Ear Abnormalities

Sana L. Bhatti, MD,*† Lauren T. Daly, MD,‡ Martha Mejia,* Chad Perlyn, MD, PhD*†

*Division of Plastic Surgery, Nicklaus Children's Hospital, Miami, FL
†Division of Plastic Surgery, Florida International University College of Medicine, Miami, FL
‡Division of Plastic Surgery, University of Massachusetts, Worcester, MA

PRACTICE GAPS

1. To facilitate prompt treatment for congenital ear abnormalities, pediatric health-care providers should be able to identify common ear deformities, some of which can be treated nonsurgically only if recognized within the neonatal period.
2. Clinicians should know when to refer patients with ear abnormalities to specialists for timely intervention.

OBJECTIVES *After completing this article, readers should be able to:*

1. Understand the normal anatomy of the ear.
2. Identify common congenital ear abnormalities as they present in the neonatal period.
3. Recognize the psychosocial impact of ear differences on pediatric patients.
4. Facilitate prompt diagnosis of congenital ear abnormalities and refer patients to specialists so that nonsurgical treatment can be initiated in the neonatal period.

ABSTRACT

Congenital ear abnormalities present an aesthetic and psychosocial concern for pediatric patients and their parents. Diagnosis of external ear deformities is based on clinical examination and is facilitated by an understanding of normal ear anatomy. Ear anomalies can be categorized as malformations or deformations. Malformations are characterized by absent anatomical structures of the ear (or absence of the ear itself), as exemplified by microtia and anotia. Ear deformations are characterized by ear anatomical landmarks that are present but are distorted or abnormal, with Stahl ear, constricted ear, and prominent ear being common presentations. Ear malformations will not improve with growth of the patient and uniformly require surgical intervention to recreate an anatomically typical ear. Although a small percentage of ear deformations can self-resolve, most patients with ear deformations will require nonsurgical or surgical reconstruction to achieve a normal or more aesthetic ear. In recent decades the use of nonsurgical ear splinting or molding has been recognized as a highly effective method in correcting a variety of congenital

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ear deformations when treatment is initiated in the first 8 weeks of life. The urgency in initiating nonsurgical treatment of ear deformations at an early age makes prompt recognition of these ear deformations essential because surgical correction remains the only viable reconstructive option in older infants and children.

INTRODUCTION

Congenital ear abnormalities are the result of an absence or malformation of the skin and/or cartilage of the neonatal ear. Auricular anomalies can be categorized as either malformations or deformations. Malformations are due to disrupted embryogenesis, which results in deficient growth of structures. Examples of malformations include anotia (absence of external ear), microtia (underdeveloped, usually malformed ear), cryptotia (ear cartilage partially buried beneath the skin), and preauricular sinuses and remnants. (1) Deformational auricular anomalies have an intact but distorted chrondrocutaneous framework. Ear deformations are thought to be due to external forces in utero or ex utero, thus leading to abnormal ear architecture. (1) A variety of different ear deformations have been described, including Stahl ear, constricted ear, and prominent ear. (2) Along with external ear abnormalities there can be hearing loss; this is most commonly seen with microtia.

Epidemiology and Pathophysiology

Although the true incidence of congenital ear abnormalities is not known, estimates range from 15% to 20% of newborns. (3) Patients with an ear malformation, such as microtia or anotia, should not be expected to have any spontaneous improvement. On the contrary, up to 30% of patients with a recognized ear deformation at birth will experience self-resolution by 4 to 6 weeks of age. (3)(4)(5) However, because of the small window of opportunity to treat these deformations nonsurgically, early referral to a specialist is preferred. Auricular malformation or deformity can cause significant psychological and social morbidity, including issues with poor self-esteem, social avoidance, anxiety, depression, and behavioral problems. (6)(7)(8) Fortunately, these symptoms significantly improve after reconstruction of the ear anomaly. (6)(7)

Anatomy

The external ear, or auricle, is the most peripheral component of the auditory mechanism (Fig 1). The external ear acts to funnel sounds to the tympanic membrane in a way that boosts sound frequencies associated with the human voice and aids in differentiating the spatial origin of sounds. (9) In addition to its role in hearing, the ear is an important component of craniofacial aesthetics, with minor ear differences attracting biased visual attention from viewers. (10)

The external ear is composed of elastic cartilage covered with hairless skin that is tightly adherent anteriorly and more lax posteriorly. The cartilaginous framework of the ear can be viewed as a topographical map, with the helix and lobule as the most elevated structures, the antihelix and tragus located midlevel, and the concha presenting as the deepest aspect of the ear. (1) The cartilaginous ridges have associated depressions, or scapha (Fig 1).

Embryologic development of the external ear begins during the fifth week of gestation, with the fetal auricle resembling the adult ear by the ninth week of gestation. By age 3 years, approximately 85% of adult ear growth has been attained. (11)

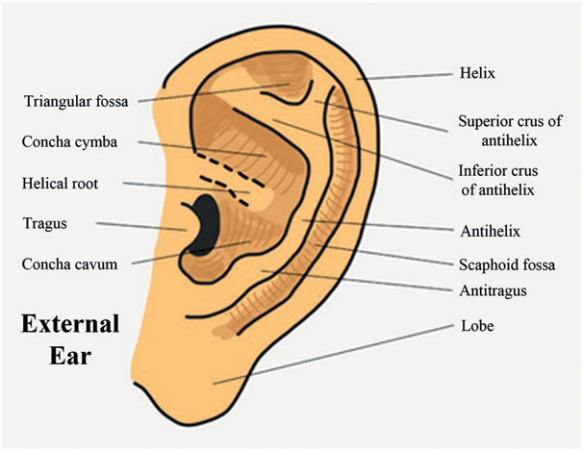

Figure 1. External ear.

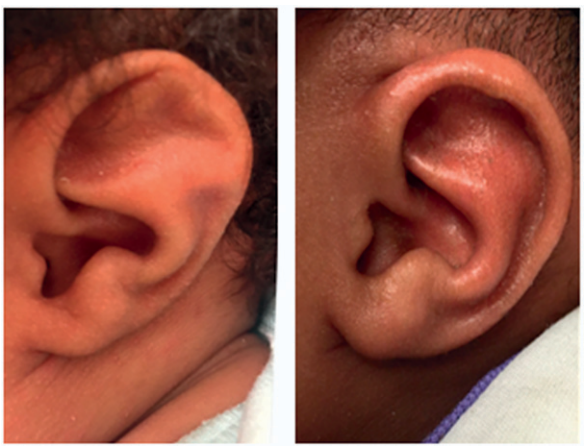

Figure 2. Stahl ear. Left, before. Right, 6 weeks after ear molding.

CLINICAL ASPECTS OF EAR DEFORMATIONS

Stahl Ear

Stahl ear is characterized by an additional abnormal vertical cartilage band crossing the scaphoid fossa from the antihelix to the helix and creating a pointed, elflike appearance to the ear (Fig 2). (12) The etiology of Stahl ear is unclear, but an anomalous insertion of the transverse auricular muscle has been implicated. (13) Similar to other congenital ear deformities, Stahl ear that is recognized early can be treated successfully in the neonatal period with splinting and molding. (14)(15) If treated later in childhood, Stahl ear can be corrected with surgery by resecting or repositioning the abnormal vertical cartilage band and reconstructing a more normal-appearing antihelical superior crus. (12)

Prominent Ear

Prominent ear can be caused by an underdeveloped antihelix, an overdeveloped conchal bowl, or a combination of the 2 cartilaginous deformations (Fig 3). (16) A prominent ear is characterized by an external ear that projects more than 2 cm from the side of the head and generally has an appreciably large surface area. (17) It is estimated that prominent ear is present in approximately 5% of the white population. (18) Prominent ear can be associated with a large psychological burden for both children and adults. (19)(20)(21) Multiple different operative techniques, including suture-based techniques, cartilage scoring, or cartilage excision, have been described to surgically correct prominent ear. (22)(23) When prominent ear is recognized and treated at a very young age, ear molding and splinting are successful methods of reconstruction. (24)

Constricted Ear

Constricted ear refers to deformities of the superior third of the auricle; presentation can be diverse (Fig 4). A constricted ear has been described by several terms, including *cup ear, lidded ear, lop ear, canoe ear,* and *cockleshell ear.* Cosman (25) described 4 fundamental features of the constricted ear: lidding caused by helical overhang and flattening of the antihelix, protrusion associated with deepened conchal fossa, decreased ear size due to the superior third deficiencies, and low ear position seen in severe cases.

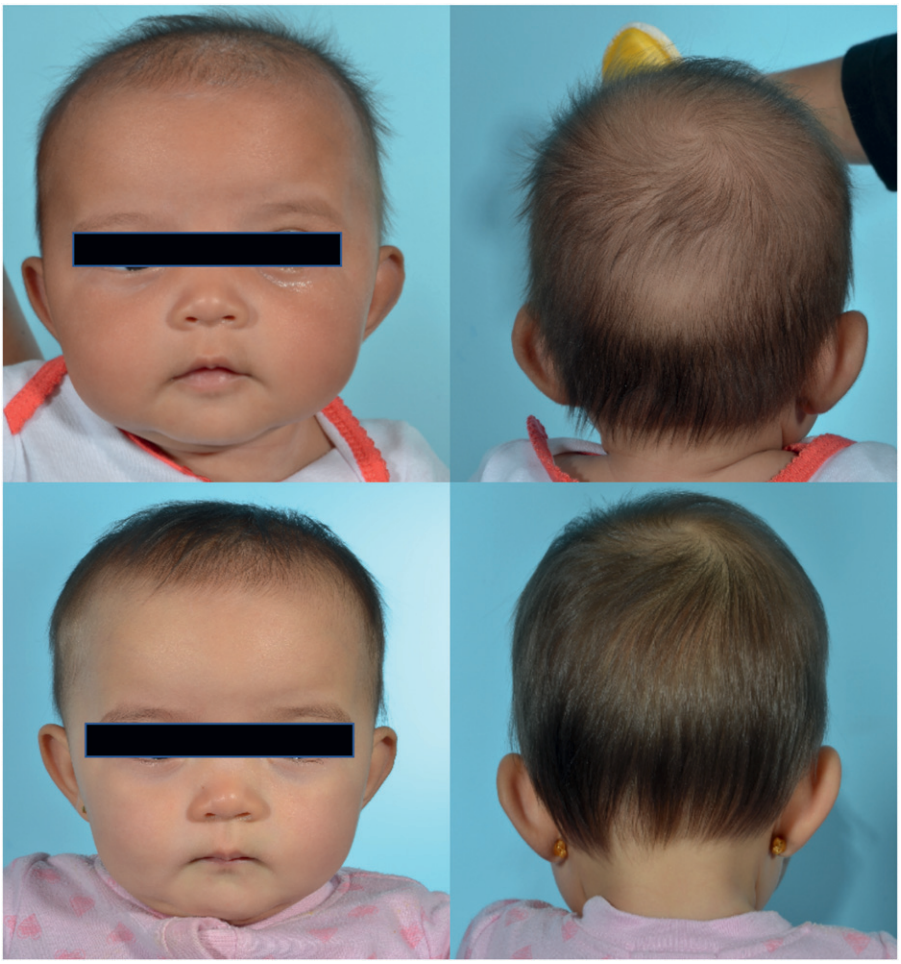

Figure 3. Prominent ear. Top, before. Bottom, after 6 weeks of ear molding.

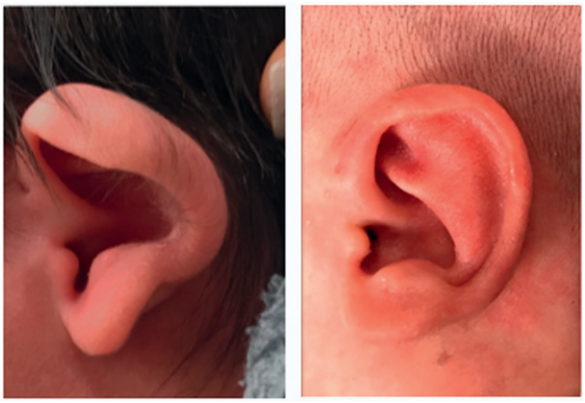

Figure 4. Constricted ear. Left, before. Right, 6 weeks after ear molding.

Mild deformities can be treated with nonsurgical molding with ear splints if initiated in the neonatal period. Moderate to severe cases, or cases in older children, require surgical correction.

CLINICAL ASPECTS AND MANAGEMENT OF EAR MALFORMATIONS

Microtia

Microtia is the term used for an external ear with absent skin or cartilage that is small, collapsed, or only has an earlobe present (Fig 5). Microtia can occur as an isolated birth defect (the most common presentation), as a part of a spectrum of anomalies, or as a component of a syndrome. Treacher Collins and Goldenhar syndromes are 2 of the most commonly associated syndromes. In most cases of microtia there is also agenesis of the external auditory canal. Microtia is most commonly associated with conductive hearing loss, which is due to a malformed middle and external ear. In unilateral microtia, the contralateral ear has normal hearing. Microtia prevalence varies geographically and is reported to be 0.83 to 17.4 per 10,000 births. It occurs most frequently in boys (2 or 3:1) and is predominantly unilateral (70%–90%), with a right-left-bilateral ratio of 6:3:1. (26)

Figure 5. Microtia.

The cause of microtia is still poorly understood. There is evidence for environmental and genetic causes of microtia. The most common anomalies associated with microtia include vertebral anomalies, macrostomia (a form of lateral facial cleft extending from the corners of the mouth, resulting in a wide oral aperture), oral clefts, facial asymmetry, renal abnormalities, cardiac defects, microphthalmia, holoprosencephaly, and polydactyly. (27)

Microtia can present in varying degrees of severity (Fig 5). Although several classification systems of microtia have been described, none are universally used clinically. As such, the standard should be to document, at the very least, a detailed description of the malformation of the ear, including each anatomical component of the ear, in addition to taking photographs.

Microtia treatment involves restoration of hearing and reconstruction of the external ear, often with a multidisciplinary approach that includes genetics, otolaryngology, audiology, and plastic surgery. Hearing screening is necessary for these patients on initial evaluation. An auditory brainstem response test is recommended soon after birth to evaluate both ears for inner ear function. Frequent ear examinations are also advised because these patients have a higher risk of ear infections and drainage. Screening with renal ultrasonography is recommended for all patients with microtia given the fairly high rate of associated abnormalities and high percentage of findings requiring renal follow-up. (28) A study by Koenig et al (28) found that syndromic children with microtia demonstrated a higher rate of renal ultrasonography abnormalities (22%) than children with isolated microtia (7%). Of these patients, 69% required specialist follow-up. Varying degrees of renal abnormalities can be found, such as agenesis, hypoplasia, ectopia, hydronephrosis, ureteral abnormalities, and vesicoureteral reflux. (28)

Early consultation with a surgeon helps develop a trusting relationship with the family as the surgeon guides them through a discussion of treatment options while managing expectations. Ear reconstruction is considered for aesthetic, psychological, and functional reasons. The external ear serves a functional structural purpose and allows children to use glasses, wear earrings, and normalize appearance.

The age at which microtia reconstruction should begin depends on psychological and physical considerations. Patients with microtia have a high prevalence of mood disorders, with depression in 20.2%, interpersonal sensitivity/social difficulties in 36%, and hostility/aggression in 26.3%. Studies have shown that correction of microtia improves psychosocial abilities postoperatively. (29)

Ear reconstruction would ideally be performed before the child enters school. However, surgery is delayed until rib cartilage is substantial enough to allow for quality auricular framework construction, which is 6 years and older. Surgery at this time also allows for the reconstructed ear to be created at adult size. In recent years, the preferred age of reconstruction has shifted even later, with some surgeons recommending 10 years of age as the ideal time. Worldwide, the predominant method of microtia reconstruction in children is autologous reconstruction, although several options exist for reconstruction of microtia.

Autologous reconstruction uses the patient's own rib cartilage. This technique typically requires 3 stages and begins with harvest of autologous cartilage rib grafts and creation of the ear framework. Additional elements include elevation of the framework, creation of a retroauricular sulcus, lobule transposition, and tragus formation (Fig 6). Composite reconstruction combines autologous tissues, such as local fascial flaps and skin grafts, for coverage and alloplastic materials, such as porous polyethylene, for an auricular framework, thus avoiding chest donor site morbidity. Osteointegrated prosthetic reconstruction involves implanting 2 to 3 magnetic posts into the temporal bone region to allow for a stable attachment of a prosthetic ear (Fig 6). Historically for patients with congenital microtia, prosthetic ears were often reserved as an alternative to failed autologous reconstruction. However, with recent advances, the osteointegration process has gained some popularity given the relative ease of the procedure for the patient. The downside is that the prosthesis needs to be replaced frequently, and there is no adaption of color to skin temperature or pigment changes due to weather or sun exposure.

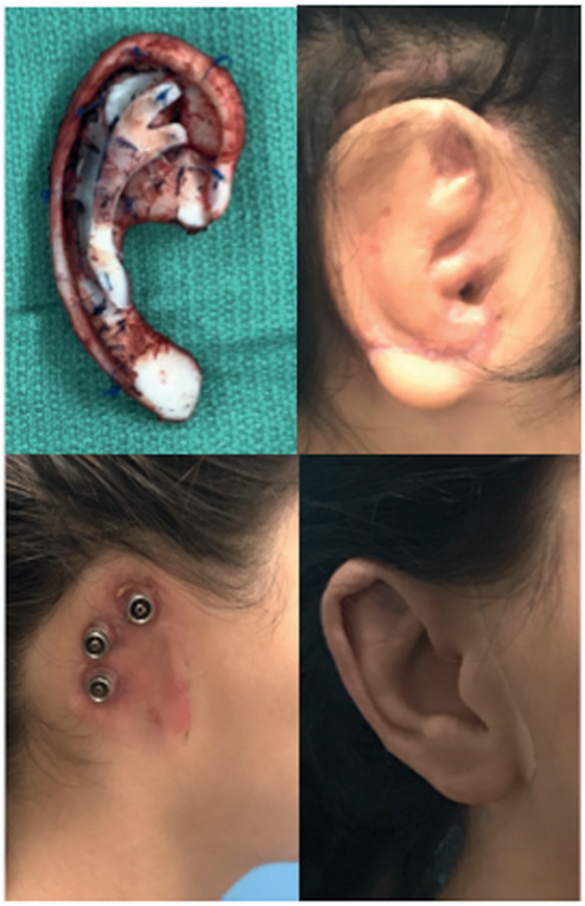

Figure 6. Ear reconstruction.

Cryptotia

Cryptotia, known as hidden ear, is identified as the absence of the superior auriculocephalic sulcus, which is due to the superior third of the auricle being buried underneath the temporal skin (Fig 7). Cartilage malformation may also be present. Correction can be attempted with application of a nonsurgical molding appliance in the first few weeks of life. Surgical treatment for older children or more recalcitrant cases involve division of the abnormally attached skin and placement of a full- or split-thickness skin graft to create a new sulcus.

Preauricular Anomalies

Preauricular anomalies include remnants with or without associated ectopic cartilage and sinuses (Fig 8). Screening renal ultrasonography is not indicated for isolated preauricular anomalies. (30) Preauricular remnants do not regress over time, and although there is no physiologic impact, it is a stigmatizing lesion, and, thus, treatment entails surgical excision. Preauricular sinuses are typically asymptomatic; however, they can become infected. Infected preauricular sinuses require antibiotics, and surgical excision is necessary. (31)

Management of Ear Deformations

Newborn ear molding has been used for nonsurgical correction of auricular deformations since the 1980s. (32) Multiple studies have demonstrated that satisfactory nonsurgical correction can be made by forcing the ear into a

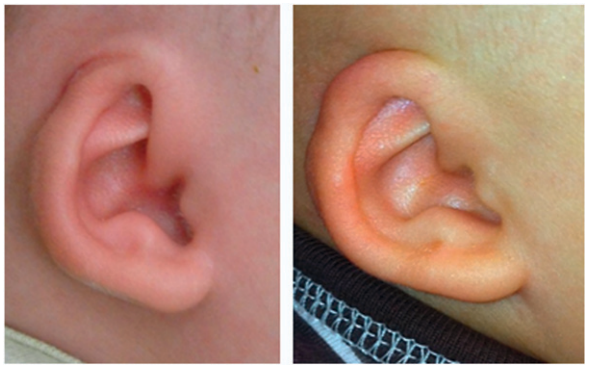

Figure 7. Cryptotia. Left, before treatment. Right, 6 weeks after ear molding.

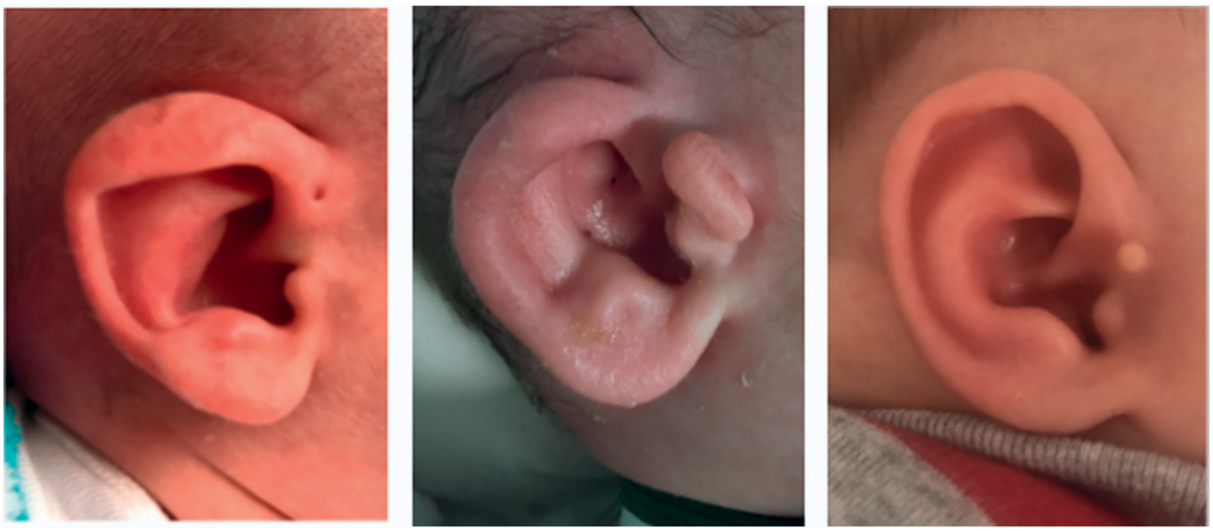

Figure 8. Preauricular anomalies.

proper position and maintaining it there for several weeks. (32) Permanent results can potentially be achieved if ear molding is started immediately after birth, preferably before the third day after birth, and can be continued until the infant is 3 months old. The impetus to start nonsurgical correction early is the role of circulating maternal estrogen levels in the pliability of ear tissue. It is hypothesized that retained circulating maternal estrogen decreases the structural density of collagen. Cartilage elasticity is dependent on the concentration of proteoglycan aggregate, of which hyaluronic acid is a major component, and the presence of hyaluronic acid is increased by estrogens. Maternal estrogen levels peak in neonates at 3 days of life, then taper significantly and normalize at approximately 6 weeks of age. (33)

In ideal circumstances, the infant is seen in a plastic surgery clinic on day 3 after birth and, depending on the physician's preference, either a commercial or a custom molding system can be used. Depending on the auricular deformity, a customized mold or prothesis can be fabricated from either plastic or acrylic and held in place with liquid adhesive and tape (Fig 9). Average treatment duration is 4 to 8 weeks, with biweekly follow-up visits for reevaluation and any necessary splint adjustments. Newborns are frequently seen by their pediatrician during this period, and coordination of appointments for families is encouraged. Complications associated with nonsurgical correction are rare and usually minor and can include minor superficial excoriations and skin loss due to pressure necrosis. (32)

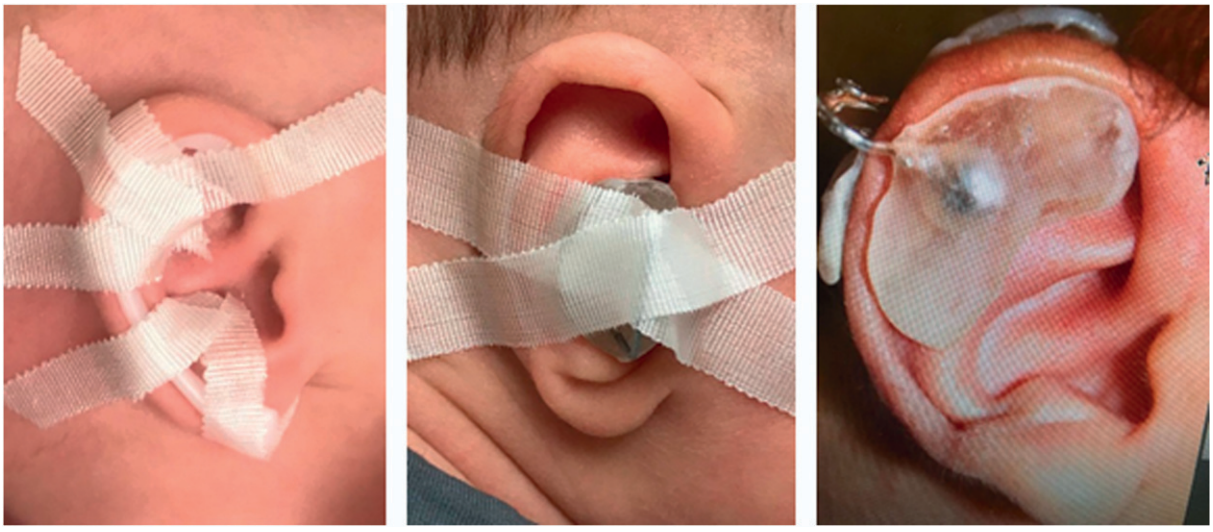

Figure 9. Custom molding system.

Table. Ear Malformations and Deformations: Recommended Diagnostics and Treatment Options

EAR ABNORMALITY	DIAGNOSTIC HEARING	RENAL ULTRASONOGRAPHY	SURGERY	NONSURGICAL CORRECTION
Malformations				
Microtia	+	+	+	–
Cryptotia	–	–	Possible	+
Preauricular remnant	–	–	+	–
Preauricular sinus	–	–	If infected	–
Deformations				
Prominent ear	–	–	If not corrected with molding	+
Stahl ear	–	–	If not corrected with molding	+
Constricted ear	–	–	If not corrected with molding	+

Summary

- Based on strong research evidence, the external ear is a complex 3-dimensional anatomical landmark with raised cartilaginous crura and depressed scapha. (9)(10)
- Based on some research evidence as well as consensus, ear differences are associated with psychosocial stresses and self-awareness in school-age children and adults. (20)(21)
- Based primarily on consensus due to lack of relevant clinical studies, ear anomalies can be divided into either malformations or deformations. Malformations include anotia, microtia, cryptotia, and preauricular anomalies. Deformations include Stahl, prominent, and constricted ears. (1)
- Based on strong research evidence, early referral is mandatory for nonsurgical correction in the neonatal period for most congenital ear anomalies. (3)(4)(15)(32)(33)
- Based on strong research evidence, if recognized in the neonatal period, most congenital ear anomalies can be treated successfully with nonsurgical intervention. However, if diagnosis is delayed, surgical intervention is required to reconstruct congenital ear deformities (Table). (16)(18)
- Based on some research evidence as well as consensus, hearing screening and renal ultrasonography are typically not required except for cases of microtia. (28)(30)

References for this article can be found at http://pedsinreview.aappublications.org/content/42/No. 4/180.

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1. You are making rounds in the newborn nursery. As you perform the physical examination of a term male newborn you notice an ear deformity possibly consistent with a Stahl ear. The remainder of the physical examination findings are normal. The baby is feeding well and is ready for discharge. Which of the following is the best next step in management of the ear deformity?

   A. Advise the parents to tape both ears backward.
   B. Follow up with his primary care provider at his 2-month health supervision visit.
   C. No follow-up is needed as this will spontaneously resolve.
   D. Recommend referral to plastic surgery if no improvement by 6 months of age.
   E. Schedule him an outpatient appointment with plastic surgery in 48 hours.

2. A term female newborn was noted on physical examination in the newborn nursery to have an isolated microtia on the left side. The remainder of the physical examination findings are normal, with no evidence of dysmorphic features. Which of the following is the most appropriate immediate next step in testing in this patient?

   A. Auditory brainstem response test.
   B. Chromosomal microarray.
   C. Echocardiography.
   D. Magnetic resonance imaging of the brain.
   E. Upper endoscopy.

3. A 2-year-old boy with unilateral microtia is followed in your practice. The patient has been followed by plastic surgery since birth. The parents are seeking your opinion as to when their child's surgical reconstruction of the ear should be performed. They have read on the Internet conflicting information. Which of the following is the optimal age to complete the surgical intervention in this patient?

   A. As soon as possible.
   B. At 4 years of age, before starting school to avoid him being subject to bullying.
   C. At 10 years of age.
   D. At mid-adolescence after the child goes through his growth spurt.
   E. The timing is variable with every child depending on the child's size.

4. A 4-year-old boy is brought to the clinic for a health supervision visit. The patient is healthy and has had normal growth and development. He has no recent illnesses. On physical examination he is noted to have a preauricular tag on the right. Otherwise his physical examination findings are normal. The parents inquire about the prognosis of the tag. Which of the following is the best approach to the management of this patient?

   A. Renal ultrasonography because preauricular tags are usually associated with renal anomalies.
   B. Excision and antibiotics if preauricular tags become infected.
   C. No intervention required because such tags regress overtime.
   D. No surgical excision required because the tag does not have an associated preauricular sinus tract.
   E. Surgical excision for cosmetic reasons because preauricular tags can potentially grow over time.

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5. Because primary care clinicians diagnose auricular deformations on initial physical examination in the newborn nursery, the success of ear molding nonsurgical correction in achieving permanent results depends on which of the following factors?

   A. Absence of associated renal anomalies.
   B. Absence of family history of ear deformations.
   C. Early intervention, before the third day after birth.
   D. Female sex.
   E. First born.

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