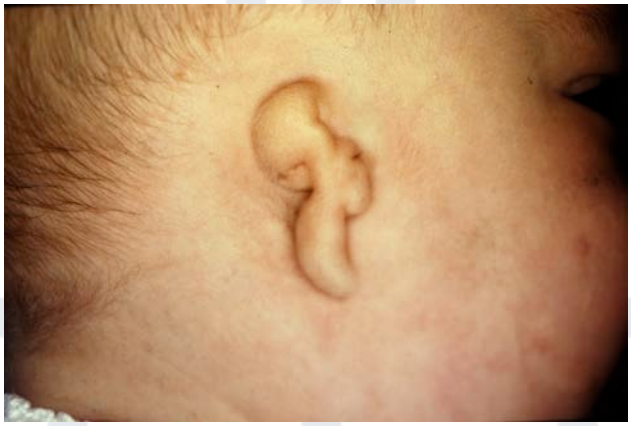
MICROTIA

Underdevelopment /deformity of the auricle (pinna) varies from subtle deformities and small pre-auricular rudiments to gross developmental failure, distortion or malpositioned remnants. The external appearances are commonly associated with external canal atresia, middle ear deformity and disorganisation, inner ear malformations, aberrations of the course of the facial nerve, and sometimes auditory nerve abnormalities. Poor auricular development may present as a syndromal component, bilateral or unilateral, or may be associated with unilateral facial deformity (hemifacial microsomia).

The condition is a complex mix of cosmetic, functional, and often psychological difficulties.



Microtia: Not only the ear.

The cause is either genetic or developmental, but generally not a repetitive genetic pattern unless part of a syndromal pattern. Genetic causes are generally the more advanced patterns. Lesser examples, especially unilateral, are developmental – failure of the normal tissue combinations that are complex in the ear. The cause of this may be an ill-defined, otherwise innocuous event (e.g. viral), or a clearly recognisable noxious agent e.g. thalidomide.

The auricle develops from six “hillocks” of tissue derived from two folds of the developing head and neck: the first and second brachial arches. Failure of the first arch often produces lesser problems such as pre-auricular rudiments or relatively slight auricular malformations. Canal atresia3. 1. 22 may not be present in these cases. Failure of both arches is accompanied by advanced malformations, perhaps with associated syndromal attributes.

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Characteristics

Commonly, external ear deformity (Pinna) is mild, presenting as tiny pre-auricular skin tags or shallow pre-auricular sinuses of little clinical or cosmetic importance, although both may require surgical or other intervention in more extreme cases.

Lesser deformities of the auricle likewise may cause little concern.

Alternatively, auricular deformities may coincide with major hearing loss in the event of associated canal atresia or middle ear deformities.

Advanced pinna malformations are usually more problematic, as atresia and other cosmetic problems are common in this group. Both difficult cosmetic correction and advanced hearing restoration techniques will be needed for resolution of the combined difficulties. Also, the commonly associated syndromal changes in these children often necessitate other general medical or specific managements (renal, airway, cardiac, psychological).

The degree of hearing loss is unpredictable from the appearance of the auricle, as only minor anatomical derangement may result in substantial conductive deafness. This may be due to a narrow external canal, with ossicles that are fused to the canal wall or malformed and dysfunctional. A narrow canal may thwart accurate visual evaluation of the drum and chain, or debris may accumulate in the deeper harder-to-access reaches. Ossicular repair may be impossible if the facial nerve is abnormally positioned. Syndromal cases such as CHARGE association are notorious for inner ear malformations.

Investigation

Assessments are undertaken with audiology and radiology. Conductive deafness will be maximal in severe microtia with canal atresia, but variable in lesser cases.

High resolution CT scans are used to assess the temporal bone anatomy as a guide to the extent of deeper abnormalities and the surgery required to correct this. 3D reconstructions may also be employed.

Importantly, the scans also show the status of the temporomandibular joint. In severe microtia and atresia cases, the joint may be displaced into a position filling the site of the external canal. This invalidates the possibility of canalplasty restoring an external canal; the surgeon would then need to resort to implant surgery to regain hearing

Management

Treatment of the deformed ear may focus on the appearance, the function, or a combination of both aspects.

i. Cosmetic

Several methods address the cosmetic aspects. The least surgery removes rudiments or skin tags to “tidy up” the site. In the past, this has been supplemented by auricular prostheses, adhesive or attached via osseo-integrated mounts attached through the skin.

Reconstruction of the pinna is much more complex and requires specific skills. Rib cartilage grafts may be used, but require tissue removal via an abdominal incision, then shaping to form a scaffold, implantation, plus secondary skin grafting. Residual external ear elements may need to be included in the repair, especially the ear lobe, if present (this is difficult to create with grafts). Scarring and other tissue dynamics may prove unsatisfactory.

Alternatively, a framework of MedPor (polyethylene) may be utilised to avoid abdominal trauma and to have a ready-made shape available, albeit with some risk of biomaterial reactions.

ii. Functional

Restoring hearing may be achieved via a canal reconstruction, then tympanoplasty techniques to rebuild the drum and chain. However, split skin grafting is needed to re-line the external canal. These grafts are taken from the medial upper arm, for cosmetic reasons. Split skin is not self-cleaning: periodic removal of external canal keratin is needed on a 6-12 month basis in the long term. Canal and chain surgery may be combined with auricular reconstruction in one or two stages.

If the ear is anatomically unsuited to tympanoplasty surgery because of the extent of deformity, technological aiding is used. From diagnosis, an infant will be fitted with a bone conduction headband aid to stimulate the auditory pathways of the ear, and in bilateral cases, to promote hearing and speech development.

Later (5 years+), electronic implants may restore hearing. Current options include the Med EL Vibrant Soundbridge and Bonebridge devices, or Cochlear BAHA models.

Soundbridge devices are attached to the ossicular chain to vibrate this in order to create hearing. The Bonebridge is an active bone conduction implant, similar in shape to a cochlear implant. BAHA devices are also bone conductors, functioning via a titanium screw fixed in the skull through the skin, or held in position by magnetic force. Bonebridge devices may be implanted via a small CI “thumbnail” incision on the rear of the ear, or by other limited approaches. Conversely, BAHA models require larger incisions to site the fixation screw.

The Bonebridge external processor is also less bulky than the current BAHA equivalent.

Cochlear implantation is necessary in some cases, but others may have concurrent absence of the auditory nerve, possibly necessitating the use of an auditory brainstem implant.

Outcomes

In less deformed cases, and in expert hands, 80% of cases may achieve good hearing levels from canalplasty and chain reconstruction. Surgical success is not guaranteed, and delayed canal stenosis may occur. Periodic debris accumulation may cause temporary occlusion.

Bone conduction aids are cosmetically evident and may cause uncomfortable pressure effects.

Soundbridge implants are attached to the chain or sited on the round window. Middle ear disorganisation invalidates their use in more advanced pathology.

Bonebridge devices can be used only when the skull has matured to sufficient thickness, but are effective once in position.

BAHA devices mounted on screws placed through the skin commonly incur local tissue reactions require local clearing, and may entangle hair. The magnet-stabilised intact skin models avoid these problems, but remain somewhat bulky and prone to displacement

CONCLUSIONS

Microtia and the attendant canal atresia vary from limited aberrations to complex difficulties. For the surgeon, the variability of pathology encountered can be a major difficulty. Hearing restoration is a prime concern, especially in the bilaterally afflicted. From the family aspect however, the importance of cosmesis should not be underestimated.

Above all, the family needs realistic advice on the origins of the condition, the management options, the expected outcomes, and the difficulties faced to achieve these gains.

More information

- Inherited Conditions
- Congenital Ears

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