

## PAEDIATRIC EIGHTH NERVE DISORDERS

The eighth cranial nerve connects the ear to the brain, for hearing and balance sensation. In childhood, several problems may afflict the nerve

Failure of development may result in a totally (aplastic) or partially (dysplastic) absent nerve, or one that is deformed in structure. The abnormalities may be unilateral or bilateral. Poor nerve development may be found in isolation, but unfortunately is often associated with other difficulties. Inner ear deformities may or may not be present, but other major syndromal or developmental aberrations may also be present. Nerve dysplasia is relatively common in CHARGE disorders. Associated nerve deafness may be partial or profound.

An intact nerve may not, however, function normally. In Auditory Dyssynchrony (otherwise described by the misnomer “auditory neuropathy spectrum disorder”), the auditory stimuli fail to transmit from the receptor hair cells of the cochlea into the auditory nerve itself. The problem is noted in cases of premature birth, but may improve with time in a minority of cases. These cases respond well to cochlear implantation, as the implant stimulates the normally functioning eighth nerve directly, bypassing the transmission site. Normal cochlear hair cell responses often aid this diagnosis.

Alternatively, in true auditory neuropathy, the nerve itself may be damaged by disease. Cytomegalovirus, rubella and meningitis are common causes, frequently with concurrent hair cell damage. In neuropathy cases the damage to the nerve precludes management by cochlear implantation.

From the implant surgeon’s aspect, it is essential to differentiate the above conditions, to determine suitability for implantation. Retention of partial hearing, and a normally appearing eighth nerve on MRI scanning are encouraging signs. Implantation of the profoundly deaf ear without an MRI scan is especially unwise, for this reason.

In doubtful cases, where the hearing is lost and the nerve is suspect on an MRI scan, Electric Auditory Brainstem Response testing (EABR) may be beneficial to ascertain nerve function, although false negative outcomes are possible, and the test poorly predicts outcomes where partial responses are noted.

It should be reiterated that cases of congenital profound sensorineural deafness, unilateral or bilateral, should be carefully assessed to exclude vestibular nerve pathology, due to the adverse effect of this on cochlear implantation.

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### Other Locations

Beenleigh

Sunnybank

Mt Ommaney

Caboolture

More Information:

[Meningitis-related Deafness](#)

[Inner Ear Deformities](#)

[Acoustic Schwannoma](#)



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