CONDITIONS OF THE INNER EAR Congenital sensorineur

CONDITIONS OF THE INNER EAR Congenital sensorineural hearing loss

Half of congenital sensorineural hearing loss is genetic and half is acquired. Of the genetic hearing loss 75% is non-syndromic, of which the most common is a connexin 26 gene mutation. Syndromic causes include Usher, Pendred, Jervell and Lange-Nielsen, Waardenburg, Treacher Collins, Alport, Stickler, neurofibromatosis type 2 and branchio-oto renal syndromes. Acquired causes are intrauterine infections, including rubella, toxoplasmosis and cytomegalovirus infection; perinatal hypoxia, jaundice and prematurity; and postnatal meningitis. All newborn babies in the UK are now screened at birth for deafness b y measuring otoacoustic emissions in response to ‘clicks’ in the ear. Children failing this are referred for auditory brainstem response to establish hearing thresholds (Figure 51.26). If some hearing is present, the early fitment of hearing aids can maximise the neural plasticity that is pres ent in the developing brain. If a child has a profound hearing loss, early intervention with a cochlear implant is essential for the development of the auditory cortex (Figure 51.27). Most cases of prof ound sensorineural hearing loss are due to loss of cochlear hair cells, so an implant inserted through the round window can selectively stimulate the cochlear neurones, which usually remain intact.

Figure 51.26 Evoked-response audiometry. A simple non-invasive objective test of hearing thresholds. (Reproduced with permission from O’Donoghue GM, Bates GJ, Narula A. Clinical ENT: an illustrated textbook . Oxford: Oxford University Press, 1991.)

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