Evaluation and Treatment of the Deformed and Malformed Auricle

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Introduction

Malformations of the auricle are uncommon problems encountered by pediatricians. Abnormalities in ear form range from minor alterations of the cartilage, to absence of the entire appendage. When all anatomic elements of the anomalous pinna are present, nonsurgical correction can, and must be, started within the first few days after delivery.[1] Ears with more complex malformations must also be evaluated early in the neonatal period, even though nonsurgical correction may not be helpful. Concomitant malformations in other organ systems are occasionally present with severely malformed or microtic auricles, and an effort must be made initially to determine the overall nature of the problem, to decide the management, to predict prognosis, and to counsel the family.[2]

Embryology

The auricular anlage can first be identified during the fourth intrauterine week as six swellings (hillocks) of mesodermal tissue around the first and second branchial arches, surrounding the pharyngeal groove lying between them (Fig. 1).[3] These grooves and arches represent what were originally the primordial "gill slits" of a fish. These six hillocks fuse and differentiate into an auricle. By the second month, the primordial ear tissue begins migrating from an inferior medial position beneath the mandible to a posterior superior location on the temporal bone. Adverse genetic or environmental factors can interfere with this rapid sequence of developmental changes causing subtle to catastrophic abnormalities. More than 50% of children with severe ear anomalies have associated preauricular appendages, facial asymmetry, facial paresis or paralysis, deafness, and cervical vertebral anomalies.[4] These should all be carefully evaluated early, but it is critical to evaluate for hearing loss in the normal ear within the first two months of life. Brain stem evoked response audiometry (BSERA) is the most reliable and noninvasive test for hearing at this age. This noninvasive test can be administered without sedation if it is administered before the infant is three months old.

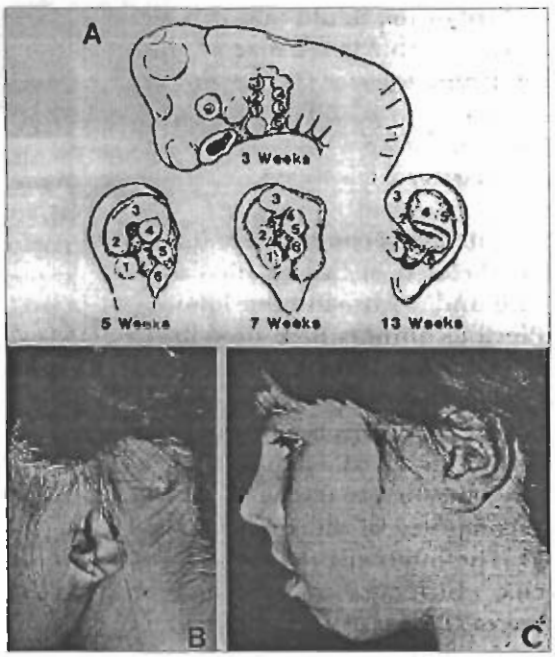

Figure 1. A. The auricle develops from six hillocks on the first and second branchial arches, and the groove that lies between them. **B.** This auricle failed to migrate from an inferior-medial position to its proper position on the mastoid. **C.** The mature auricle is more ovoid than round, not vertical but posteriorly inclined, and protrudes from the scalp.

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The pinna reaches its adult configuration by the beginning of the second trimester. However, late gestational mechanical forces may also affect auricular shape and form. Abnormal intrauterine fetal positioning or posterior cervical cystic hygromas may displace the ear, causing excessive protrusion or constriction. At birth the ear is at a lower position on the face than the adult, and it is almost two thirds the size of the adult auricle.

Anatomy

The pinna consists of an internal skeleton of fibroelastic cartilage and a fibroadipose lobule. Previous authors have described measurements and relationships between various parts of the auricle.[5] It is perhaps best to conceptualize the ear as a flap-like structure, attached to the head, which is composed of three layers (Fig. 2). The internal layer is the concha, which surrounds the opening of the ear canal. The second layer is the scapha, which is the main supporting buttress and site where most anomalies occur. The helix is the outer layer, silhouetting the perimeter of the auricle.

Deformed Ears

Auricular anomalies have been categorized and graded in a variety of ways. These definitions have been further subcatergorized, with descriptive terms such as prominent, protruding, lopped, hooded, cryptotic, macrotic, and microtic auricles. We have used a grading system based on three levels of severity. Grade I anomalies include ears with all anatomic subunits present, but misshapened (Fig. 3).

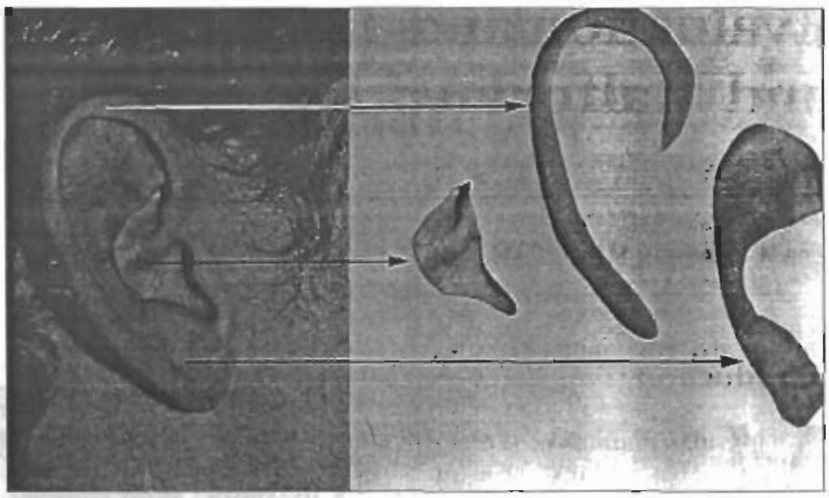

Figure 2. The auricle is composed of three layers. The internal layer is the concha, which acts as an "entrance" to the canal. The scapha is the middle layer and the main structural support of the pinna. The helix delineates the outer margins.

Using one's fingers as "measuring tools," an accurate diagnosis can be easily made. These seemingly severe deformities can often be corrected at birth with nonsurgical methods.

The pediatrician can take advantage of the excessive "plasticity" of the ears during the first few days after birth. During the first few days of life, the high maternal estrogen levels circulating in the neonate cause the hyaluronic acid content and mucopolysaccharide matrix of the ear cartilage to remain soft and pliable.[6] Even strikingly dysmorphic deformities can be resculptured with tape and molding.[7-10]

Technique

The ears should be evaluated

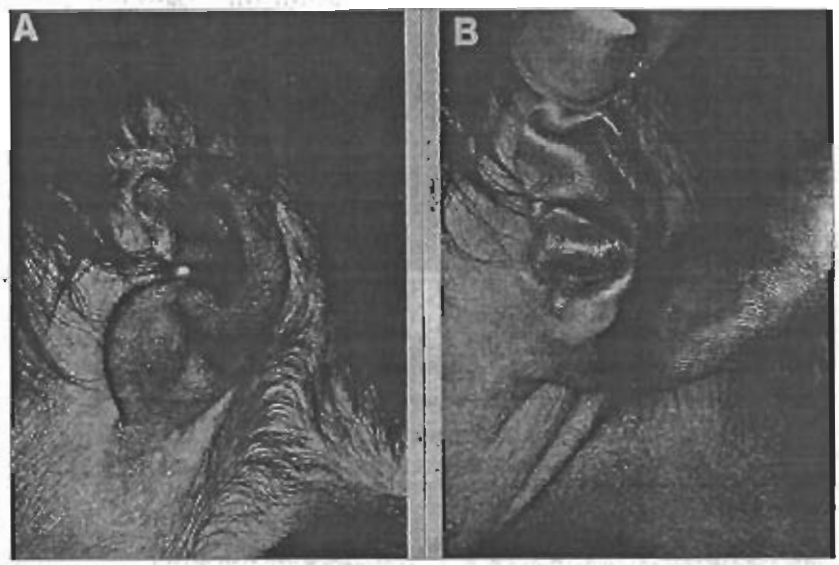

Figure 3. Dysmorphic ears have all their "anatomic subunits" present but are misshapen. Using one's fingers as a measuring tool, deformities can easily be identified.

and remolded immediately after birth. Using one's own fingers, the deformed ear is pressed backward against the skull to identify each minor deformity. The scalp is shaved 2 cm postauricularly. Wetted cotton or any molding clay is used to resculpture the abnormal auricular folds. Benzoin adhesive is applied on the skin, and ½ inch Steri Strip tape is placed to secure the ear molds in proper position against the mastoid. The tape and the molding material is changed weekly for three weeks. If molding is started within the first few days, two to three weeks of resculpturing is usually adequate (Fig. 4). We have been unsuccessful remolding "prominent" ears *after* the first year. These children must wait until age 5 or 6 when the auricle attains 80% of its adult size. At this age a formal surgical otoplasty can readily improve both types of problems causing protrusion.

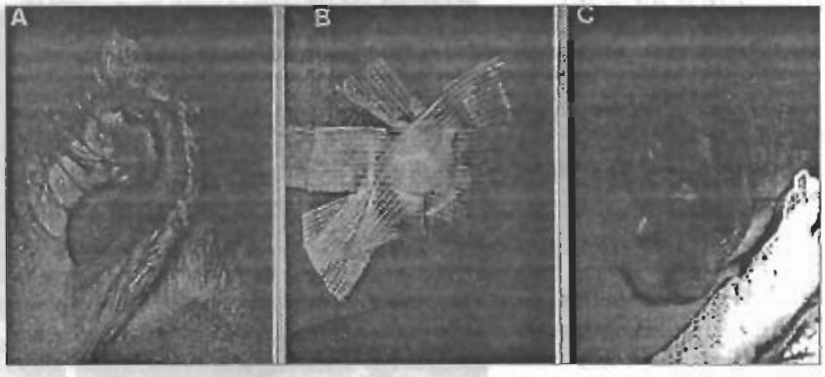

Figure 4. Molding of the ear is accomplished with moistened cotton placed in the deformed areas for two weeks. Steri strip tape secures the cotton in place. Patient before (B) and after (C) nonsurgical molding of the auricle.

Protruding Ears

The "protruding" or "prominent" ear presents a special problem (Fig. 5). Although many minor anomalies spontaneously improve in the first year, excessive ear protrusion can be an acquired deformity that may become more prominent until one year of age.[11] Hence, there is a greater incidence of excessively protruding ears at one year than at birth. Two different mechanisms may explain the excessive "unfurling" present in the prominent ear: (1) weakness of the intrinsic auricular muscles; and (2) the forward displacement of one ear when the infant lies with the head turned to one side for an excessive period of time (e.g., plagiocephaly-torticollis deformation sequence).[12,13] Whenever the rim of the helix protrudes more than 2 cm from the mastoid, one should consider placing a tubular elastic net headband over the pinna when the child is lying supine. Such "head banding" must be continued throughout the entire first year. If ear molding is undertaken immediately after birth, such prolonged head banding may be avoided in some cases.[14,15]

Dysplastic or Malformed Auricles

This more severe type of congenital anomaly results when the

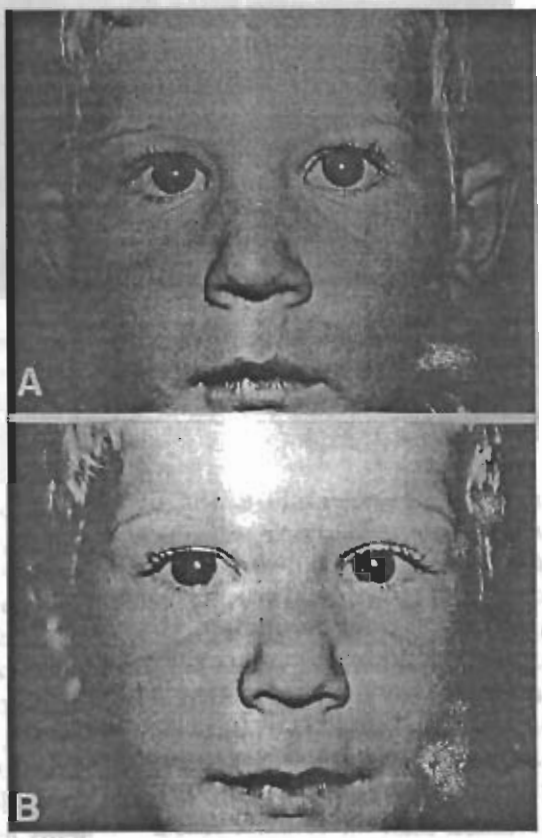

Figure 5. The "prominent" ear may not be protruding at birth. All components are present; however, there is increased "unfolding of the antihelical fold and increased angulation of the concha. Prominent "protruding" ears not corrected by early nonsurgical molding will need formal surgical otoplasty.

primordial tissues that form the auricle fail to develop. These deformed ears are small and are constricted in size and configuration. Such defects do not respond to the molding approaches described above.

The Constricted Ear

These Grade II anomalies result from dysplastic or aplastic development of the auricular hillocks of the first two branchial arches. One or more anatomic aspects of the three-layered auricle fails to develop (Fig. 6). Unlike the overly prominent ear, dysplastic or constricted ears do not worsen after birth. Most commonly, the scapha (which is the main supporting buttress of the ear) is weak and deficient. Without its support, the superior aspect of the pinna falls over upon itself, causing "lidding" and "foreshortening." These defects must be surgically reconstructed with grafts of cartilage and skin usually at age six years.

Microtia

The microtic auricle (Grade III) results from arrested development of the branchial arches after the fifth intrauterine week. Classically, the microtic ear (Fig. 7) is a vestigial anlage of the six hillocks represented as a vertically oriented flap of tissue with cartilage remnants superiorly and a fibroadipose nubbin inferiorly. In most patients the external canal also fails to develop. Such atretic ear canals are often associated with a moderate to profound unilateral hearing loss. Thirty percent of these patients have bilateral microtia. They require early hearing evaluation with BSERA. Infants with bilateral hearing loss must be fitted with hearing amplification by three months to enhance processing in the auditory

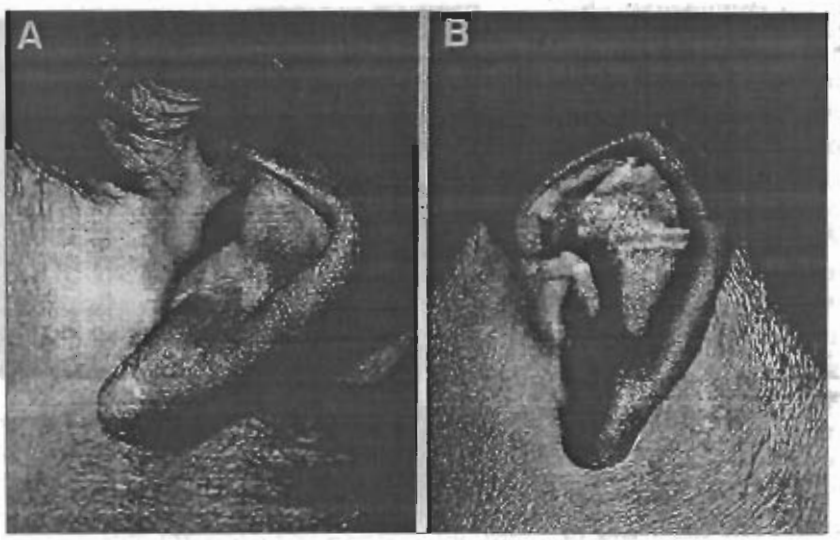

Figure 6. The constricted ear has deficient support from the scapha and appears to be "lidded and fall over upon itself." Many "anatomic subunits" are missing. Repair of a constricted ear with grafts taken from the opposite auricle.

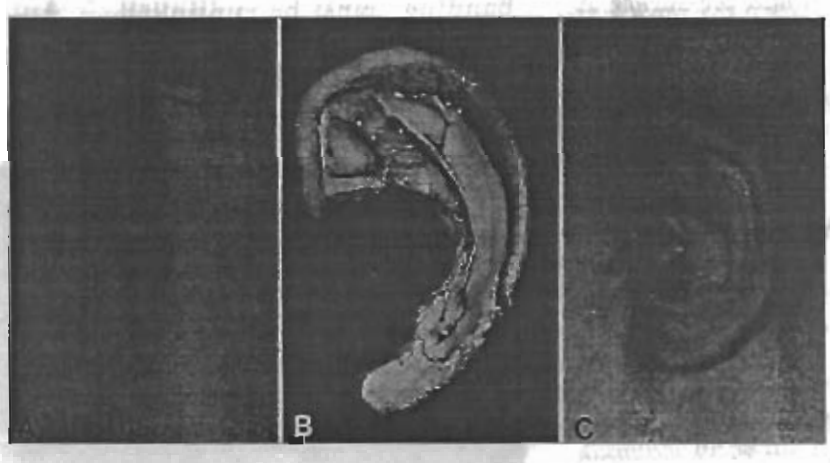

Figure 7. Microtia is often associated with atresia of the canal and extreme dysplasia of its cartilaginous structure. Ribs are harvested and sculptured into an neoauricle. This framework is placed beneath the microtic nubbin.

nuclei and in the cerebral cortex.[16] Previously, many clinicians felt that these patients with microtia should be screened for urinary tract anomalies. However, syndromes in which the ear and renal anomalies are associated together on a genetic basis (e.g., branchio-oto-renal syndrome) are quite rare and are considerably less common than the isolated association of external and middle ear (conductive hearing loss) deformities.[17] The National Collaborative Perinatal Project followed up more than 50,000 mothers and their children from pregnancy through age 8. Of this group, 591/53,257 children had at least one malformed auricle (incidence 0.0111%). At least 42 patients in this group had dysmor-

phic syndromes. Among patients with isolated ear malformations, 6% had a conductive hearing loss, while 0.4% had dysfunctional anomalies of the urinary tract.[18] This suggests that evaluation for hearing loss is much more important in this group of patients than evaluation for urinary tract anomalies.

The surgery for complex auricular reconstruction is begun at age six when the chest wall is large enough to harvest the cartilaginous portion of the 7th, 8th, and 9th ribs. These rib grafts are sculptured into an auricle (Fig. 7) and are placed in a skin pocket beneath the microtic nubbin. A four-stage auricular reconstruction is performed at three-month intervals and completed within one year. At the final stage the otologist can also create an external canal and ear drum to enhance hearing.[19,20]

Summary

Auricular malformations begin within the first few weeks of intrauterine growth. Less severe deformational abnormalities occur from abnormal fetal positioning. They can be corrected early after birth by simple reshaping and molding. No longer should the pediatrician wait for minor deformities to resolve spontaneously. The protruding ear may not be present at birth but may develop and worsen during the first year. The most severe anomalies require complex multistage reconstructions after other concomitant anomalies are excluded. The pediatrician must be an integral part of this reconstructive team.

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