related infections can jeopardize wound healing and expose the cartilage graft. Although quite rare, a hematoma is devastating, requiring immediate drainage. The altered shape of the graft framework can decrease the stability of the reconstructed ear from several months to years following an initially successful surgery, resulting in delayed worsening of hearing.¹⁶ A revision procedure may also be performed for malposition, asymmetry, and scar revision to improve aesthetic results.¹⁰

Conclusion

Microtia repair and atresioplasty require a multidisciplinary approach to address the otologic and audiology concerns of ear deformities. Autologous microtia reconstruction improves cosmetic outcomes, but does not necessarily address functional hearing issues in patients with associated atresia. Atresioplasty and implanted bone-conducting devices are important mechanisms of hearing restoration, which must be used in concert with auricular reconstruction. Through this comprehensive approach, surgeons can optimize acoustic functionality while preserving the aesthetic components of reconstruction in patients with microtia and atresia.

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