**Exostosis of the external auditory canal**

**DEFINITION**

 An exostosis is a benign growth of periosteal bone, which forms a smooth, sessile, hemispherical swelling in the deep part of the meatus, adjacent to the tympanic membrane.

 Exostoses are usually multiple, occurring in a group of three, and are bilateral. They may arise from the anterior and posterior walls of the external auditory canal. They are usually found as an incidental finding during the examination of an asymptomatic patient.

 There is evidence that prolonged exposure to cold water in the external auditory canal leads to increasing size and incidence of exostoses.

**DIAGNOSIS**

 The diagnosis of exostoses is usually made clinically from the characteristic otoscopic appearances of multiple and usually bilateral sessile, hemispherical, bony swellings arising deep in the external auditory canal, adjacent to the tympanic membrane. It is usually straightforward to differentiate them from **an osteoma**, which is generally a unilateral, solitary, discrete, pedunculated mass, arising from the lateral part of the bony external auditory canal.

 Exostoses are generally an incidental finding and asymptomatic when small.

**MANAGEMENT OPTIONS**

 In the majority of cases, no treatment is required as the patient is asymptomatic. However, general advice about **avoidance of cold water** should be given, with a recommendation to use earplugs or a wet suit hood, for water sports.

 Hearing loss due to debris collection, or otitis externa, can be managed **medically**, by meticulous aural toilet using a microscope and suction. Topical application of steroid, antibiotic, antifungal or combination ear drops, may be necessary to treat inflammation or infection.

**Surgery**

 Surgery to remove the exostoses and enlarge the meatus by a meatoplasty procedure is indicated for cases refractory to medical treatment, causing recurrent or persisting otitis externa, frequent cerumen obstruction causing hearing loss and cases where wider access is required for middle ear surgery.

**otalgia**

 A central common pathway for otalgia, whether primary or referred, is probably the spinal tract nucleus of the trigeminal nerve. Fibers from cranial nerves V, VII, IX and X and cervical nerves C2, C3 converge here and all play some role in sensory supply of the ear and temporal bone.

 Pain may be **primary** from the ear like otitis externa, furunculosis ,otitis media , bullous myringitis, etc or it may be **referred** from any site of the head and neck like temporomandibular joint diseases, sinusitis ,dental cause, tonsillitis, post tonsillectomy ,cervical spondylosis ,tumors of mouth ,pharynx , etc.

**The structure of the cochlear duct**

 The cochlear duct is subdivided by two longitudinally running membranes that separate three chambers, the **scala tympani**, **scala media** and **scala vestibule**.

 **The organ of Corti** runs in a spiral along the floor of the scala media , situated on its lower boundary, an acellular layer called the **basilar membrane**. The scala media is triangular in section, the other boundaries represented by Reissner's membrane, which runs obliquely with respect to the basilar membrane from the modiolus, to the lateral wall that runs along the inside of the bony wall.

 The composition of the fluid within the scala media, the endolymph, is containing high potassium but low sodium levels. This contrasts with the scala tympani and scala vestibuli, both of which are filled with perilymph that has high sodium content.

 The **sensory region** consists of two types of sensory hair cell that are characterized by an apical bundle of hairs called stereocilia : ***Inner hair cells*** usually form a single longitudinal row running along the inner side of the sensory region , whilst ***outer hair cells*** form three rows running along the outer side of the epithelium.

 *The functions* of the organ of Corti are to detect sounds and decompose them into their component frequencies, in the process converting the physical vibrations into an electrical response (mechanoelectrical transduction) and then causing neural signals to be transmitted along the auditory nerve and higher auditory pathway for central processing. Sound pressure waves in cochlear fluids set up waves of motion along the basilar membrane (called travelling waves) which peak in different parts of the spiral according to frequency, The hair cells detect the basilar membrane motion, being stimulated more strongly at the point coinciding with the peak of the travelling wave than elsewhere.

 The innervation pattern of the organ of Corti strongly suggests that inner hair cells contribute most to the neural signaling representing sensory transduction and processing by the cochlea. However, the cochlea is also known to contain an active amplifier that enhances the ability to detect and separate frequencies in sound. The outer hair cells are thought to represent this amplifier.

**INNERVATION OF THE ORGAN OF CORTI**

 Acoustic information from the hair cells is transferred by the auditory portion of the VIIIth cranial nerve (the vestibulocochlear nerve) to the ipsilateral cochlear nuclear complex in the brain stem. The auditory nerve is composed of afferent fibres projecting from **spiral ganglion neurons**, the cell bodies of which reside in the modiolus.

 The neurons are of two types, type I that innervate the inner hair cells and type II that innervate the outer hair cells. The majority of spiral ganglion neurones (up to 95 percent) are type I and innervate the inner hair cells while the remaining 5 percent innervate the outer hair cells. The relatively small number of fibres to the outer hair cells, which are three times more numerous than inner hair cells, suggests they are not the primary signalling pathway from the cochlea. What they signal to the brain is uncertain, although there is evidence that they respond to very loud sounds.

**Ototoxicity**

 Ototoxicity is chemical injury to the labyrinth occurring as a side effect of pharmacotherapy. An ototoxic insult may affect the hearing, vestibular function or both.

 The most widely used drugs causing irreversible ototoxicity are the aminogIycosides and the chemotherapeutic agent, cisplatin. For these drugs the toxicity to the labyrinth may be dose-limiting. Other drugs, particularly the salicylates and the loop diurectics, tend to cause a temporary hearing loss when used at therapeutic doses.

**AMINOGLYCOSIDES**

 Millions of people throughout the world are at risk of developing amino glycoside ototoxicity. The major target for aminoglycoside ototoxicity is the sensory neuroepithelium of the inner ear. Within the cochlea, the **outer hair cells** are more susceptible than the inner hair cells. Loss of the cochlear hair cells results in a secondary degeneration of the auditory nerve.

There is a genetic susceptibility to amino glycoside ototoxicity ,( mitochondrial). The phenotypic expression is of a maternally transmitted, nonsyndromic susceptibility to aminoglycosides.

 Loop diuretics (ethacrynic acid or frusemide) potentiatethe otoxicity of aminoglycosides by increasing thepermeability of strial blood vessels which leads to anincreased concentration of aminoglycosides within thescala media.

 Randomized, prospective trials **have not** revealed significant differences in ototoxicitywhen comparing individual aminoglycosides, includinggentamicin, amikacin, tobramycin, netilmicin, isepamicin.

 Aminoglycoside ototoxicity may occur following either systemic administration, peritoneal dialysis or topical application to the tympanic cavity. The latter route of administration may result from the application of aminoglycoside- containing ear drops in the presence of an aural perforation or a ventilation tube.

**Natural history**

 Labyrinthine injury will usually be gradual, progressive, symmetrical *bilaterally* and *permanent*, but unilateral hearing loss and cases of partial recovery have been reported. Ototoxicity develops once a threshold dose is exceeded, but this threshold varies between individuals. The basal regions of the cochlea are more extensively injured than apical regions.

 Although ototoxicity is usually gradual, a sudden profound sensorineural hearing loss may follow a short duration of treatment, or even single dose, of aminoglycoside administered systemically or topically to the round window.

 ***Risk factors*** for ototoxicity include the cumulative drug dose, the duration of treatment, bacteraemia and renal or liver failure. genetic susceptibility also play some role. Premature babies treated in a neonatal intensive care are at a higher risk than normal children of developing a sensorineural hearing loss.

**CISPLATIN**

 Cisplatin is a chemotherapeutic agent effective against solid tumours , including head and neck carcinoma. Therapeutic doses are limited by its ototoxicity. The most frequent pattern of hearing loss is a bilateral, symmetric, progressive, high frequency sensorineural loss, caused by a loss of cochlear outer, and to a lesser extent inner hair, cells.

**Loop diuretics**

 The potentiation of aminoglycoside ototoxicity by the loop diuretics ethacrynic acid and frusemide has already been discussed, but these diuretics can cause a hearing loss when used as sole agents. The hearing impairment is usually a ***reversible***, flat sensorineural hearing loss, but a permanent profound loss can occur.

**Salicylates & Quinine**

 usually cause a reversible sensorineural hearing loss

**Erythromycin**

 Erythromycin can cause a transient ototoxicity when used in high doses systemically. There are reports of reversible ototoxicity with other macrolide antibiotics, including azithromycin and clarithromycin.

**MANAGEMENT**

* **Prevention and protection of the inner ear**

 Recognition of at-risk groups allows the modification of therapeutic regimes to reduce drug dosages and/or more closely monitor individuals in the hope of avoiding ototoxicity. the coadministration of antioxidants or iron-chelators may also help to decrease ototoxicity.

* **Early recognition of ototoxicity**
* **clinical presentation**

For both aminoglycosides and cisplatin, the earliest symptoms of ototoxicity are tinnitus and/or reduced hearing.

* **Objective auditory monitoring**

 It is recommended that objective auditory monitoring should be undertaken for all patients, and that it is mandatory when symptomatic, or in children, as the latter group cannot be relied upon to report auditory symptoms when they do occur.

 Early ototoxicity can be recognized by a high-frequency sensorineural hearing loss which progresses to lower frequencies over time. Therefore, the key is to monitor the highest frequencies possible either by ultra-high (up to 12 kHz) frequency pure-tone audiometry or otoacoustic emissions.

* **Vestibular monitoring**

**Treatment :**

Hearing loss should be treated with a hearing aid or cochlear implant. Vestibular dysfunction should be treated with vestibular rehabilitation.