CONGENITAL DRUM AND OSSICLE DEFORMITY

In a variety of situations, the eardrum and Ossicular chain may be deformed at birth. The problem is usually related to microtia (Microtia) and/or canal atresia (External Canal Atresia) (Failed normal development of the external ear auricle or ear canal), but not invariably.

The cause may be genetic or developmental; lesser abnormalities are thought to be the latter, perhaps induced by viral or other influences.

Characteristics

The drum may be small or deformed, lacking the normal loudspeaker shape, or the chain of ossicles (the small hearing bones of the middle ear) may be misshapen, or fused to the surrounding bone.

The problem usually presents in infancy, particularly when routine neonatal hearing testing is performed. Inspection of the external canal and drum then shows a narrowed canal, or, if this is of adequate diameter, a misshapened drum. More subtle ossicular abnormalities may require a specialist otologist’s opinion.

The audiological outcome will depend on the degree of deformity. Mild chain abnormalities may exhibit only slight losses, whereas more extensive pathology may produce very severe losses, or even a profound nerve loss should a concurrent inner ear abnormality be present.
Treatment

Management will likewise depend on the degree of the problem. If an adequate canal is present, a conventional air conduction aid may be preferred.

Drum deformity may be due to narrowing of the deep external canal; this may need to be drilled wider, for surgical exposure and to create an adequately large drum vibration surface. Widening may in turn necessitate skin grafting of denuded deep canal surfaces.

Recreating a new drum generally employs soft tissue grafts, but if the chain is repaired, special cartilage and soft tissue composite grafts are used.

Chain repair can be achieved by mobilising the existing structures, or by using either reshaped patient’s bones or micro-prostheses, usually titanium or ceramic.

The combination of complex repair steps predisposes the procedures to a significant risk of subtle or severe adverse influences on the success of the combined canal-drum-chain repair. Some residual deafness is therefore not uncommon with these procedures, but may be recovered with revision surgery.

If split skin grafting is used, this will require occasional cleaning in the long term.

The alternative is to use emerging hearing implant technology.

If the Ossicular chain is not badly deformed, a Med El Vibrant Soundbridge (Sound bridge animation) may be attached to the chain. The device is slightly similar to a cochlear implant, producing sound via a tiny vibrating electromagnet. Alternatively, the Soundbridge may be attached to the otic capsule entrance for sound effect.

If the ear is disorganised, but with good nerve function, bone fixation implants are suitable (Med El Bonebridge (Bonebridge animation 2. 1. 9. 6 – can’t find animation), Cochlear BAHA (Implantable Hearing Technology). These vibrate the skull for sound effect and are also useful if the nerve is damaged, by stimulating the contralateral normal hearing ear, to dispel the hearing “dead spot” on the affected side.

Lastly, if the cochlear function is lost, but the cochlear anatomy and VIIth nerve are undamaged, a cochlear implant supplies very serviceable hearing. These implants are increasingly used for single sided total congenital deafness.

More Information

- [Congenital Ears](#)