COCHLEAR IMPLANTS
Aetiology of Deafness
Heterochromia iridis. Cases may be healthy or associated with a variety of conditions, e.g. Waardenburg syndrome.
Waardenburg syndrome. Note the snowy lock of hair and the heterochromia iridis. Usually carries a good CI prognosis.
Heterochromia iridis and associated profound SND.
Kabuki syndrome. Good CI prognosis in the absence of intellectual or other problems.
Osteogenesis imperfecta. Associated otosclerosis and ossified cochleas. Caution re CI advisable.
The blue sclerae of osteogenesis imperfecta. Profound SND due to cochlear otosclerosis.
Cochlear otosclerosis. Heavy ossification of the otic capsule to the left.
Gross cochlear otosclerosis, near-total obliteration of both cochlear spirals.
Similarly heavily ossified otic capsule due to otosclerosis.
Very guarded CI prognosis.
Bilateral otosclerotic cochlear basal turn obliteration, regrettably precluding optimal CI results. A partial spiral drill-out would seem likely, lessening prospects.
Previous case, showing partial basal turn obliteration. Stapedectomy piston on the left.
Post CMV child with profound SND, developmental and intellectual degradation. Guarded CI prognosis due to CNS disease.
Right labyrinthitis ossificans after pneumococcal meningitis. The cochlea, left, is severely narrowed secondary to intra-labyrinthine infection and ossification.
Post-meningitic labyrinthitis ossificans on the left. Cochlear spiral severely occluded. CI to the right, but the prognosis in the non-operated ear would be poor.
Post-meningitic cochlear obliteration. The spiral to the left is partially occluded, when compared to the less affected spiral on the right.

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Bilateral large ventricular aqueducts (LVAs); progressive sensorineural losses leading to profound bilateral deafness and bilateral CIs. Good CI prognosis, ? Perilymph “gusher” at surgery.
Bilateral enlarged ventricular aqueducts. Progressive asymmetrical sensorineural losses requiring sequential bilateral cochlear implantation.
An LVA extending into the vestibule. Profound loss present.
Contralateral view of the previous case. Similar course and Rx. LVAs have a good implant prognosis, in the absence of other difficulties.
Large right ventricular aqueduct in continuity with the vestibule. Axial view. Profound loss from birth. Managed with a CI.
Bilateral ventricular duct enlargement, seen to extend to and connect into the vestibule on each side. Profound losses at birth. Bilateral simultaneous CI.
Left cochlear deformity. Aberrant spiral with only 1 turn. Associated VIII deficiency.
Severe Mondini deformity, right cochlea. Loss of partitions and IAC open into the cochlea, causing a risk of an electrode array penetrating the posterior fossa.

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Previous case, showing the cochlear abnormalities in both ears.

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Bilateral dilated cochlear spirals. Profound losses detected on neonatal hearing screening tests. Good responses to bilateral CI.
Moderately advanced cochlear dysplasia. A “comma” deformity is present. CI achieved limited success due to reduced numbers of inserted electrodes.
Severe bilateral inner ear deformities with globular cochleas and dilated vestibule malformations.
Bilateral cochlear and vestibular dysplasia dysplasia. The cochlear spirals are dilated and there is expansion of the vestibule on the right.
Lower axial cut of the previous case showing the dilated cochlea and vestibule on the right.
Lower level view, normal left basal cochlear turn, dilated on the right.

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Coronal CT. Bilateral congenital cochlear deformities exhibiting partial globular cochlear malformations, both ears.

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Bilateral globular cochlear deformities. Profound loss at birth, moderate responses to bilateral early CI.
Right globular cochlea in a Klippel-Feil syndrome case.
Axial view detail of the previous case.
Cochleo-vestibular hypoplasia, with IAC atresia. Angled view due to Klippel-Feil neck abnormalities. Right EAC atresia.
Lower cut of the previous case. Bilateral EAC absence.
Bilateral IAC atresia, axial view. EABR: no response. Auditory brainstem implant required.
Narrowed left IAC. Absent cochlear nerve.
Right IAC atresia, coronal view. A very narrow canal leads to a normal middle ear.
Atresia of the right IAC. A rudimentary passage remains. Undersized, malformed cochlea. The attic ossicles appear normal.
Left IAC atresia. Tiny canal leading to a normal middle ear. No EABR response. Managed with a Bonebridge bone conduction active implant.