CONGENITAL STAPES FIXATION

Congenital stapes fixation is due to bone formation around the base of the third bone of the middle ear chain. The stirrup-shaped stapes is the smallest bone in the body, sited at the entrance of the inner ear (cochlea). The “footplate” of the stapes fits exactly into the entrance (oval window), transmitting vibrations into the cochlea, where the hair cells convert the mechanical energy into electrical. The fine gap around the stapes is normally sealed by a fine ligament, which is attached to the surrounding bone (the otic capsule, that encloses the inner ear).

Congenital stapes fixation: Immobile and atypical third middle ear ossicle.

Characteristics

In congenital stapes fixation, the surrounding ligament fails to develop, the stapes footplate being in continuity with the otic capsule. Being rigidly fixed in position, the sound transmitted to the stapes cannot pass into the cochlea. Conductive (mechanical) deafness results, but the nerve remains capable of good function if the obstruction is overcome. The eardrum and the rest of the Ossicular chain may be normal, or there may be fine aberrations of the rest of the stapes structure. Uncommonly, the facial nerve that normally passes close by may be abnormal in position.
The situation is almost identical with a maturity-onset conductive deafness – otosclerosis (Otosclerosis and Stapedectomy). The latter appears in late teens or later, but the resulting audiological pattern is commonly identical. Audiology produces a characteristic conductive pattern with a slight nerve deafness at 2000 cps – a “Carhart’s Notch”.

Treatment

When a conductive loss is present in a child, with a normal drum and without other evidence of cause, hearing aids are the usual initial management, until the child is more mature.

Congenital stapedial fixation is diagnosed by exclusion and confirmed by exploratory surgery and stapedectomy surgery (Middle Ear Surgery). The eardrum is lifted via the external canal, using an operating microscope, and the arch of the fixed stapes is removed. The footplate is then perforated using a micro-drill or laser technique, A specialised piston-shaped prosthesis will generally restore hearing, but some cases incur a degree of continuing conductive loss for uncertain reasons. The surgery is brief and rarely troublesome for the child.

The results are generally very good, but a small proportion of cases suffer a continuing partial deafness, the cause of which is poorly understood. In these cases a hearing aid is suggested, if the remaining loss is significant.

More information

- Otosclerosis
  - General
  - Surgery
  - Atypical Situations
  - Surgical Complications