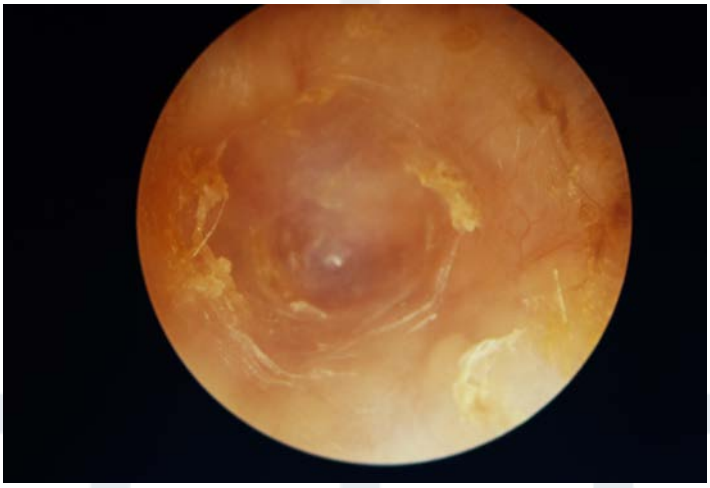
EXTERNAL CANAL ATRESIA

Atresia is defined as the failed development of the external auditory canal. The problem results from malfunction of the development of the cells between the folds of a developing pinna. These normally form a skin-lined canal, beginning from the exterior, then hollowing out down to the site of the eardrum, and forming the exterior layer of the drum. Atresia (Inherited Conditions) may arise from failure of the first branchial arch tissue, or, in the more severe deformities, from combined failure of the first and second arches.



Canal Atresia: Profound conductive deafness.

Characteristics

Failed development covers a range of canal abnormalities. Some mild aberrations occur in isolation. Down syndrome commonly shows narrowing of the mid canal forming an “hour-glass” canal, but with a normal drum and middle ear.

Other narrowing commonly occurs in the presence of microtia. The canal may be very narrow, leading to a distorted, partially developed drum. Others exhibit either a blind pit, or total absence, as in most cases of advanced microtia.

When atresia is present, the drum may be vestigial, partially formed or disorganised in structure. The underlying ossicles are usually malformed; the malleus and incus with first arch problems. Combined arch deformities result in gross chain problems that are difficult to correct, and which may be associated with facial nerve abnormalities or syndromal conditions.

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With atresia, there is also a significant risk of inner ear abnormalities, which may cause substantial nerve deafness.

Investigation

When atresia is present, hearing assessment is important, to gauge the extent and nature of hearing loss, as this will determine treatment. Pure tone tests and computer-assisted investigations determine the degree of conductive (mechanical) and nerve deafness respectively. CT and MRI scans assess the anatomical status.

Treatment

Aiding. The management of atresia is directed at hearing restoration. It is important to stimulate the unilateral case, to activate the connections to the brain, that otherwise risk becoming inactive with time.

Where possible, air conduction (standard) hearing aids are used, or alternatively, headband bone conduction aids that act via skull vibration (until more definitive surgical management).

Surgery aims to reconstruct hearing by re-creating the external canal (meatoplasty, canalplasty), the eardrum (myringoplasty), and the chain of ossicles (ossiculoplasty). In advanced disease, this may not be effective, and one of a range of surgical hearing implants may be required.

Several **implants** are available. The Med El Vibrant Soundbrige is attached to the ossicular chain or the cochlea itself, acting via a tiny electromagnetic vibrator that produces sound. These can be used in the first years of life and can be implanted via minimal incision surgery.

The Med El Bonebridge is a bone conduction device, implanted similarly, but requires a thicker skull for fixation – usually 5+ years.

The Cochlear BAHA (bone anchored hearing aid) is likewise a bone conduction device fixed to the skull, again around 5 years, but requires a larger incision and is somewhat more bulky.

When sensorineural deafness is present, cochlear implantation is required. The structure of a concurrently deformed cochlea may impede the use of these implants, permitting only partial success. Also, an underdeveloped nerve to the brain may likewise impede or prevent their use.

Conclusion

The overriding consideration with atresia is restoration of hearing, even if the contralateral ear appears normal. Failure to ensure stimulation of the deformed ear from an early age causes significant disadvantage, and may prevent optimal later hearing restoration.

More information

- Microtia and Atresia

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