Chapter 34
Conductive and Cochlear Hearing Loss

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Keypoints

1. Any kind of hearing loss may be accompanied by tinnitus.
2. This chapter describes possible causes of conductive hearing loss located in the external ear and middle ear.
3. Pathologies of this area include neoplastic changes (e.g., tumors), inflammatory disease (e.g., otitis media), or disorders of unknown origin (e.g., otosclerosis).
4. Cochlear hearing loss of genetic origin can be classified in to syndromic and non-syndromic forms.
5. Labyrinthitis can occur due to bacterial or viral infection or in the context of immunological disease.

Keywords Hearing loss • Inner ear • Middle ear • Otitis media • Otosclerosis • Tinnitus

Abbreviations

AIED	Autoimmune inner ear disease
NSHL	Non-syndromic hearing loss
SHL	Syndromic hearing loss

Introduction

Virtually any pathology involving the ear appears to have the capacity to cause hearing loss and tinnitus may accompany the hearing loss at any time. Pathology involving the external ear and middle ear leads to conductive hearing loss, whereas a pathological change in the cochlea causes cochlear hearing loss. In most cases, tinnitus cannot be regarded as a direct consequence of the pathological changes, but rather hearing loss causes deprivation of input to the central auditory pathway activating neural plasticity (see Chap. 10). The tinnitus that occurs together with hearing loss is, in most cases, subjective tinnitus. Only very rarely can conductive hearing loss cause objective tinnitus, generally because of vascular turbulence. The effect of conductive hearing loss is the same as that of an earplug, and tinnitus might be interpreted as intensified perception of body sounds that occurs because sounds from the outside are reduced.

If the hearing loss is reduced or eliminated, tinnitus may also disappear after a certain period. Many forms of conductive hearing loss, in particular, can be treated successfully by surgical interventions (Chap. 83), leading to improvement of hearing and disappearance of tinnitus. If hearing loss persists, the accompanying tinnitus also usually persists. Often, the frequency of maximum hearing loss coincides with the frequency of the tinnitus.

Causes of Conductive Hearing Loss

Changes in the Territory of the External Auditory Canal

Pathologies leading to conductive hearing loss with subsequent tinnitus can be of a mechanical, inflammatory, or neoplastic nature. It may affect the ear canal or the middle ear. Genetic factors as well as exogenous noxious agents (thalidomide embryopathy [1]) may be involved in the development of auditory canal anomalies.

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Obstruction of the external auditory canal by wax is a mundane reason for sudden onset of hearing loss and tinnitus. Inflammation of the external auditory canal (swimmer's otitis) may cause very painful swelling, redness, and discharge from the external auditory canal due to bacterial (*Pseudomonas aeruginosa*, *Staphylococcus aureus*) or, more rarely, fungal infection. When the external auditory canal becomes blocked, tinnitus may develop together with the hearing loss [2]. Neoplastic disorders leading to increasing stenosis and finally occlusion of the external auditory canal may be benign or malignant. Exostoses[1] of the external auditory canal are benign new bone formations that occur with an incidence of 3–6% [3]. They may cause recurrent inflammation of the external ear canal with conductive hearing loss and transient tinnitus [4]. Apart from the constitutional factors, repeated thermal irritation of the external auditory canal by frequent contact with cold water (swimmer's ear, surfer's ear) has long been regarded as a predisposing factor [5].

Malignant neoplasms of the external auditory canal are far less common. These may arise from skin cells (basal cell carcinoma, squamous cell carcinoma, malignant melanoma) or in the ceruminous glands (adenocarcinoma, adenoid cystic carcinoma) [6]. Congenital changes resulting in partial occlusion or atresia of the external auditory canal can cause hearing loss and possibly tinnitus. Severe forms may also be accompanied by an auricular anomaly in addition to complete atresia of the external auditory canal. As syndromic components, external ear anomalies may also occur in association with further dysmorphologies, for example, mid-facial dysplasia (mandibulofacial dysostosis, Treacher-Collins syndrome [7]), or craniofacial dysostoses (Crouzon syndrome [8]). These anomalies may be accompanied by additional anomaly of the middle ear, which can be detected by high-resolution computed tomography [9].

Pathological Conditions of the Middle Ear

Eustachian tube dysfunction leads to impairment of the function of the middle ear. Disturbances affecting opening and closure of the Eustachian tube are an important factor in the pathogenesis of many middle ear conditions because the ventilation and drainage of the middle ear no longer function properly. The sequelae may include chronic mucoid otitis media (glue ear), recurrent acute otitis media, or chronic otitis media. Disturbances affecting opening and closure of the Eustachian tube may be caused by mechanical blockage of the tubal orifice (adenoids, tumor), by inflammatory swelling of the tubal mucosa, or by muscular insufficiency such as may occur in individuals with cleft palate [10, 11]. Conductive hearing loss, sometimes accompanied by tinnitus, may develop subsequent to Eustachian tube dysfunction.

A patulous Eustachian tube is a special condition in which there is a permanently open connection between the tube and the nasopharynx. This condition may entail a variety of symptoms, such as autophony, aural fullness, and the unpleasant sensation of hearing one's own respiratory sounds. Reduced muscle tone and weight loss are the main factors predisposing to the development of patulous Eustachian tube.

Acute otitis media often occurs secondary to rhinitis or pharyngitis. The common organisms that cause otitis media are streptococci, *Haemophilus influenzae*, and staphylococci. The main symptoms are earache and hearing loss. In many cases tinnitus may occur as an additional symptom. The tinnitus may be objective, having a vascular origin. It may be perceived as pulsatile pounding and buzzing sounds that occur in acute inflammatory stage by dilatation of vessels and high pulse amplitude of blood flow. Chronic otitis media is an umbrella term that covers several different middle ear pathologies. If it lasts for several years, there may be different extents of irreversible tissue destruction in the middle ear. In addition to perforation of the eardrum and defects in the ossicular chain, enzymatic degradation processes associated with cholesteatoma[2] may occur and may, in particular, lead to destructive erosion of the bony walls of the middle ear toward the cranial base. The common consequence of these

[1] Exostosis: A cartilage-capped bony projection arising from any bone that develops from cartilage. Stedman's Electronic Medical Dictionary.

[2] Cholesteatoma: Squamous metaplasia or extension of squamous cell epithelium inward to line an expanding cystic cavity that may involve the middle ear or mastoid, erode surrounding bone, and become filled with a mass of keratinized squamous cell epithelial debris, usually resulting from chronic otitis media. The lesion often contains cholesterol clefts surrounded by inflammatory and foreign body giant cells, hence the name *cholesteatoma*. Stedman's Electronic Medical Dictionary.

pathologies is increasing conductive hearing loss with recurrent mucous or purulent discharge. Individuals in whom cholesteatoma is associated with erosion of the bony walls of the structures of the labyrinth, may also have sensorineural hearing loss and even deafness because the pathologies have spread to the cochlea.

There are three different causes of traumatic eardrum perforation: direct mechanical or thermal injury, a pressure wave in the external auditory canal, or an otobasis fracture. Depending on the extent of hearing loss, the possibility of a concomitant injury to the ossicular chain or the cochlea must also be considered.

Otosclerosis causes stapedial ankylosis, which in turn causes conductive hearing loss. Otosclerosis can also affect the bony labyrinth characterized by bone resorption and remodeling processes (otospongiosis). Otosclerosis is responsible for 5–9% of all hearing losses and 18–22% of all conductive hearing losses [12]. The condition is encountered almost exclusively in Caucasians, very rarely occurring in Asians and almost never in Blacks [13]. The female-to-male ratio is approximately 2:1. The precise etiology remains unclear [14]. Alongside genetic factors [15], the role of inflammatory processes (localized measles virus infection of the otic capsule [16]), endocrine factors [17], and immunological disease [18] have been considered in the etiopathogenesis.

Independently of sex and age, tinnitus is a concomitant symptom in 65–91% of individuals with otosclerosis [19, 20]. Tinnitus already develops in many individuals with otosclerosis years before the onset of noticeable hearing loss. Tinnitus sometimes persists despite an optimal hearing outcome from surgical management (see Chap. 83).

Otosclerotic processes may spread to the cochlea and that may be responsible for persisting tinnitus. This condition, known as "cochlear otosclerosis" by many authors, causes signs of sensorineural hearing loss [21–23]. It is also associated with changes in the stria vascularis, the organ of Corti and the spiral ligaments, as demonstrated in histopathological and radiological studies [24–26].

Tumors affecting the middle ear are rare. The glomus tumor (synonyms: paraganglioma, chemodectoma) is one kind of benign tumor of the middle ear, displaying destructive growth. Two locations of glomus tumors occur: glomus tympanicum tumors (which are limited to the middle ear) and glomus jugulare tumors (lesions that affect both the middle ear and the bulb of the jugular vein) [27]. In histological terms, the tumors consist of non-chromaffin paraganglionic cells along the course of cranial nerves IX and X.

The majority of glomus tumors occur in adulthood, with a female-to-male predominance of 6:1 [28]. Glomus tumors can generally be diagnosed clinically on the basis of the symptom triad of conductive hearing loss, pulsatile tinnitus, and a red middle-ear tumor that can be seen through the eardrum using otoscopy [29]. In some patients, pulsatile objective tinnitus can be detected objectively by inserting a stethoscope or microphone into the external auditory canal. The sound is probably produced by the formation of microvascular shunts within the tumor mass [30]. With increasing tumor infiltration into the jugular foramen, additional deficits related to caudal cranial nerves IX–XI may become evident.

Facial nerve schwannoma is another – very rare – benign tumor affecting the middle ear, characterized by slowly progressive facial paralysis as well as conductive hearing loss. Wegener's granulomatosis,[3] Langerhans cell histiocytosis,[4] and sarcoidosis[5] are among the tumor-like lesions that potentially involve the middle ear and are also accompanied by tinnitus, in addition to conductive hearing loss [31].

Malignant tumors that may involve the middle ear are squamous cell carcinoma and adenoid cystic carcinoma.

[3] Wegener's granulomatosis: a disease, occurring mainly in the fourth and fifth decades, characterized by necrotizing granulomas and ulceration of the upper respiratory tract, with purulent rhinorrhea, nasal obstruction, and sometimes with otorrhea, hemoptysis, pulmonary infiltration and cavitation, and fever; exophthalmos, involvement of the larynx and pharynx, and glomerulonephritis may occur; the underlying condition is a vasculitis affecting small vessels, and is possibly due to an immune disorder. Stedman's Online Medical Dictionary.

[4] Langerhans cell histiocytosis: a set of closely related disorders unified by a common proliferating element, the Langerhans cell. Three overlapping clinical syndromes are recognized: a single site disease (eosinophilic granuloma), a multifocal unisystem process (Hand-Schuller-Christian syndrome), and a multifocal, multisystem histiocytosis (Letter-Siwe syndrome.) Formerly this process was known as histiocytosis X. Stedman's Online Medical Dictionary.

[5] Sarcoidosis: a systemic granulomatous disease of unknown cause, especially involving the lungs with resulting interstitial fibrosis, but also involving lymph nodes, skin, liver, spleen, eyes, phalangeal bones, and parotid glands; granulomas are composed of epithelioid and multinucleated giant cells with little or no necrosis. Stedman's Online Medical Dictionary.

Causes of Sensorineural Hearing Loss

The causal factors responsible for the development of sensorineural hearing loss can be many and varied. Apart from congenital factors, the etiology may include infectious diseases, autoimmune diseases, toxic lesions (see Chap. 42), noise-related injury (see Chap. 37), traumatic damage (see Chap. 67), or age-induced changes (presbycusis, see Chap. 36). Furthermore, fluctuating sensorineural hearing loss is one of the three signs of Ménière's disease (see Chaps. 38 and 60) and sudden hearing loss (Chap. 56). Depending on the cause, the form of sensorineural hearing loss can be different and have varying severity. All forms of hearing loss may be accompanied by tinnitus of varying severity. The following discussion will deal with those forms of sensorineural hearing loss that are not covered in separate chapters of their own.

Sensorineural Hearing Loss of Genetic Origin

Impairment of hearing is the most common sensorineural pathology affecting humans. Approximately, one-half of all the cases of prelingual hearing impairment have a genetic cause [32–34]. A distinction is made between genetic hearing loss occurring as a component of a specific (genetic) syndrome (30%) and non-syndromic hearing loss that occurs in the absence of any other genetic diseases or developmental anomalies [35]. Syndromic hearing loss (SHL) can be inherited in an autosomal dominant, autosomal-recessive, or X-linked manner [34], and can be associated with developmental anomalies of the inner ear or petrous portion of the temporal bone like Mondini[6] or Scheibe[7] dysplasia; frequently there is also a link with other organic disorders, such as thyroid disease (Pendred syndrome), renal dysfunction (Alport syndrome), or eye disease (Usher syndrome) [34]. Children with Down's syndrome are more likely to have congenital permanent inner ear hearing loss than the general population (which has an incidence of 1:1,000). From teenage years onward, they are likely to develop degenerative cochlear changes, and most will have significant hearing loss by the age of 40 years [34, 36]. Of non-syndromic hearing loss (NSHL), 80% have autosomal recessive, 18% an autosomal dominant, and 2% an X-linked or mitochondrial inheritance pattern [32]. In the majority, a single gene defect leads to the phenotypical development of hearing loss, which may not have its onset until later in life. In recent years, many gene loci and mutations have been described that are responsible for various forms of hearing loss. For example, known mutations affect the GJB2 gene – which codes for connexin-26 – and the GJB6 gene (connexin-30) [37] (see also Chap. 7).

Infections

Inflammation of the cochlea may develop together with acute or chronic otitis media when bacteria enters the cochlea through the round or oval window, or may develop together with meningitis where bacteria enters the cochlea and the vestibular apparatus via the internal auditory canal, the cochlear aqueduct, or the vestibular aqueduct. The resulting sensorineural hearing loss is often accompanied with tinnitus. Because of the involvement of the vestibular apparatus, the symptoms are often dominated by pronounced rotatory vertigo accompanied by nausea and vomiting. Meningitis is often followed by the cochlea being filled with bone (labyrinthitis ossificans or "white cochlea"), with complete obliteration of the membranous labyrinth [38]. The bacteria in borreliosis or syphilis can spread via blood to the inner ear. Many kinds of infections can cause damage to the cochlea or the inner ear as a whole. Especially serious ones are congenital rubella or cytomegalovirus infections, which may lead to severe sensorineural hearing loss or to deafness. Of the postnatal viral infections of the inner ear, epidemic parotitis (mumps) typically causes unilateral deafness without vestibular involvement [39]. Herpes zoster oticus that is caused by reinfection with the varicella zoster virus can cause blisters in the external auditory canal and the pinna in addition to sensorineural hearing loss and

[6] Mondini dysplasia: Congenital anomaly of osseus and membranous otic labyrinth characterized by aplastic cochlea and deformity of the vestibule and semicircular canals with partial or complete loss of auditory and vestibular function; may be associated with dilated vestibular aqueduct and spontaneous cerebrospinal fluid otorrhea resulting in meningitis.

[7] Scheibe dysplasia: Hearing impairment due to cochleosaccular dysplasia; usually autosomal recessive inheritance. Stedman's Online Medical Dictionary.

tinnitus, vestibular symptoms, and facial nerve palsy. When the facial nerve is involved it is known as the Ramsey Hunt syndrome [40]. In addition to infection of the nerve sheaths, the symptoms may also be caused by secretion of toxins into the perilymph spaces of the inner ear [41].

Immunogenic Labyrinthitis

Sensorineural hearing loss may occur together with immunological diseases as a heterogeneous group of sensorineural hearing loss types under the heading "autoimmune inner ear disease" (AIED) [42]. Possible target structures for antibodies are the stria vascularis in the organ of Corti and the blood vessels supplying the inner ear [43]. AIED is characterized by progressive, often fluctuating and usually bilateral, sensorineural hearing loss with tinnitus and vertigo, more often in women. Progression over time is too rapid to suggest presbycusis and too slow for a diagnosis of sudden deafness. AIED patients respond well to immunosuppressant corticosteroid therapy. Some patients with AIED present with a systemic autoimmune disease, such as Wegener's granulomatosis (see footnote 3), Cogan syndrome,[8] or relapsing polychondritis[9] [44]. No specific test battery that will unequivocally show the presence of an autoimmune reaction to structures of the inner ear has yet been described. The recommendation is to use general laboratory tests (antinuclear antibodies, antineutrophil cytoplasmatic antibodies, etc.) to screen patients who are suspected of having an autoimmune disease for the presence of systemic signs [45].

[8] Cogan syndrome: Typical Cogan syndrome is characterized by interstitial keratitis and vestibuloauditory dysfunction. There is generally a brief episode of inflammatory eye disease (interstitial keratitis) followed by bilateral audiovestibular symptoms. The interstitial keratitis usually occurs with sudden onset and is characterized by photophobia, lacrimation, and eye pain. The vestibuloauditory dysfunction is usually bilateral, presenting with tinnitus, sensorineural hearing loss, and acute episodes of vertigo.

[9] Relapsing polychondritis: a degenerative disease of cartilage producing a bizarre form of arthritis, with collapse of the ears, the cartilaginous portion of the nose, and the tracheobronchial tree; death may occur from chronic infection or suffocation because of loss of stability in the tracheobronchial tree; of autosomal origin. Stedman's Online Medical Dictionary.

Age-Related Hearing Loss

Age-related hearing loss is the commonest of all forms of hearing loss (see Chap. 36). It affects more than 40% of people over the age of 65 [46]. Apart from physiological age-related processes, endogenous and exogenous factors such as hypoxia, exposure to loud noise, hypertension, hypercholesterolemia, or diabetes mellitus may cause or contribute to hearing loss in old age [47]. Consequently, excessive noise exposure and atherosclerosis contribute to the development of presbycusis in industrialized countries. The reported presence of tinnitus together with presbycusis varies between 8 and 72% [48–50]. The risk for the development of tinnitus rises with increasing age and with increasing exposure to noise [51].

References

1. Takemori S, Tanaka Y, Suzuki JI. Thalidomide anomalies of the ear. Arch Otolaryngol, 1976;102(7):425–7
2. Ostrowski VB, Wiet RJ. Pathologic conditions of the external ear and auditory canal. Postgrad Med, 1996;100(3):223–8, 233–7
3. Adams WS. The aetiology of swimmer's exostoses of the external auditory canals and of associated changes in hearing. J Laryngol Otol, 1951;65(4):232–50; concl.
4. Mlynski R, Radeloff A, Brunner K, Hagen R. [Exostoses of the external auditory canal Is the cold water hypothesis valid for patients in continental areas?] HNO, 2008; 56(4):410–6. German
5. Van Gilse, PHG. Des observations ultérieures sur la genèse des exostoses du conduit externe par l'irritation d'eau froide. Acta Otolaryngol, 1938;2:343–352
6. Breau RL, Gardner EK, Dornhoffer JL. Cancer of the external auditory canal and temporal bone. Curr Oncol Rep, 2002;4(1):76–80
7. Horbelt CV. A review of physical, behavioral, and oral characteristics associated with Treacher Collins syndrome, Goldenhar syndrome, and Angelman syndrome. Gen Dent, 2008;56(5):416–9
8. Rice DP. Clinical features of syndromic craniosynostosis. Front Oral Biol, 2008;12:91–106
9. Jahrsdoerfer RA, Yeakley JW, Aguilar EA, Cole RR, Gray LC. Grading system for the selection of patients with congenital aural atresia. Am J Otol, 1992;13(1):6–12
10. Timmermans K, Vander Poorten V, Desloovere C, Debruyne F. The middle ear of cleft palate patients in their early teens: a literature study and preliminary file study. B-ENT, 2006;2(Suppl 4):95–101
11. Phua YS, Salkeld LJ, de Chalain TM. Middle ear disease in children with cleft palate: protocols for management. Int J Pediatr Otorhinolaryngol, 2009;73(2):307–13

12. Chole RA, McKenna M. Pathophysiology of otosclerosis. Otol Neurotol, 2001;22(2):249–57
13. Altmann F, Glasgold A, Macduff JP. The incidence of otosclerosis as related to race and sex. Ann Otol Rhinol Laryngol, 1967;76(2):377–92
14. Markou K, Goudakos J. An overview of the etiology of otosclerosis. Eur Arch Otorhinolaryngol, 2009;266(1):25–35
15. Ali IBH, Thys M, Beltaief N, Schrauwen I, Hilgert N, Vanderstraeten K, Dieltjens N, Mnif E, Hachicha S, Besbes G, Arab SB, Van Camp G. A new locus for otosclerosis, OTSC8, maps to the pericentromeric region of chromosome 9. Hum Genet, 2008;123(3):267–72
16. Niedermeyer HP, et al., Measles virus and otosclerosis. Adv Otorhinolaryngol, 2007;65:86–92
17. Lippy WH, Berenholz LP, Schuring AG, Burkey JM. Does pregnancy affect otosclerosis? Laryngoscope, 2005;115(10):1833–6
18. Dahlqvist A, Diamant H, Dahlqvist SR, Cedergren B. HLA antigens in patients with otosclerosis. Acta Otolaryngol, 1985;100(1–2):33–5
19. Gristwood RE, Venables WN. Otosclerosis and chronic tinnitus. Ann Otol Rhinol Laryngol, 2003;112(5):398–403
20. Sobrinho PG, Oliveira CA, Venosa AR. Long-term follow-up of tinnitus in patients with otosclerosis after stapes surgery. Int Tinnitus J, 2004;10(2):197–201
21. Sellari-Franceschini S, Ravecca F, De Vito A, Berrettini S. [Progressive sensorineural hearing loss in cochlear otosclerosis]. Acta Otorhinolaryngol Ital, 1998;18(4 Suppl 59):59–65
22. Shinkawa A, Sakai M, Ishida K. Cochlear otosclerosis 30 years after stapedectomy confirmed by CT, MRI. Auris Nasus Larynx, 1998;25(1):95–9
23. Linthicum F, Jr. Post-stapedectomy cochlear otosclerosis. Ear Nose Throat J, 2009;88(4):872
24. Hinojosa R, Marion M. Otosclerosis and sensorineural hearing loss: a histopathologic study. Am J Otolaryngol, 1987;8(5):296–307
25. Ramsden R, Rotteveel L, Proops D, Saeed S, van Olphen A, Mylanus E. Cochlear implantation in otosclerotic deafness. Adv Otorhinolaryngol, 2007;65:328–34
26. Mafee MF. Use of CT in the evaluation of cochlear otosclerosis use of CT in the evaluation of cochlear otosclerosis. Radiology, 1985;156(3):703–8
27. Alford BR, Guilford FR. A comprehensive study of tumors of the glomus jugulare. Laryngoscope, 1962;72:765–805
28. Gulya AJ. The glomus tumor and its biology. Laryngoscope, 1993;103(11 Pt 2 Suppl 60):7–15
29. Horn KL, Hankinson H. (1994) Tumors of the jugular foramen, in Neurotology, RK Jackler, DE Brackmann, Editors, Mosby: New York, 1059–64
30. Sismanis A. (1997) Pulsatile tinnitus, in Clinical otology, GB Hughes, ML Pensak, Editors, Thieme: New York, 445–60
31. Wackym PA, Friedman I. (2000) Unusal tumors of the middle ear and mastoid, in Tumors of the ear and temporal bone, RK Jackler, CLW Driscoll, Editors, Lippincott Williams & Wilkins: Philadelphia, 128–45
32. Birkenhager R, Aschendorff A, Schipper J, Laszig R. [Nonsyndromic hereditary hearing impairment]. Laryngorhinootologie, 2007;86(4):299–309; quiz 310–3
33. Kitamura K, Takahashi K, Tamagawa Y, Noguchi Y, Kuroishikawa Y, Ishikawa K, Hagiwara H. Deafness genes. J Med Dent Sci, 2000;47(1):1–11
34. Bayazit YA, Yilmaz M. An overview of hereditary hearing loss. ORL J Otorhinolaryngol Relat Spec, 2006;68(2):57–63
35. Smith RJ, Bale JF, Jr, White KR. Sensorineural hearing loss in children. Lancet, 2005;365(9462):879–90
36. Hess C, Rosanowski F, Eysholdt U, Schuster M. [Hearing impairment in children and adolescents with Down's syndrome]. HNO, 2006;54(3):227–32
37. Batissoco AC, Abreu-Silva RS, Braga MC, Lezirovitz K, Della-Rosa V, Alfredo T, Jr, Otto PA, Mingroni-Netto RC. Prevalence of GJB2 (connexin-26) and GJB6 (connexin-30) mutations in a cohort of 300 Brazilian hearing-impaired individuals: implications for diagnosis and genetic counseling. Ear Hear, 2009;30(1):1–7
38. Douglas SA, Sanli H, Gibson WP. Meningitis resulting in hearing loss and labyrinthitis ossificans – does the causative organism matter? Cochlear Implants Int, 2008;9(2):90–6
39. Hviid A, Rubin S, Muhlemann K. Mumps. Lancet, 2008;371(9616):932–44
40. Sweeney CJ, Gilden DH. Ramsay Hunt syndrome. J Neurol Neurosurg Psychiatry, 2001;71(2):149–54
41. Sugiura M, Naganawa S, Nakata S, Kojima S, Nakashima T. 3D-FLAIR MRI findings in a patient with Ramsay Hunt syndrome. Acta Otolaryngol, 2007;127(5):547–9
42. McCabe BF. Autoimmune sensorineural hearing loss. Ann Otol Rhinol Laryngol, 1979;88(5 Pt 1):585–9
43. Ruckenstein MJ. Autoimmune inner ear disease. Curr Opin Otolaryngol Head Neck Surg, 2004;12(5):426–30
44. Broughton SS, Meyerhoff WE, Cohen SB. Immune-mediated inner ear disease: 10-year experience. Semin Arthritis Rheum, 2004;34(2):544–8
45. Bovo R, Ciorba A, Martini A. The diagnosis of autoimmune inner ear disease: evidence and critical pitfalls. Eur Arch Otorhinolaryngol, 2009;266(1):37–40
46. Cruickshanks KJ, Wiley TL, Tweed TS, Klein BE, Klein R, Mares-Perlman JA, Nondahl DM. Prevalence of hearing loss in older adults in Beaver Dam, Wisconsin The Epidemiology of Hearing Loss Study. Am J Epidemiol, 1998;148(9):879–86
47. Mazurek B, Stöver T, Haupt H, Gross J, Szczepek A. [Pathogenesis and treatment of presbyacusis current status and future perspectives]. HNO, 2008;56(4):429–32; 434–5
48. Rosenhall U, Karlsson AK. Tinnitus in old age. Scand Audiol, 1991;20(3):165–71
49. Nondahl DM, Cruickshanks KJ, Wiley TL, Klein R, Klein BE, Tweed TS. Prevalence and 5-year incidence of tinnitus among older adults: the epidemiology of hearing loss study. J Am Acad Audiol, 2002;13(6):323–31
50. do Carmo LC, Médicis da Silveira JA, Marone SA, D'Ottaviano FG, Zagati LL, Dias von Söhsten Lins EM. Audiological study of an elderly Brazilian population. Braz J Otorhinolaryngol, 2008;74(3):342–9
51. Ahmad N, Seidman M. Tinnitus in the older adult: epidemiology, pathophysiology and treatment options. Drugs Aging, 2004;21(5):297–305