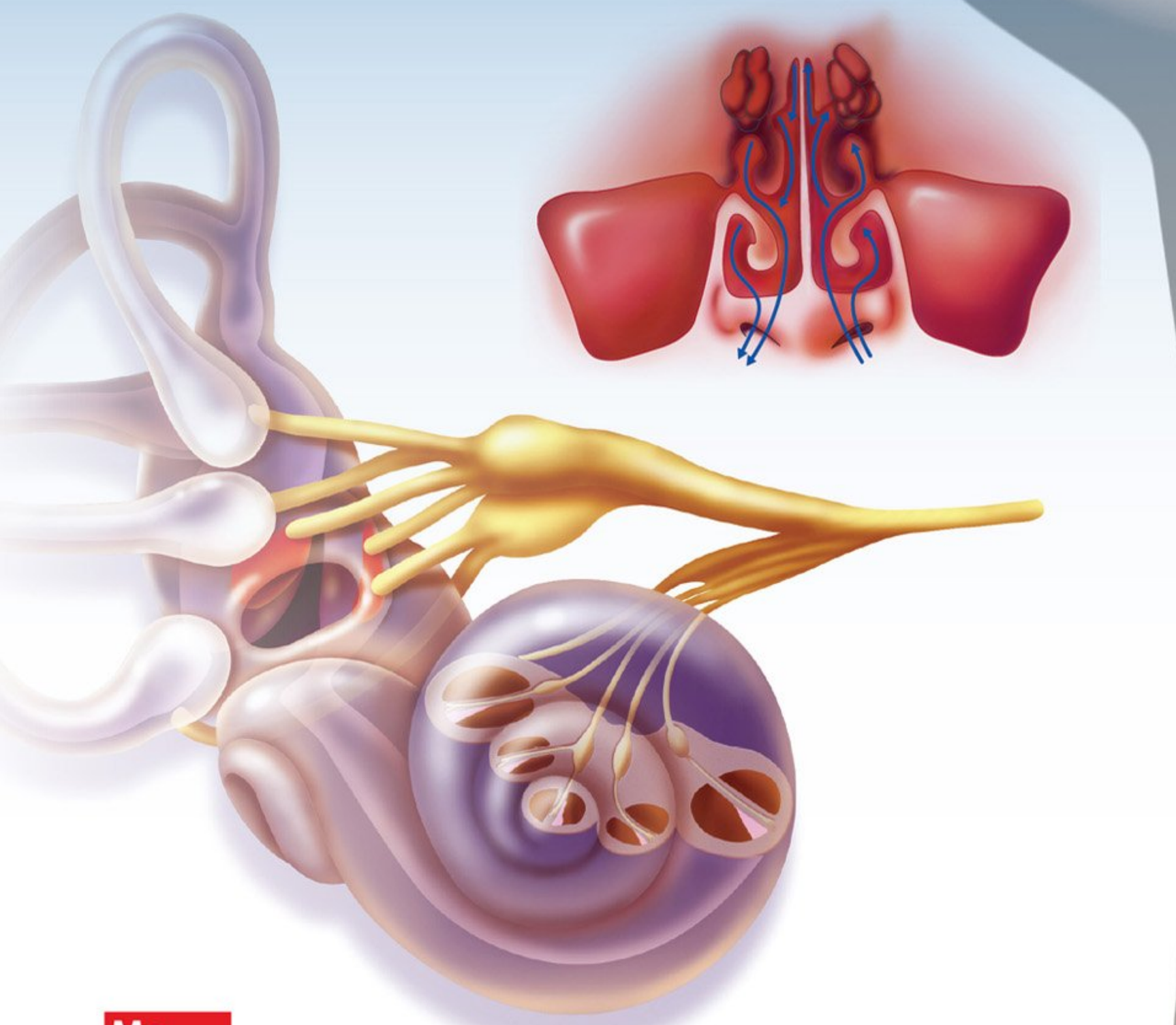
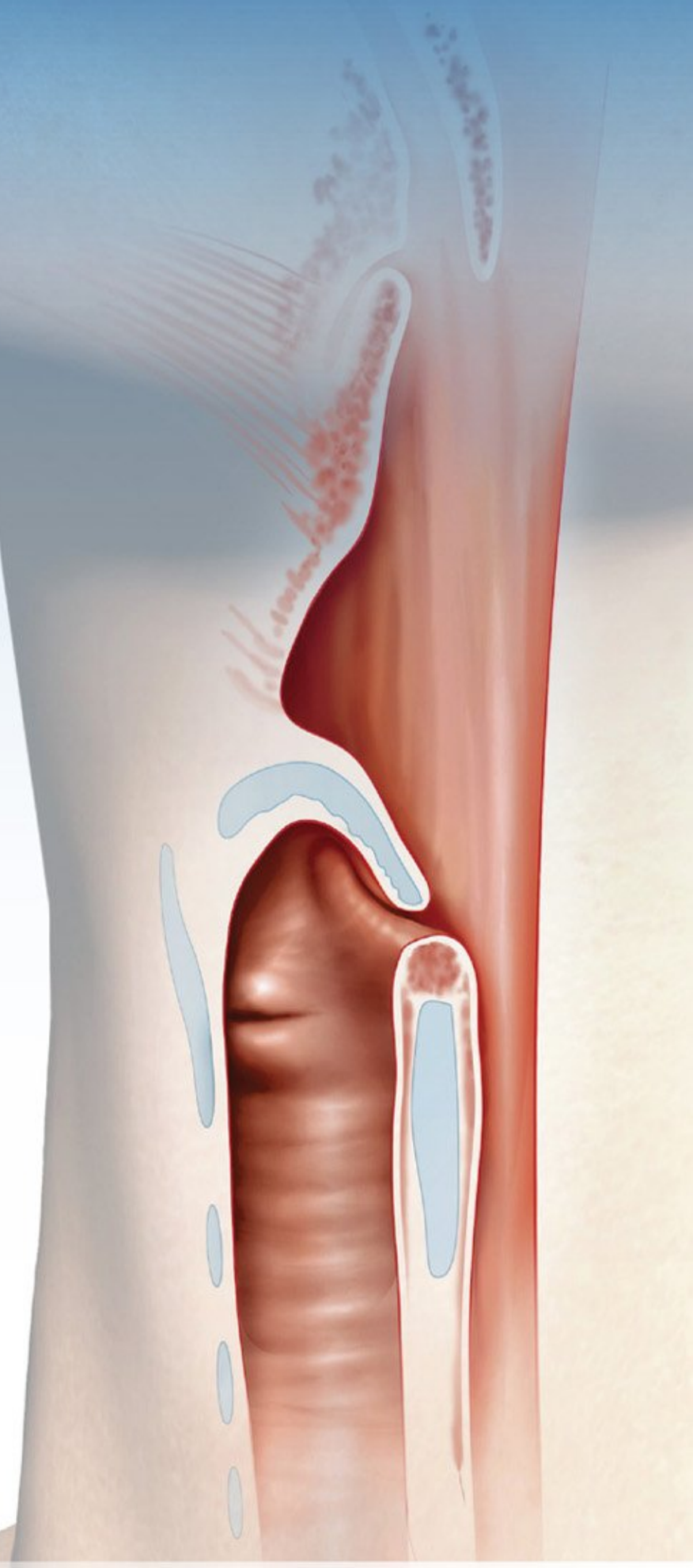
K.J. LEE's ESSENTIAL Otolaryngology

ELEVENTH EDITION

HEAD AND NECK SURGERY





Mc Graw Hill Education

EDITORS: YVONNE CHAN & JOHN C. GODDARD

K.J. Lee's Essential Otolaryngology

Notice

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K.J. Lee's Essential Otolaryngology

Head & Neck Surgery

Eleventh Edition

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Preface

It is a privilege and great pleasure for me to introduce two excellent editors for the 11th edition of K. J. Lee's Essential Otolaryngology—Head and Neck Surgery, Dr. Yvonne Chan and Dr. John Goddard.

T e first edition of Essential Otolaryngology, published in 1973, was based predominantly on my own notes that had helped me through my Board examination. Because of the enthusiastic reception among practicing clinicians and the universal acceptance of this book among residents in the United States and abroad, I have found keeping this book current a most satisfying endeavor. Dr. Anthony Maniglia arranged for the 6th edition to be translated into Spanish by Drs. Blanco, Cabezas, Cobo, Duque, Reyes, and Santamaria. T e 7th edition was also translated into Spanish by Drs. Rendón, Araiza, Pastrana, Enriquez, and González. T e 8th edition was translated into Turkish by Professor Metin Onerci and Dr. Hakan Korkmaz and translated into Chinese by Professor Chen and her colleagues. A previous edition was also translated into Turkish by Professor Vecdet Kayhan, Doc. Dr. Tayfun Sunay, and Uz. Dr. Cetin Kaleli. It has also been translated into other languages without our knowledge.

Although the original material still forms a significant portion of the book, Dr. Chan and Dr. Goddard have assembled a broad panel of authorities in several subspecialities to present additional information, which is considered the most current in their areas of expertise.

Neither a complete review of otolaryngology nor a comprehensive textbook on the subject, K.J. Lee's Essential Otolaryngology, 11th edition, remains true to its original intent—to serve as a guide for Board preparation as well as a practical and concise reference text ref ecting contemporary concepts in clinical otolaryngology. Senior medical students, residents, and fellows; Board-eligible, Board-certified otolaryngologists; primary care physicians; and specialists in other fields will all find this edition to be an even more useful and indispensable resource.

I thank Dr. Chan and Dr. Goddard.

K. J. Lee

Acknowledgments

For this 11th edition, I want to thank our two editors, Dr. Yvonne Chan and Dr. John Goddard, who have done a tremendous job. Particularly, I would like to thank the one person who has been by my side even before the appearance of the very 1st edition of this book in 1973, my lovely and devoted wife of 49 years, Linda. Our three sons, Ken, Lloyd, and Mark, used to help with editorial assistance, but are now busy with their respective professions, law, private equity, and movie production; Jeannie Grenier, my nurse for over 31 years and editorial associate, worked hard on previous editions. I thank the McGraw-Hill staff for their diligence, great work, and congeniality.

I thank my parents for the gene and nurturing environment that allowed me to develop a passion for hard work, a sense for organization, and an ability to distill complex materials into simple facts. T ese are the three cornerstones that have shaped this book from the 1st edition. I am thrilled to see these traits are also inherent in Yvonne and John.

And to those newcomers at the frontiers of medical science who have contributed to this edition, I also extend my thanks for taking the time to share their own expertise and, by doing so, helping to keep this book up-to-date.

K. J. Lee

Part 1
General Otolaryngology

Chapter 1
Syndromes and Eponyms

Syndromes and Diseases

Adult Respiratory Distress Syndrome

Adult respiratory distress syndrome (ARDS) is characterized by a delay in onset (12-24 hours) following injury, shock, and/or successful resuscitative effort. Septic shock, extrathoracic trauma, central nervous system (CNS) pathology, fat embolism, oxygen toxicity, head and facial injuries, and massive blood transfusions can lead to ARDS. It is characterized by hypoxia and pulmonary infiltrates secondary to increased pulmonary vascular permeability, microvascular hemorrhage, or both.

Aide Syndrome

Aide syndrome is characterized by decreased pupillary reaction and deep tendon reflex. T e etiology is unknown.

Alagille Syndrome

Alagille syndrome is marked by cardiovascular abnormalities, characteristic facial appearance, chronic cholestasis, growth retardation, hypogonadism, mental retardation, vertebral arch defect, temporal bone anomalies in the cochlear aqueduct, ossicles, semicircular canals (SCCs), and subarcuate fossa. Liver transplantation is a possible treatment.

Albers-Schönberg Disease

A genetic disorder also known as osteopetrosis, Albers-Schönberg disease results in progressive increase in the density (but also increase in weakness) of the bones in the skeletal system. Vascular nutrition to affected bones is also decreased by this disease. Broken down into three categories, there is osteopetrosis with precocious manifestations, osteopetrosis with delayed manifestations, and pyknodysostosis. In the mandible long-term antibiotic therapy, multiple debridements, sequestrectomies, or even resection are possible treatments.

Albright Syndrome

Polyostotic fibrous dysplasia usually manifests early in life as multicentric lesions involving the long bones and bones of the face and skull with scattered skin lesions similar to melanotic café au lait spots and precocious puberty in female patients. Frequently, there is an elevation of serum alkaline phosphatase as well as endocrine abnormalities.

Aldrich Syndrome

T rombocytopenia, eczema, and recurrent infections occur during the first year of life. It is inherited through a sex-linked recessive gene. T e bleeding time is prolonged, the platelet count is decreased, and the bone marrow megakaryocytes are normal in number.

Amalric Syndrome

Granular macular pigment epitheliopathy (foveal dystrophy) is associated with sensorineural hearing loss. Visual acuity is usually normal. Amalric syndrome may be a genetic disorder, or it may be the result of an intrauterine rubella infection.

Aortic Arch Syndrome

See Takayasu Disease.

Apert Syndrome

Apert syndrome is not to be confused with Pfeiffer syndrome, which has different types of hand malformations.

Ascher Syndrome

Ascher syndrome is a combination of blepharochalasis, double lip, and goiter.

Auriculotemporal Syndrome (Frey Syndrome)

Auriculotemporal syndrome is characterized by localized flushing and sweating of the ear and cheek region in response to eating. It usually occurs after parotidectomy. It is assumed that the parasympathetic fibers of the ninth nerve innervate the sweat glands after parotidectomy. It has been estimated that 20% of the parotidectomies in children result in this disorder.

Avellis Syndrome

Unilateral paralysis of the larynx and velum palati, with contralateral loss of pain and temperature sensitivity in the parts below the larynx characterize Avellis syndrome. T e syndrome is caused by involvement of the nucleus ambiguus or the vagus nerve along with the cranial portion of the ninth nerve.

Babinski-Nageotte Syndrome

Babinski-Nageotte syndrome is caused by multiple or scattered lesions, chiefly in the distribution of the vertebral artery. Ipsilateral paralysis of the soft palate, larynx, pharynx, and sometimes tongue occurs. T ere is also ipsilateral loss of taste on the posterior third of the tongue, loss of pain and temperature sensation around the face, and cerebellar asynergia. Horner syndrome with contralateral spastic hemiplegia and loss of proprioceptive and tactile sensation may also be present.

Baelz Syndrome

Painless papules at the openings of the ducts of the mucous glands of the lips with free exudation of mucus are characteristic. Congenital and familial forms are precancerous. Acquired forms are benign and caused by irritating substances.

Bannwarth Syndrome (Facial Palsy in Lymphocytic Meningoradiculitis)

A relatively benign form of acute unilateral or bilateral facial palsy that is associated with lymphocytic reactions and an increased protein level in the cerebrospinal fluid (CSF) with minimal, if any, meningeal symptoms is known as Bannwarth syndrome. Neuralgic or radicular pain without facial palsy and unilateral or bilateral facial palsy of acute onset are symptoms of this syndrome. A virus has been suggested as a possible etiology. Males are more often affected than females, with the greatest number of cases occurring in the months of August and September.

Barany Syndrome

Barany syndrome is a combination of unilateral headache in the back of the head, periodic ipsilateral deafness (alternating with periods of unaffected hearing), vertigo, and tinnitus. T e syndrome complex may be corrected by induced nystagmus.

Barclay-Baron Disease

Vallecular dysphagia is present.

Barre-Lieou Syndrome

Occipital headache, vertigo, tinnitus, vasomotor disorders, and facial spasm due to irritation of the sympathetic plexus around the vertebral artery in rheumatic disorders of the cervical spine are characteristic. It is also known as cervical migraine.

Barrett Syndrome

Barrett syndrome is characterized by esophagitis due to change in the epithelium of the esophagus.

Barsony-Polgar Syndrome

A diffuse esophageal spasm, caused by disruption of the peristaltic waves by an irregular contraction resulting in dysphagia and regurgitation, is evidence of this syndrome. It most commonly affects excitable elderly persons.

Basal Cell Nevoid Syndrome

T is familial syndrome, non–sex-linked and autosomal dominant with high penetrance and variable expressivity, manifests early in life. It appears as multiple nevoid basal cell epitheliomas of the skin, cysts of the jaw, abnormal ribs and metacarpal bones, frontal bossing, and dorsal scoliosis. Endocrine abnormalities have been reported and it has been associated with medulloblastoma. T e cysts in the jaw, present only in the maxilla and mandible, are destructive to the bone. T e basal cell epitheliomas are excised as necessary, and the cysts in the jaw rarely recur after complete enucleation.

Bayford-Autenrieth Dysphagia (Arkin Disease)

Dysphagia lusoria is said to be secondary to esophageal compression from an aberrant right subclavian artery.

Beckwith Syndrome

T is is a congenital disorder characterized by macroglossia, omphalocele, hypoglycemia, pancreatic hyperplasia, noncystic renal hyperplasia, and cytomegaly of the fetal adrenal cortex.

Behçet Syndrome

Of unknown etiology, this disease runs a protracted course with periods of relapse and remission. It manifests as indolent ulcers of the mucous membrane and skin, stomatitis, as well as anogenital ulceration, iritis, and conjunctivitis. No definitive cure is known, though steroids help.

Besnier-Boeck-Schaumann Syndrome

Sarcoidosis is present.

Bloom Syndrome

An autosomal recessive growth disorder, Bloom syndrome is associated with chromosomal breaks and rearrangements. It is also associated with an unusually high rate of cancer at an early age. Associated with facial erythema, growth retardation, immunodeficiency, infertility, and sun sensitivity, diagnosis is confirmed by chromosome analysis. Anomalous numbers of digits or teeth, asymmetric legs, heart malformation, hypopigmented spots in blacks, protruding ears, sacral dimple, simian line, and urethral or meatal narrowing are less common characteristics. For head and neck tumor patients, there is an increased chance of secondary and primary tumors.

Bogorad Syndrome

Bogorad syndrome is also known as the syndrome of crocodile tears, characterized by residual facial paralysis with profuse lacrimation during eating. It is caused by a misdirection of regenerating autonomic fibers to the lacrimal gland instead of to the salivary gland.

Bonnet Syndrome

Sudden trigeminal neuralgia accompanied by Horner syndrome and vasomotor disorders in the area supplied by the trigeminal nerve are manifestations of this syndrome.

Bonnier Syndrome

Bonnier syndrome is caused by a lesion of Deiters nucleus and its connection. Its symptoms include ocular disturbances (eg, paralysis of accommodation, nystagmus, diplopia), deafness, nausea, thirst, and anorexia, as well as other symptoms referable to involvement of the vagal centers, cranial nerves VIII, IX, X, and XI, and the lateral vestibular nucleus. It can simulate Ménière disease.

Bourneville Syndrome

Bourneville syndrome is a familial disorder whose symptoms include polyps of the skin, harelip, moles, spina bifida, and microcephaly.

Bowen Disease

Bowen disease is a precancerous dermatosis characterized by the development of pinkish or brownish papules covered with a thickened horny layer. Histologically, it shows hyperchromatic acanthotic cells with multinucleated giant cells. Mitoses are frequently observed.

Branchio-Oto-Renal Syndrome

Branchio-Oto-Renal syndrome is an autosomal dominant disorder characterized by anomalies of the external, middle, and inner ear in association with preauricular tissues, branchial cleft anomalies, and varying degrees of renal dysplasia, including aplasia. Many of the following symptoms (but not necessarily all) are present:

A. Conductive or mixed hearing loss
B. Cup-shaped, anteverted pinnae with bilateral preauricular sinuses
C. Bilateral branchial cleft fistulas or sinuses
D. Renal dysplasia

T is syndrome is among a group of syndromes characterized by deformities associated with the first and second branchial complexes. T e precise incidence of the disorder is unknown.

Briquet Syndrome

Briquet syndrome is characterized by a shortness of breath and aphonia due to hysteric paralysis of the diaphragm.

Brissaud-Marie Syndrome

Unilateral spasm of the tongue and lips of a hysteric nature are characteristic.

Brown Syndrome

Brown syndrome is a congenital or acquired abnormality of the superior oblique muscle tendon characterized by vertical diplopia and the inability to elevate the eye above midline or medial gaze. T is syndrome is of two types: true and simulated. True Brown syndrome is always congenital. Simulated Brown syndrome is either congenital or acquired. T e congenital simulated type may be caused by thickening of an area in the posterior tendon or by the firm attachment of the posterior sheath to the superior oblique tendon. T e acquired simulated type may be caused by inflammation extending from the adjacent ethmoid cells to the posterior sheath and tendon, an orbital floor fracture, frontal ethmoidal fracture, crush fracture of nasal bones, sinusitis, frontal sinus surgery, or surgical tucking of the superior oblique tendon.

Brun Syndrome

Vertigo, headache, vomiting, and visual disturbances due to an obstruction of CSF flow during positional changes of the head are seen. T e main causes of this syndrome include cysts and cysticercosis of the fourth ventricle as well as tumors of the midline cerebellum and third ventricle.

Burckhardt Dermatitis

Burckhardt dermatitis appears as an eruption of the external ear. It consists of red papules and vesicles that appear after exposure to sunlight. T e rash usually resolves spontaneously.

Caffey Disease (Infantile Cortical Hyperostosis)

Of familial tendency, its onset is usually during the first year of life. It is characterized by hyperirritability, fever, and hard nonpitting edema that overlie the cortical hyperostosis. Pathologically, it involves the loss of periosteum with acute inflammatory involvement of the intratrabecular bone and the overlying soft tissue. Treatment is supportive, consisting of steroids and antibiotics. T e prognosis is good. T e mandible is the most frequently involved site.

Caisson Disease

T is symptom complex occurs in men and women who work in high air pressures and are returned too suddenly to normal atmospheric pressure. Similar symptoms may occur in fliers when they suddenly ascend to high altitudes unprotected by counterpressure. It results from the escape from solution in the body fluids of bubbles (mainly nitrogen) originally absorbed at higher pressure. Symptoms include headache; pain in the epigastrium, sinuses, and tooth sockets; itchy skin; vertigo; dyspnea; coughing; nausea; vomiting; and sometimes paralysis. Peripheral circulatory collapse may be present. Nitrogen bubbles have been found in the white matter of the spinal cord. It also can injure the inner ear through necrosis of the organ of Corti. T ere is a question of rupture of the round window membrane; hemotympanum and eustachian tube obstruction may occur.

Camptomelic Syndrome

T e name is derived from a Greek word meaning curvature of extremities. T e syndrome is characterized by dwarfism, craniofacial anomalies, and bowing of the tibia and femur, with malformation of other bones. T e patient has cutaneous dimpling overlying the tibial bend. Respiratory distress is common, and the patient has an early demise in the first few months of life. In the otolaryngologic area, the patient exhibits a prominent forehead, flat facies with a broad nasal bridge and low-set ears, cleft palate, mandibular hypoplasia, and tracheobronchial malacia that contributes to the respiratory distress and neonatal death. Histologically, two temporal bone observations showed defective endochondral ossification with no cartilage cells in the endochondral layer of the otic capsule. T e cochlea was shortened and flattened, presenting a scalar communis. T e vestibule and the SCC were deformed by bone invasion.

T is syndrome is often of unknown etiology, although some believe it is autosomal recessive. Others believe it may be due to an exogenous cause.

T is syndrome is not to be confused with Pierre Robin syndrome, which presents with very similar clinical features.

Cannon Nevus

T is is an autosomal dominant disorder characterized by spongy white lesions of the oral and nasal mucosa. T e lesions are asymptomatic and may be found from the newborn period with increasing severity until adolescence. T e histologic picture is that of keratosis, acanthosis, and parakeratosis.

Carcinoid Syndrome

T e symptoms include episodic flushing, diarrhea, and ascites. T e tumor secretes serotonin. Treatment is wide excision. T e tumor may give a positive dopa reaction.

Carotid Sinus Syndrome (Charcot-Weiss-Barber Syndrome)

When the carotid sinus is abnormally sensitive, slight pressure on it causes a marked fall in blood pressure due to vasodilation and cardiac slowing. Symptoms include syncope, convulsions, and heart block.

Castleman Disease

Castleman disease was first described by Castelman et al in 1954. It is a benign lymphoepithelial disease that is most often mistaken for lymphoma. It is also known as localized nodal hyperplasia, angiomatous lymph node hyperplasia, lymphoid hamartoma, and giant lymph nodal hyperplasia. Symptoms include tracheobronchial compression, such as cough, dyspnea, hemoptysis, or dysphagia. Masses in the neck are also not uncommon. T ere are two histologic types: the hyaline vascular type and the plasma cell type. Follicles in the hyaline vascular type are traversed

by radially oriented capillaries with plump endothelial cells and collagenous hyalinization surrounding the vessels. The follicles in the plasma cell type are normal in size without capillary proliferation or hyalinization. Intermediate forms exist but are rare. Treatment entails complete excision of the mass. Etiology is unknown.

Cavernous Sinus Syndrome

The cavernous sinus receives drainage from the upper lip, nose, sinuses, nasopharynx, pharynx, and orbits. It drains into the inferior petrosal sinus, which in turn drains into the internal jugular vein. The cavernous sinus syndrome is caused by thrombosis of the cavernous intracranial sinus, 80% of which is fatal. The symptoms include orbital pain (V_1) with venous congestion of the retina, lids, and conjunctiva. The eyes are proptosed with exophthalmos. The patient has photophobia and involvement of nerves II, III, IV, and V_1. The treatment of choice is anticoagulation and antibiotics. The most common cause of cavernous sinus thrombosis is ethmoiditis. The ophthalmic vein and artery are involved as well. (The nerves and veins are lateral to the cavernous sinus, and the internal carotid artery is medial to it.)

Cestan-Chenais Syndrome

Cestan-Chenais syndrome is caused by occlusion of the vertebral artery below the point of origin of the posteroinferior cerebellar artery. There is paralysis of the soft palate, pharynx, and larynx. Ipsilateral cerebellar asynergia and Horner syndrome are also present. There is contralateral hemiplegia and diminished proprioception and tactile sensation.

Champion-Cregah-Klein Syndrome

This is a familial syndrome consisting of popliteal webbing, cleft lip, cleft palate, lower lip fistula, syndactyly, onychodysplasia, and pes equinovarus.

Chapple Syndrome

This disorder is seen in the newborn with unilateral facial weakness or paralysis in conjunction with comparable weakness or paralysis of the contralateral vocal cord, the muscles of deglutition, or both. The disorder is secondary to lateral flexion of the head in utero, which compresses the thyroid cartilage against the hyoid or cricoid cartilages or both, thereby injuring the recurrent or superior laryngeal nerve, or both.

Charcot-Marie-Tooth Disease

This is a hereditary and degenerative disease that includes the olivopontocerebellar, cerebelloparenchymal, and spinocerebellar disorders and the neuropathies. This is disease is characterized by chronic degeneration of the peripheral nerves and roots; and distal muscle atrophy in feet, legs, and hands. Deep tendon reflexes are usually nil. It is also associated with hereditary cerebellar ataxia features, optic atrophy, and other cranial involvement. Some suggest that this disease is linked to auditory dysfunction and that it is also linked to other CNS dysfunctions. This is disease can be progressive, and it can also spontaneously arrest.

CHARGE Syndrome

CHARGE syndrome (Coloboma of the eye, Heart defects, Atresia of the choanae, Retardation of growth and development, Genital and/or urinary abnormalities and Ear abnormalities and deafness) is a genetic pattern of birth defects which occur one in 10,000 births worldwide, without any family history. It involves heart defects, breathing and swallowing difficulties, hearing loss, vision loss, and balance problems.

Major Features of CHARGE Syndrome (Very Common in CHARGE and Relatively Rare in Other Conditions)

Feature	Includes	Frequency (%)
Coloboma of the eye	Coloboma (sort of like a cleft) of the iris, retina, choroid, macula, or disc (not the eyelid); microphthalmos (small eye) or anophthalmos (missing eye): Causes vision loss	80-90
Choanal atresia or stenosis	T ey can be stenosed or atretic. It can be unilateral or bilateral, bony, or membranous.	50-60
Cranial nerve abnormality	I—Missing or decreased sense of smell	90-100
	IX/X—Swallowing difficulties, aspiration	70-90
	VII—Facial palsy (one side or both)	40
CHARGE outer ear	Short, wide ear with little or no lobe, "snipped off" helix, prominent antihelix which is discontinuous with tragus, triangular concha, decreased cartilage (floppy), often stick out, usually asymmetric	> 50
CHARGE middle ear	Malformed bones of the ossicles: Conductive hearing loss	Common
CHARGE inner ear	Mondini defect; small or absent semicircular canals: Balance problems and sensorineural loss	90

Minor Characteristics of CHARGE: Significant, But More Difficult to Diagnose or Less Specific to CHARGE

Feature	Includes	Frequency (%)
Heart defects	Can be any type, but many are complex, such as tetralogy of Fallot	75
Cleft lip ± cleft palate	Cleft lip with or without cleft palate, cleft palate, submucous cleft palate	20
TE fistula	Esophageal atresia, tracheoesophageal fistula (TEF), H-shaped TEF	15-20
Kidney abnormalities	Small kidney, missing kidney, misplaced kidney, reflux	40
Genital abnormalities	Males: small penis, undescended testes Females: small labia, small or missing uterus Both: lack of puberty without hormone intervention	50 25 90
Growth deficiency	Growth hormone deficiency Other short stature	15 70
Typical CHARGE face	Square face with broad prominent forehead, arched eyebrows, large eyes, occasional ptosis, prominent nasal bridge with square root, thick nostrils, prominent nasal columella, flat midface, small mouth, occasional small chin, larger chin with age. Facial asymmetry even without facial palsy	
Palm crease	Hockey-stick palmar crease	50
CHARGE behavior	Perseverative behavior in younger individuals, obsessive compulsive behavior (OCD) in older individuals	> 50

Chédiak-Higashi Syndrome

Chédiak-Higashi syndrome is the result of an autosomal recessive trait. It is characterized by albinism, photophobia, nystagmus, hepatosplenomegaly, anomalous cellular granules, and development of lymphoma. T ese patients usually die during childhood of fulminant infections.

Cleft Lip Palate and Congenital Lip Fistulas

T is syndrome is transmitted in an autosomal dominant manner with 80% penetrance; it occurs in 1 per 100,000 live births. Usually bilateral, symmetrically located depressions are noted on the vermilion portion of the lower lip and communicate with the underlying minor salivary glands. T e lip pits may be an isolated finding (33%) or be found with cleft lip palate (67% of cases). Associated anomalies of the extremities may include talipes equinovarus, syndactyly, and popliteal pterygia. Congenital lip pits have also been seen in association with the oral-facial-digital syndrome.

Cockayne Syndrome

Cockayne syndrome is autosomal recessive, progressive bilateral sensorineural hearing loss, associated with dwarfism, facial disharmony, microcephaly, mental deficiency, retinitis pigmentosa, optic atrophy, intracranial calcification, and multiple dental caries. Patients succumb to respiratory or genitourinary infection in the teens or twenties.

Cogan Syndrome

Nonsyphilitic interstitial keratitis and vestibuloauditory symptoms are characteristics of Cogan syndrome. Interstitial keratitis gives rise to rapid visual loss. Symptoms include episodic severe vertigo accompanied by tinnitus, spontaneous nystagmus, ataxia, and progressive sensorineural hearing loss. T ere are remissions and exacerbations. It is believed to be related to periarteritis nodosa. Eosinophilia has been reported in this entity. Pathologically, it is a degeneration of the vestibular and spiral ganglia with edema of the membranous cochlea, SCCs, and inflammation of the spiral ligament. Treatment with steroids has been advocated.

Cyclophosphamide and azathioprine have been used in addition to prednisone (40 mg daily). T is syndrome is not to be confused with Ménière disease despite vertiginous symptoms and fluctuating hearing loss. Vogt-Koyanagi-Harada syndrome is also similar but involves alopecia, poliosis, and exudative uveitis. Syphilis is also confused with this syndrome, but in syphilis, the interstitial keratitis is old and usually does not demonstrate active inflammatory changes. Syphilitic involvement of the cornea is often centrally located. Follow-up treatment of patients must be thorough in order to detect more extensive involvement, such as systemic vasculitis or aortitis.

Collet-Sicard Syndrome

T e 9th, 10th, and 11th nerves are involved with normal sympathetic nerves. T e etiology is usually a meningioma or other lesion involving the nerves in the posterior cranial fossa.

Conradi-Hünermann Syndrome

T e most common variant of chondrodysplasia punctata; this syndrome is characterized by punctate epiphyseal calcifications. Clinical features include saddle nose deformity, micromelia, rhizomelia, short stature, flexion contractures, and dermatoses. T is syndrome is also known as chondrodystrophia epiphysialis punctata, stippled epiphysis disease, dysplasia epiphysialis punctata, chondroangiopathia calcarea punctata, and Conradi disease. Some cases point to sporadic mutations and others to autosomal dominant patterns of inheritance. T e clinical features