INNER EAR DEFORMITIES

The inner ear is an organ of both hearing and balance. Deformities may afflict either the hearing mechanism (cochlea), the balance (vestibular) sections, or both.

Cochlear deformities may be minor or gross. Minor deformities frequently affect the internal structure of the cochlea (Mondini deformities) that may result in partial or total losses. Alternatively, the snail-shell cochlear may suffer more substantial globular, undersized or total absence abnormalities. The more advanced problems are almost invariably associated with severe or total deafness.

The practical implications of cochlear deformities are the extent to which they affect recovery of hearing by cochlear implantation. In the absence of other factors, the mild abnormalities frequently respond optimally to implantation, whereas more severe structural deficiencies may permit only partial response to implantation, or, as in complete atresia, may prevent implantation. Brainstem implants may be appropriate in the latter cases.

Conversely aberrations of the balance mechanisms may produce only slight hearing difficulties, and may not intrude on the efficacy of implantation. A common example is the large/dilated vestibular aqueduct. This channel between the inner ear and the brain surrounds is only vestigial or absent in most cases. Larger examples however, that maintain communication between the sites, commonly cause profound losses at birth or at a later stage. In the latter instance, hearing may be good at birth, only to become unstable, fluctuant and deteriorating years later. All cases of progressive childhood deafness should be subjected to CT scanning to check for this condition. The delayed hearing losses are often triggered by minor head trauma.
Fortunately, the large ventricular aqueduct patient usually responds well to cochlear implantation. Cases of later onset, who have mastered speech partially or fluently, generally offer some of the best responses to implantation.

Regrettably, inner ear deformities may be part of more major head and neck congenital syndromes, that may incur concurrent other major disabilities. These aspects need to be considered when implantation is contemplated.

More Information:

Inner Ear