

Contents

1	Introduction and Definitions	1
2	General Features	2
2.1	Essential Physiological Data	2
2.2	Differential Audiometry	7
2.3	The Differentiation of Various Types of Inner Ear Deafness by Means of the Sound Threshold Curves	13
3	Special Features	17
3.1	Traumatic Inner Ear Deafness	17
3.1.1	Noise and Blast Injury	17
3.1.2	Explosion and Cranial Trauma	22
3.1.3	Rupture of the Windows (Round and Oval)	24
3.1.3.1	Round Window Membrane Rupture	24
3.1.3.2	Ruptures in the Oval Window	28
3.2	Ototoxic Deafness	30
3.2.1	Aminoglycoside Antibiotics (AA)	30
3.2.2	Loop Diuretics	34
3.2.3	Salicylates	36
3.2.4	Atoxyl	38
3.2.5	Other Ototoxic Substances	38
3.3	Infections	40
3.3.1	Inner Ear Deafness in Syphilis	44
3.4	Heredity	44
3.4.1	Monosymptomatic Hereditary Hearing Impairments	45
3.4.2	Syndromes Associated with Hereditary Inner Ear Deafness	49
3.4.2.1	Alport's Syndrome	49
3.4.2.2	Alport-like Syndromes	52
3.4.2.3	Renal Tubular Acidosis (RTA)	54
3.4.2.4	Pendred's Syndrome	55
3.4.2.5	Syndromes Resembling Pendred's Syndrome	55
3.4.2.6	Disturbances of Parathyroid Hormone Metabolism	57
3.4.2.7	Refsum's Syndrome	61
3.4.2.8	Other Storage Diseases	62
3.4.2.9	Diabetes Mellitus	63
3.4.2.10	Sickle Cell Anemia	63

3.4.2.11	Skin Diseases	64
3.4.2.12	Other Hereditary Syndromes	64
3.5	Deafness in Childhood	65
3.5.1	Rubella	69
3.5.2	Cytomegaly	70
3.5.3	Toxoplasmosis	70
3.5.4	Erythroblastosis	71
3.5.5	Perinatal Asphyxia	71
3.5.6	Postnatal Deafness	71
3.6	Vascular and Metabolic Disturbances	73
3.6.1	Acute Inner Ear (Sensori-Neural) Deafness	73
3.6.1.1	Symptomatic Acute Deterioration of Hearing	74
3.6.1.2	Idiopathic "Sudden Hearing Loss"	81
3.6.2	Chronic Inner Ear Deafness	89
3.6.2.1	Disturbances of Renal Function	89
3.6.2.2	Diabetes Mellitus	91
3.6.2.3	Disturbances of Fat Metabolism	93
3.6.2.4	Disturbances of Liver Function	94
3.6.2.5	Disturbances of Thyroid Function	95
3.6.2.6	Vascular Disturbances	96
3.7	Deafness in the Aged	102
3.8	Low Tone Deafness	108
3.9	Middle Ear Causes of Inner Ear Deafness	116
3.10	Immunological Diseases	119
References		125
Subject Index		165