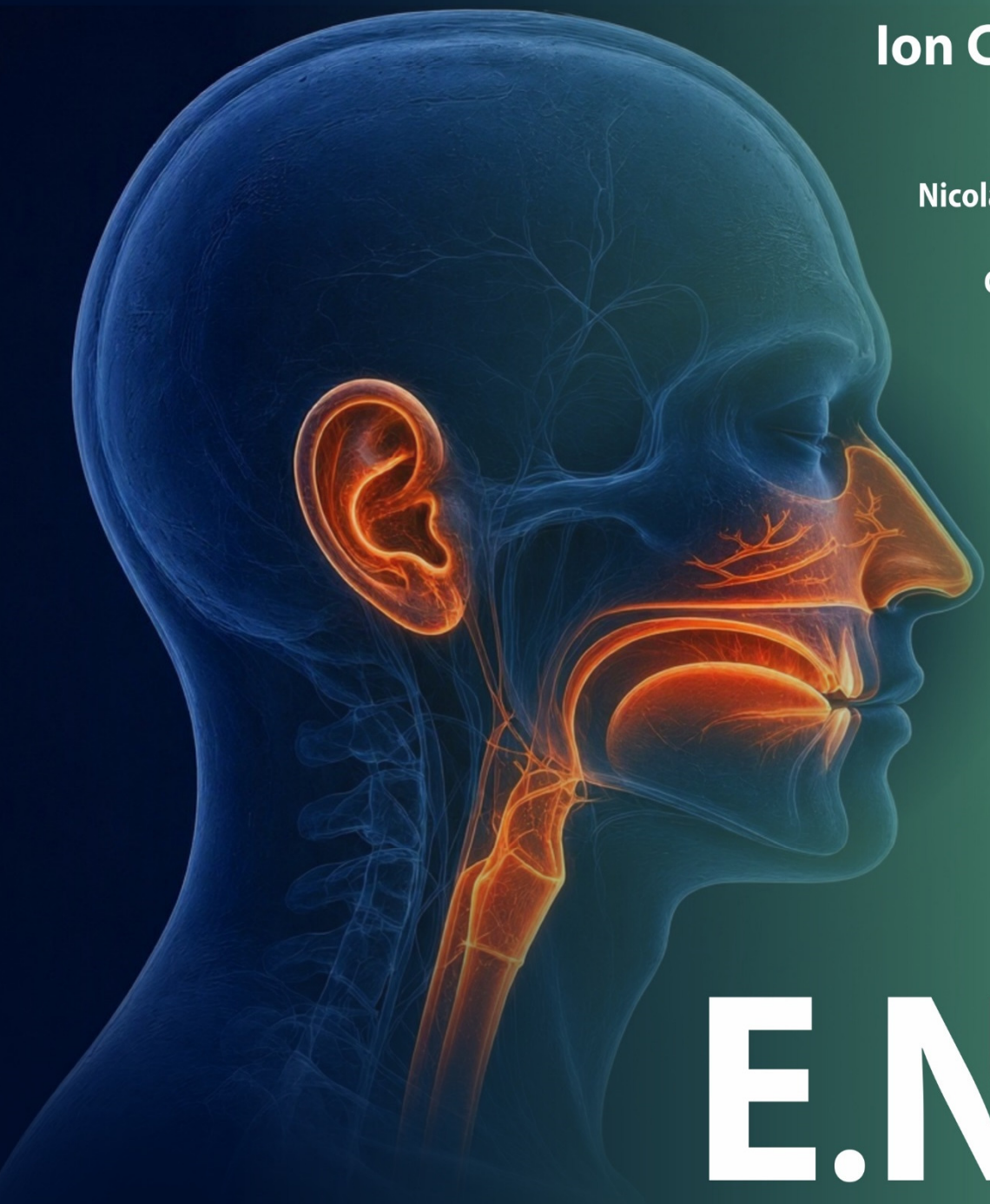
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CHAPTER I — NOSE AND PARANASAL SINUSES PATHOLOGY

ANATOMY OF THE NASAL CAVITY AND PARANASAL SINUSES

Sphenoid bone:

- the sphenoid (os sphenoidale) is a bone situated in the posterior part of the nasal cavity
- it articulates with:
  - ethmoid bone
  - palatine bone
  - vomer bone
  - maxillary bone
  - frontal bone
  - corpus sphenoidalis
- it forms the central part and contains the sphenoid sinuses
- pneumatization **may extend** into the clinoid and pterygoid processes

Frontal bone:

- **posterior wall** – thin and in contact with the anterior cranial fossa
- **inferior wall** – part of the orbital roof; the frontal sinuses are separated by a thin, oblique bony wall, responsible for their asymmetry
- **the posterior border** (margo sphenoidalis) articulates with the lesser wings (ala minor) and greater wings (ala major) of the sphenoid bone
- **the nasal part** (pars nasalis) lies between the orbital parts (pars orbitalis) and presents the nasal border (margo nasalis)
- **the nasal spine** (spina nasalis) is an extension in the midline

Maxillary bone

- body of the maxilla (corpus maxillae)
- medial – the frontal process
- posterior of the frontal process
- lacrimal crest (sulcus lacrimalis)

Maxillary sinus (sinus maxillaris)

- pyramidal shape
- **a base** that corresponds to the **medial face**
- the apex points towards the zygomatic process of the maxilla (***processus zygomaticus***)

Ethmoidal bone

- superior face
- the horizontal plate (cribriform plate) separates the anterior cranial fossa from the nasal cavity
- **the crista galli**, perpendicular to the cribriform plate, serves as the attachment of the ***falx cerebri***
- **perpendicular lamina,** lamina perpendicularis
- **lateral**, ethmoidal cells (***cellulae ethmoidales***)
- **the height of the ethmoid roof** relative to the **cribriform plate** may vary:
  - **Keros I**
    - the difference is 1-3 mm, i.e. the ethmoid roof and the cribriform plate are at almost the same height

- **Keros II**
  - the difference is 4-7 mm
- **Keros III**
  - the difference is 8-16 mm – **the medial wall of the ethmoid can be extremely thin**
  - significant risk of injury to the anterior cranial fossa

Inferior concha

- the inferior concha (concha nasalis inferior) is an **independent bone** located in the lateral wall of the nasal cavity, with **three major processes:**
  - **the lacrimal process** (processus lacrimalis) **completes the nasolacrimal canal** by articulating with the lower edge of the lacrimal bone
  - **the maxillary process** is attached to the maxilla, in the lower part of the maxillary hiatus, and to the conchal crest of the palatine bone
  - **the ethmoidal process** (processus ethmoidalis) articulates with the lower end of the uncinate process

Uncinate process (processus uncinatus)

- a thin, oblique, **sickle-shaped plate**
- sagittal oriented
- with many variations of insertion:
  - on the base of the skull
  - on the lamina papyracea
  - the anterior part of the lacrimal bone
  - the perpendicular (vertical) plate of the palatine bone
  - the ethmoidal process of the inferior concha (concha nasalis inferior)
- curved posteriorly, parallel to the ethmoidal bulla (bulla ethmoidalis)
- **represents the anterior limit of the inferior semilunar hiatus (hiatus semilunaris inferior) and of the ethmoidal infundibulum (infundibulum ethmoidale)**

Infundibulum ethmoidale

- the most important nasal infundibulum
- ***the frontal and maxillary infundibula*** drain into the ethmoidal infundibulum
- it is bordered by:
  - medially – uncinate process
  - laterally – lamina papyracea
  - posteriorly – ethmoidal bulla
- it may drain directly into the ***frontal recess*** or it may end in the recessus terminalis

Frontal recess

- ***the most anterior and superior part*** of the middle meatus
- It is bordered by:
  - medially and superiorly - middle concha
  - laterally - lamina papyracea
  - posteriorly - ethmoidal bulla basal lamina

PHYSIOLOGY OF THE NOSE

Nasal fossa functions:

- respiratory function
- defense function
- phonatory function
- olfactory function
- reflex function

Respiratory function:

- its vasculo-erectile function
- adjusting the air flow
- air conditioning function by:
  - warming
  - humidification
  - filtering

Defense function:

- mucociliary apparatus – self-cleaning process
- specific defense factors
  - IgA, IgM, IgG
- non-specific defense factors
  - mucus, lysozyme, IFN, protease inhibitors, complement

Phonatory function

- nasal fossae and paranasal sinuses – **resonance cavity**
- change in the quality of the voice through changes in the volume and patency of the nasal passages

Olfactory function

- **olfactory nerves** (olfactory mucosa in the upper part of the nasal fossa)

Reflex function

- nasal mucosa receptors (thermal, tactile, olfactory) – maintain the functional respiratory balance
- **trigeminal endings** - changes in **pulmonary ventilation**

Nasal pathophysiological syndromes

- I. Nasal respiratory insufficiency
- Etiologic classification of **nasal obstruction**:
  - congenital:
    - choanal atresia
    - posterior choanal stenosis
    - anterior nasal stenosis
    - congenital malformations
    - midline tumors
  - traumatic
  - iatrogenic
  - inflammatory

  - tumoral
- II. Secretory syndromes
  - serous rhinorrhea or hydrorrhea
  - mucopurulent or purulent rhinorrhea, unilateral or bilateral
- III. Sensory syndromes
  - hyperosmia
  - parosmia
  - hyposmia
  - anosmia
- IV. Sensory and reflex syndromes
  - anesthesia
  - nasal hyperesthesia
- V. Vascular syndromes – Epistaxis

EXAMINATION OF THE NASAL CAVITY AND PARANASAL SINUSES

- Inspection and palpation
- Anterior rhinoscopy
- Posterior rhinoscopy
- Nasal endoscopy
- **The gold standard** in nasal examination; **it allows visualization of**:
  - spheno-ethmoidal recess
  - ostiomeatal complex
  - the necessary equipment consists of a light source, a fiber-optic cable, an endoscope, a video camera and a professional monitor
  - rigid endoscope
  - fiber-optic endoscopes
- Paranasal sinus examination:
  - palpation points for the sinuses:
    - maxillary – infraorbital region
    - the frontal and supraorbital region
    - ethmoidal – medial orbital suture (Ewing's point)

Functional investigations of the nose

Rhinomanometry

- simultaneous measurement of the flow and pressure variations of an air current passing through the nasal cavities
- the choanal pressure is measured:
  - in the contralateral nostril, during **active anterior rhinomanometry**
  - in the oral cavity during **active posterior rhinomanometry**

Acoustic rhinomanometry

- acoustic scanning
- provides an **objective geometric study** of the nasal cavities
- **computerized analysis** of the reflection of sound waves in the nasal cavities is used to assess them

Mucociliary function exploration

- **is not** performed in daily clinical practice
- **the best clinical indicator of mucociliary clearance** is the **saccharin transit time** test – saccharin powder is placed on the inferior turbinate
- the time it takes for the patient to feel the taste of saccharin is usually **less than 20 min**

Cellular exploration: Nasal cytology

- may involve taking a smear or nasal brushing

Exploring olfaction

- "sniff bottles"
- perfume testing
- microcapsules that release odorant molecules when scratched

CONGENITAL MALFORMATIONS OF THE NASAL PYRAMID

- Mediofacial closure defects
- Unilateral or bilateral narial atresia
- Stenosis and atresia of the anterior portion of the nasal fossa
- Saddle nose associated with hypertelorism
- Velopalatal cleft
- Unilateral or bilateral choanal atresia
- Encephaloceles
- Meningoceles
- Glioma
- Dermoid cyst
- Haemangioma

NASAL TRAUMA

Lesions of musculoskeletal and skin structures:

- traumatic injuries **without** break in continuity
- contusions / edema / bruising
- traumatic injuries **with** break in continuity
- wounds

Lesions of the cartilaginous skeleton:

- fractures of the quadrangular septal cartilage
- sprain of the septal cartilage / septal cartilage hematoma

Lesions of the bony skeleton:

- cracks or bone fractures
- fractures with or without displacement, depression or dislocation
- comminuted or burst fractures
- closed fractures or fractures open to the outside or into the nasal passages

DISEASES OF THE OUTER AND INNER NOSE

Rhinoliquorrhea

- leakage of cerebrospinal fluid (CSF) through the nasal passages

- originated in:
  - the cribriform plate of the ethmoid bone
  - the frontal, ethmoid, sphenoid sinus
  - middle ear via Eustachian tube
- **Etiology:**
  - post-traumatic (most common) – craniofacial trauma
  - fracture of the anterior skull base
  - radiating fractures – frontal, ethmoid and sphenoid sinuses
  - isolated fractures of the sinuses
  - fractures of the cribriform plate of the ethmoid bone
  - craniofacial fractures / postoperative iatrogenic
  - secondary (tumors)
  - idiopathic

Septal hematoma

- collection of blood under the perichondrium of the septum
- may cause **cartilage necrosis in 3 days**
- **Diagnosis**
  - intense pain
  - complete nasal obstruction
  - inflammation
  - hematoma of the upper lip and philtrum
- **Treatment**
  - hematoma drainage – **mucoperichondrial incision**
  - oral antibiotic therapy
  - immediate treatment – to prevent long-term complications:
    - saddle nose
    - perforation of the nasal septum
    - columellar retraction

Intranasal foreign bodies

- usually introduced voluntarily:
- child, 1 to 6 years
- animated
  - insects
  - larvae
  - parasites
- inert
  - organic
  - inorganic
- most often **unilateral**, less often bilateral
- can be single or multiple
- Symptomatology:
  - ***unilateral*** nasal complaints:
    - unilateral nasal obstruction
    - epistaxis
    - repeated sneezes

        - abundant rhinorrhea, serous and seromucous at first, purulent in the late stages
        - local pain
        - hyposmia
    - ***the severity*** of the symptoms depends on:
        - nature
        - foreign body shape
        - the time elapsed since introduction into the nasal passages
- Diagnosis:
    - anamnesis (child or parent)
    - objective clinical exam
    - narinoscopy
    - anterior rhinoscopy
    - secretions aspirations
- Complications:
    - **are rare:** rhinitis, sinusitis, rhinolithiasis

Rhinolithiasis

- calcium deposition around a foreign body or a nasal blood clot
- hard, tumor-like mass
- dark or dark-gray
- consisting of calcium and magnesium phosphates or carbonates, arranged in concentric layers
- organic foreign bodies may release irritating substances
- Treatment:
    - extraction using **appropriate instruments** (hooks, curettes, special forceps) under local or general anesthesia
    - prior crushing and fragmentation
    - surgical extraction

Erysipelas

- infection affecting the epidermis
    - etiological agent: **Streptococcus pyogenes (group A)**
    - on the **extremities and face**
    - **less often** on the scalp and genitals
- Epidemiology/etiology
    - the elderly / immunocompromised / diabetic patients, children and the malnourished
- Predisposing factors:
    - alcoholism
    - skin ulceration
    - penetrating wounds
    - mycotic infections
    - chronic lymphatic and venous obstructions
    - burns
- Signs and symptoms:
    - fever, chills and asthenia
    - red, indurated and raised skin
    - well-defined margin

  - rapid expansion
  - painful, warm on palpation
  - variable edema
  - bullae (2nd or 3rd day)
  - skin peeling (within the next 10 days)
  - regional adenitis
- Complications:
  - recurrence, with a predilection for women, less often men
  - necrotizing fasciitis (very rarely)
  - glomerulonephritis: Streptococcus pyogenes (group A)
- Differential diagnosis
  - contact dermatitis
  - angioneurotic edema
  - osteomyelitis
- Therapy
  - Conservative therapy:
    - the antibiotic of choice is **penicillin**
    - in case of penicillin allergy – azithromycin, clarithromycin or clindamycin
    - in moderate and severe forms with systemic signs and symptoms: hospitalization and injectable treatment; adjuvant: **corticotherapy**
    - warm, moist compresses
  - **Surgical treatment:** incision / debrided / drainage

Impetigo

- superficial infection of the **nasal vestibule**
  - formation of blisters and pustules that turn into yellow-brown crusts
  - the crusts crack and peel off, uncovering an epidermis with superficial erosive lesions
- Pathogen:
  - Group A hemolytic streptococcus
- Treatment:
  - local toilet
  - antibiotic ointment
  - general antibiotic therapy

Furuncle/Carbuncle

- infection starting from the **hair follicle**
- extension into the dermis and subcutaneous cellular tissue
- forms a firm, painful, well-delineated nodule
- develops into a **red, painful and fluctuating abscess**
- the involvement of several grouped follicles leads to a **carbuncle**
- Epidemiology/etiology
  - Staphylococcus aureus
  - multi-microbial
- Signs and symptoms
  - neck, armpit, groin and thighs: **warm, moist or hairy areas**
  - carbuncles frequently appear **on the back of the neck**
  - fever

  - altered general condition
  - surrounding cellulitis
- Complications

Furunculosis – multiple areas of purulent ulceration, which heal with visible scars

- Diagnosis
  - clinical inspection
  - microbial culture in case of fever, cellulitis
  - **hospitalization – methicillin-resistant Staphylococcus aureus (MRSA)**
- Treatment
  - wet and warm compresses stimulate drainage
  - surgical incision and drainage
  - antiseptic treatment / systemic antibiotic treatment

Tuberculosis lupus

- tuberculous **infection of attenuated virulence**
- damage and deformities
- polymorphism
  - lupus nodules
  - ulceration
  - scarring
  - dermal and cartilage damage
- spares the bone
- exo- and endonasal retractions
- characteristic deformities **(parrot nose)**
- Treatment:
  - anti-tuberculosis
  - surgical - aesthetic and functional deformities

Nasal syphilis

- **Treponema pallidum**, with predominantly sexual transmission, but also through the mucosa, oropharyngeal, transcutaneous by scratching or through contaminated objects

Primary syphilis:

- occurs **3–4 weeks after contact**
- **painful infiltration** of the nose, unilateral or as a crateriform ulceration with a dirty base and regular edges, with surrounding induration
- **retro-angulo-mandibular adenitis**, disproportionate to the local lesion
- positive diagnosis: **bacteriological examination**

Secondary syphilis:

- persistent acute rhinitis
- syphilitic rosary
- vestibular fissures, pale mucous patches on the mucosa
- diagnosis of certainty: **serological examination** (BWR strongly positive)

Tertiary syphilis:

- occurs **3–4 years after** contamination
- due to incorrect treatment

- **nasal syphilitic gumma** (bony septum, floor and nasal bones)
- ulceration and perforation of the nasal septum
- functional and aesthetic deformity of the nasal pyramid (**saddle nose**)
- treatment:
- dermato-venerologic
- the antibiotic of choice is **penicillin**

Recurrent polychondritis

- rare connective tissue disorder
- recurrent inflammatory episodes of the cartilaginous structures
- most commonly affected are:
  - auricular cartilage
  - articular cartilage
  - nasal cartilage
- Positive diagnosis:
  - anamnesis (deformity of the nasal pyramid – **saddle nose**)
  - biopsy
- **Treatment:**
  - symptomatic, anti-inflammatory (non-steroidal drugs and corticotherapy)
  - sometimes cytostatic

ACUTE RHINITIS

- acute inflammation of the mucous membrane of the nasal fossa
- common in children
- Etiology:
  - rhinoviruses
  - myxoviruses: type A, B and C, parainfluenza
  - adenovirus
  - enteroviruses, which also cause respiratory infections
  - viruses promote **secondary bacterial infection**
  - contagion occurs through **Flügge's droplets**
- **Favoring factors:**
  - low immunity / decreased Ig E
  - **immunological gap** in newborns between the transmission of maternal antibodies and the production of their own antibodies, within the **first 6-8 weeks of life**
  - temperature and humidity variations
  - smoking
  - environmental pollution (various dusts and toxic gases)
  - acute or chronic infections of neighboring structures
  - nasal obstruction
- **Classification:**
  - 1. nonspecific
  - 2. specific

1. Nonspecific acute rhinitis:

- common acute rhinitis
- septic rhinitis of the newborn

- b1. infectious rhinitis with Staphylococcus aureus
- b2. streptococcal rhinitis
- b3. pseudomembranous rhinitis (streptococcus, pneumococcus)
- b4. gonococcal rhinitis
- b5. rhinitis from early congenital syphilis

a) Common acute rhinitis

- **Etiology**: ***viral***
- **Favoring factors**:
  - cold
  - moisture
  - adenoid hypertrophy
  - septal deviation
  - reduced immunity
- **Diagnosis:**
  - fever, tiredness
  - myalgia, arthralgia
  - frontal-orbital headache
  - pharyngeal burning sensation
  - common sneezes
  - nasal obstruction
  - aqueous rhinorrhea
  - hyposmia, anosmia
  - anterior rhinoscopy:
    - the turbinates are red and turgid
    - serous or mucopurulent secretions
- **Complications:**
  - otitis media
  - acute sinusitis, vestibulitis
  - subglottic laryngitis, laryngeal spasms, laryngo-tracheo- bronchitis
  - conjunctivitis, dacryocystitis
  - gastrointestinal disorders
- **Treatment:**
  - bed rest
  - nasal disinfectants
  - nasal toilet
  - non-steroidal anti-inflammatory drugs
  - antibiotic therapy in case of bacterial infection
  - removal of all predisposing factors

1. Non-specific acute rhinitis:

b) Septic rhinitis of the newborn (primary bacterial)

b1) Infectious rhinitis with Staphylococcus aureus

- in the first week after birth
- infection **from the mother**, during passage through the birth canal or in puerperal infection, or from medical staff carrying Staphylococcus aureus

- rhinorrhea – lemon-yellow
- possible progression to **toxic septic shock**

b2) Streptococcal rhinitis

- altered general condition
- fever 39-40
- seizures
- infection from the mother or from streptococcus-carrying staff with whom the child comes into contact

b3) Pseudomembranous rhinitis

- **microbial polymorphism** (streptococcus, staphylococcus, pneumococcus)
- anterior rhinoscopy: yellowish-white pseudomembrane
- nasal obstruction
- **treatment:** nasal toilet, nasal disinfectants, antibiotic therapy

b4) Gonococcal rhinitis

- **Neisseria gonorrhoeae** (passage through the infected maternal birth canal)
- **Diagnosis:**
  - within the first 24 hours, the following appear:
    - swelling of the upper lip
    - gonococcus ophthalmia
    - nasal obstruction
    - ***abundant, creamy, greenish rhinorrhea***
    - severe toxic-septic syndrome
    - certainty: **bacteriological examination**
- **treatment:**
  - antibiotics
  - nasal and ocular disinfection with 0.5% **protargol**
  - nasal and ocular instillations in the newborn with **silver nitrate solutions**
  - control of vaginal secretions in pregnant women

b5) Rhinitis from early congenital syphilis

- between 15 and 40 days after birth
- **Diagnosis:**
  - nasal obstruction
  - muco-purulent-bloody rhinorrhea
  - blackish crusts
  - ulceration, necrosis of septal cartilage
  - dermatological signs
  - bacteriological examination
  - BWR (newborn and mother)
- **Complications:**
  - chronic atrophic rhinitis
  - deformities of the nasal pyramid (synechiae, scar retractions)
- **Treatment: antisyphilitic drugs** for the newborn and the mother

2. Specific acute rhinitis:

a) Measles rhinitis

- nasal obstruction
- sneezing
- epistaxis
- Koplik's sign
- characteristic rash

b) Scarlet fever rhinitis

- nasal obstruction
- mucous, then purulent rhinorrhea
- raspberry tongue
- characteristic rash
- febrile state

c) Diphtheria rhinitis

- Löffler bacillus
- **Diagnosis:**
  - febrile state
  - pallor
  - adynamia, asthenia
  - serous-bloody rhinorrhea
  - **false membranes,** whitish-black, thick, adherent
  - ulceration of the nasal mucosa
  - subangulomandibular adenitis
  - bacteriological examination
- **Treatment**
  - **antidiphtheria treatment** (antidiphtheritic serotherapy, toxoid)
  - penicillin

CHRONIC RHINITIS

Specific chronic rhinitis

Nasal tuberculosis

- primary or secondary localization of **Koch's bacillus** in the nasal passages
- occurs only **exceptionally**
- **Clinical symptomatology:**
  - muco-purulent rhinorrhea / crusts
  - local pain / nasal obstruction
  - involves the anterior septum or the inferior turbinates
  - lesions – polymorphism
  - simple ulceration / papillomatous masses
  - perforation of the nasal septum
  - cartilage damage (nasal lupus)
- **Diagnosis:**
  - clinical signs

  - culture
  - biopsy
- **Treatment:**
  - identical to that of pulmonary tuberculosis: **tuberculostatic treatment**

Rhinoscleroma

- specific chronic rhinitis
- etiological agent: **Klebsiella rhinoscleromatis (Frisch bacillus)**
- conditions of occurrence - hygienic deficiencies
- **Symptoms:**
  - any segment of the upper respiratory tract
  - chronic inflammation
  - limited to the mucosa
  - inflammatory infiltrate rich in lymphocytes (**Mikulicz vacuolar cells**)
- **Evolution stages:**
  - **1. Catarrhal stage** - muco-purulent rhinitis - several weeks
  - **2. Atrophic stage** – extensive crusts – in both nasal fossae
  - **3. Granulomatous stage**: nodules in the nasal fossae, pharynx, larynx, trachea and bronchi; the granulomatous nodules cause stenosis of the nasal fossae, nasopharynx and, exceptionally, the trachea
- **Diagnosis**
  - identification of typical cells (**Mikulicz cells**)
  - bacterial cultures identifying the etiological agent (**Frisch bacillus**)
  - complement fixation test
- **Treatment**
  - chemotherapy
  - radiotherapy
  - antibiotic therapy according to antibiogram
- **Prognosis**
  - reserved
  - high mortality due to **laryngeal involvement**

Wegener Granulomatosis

- chronic systemic vasculitis of small and medium vessels
- autoimmune etiology
  - affects only certain organs, with **variable predilection**
- **Signs and symptoms**
  - **Disseminated form** involving:
    - upper respiratory tract, lungs, kidneys
      - cough
      - dyspnea
      - hemoptysis
      - pleural pain
      - general condition affected
      - fever
      - lethargy
      - fatigue

        - weight loss
        - sweats
        - migratory arthralgia
    - **Localized form**
        - affects the upper airways
        - nasal obstruction / crusts / purulent or bloody rhinorrhea
        - painful or painless ulceration
        - perforated nasal septum – "saddle nose"
        - dysphonia / stridor due to subglottic stenosis
        - otorrhea, otalgia / conductive or sensorineural hearing loss
        - scleritis, episcleritis, uveitis, conjunctivitis
- **Diagnosis:**
    - clinical examination – lung, kidney and upper respiratory tract involvement
    - biological analyses:
        - c-ANCA / proteinase 3 (PR3)-ANCA / myeloperoxidase (MPO)-ANCA
    - biopsy – essential
- **Therapy:**
    - high-dose corticosteroids (prednisone 1 mg/kg/day for 1 month)
    - cyclophosphamide
    - antibiotic therapy – Trimethoprim-sulfamethoxazole
    - topical steroids and nasal irrigation
- **Differential diagnosis:**
    - microscopic polyangiitis
    - other vasculitis
- **Prognosis:**
    - relapses can be prolonged and problematic
    - long-term complications: renal failure and hearing loss

NASAL OBSTRUCTION

- "blocked" nose
- nasal blockage or nasal congestion
- unilateral or bilateral
- transient or permanent

Etiology / Epidemiology:

- **Mechanical (Structural):**
    - alar collapse caused by insufficient rigidity of the alar cartilage
    - vestibular synechia or scars
    - septal deviations
    - septal perforations
    - turbinates hypertrophy
    - nasal malformations
    - atresia
    - hematoma
    - nasal polyposis, antrochoanal polyp
    - benign tumors: angiofibroma, inverted papilloma
    - malignant tumors: squamous cell carcinoma, adenocarcinoma, malignant melanoma

- **Functional causes:**
  - allergic rhino-sinusitis
  - acute or chronic rhino-sinusitis
  - rhinitis or drug side effects
  - pregnancy rhinitis
  - atrophic rhinitis
  - chemical rhinitis (inhaled substances, occupational or medicinal products)
  - Wegener granulomatosis

Non-allergic rhinitis and primary ciliary dyskinesia

- rhinitis is defined by the presence of nasal symptoms:
- obstruction / pruritus / secretions / sneezing
- according to skin tests and serum immunoglobulins (IgE), chronic rhinitis has traditionally been classified as:
- "allergic"
- "non-allergic"

Non-allergic rhinitis diagnosis:

- obstruction / pruritus / secretions / sneezing
- **negative allergy work-up** (history, skin tests, specific IgE)
- diagnosis is based on the exclusion of allergy and sinusitis
- non-allergic rhinitis is actually a ***group of syndromes*** without generally accepted diagnostic criteria
- Epidemiology:
  - 2-4% of the general population suffer from chronic nasal diseases with daily symptoms and need medication
  - ***approximately 50% of patients with rhinitis do not have allergic rhinitis***
  - non-allergic rhinitis usually develops in the middle-aged population
  - allergic rhinitis tends to develop mainly in children

Occupational rhinitis:

- episode, **related to work**
- sneezing, nasal obstruction and nasal secretion
- mechanism mediated by IgE
- **non-specific inflammatory mechanism** of the nose
- triggers:
  - cereals
  - laboratory animals
  - wood dust
  - latex
  - chemicals: platinum salts, acid anhydrides, adhesives and solvents
- may be associated with occupational asthma
- **Treatment**
  - prevention
  - environmental control: adequate ventilation or use of masks
  - intranasal and systemic antihistamines
  - anticholinergic drugs
  - decongestants

Drug-induced rhinitis:

- worsening of nasal obstruction in patients who regularly use **nasal decongestants**
- of special interest in **newborns**, because rebound nasal obstruction can lead to feeding difficulties and **respiratory failure**
- **Signs and symptoms:**
  - edema and irritation of the nasal mucosa
  - turbinate hypertrophy
- **Treatment:**
  - intranasal steroids
- **Drugs known to cause drug-induced rhinitis:**
  - aspirin and other non-steroidal anti-inflammatory drugs (NSAIDs)
  - alpha-blockers, beta-blockers
  - angiotensin-converting enzyme (ACE) inhibitors
  - cocaine, guanethidine, methyldopa
  - oral contraceptives, reserpine, phentolamine

Rhinitis and pregnancy:

- nasal congestion in **the last 6 weeks of pregnancy**
- in the absence of an allergic reaction or infectious cause
- **complete resolution** of symptoms **within 2 weeks after delivery**
- affects approximately 20% of pregnant women
- **Treatment**
  - still being studied
  - **nasal decongestants**, although useful, tend to be overused by pregnant patients with rhinitis, and may lead to **drug-induced rhinitis**
  - corticosteroids are not used

Rhinitis associated with physical and chemical factors:

- diving in cold water
- cold, dry air ("skier's nose")
- **ipratropium bromide** may be a useful treatment

Food-induced rhinitis:

- spicy foods "taster rhinitis"
- the syndrome can be treated with **intranasal atropine**

Atrophic rhinitis:

- infection with **Klebsiella pneumoniae subsp. ozaenae**
- progressive atrophy of the nasal mucosa
- ozena presents with **crusts, fetor, hyposmia and enlargement of the nasal cavity,** and **paradoxically with nasal congestion**
- the incidence of **primary** atrophic rhinitis decreased significantly in the last century
- the most common type nowadays is the **secondary** form
- trauma, surgery (e.g. aggressive resection of the turbinates)
- infections
- exposure to radiation

Atrophic rhinitis (Ozena)

- rare

- chronic disease of the nasal mucosa and underlying bone structures
- abnormally wide nasal cavities
- dryness, atrophy, fetor
- paradoxical sensation of nasal obstruction
- young people in developing countries with a warm climate
- associated with environmental, endocrine, vascular, nutritional, autoimmune and genetic factors
- **Epidemiology/etiology**
  - is not known
- **Primary form**
  - Klebsiella Ozaenae
- **Secondary form**
  - aggressive surgery for nasal obstruction
  - trauma
  - granulomatous diseases
  - cocaine use
  - radiotherapy
- **Signs and symptoms**
  - fetid rhinorrhea
  - crusts
  - **paradoxical subjective sensation** of **nasal obstruction**
  - septal perforations with deformity of the nasal pyramid (**saddle nose**)
- **Diagnosis**
  - patient history
  - clinical exam, anterior rhinoscopy
  - nasal exudate
  - antibiogram
  - CT and MRI investigations – evaluation of paranasal sinuses
- **Therapy**
  - **Conservative**
    - local antibiotics, lavage and crust removal, 50% glucose
    - glycerin / streptomycin / rifampicin
  - **Surgical –** narrowing of the nasal fossa using a circular flap of vestibular skin (**Young's procedure**)

Non-allergic rhinitis with eosinophilia syndrome (NARES):

- Non-allergic rhinitis may be classified according to the type of inflammation found in the mucosa
- **Non-allergic rhinitis with eosinophilia syndrome** (**NARES**) is characterized by:
  - nasal symptoms
  - nasal eosinophilia (more than 20%)
  - negative allergy test
  - **clinical symptoms are usually more intense than in allergic or vasomotor rhinitis**
  - anosmia is common
  - **the mechanism of eosinophilic infiltration is not known**
  - Although NARES is usually an **isolated syndrome**, it can be accompanied by:
    - asthma

        - aspirin intolerance
        - nasal polyposis
        - early stage of Widal's (or Samter's) triad
- **Treatment of NARES**
    - can **sometimes be difficult**
    - it may even require **systemic steroids**
    - **intranasal steroids**, either alone, or in combination
    - with **oral antihistamines**, may be useful

Vasomotor rhinitis/Idiopathic rhinitis:

- hyperreactivity to **non-specific triggers** on exposure to:
- strong odors
- changes in air temperature or humidity
- alcohol ingestion or tobacco smoke
- **Diagnosis:**
    - exclusion of allergy
    - structural lesions
    - drug abuse or systemic diseases
    - **infection is usually excluded** if the nasal secretion is serous and not purulent
    - vasomotor rhinitis implies a **neurovascular dysfunction** of the mucosa, which has not yet been demonstrated
    - thus, the term **idiopathic** is more appropriate
- **sympathetic innervation** of the nose increases nasal patency and reduces nasal secretion, while **parasympathetic innervation** increases hypersecretion and nasal congestion
- **the balance** between the two systems **is disturbed** by **sympathetic hypofunction and parasympathetic hyperactivity**

Other possible causes of non-infectious non-allergic rhinitis:

Emotions:

- psychological stress and sexual arousal are known to worsen rhinitis through **autonomic stimulation**
- postmenopausal rhinitis and/or asthma have been confirmed
- gastroesophageal reflux: no medical study has been published
- metabolic conditions: hypothyroidism and acromegaly

Primary ciliary dyskinesia:

- all congenital abnormalities of ciliary function
- in **1933 Kartagener** published a series of 4 cases of sinusitis, bronchiectasis and dextrocardia
- male infertility has been added to the syndrome described by Kartagener
- Diagnosis:
    - **after** excluding other ***sino-pulmonary syndromes***, such as:
        - cystic fibrosis
        - immunodeficiency
        - Wegener granulomatosis
    - **a screening of ciliary motility and mucociliary transport is recommended:** nasal mucociliary transport can be studied with the **saccharin test**
- Treatment:

- antibiotics / physiotherapy
- **vaccination** against influenza virus, Streptococcus pneumoniae and Haemophilus influenzae
- pulmonary lobectomy is currently **rarely indicated**
- lung transplantation

ALLERGIC RHINITIS

Allergy – defines an immunological mechanism and not a clinical picture

- Inflammation of the nasal mucosa **mediated by IgE**, induced by exposure to allergens and characterized by episodes of:
- rhinorrhea, sneezing and nasal congestion
- persisting for at least one hour per day
- ± allergic conjunctivitis - **40%** of cases
- **40-50%** of patients **with allergic rhinitis** have **asthma**
- **75%** of patients **with asthma** have **allergic rhinitis**
- inflammatory disease of the nasal mucosa
- exacerbation of the defense mechanisms of the nasal mucosa against various stimuli
- allergy represents an immune mechanism – **antigen-antibody reaction**
- **Diagnosis** - requires **three diagnostic stages**
  - 1. **Allergens** – must be identified as etiological agents
  - 2. **The immunological pathogenic mechanism** of an allergy should be identified
  - 3. **The target organ of the allergy** should be specified
- allergic patients exhibit a **pathological immunological response**, both **humoral and cellular,** to the triggering allergens
- **the main role** is played by the **production of specific IgE antibodies** and their secondary binding to mast cells, at the level of the affected organ
- if **IgE antibodies** are shown to participate in the onset of symptoms, then the **diagnosis of allergy is certain**
- the **release of mediators** may be **induced by other mechanisms as well** – the **diagnosis cannot be established only on the basis of the clinical picture**
- the participation of a **specific allergic immune reaction** must be demonstrated
- with the release of mediators within the nasal mucosa:
  - preformed histamine
  - serotonin
  - chemoattractant substances for macrophages
- **Identify**
  - the immune mechanism
  - the allergen
- **Diagnostic scheme:**
  - 1. anamnesis
  - 2. clinical condition
  - 3. skin tests
  - 4. provocation tests
  - 5. allergen-specific IgE determinations
- from wind-pollinated plants
- types of pollen according to the pollination season:

  - early trees March - April - May
  - Poaceae (Cereals) - May - June
  - weeds (Ambrosia, Artemisia) August - September
- the pollination season differs depending on:
  - geographical location
  - vegetation of the area
  - atmospheric conditions – temperature, precipitation
  - day/night ratio
- **conditions for sensitization in atopic individuals**:
  - sufficient duration and intensity of exposure
- multi-allergic patients:
  - 25% of patients – allergic to grass (Poaceae) and weed pollen
- Pollen is a usually yellow powder consisting of ***microscopic grains*** from the anthers of the stamens.
- airborne pollen (**anemophilous**) is responsible for the vast majority of "**pollen allergies**", not the pollen harvested by bees, which is **entomophilous** (transported **from flower to flower** by various insects, such as bees, bumblebees, butterflies, etc.).

Clinical forms:

- **Seasonal** allergic rhinitis
- **Perennial** allergic rhinitis
- **Perennial-seasonal** allergic rhinitis

Comorbid affections:

- Asthma
- Otitis media
- Sinusitis
- Eczema
- Pharyngitis
- Lymphoid hypertrophy
- Obstructive sleep apnea
- Altering speech
- Conjunctivitis

ARIA Classification

Intermittent: <4 days/week or <4 weeks

Persistent: >4 days/week and >4 weeks

Mild

- normal sleep
- without impairment of daily, sport and leisure activities
- normal school/work
- without troublesome symptoms

Moderate-severe - one or more:

- abnormal sleep
- impairment of daily, sport and leisure activities
- school/work impairment

- troublesome symptoms

Diagnosis:

- allergic anamnesis
- symptoms presentation
- ENT clinical examination
- serum eosinophil
- dosing IgE
- allergic provocation tests
- radiological examination

Rhinological symptoms:

- aqueous rhinorrhea
- paroxysmal sneezing
- nasal obstruction
- nasal pruritus

Sneezing

- most characteristic symptom
- **bursts** of 10-20 sneezes in rapid succession
- occurring without prodrome or preceded by nasal pruritus
- activates the tear reflex

Rhinorrhea

- light secretion → abundant and continuous
- skin irritation of the upper lip

Pruritus

- causes frequent nose rubbing → "**allergic salute**"

Nasal obstruction

- may occasionally be a **major or the only symptom**
- if severe – interferes with the aeration and drainage of the paranasal sinuses or Eustachian tubes → headache + otalgia
- **severe and continuous congestion → loss of smell and taste**

Conjunctivitis

- frequently associated with
- pruritus and tearing of the eyes
- photophobia and sensitivity → "tired eyes"
- scleral and conjunctival injection, chemosis

Marked pruritus in the ears, palate, neck or face

- can be extremely troublesome

Non-productive cough

- due to a feeling of irritation in the throat and posterior drainage of nasal secretions

Systemic symptoms

- weakness, indisposition, irritability, fatigue and anorexia

Sleep disorder - following permanent obstruction

ENT Clinical exam:

- A. R.
- P. R.
- **Rigid and flexible endoscopy:** edematous, pale, discolored mucosa

Objective exam

- the presence of **grimaces** intended to relieve the itching
- nasal pruritus
- presence of mucous nasal secretions
- **mouth breathing** → high-arched palate and dental changes
- infra-orbital darkness
- **excessive tearing**, conjunctival and scleral hyperemia

Anterior rhinoscopy

- **abundant sero-mucous secretions** in the nasopharynx
- **pale blue coloration** of the mucosa
- hypertrophy of the inferior turbinate
- ± nasal polyposis
- ± deviation of septum

ACUTE PHASE

- degranulation of preformed and newly synthesized mediators, the most important being **histamine**
- sneezing
- aqueous rhinorrhea
- ocular, nasal and palatal pruritus

TRANSITIONAL PHASE

- asymptomatic patient
- activation and recruitment of blood leukocytes in the nasal mucosa

LATE PHASE

- local infiltration with leukocytes
- **occurs 4-6 hours after** exposure to the allergen
- **nasal congestion** is the most characteristic symptom

The goal of therapy is:

- a) **identification of sources of allergens** and, where possible, their removal
- b) if it is not possible (in whole or in part) **immunotherapy** (desensitization)
- c) when a and b fail: **symptomatic drug therapy**
- Drug therapy
  - when causal therapy is:
    - with no effect
    - when it cannot be applied
  - **Anti-allergic drugs** interfere in the molecular pathway of the ***biochemical phase*** of the pathogenic mechanism
  - These drugs are **inhibitors of the release or synthesis of mediators**

- Environmental factor control:
  - the best treatment is prevention
  - mites
    - performance filters
    - washing sheets and underwear with hot water
    - special hypoallergenic sheets
- **Pollen**
  - pollen masks
- **Avoiding irritants** such as cigarette smoke

TREATMENT:

- avoidance of allergens
- nasal toilet
- desensitization
- pharmacological treatment
- cryosurgery or CO2 laser surgery
- surgical resections

1. alpha-adrenergic agonists (Oxymetazoline, Phenylephrine)
2. inhaled cromoglicate
3. antihistamines
4. leukotriene
5. intranasal inhaled corticosteroids
6. systemic corticosteroids

anamnesis and ENT clinical examination are very important in establishing the diagnosis

Treatment objectives:

- control of environmental factors
- first-line pharmacotherapy
- intranasal and systemic steroids
- adequate patient counseling

NASAL POLYPOSIS

- from mucosal edema and solitary polyps to diffuse nasal polyposis
- 5% of the European population
- 5% of ENT consultations
- 4% of allergy consultations
- occurs in approximately 0.6% of adults, but increases to 15% in patients suffering from asthma
- up to 95% of patients with bronchospastic intolerance to analgesics will develop chronic sinusitis with polyps
- more frequent in nonasthmatic male atopic patients
- in patients with asthma there is no difference in the prevalence between men and women
- nasal polyposis may follow **asthma** and **intolerance to aspirin**
- nasal polyposis can cause **serious complications if not treated**

Origin of polyps

- from ethmoid cells

- like the tip of an iceberg, in some patients small polyps appear behind the middle turbinate, while the entire ethmoid sinus may be occupied by polyps
- the ***radiological examination*** specifies the extension of the polyps; however, **CT cannot differentiate between nasal polyposis and secretions**

Treatment

- decreased nasal blockage, symptoms of rhinitis, asthma
- improvement of sinus drainage
- ablation of nasal polyps
- **solitary and non-eosinophilic polyps** are **NOT difficult to manage** and generally **DO NOT relapse**
- **ostiomeatal surgery is the key** to the functioning of the middle nasal meatus
- **eosinophilic nasal polyposis** poses a **challenge** for both clinician and surgeon:
- improves nasal breathing, olfaction, symptoms of rhinitis and asthma
- does not always remove nasal polyps and sinus pathology

Relapse

- treatment of **eosinophilic diffuse nasal polyposis**:
  - topical and systemic corticosteroids (CS) – the best results
  - leukotriene antagonists
  - immune system stimulants and antifungals
  - desensitization to aspirin
  - no treatment provides complete healing
  - surgical treatment is an adjunct to medical treatment and should not be considered the primary treatment

Surgery

- simple polypectomy
- FESS – standard surgery
- in case of diffuse polyposis, a more extensive procedure ("pansinus surgery")
- **the basic pathology resides in the mucus – in other words, the stimulus comes from the mucus**
- purpose of surgery – **drainage of this mucus**
- the main cause of relapse – areas that have not been opened or drained

Nasal polyposis in children

- the **incidence** of nasal polyposis in children is **quite low**
- symptomatic nasal polyposis is generally bilateral and associated with a systemic
- children under 16 years of age with bilateral nasal polyposis should be evaluated for ***cystic fibrosis***
- **histopathologically,** polyposis in children is **different** from that in adults
- eosinophilic polyposis does not occur as frequently in children as in adults

Surgery

- **reserved** in patients with
  - complete nasal obstruction
  - deformity of the facial skeleton
  - enlargement of the nasal dorsum
  - ogival palate

NOSE — VASCULAR ANATOMY. EPISTAXIS

- the nasal cavities are vascularized by terminal branches of the external (ECA) and internal (ICA) carotid arteries
- vessels originating from the **external carotid artery (ECA):**
  - facial artery
  - superior labial artery
  - internal maxillary artery
  - sphenopalatine artery
  - descending palatine artery
- 3 branches of the **internal carotid artery (ICA)**:
  - Vidian artery – petrous portion of the internal carotid artery
  - anterior ethmoid arteries
  - posterior ethmoid arteries
- vascular plexus (**Kiesselbach's area** or **Valsalva's area**)
- supplied by both carotid arteries (ECA and ICA)
- septal anastomosis in **Woodruff's area**

Epistaxis

- blood loss from the nasal fossa
- disruption of the nasal mucosa and blood vessels
- depending on the site of origin, it is divided into **anterior, posterior and diffuse** epistaxis

Etiology/Epidemiology:

- causes of epistaxis: local and systemic
- **the most common causes** of epistaxis are trauma and inflammatory diseases
- endonasal surgery
- recurrent epistaxis in tumors
- medical history: hypertension / anticoagulants
- men are more affected than women up to 50 years of age
- after the age of 50, severe epistaxis is more common

Symptoms:

- nasal obstruction
- vomiting
- syncope episodes - cardiovascular problems

Clinical forms:

- epistaxis has the following clinical forms, depending on the **origin of the hemorrhage**:
  - **anterior epistaxis** – from the vascular plexus of Kiesselbach, located in the antero-inferior area of the cartilaginous septum, 1 cm above the nasal spine (**over 90% of cases**)
  - **posterior epistaxis** – from the sphenopalatine branches (the most massive and serious hemorrhages)
  - **superior epistaxis** – from the anterior and posterior ethmoid arteries;
  - **diffuse** hematological epistaxis
- depending on the **amount of blood lost**, the severity of the hemorrhage may be:
  - mild, moderate, malignant or severe epistaxis
- according to **etiology**, epistaxis may be classified into **two large groups**:

- **primary, essential** epistaxis of young people – recurrent, benign, with no detectable etiology
- **secondary or symptomatic** epistaxis represents approximately **98% of all nasal bleeding**

- occurs as the ***only presenting symptom*** or as a ***complication of local or general conditions,*** announcing a worsening course, sometimes even decompensating the underlying disease

I. Local epistaxis:

- 1. Inflammatory causes
- 2. Traumatic causes
- 3. Tumor causes

II. Epistaxis of general cause:

- 1. Hematological causes (disturbance of one of the three phases of hemostasis)
- 2. Cardiovascular causes
- 3. Liver causes
- 4. Kidney causes
- 5. Avitaminosis A, C, K, states of starvation
- 6. Infectious causes
- 7. Toxic causes
- 8. Endocrine causes
- 9. Chronic conditions
- 10. Various causes: ruptures of the internal carotid artery (trauma, aneurysms), active vascular dilation: in intense physical exertion, acute sunstroke, strong emotions

I. Local epistaxis:

1. Inflammatory causes:

- **acute rhinitis** – in infectious-contagious respiratory diseases (influenza, measles, scarlet fever, chickenpox)
- **chronic rhinitis** – trophic ulceration, Hajek ulcer, ozena, nasal tuberculosis, nasal lupus, nasal syphilis
- **local inflammation:** intranasal foreign bodies, rhinolithiasis

2. Traumatic causes:

- **accidental local trauma** (work, traffic, sport, assault) causing damage to the mucoperichondrium and nasal septum, nasal fractures, fractures of the naso-ethmoido-maxillary complex
- **local microtrauma**: physical factors (nose picking, nose blowing, sneezing, nasal foreign bodies, dust inhalation), chemical factors (potassium bichromate, mercury, arsenic)
- **iatrogenic** – surgical trauma
- **barotrauma** (aviators, divers)

3. Tumor causes:

- **benign tumors:** bleeding polyp of the nasal septum, nasal polyposis, papilloma, granuloma, angioma, nasopharyngeal angiofibroma
- **ulcerated sinonasal malignant tumors** (epithelial and connective tissue)

Epistaxis of general cause:

1. Hematological causes (disturbance of hemostasis):

- ***coagulopathies*** (disturbances of blood coagulation) which may be:
- a) **congenital** (hemophilia A, B, congenital afibrinogenemia)
- b) **acquired** (hepatic impairment; acute fibrinolysis in heat stroke, polytrauma, birth complications; anticoagulant treatments – heparin, thrombolytics, aspirin)
- ***vasculopathies*** (disturbance of the vascular phase of hemostasis): rheumatoid purpura, hereditary hemorrhagic telangiectasia (Rendu-Osler), von Willebrand disease
- ***thrombocytopathies*** (disturbance of the platelet phase of hemostasis – thrombopenic or thrombopathic): thrombocytopenia in acute leukemia, acute reticulosis, myeloma, drug-induced blood dyscrasia.

2. Cardiovascular causes:

- hypertension – mainly causes posterior epistaxis from the branches of the sphenopalatine artery
- atheromatosis and atherosclerosis
- congestive and global heart failure (mitral decompensation)

3. Liver causes

- hepatic impairment, cirrhosis of the liver, toxic hepatitis, hepatic coma

4. Kidney causes

- decompensated renal failure, diffuse chronic glomerulonephritis

5. Avitaminosis A, C, K, starvation

6. Infectious causes

- eruptive diseases (measles, scarlet fever, chickenpox)
- influenza, ictero-hemorrhagic leptospirosis

7. Toxic causes

- acute and chronic general intoxication, severe toxic-septic states

8. Endocrine causes

- puberty, menstruation, the first months of pregnancy, spontaneous or induced menopause

9. Chronic conditions

- tuberculosis, diabetes, neoplasms, collagen diseases

Complications:

- cardiovascular disease – especially when blood pressure is not monitored
- amaurosis – intraorbital bleeding from the ethmoid arteries
- ligation of the ethmoid artery – enophthalmos, diplopia and even vision loss

Diagnosis:

- ENT clinical examination
- nasal endoscopy
- inspection of the oral cavity, nasal cavity and nasopharynx
- EKG
- laboratory tests (blood count, coagulation tests)

Additional diagnostic methods:

- blood pressure monitoring
- culture of nasal secretions
- CT/MRI scan

Treatment:

Conservative:

- in young patients with minor epistaxis, the nostrils should be pinched with the fingers for approximately **10 minutes,** with the patient seated and the head tilted forward
- ***digital compression*** can stop the bleeding and prevent swallowing of blood
- chemical cauterization with ***silver nitrate*** is not effective in active nasal bleeding
- in **severe epistaxis**, treatment consists of endoscopic bipolar cauterization
- when nasal endoscopy is not possible, nasal packing is indicated
- the nasal packing should be removed after 24 hours and nasal endoscopy performed after applying mucosal decongestants, to ***identify the site of the hemorrhage***
- if the bleeding has stopped, ***nasal irrigation*** and the ***use of antibiotic ointments*** are recommended for a few days

Additional therapeutic strategies:

- avoiding NSAIDs
- vitamin C
- vascular protective
- hemostatic agents
- nasal washing
- Balloon Foley

Surgical:

- **treatment of choice**: ***bipolar cauterization*** of the bleeding site under endoscopic control
- the procedure may be performed under local or general anesthesia, depending on the general condition of the patient
- ***selective embolization*** with polyvinyl alcohol under general anesthesia is recommended only in special cases
- ***arterial ligation*** – selected cases
- ***ligation of the external carotid artery*** should be considered when the other procedures fail to stop the bleeding

Prognosis:

- depends on the underlying disease
- **favorable** for nasal bleeding due to **local causes** (excluding tumors)
- when epistaxis is a symptom of an underlying systemic disease, **treatment of the primary disease** is the main concern
- treatment may be long-term, or **no treatment may be effective**, as in **hereditary hemorrhagic telangiectasia**
- when bleeding reduces the patient's hemoglobin to **less than 7 g/dl**, **transfusion is indicated**

ACUTE SINUSITIS

Etiology/Epidemiology

- 10-15% of the population of Central Europe adults and children contract an average of 2-3 and 3-8 respiratory viral infections per year
- acute bacterial rhinosinusitis (ABRS) ~2%

Favoring factors:

- immunosuppression
- asthma
- sinonasal allergies
- cystic fibrosis
- exposure to smoke
- cocaine consumption

Etiological agents:

- Streptococcus Pneumoniae
- Haemophilus influenzae
- Branhamella catarrhalis
- Staphylococcus pyogenes
- Staphylococcus aureus
- other anaerobes

Positive diagnosis:

- signs and symptoms of ABRS **are not specific** to bacterial sinusitis
- signs and symptoms of ABRS **are difficult to differentiate** from those of viral infections
  - **lack of reliable, convenient and practical diagnostic methods**
    - nasal congestion
    - anterior and posterior rhinorrhea
    - feeling of pressure / facial pain
    - fever
    - cough
    - dental-maxillary pain
    - sensation of ear fullness accompanied by pressure

Positive diagnosis:

- **lack of reliable, convenient and practical diagnostic methods**
- symptoms persisting beyond 7 days may be a sign of bacterial infection
- the **diagnostic standard for bacterial sinusitis** is ***sinus puncture with aspiration of purulent secretions***, but this invasive method is **rarely used**
- samples obtained under endoscopic guidance
- endoscopically guided evaluation is an alternative to sinus puncture, even in acute sinusitis
- **sinus imaging**
- ***standard X-ray* –** lacks specificity; this method **cannot** differentiate between infection and other sinus abnormalities such as mucosal thickening, tumor or polyp
- CT
  - excellent view of paranasal sinuses
  - high-resolution images of ostiomeatal anatomy

  - can detect a variety of sinus abnormalities
  - lacks specificity – viral infection vs ABRS
- MRI
- ***ultrasound*** – may support the suspected diagnosis, **but has limited value** in the routine diagnosis of acute uncomplicated bacterial sinusitis
- ***transillumination*** – the clinical usefulness of this method is still **controversial**

Complications

- orbital cellulitis
- orbital abscess
- meningitis
- cerebral abscess
- subdural-epidural abscess
- ***ethmoid, sphenoid and frontal localizations*** are more commonly associated with complications than maxillary sinusitis

Medical treatment

- choice of treatment
- restoring the ***integrity*** of the ostiomeatal complex
- restoring ostiomeatal complex ***function***
  - reduction of inflammation
  - restoring sinus drainage
  - eradication of the bacterial cause of infection
  - prevention of complications
- because **viruses** make a major contribution to the etiology of sinusitis and in most cases **resolve spontaneously**, the "**Wait and See" policy** combined with **symptomatic treatment** is the usual therapeutic strategy

Medical treatment

- **First-intention antibiotics**
  - **amoxicillin** as first-line treatment
  - penicillin-allergic patients – trimethoprim or trimethoprim-sulfamethoxazole (**Biseptol**)
  - therefore the use of **cephalosporins** should **be limited** as far as possible, in order not to select resistant strains
- **Second-line antibiotics**
  - alternatives for patients who **do not respond to first-line treatment**
  - change of medication if, **after 3 days**, symptoms **do not** improve or worsen
  - 2nd-generation cephalosporins
  - macrolides
  - fluoroquinolones with anti-pneumococcal activity
- **Useful additional therapeutic strategies**
  - analgesics
    - antipyretics
    - decongestants
    - nasal irrigation
    - inhalations
    - topical steroids

Surgical treatment

- antral punctures – **rare**
- **endoscopic sinus surgery** - in cases of recurrent sinusitis after specific investigations (nasal endoscopy, CT)

Prognosis

- ***high doses of antibiotics*** administered for a ***specified period*** ensure healing in most cases
- there may be ***sensorineural hearing loss*** in the high-frequency range
- in case of **relapse**, ***further investigations*** are needed to rule out dental infections or local factors (ostiomeatal dysfunction)

CHRONIC RHINOSINUSITIS

- rhinosinusitis has been classified according to duration into:
  - acute
  - chronic
  - the definition of chronic rhinosinusitis is **difficult**, data are limited
  - the definition of chronic rhinosinusitis raises many **controversies**
  - rhinitis and sinusitis usually coexist, thus the **correct term** is **rhinosinusitis**
  - inflammation of the mucosa of the **sinuses** associated with inflammation of the **nasal** mucosa, and often associated with inflammatory changes in **adjacent bone structures**
    - prevalence and morbidity
  - affects up to ***15-16% of the adult population***
  - the factor associated with this etiology is the **ciliary insufficiency**
  - ***primary dyskinesia*** - Kartagener syndrome, cystic fibrosis
  - ***secondary dyskinesia*** – damage to the sinonasal epithelium, **usually reversible** once the sinus inflammation and infection are corrected
  - **atopy** – predisposing factor
  - dysfunction of the immune system

Anatomical variations:

- concha bullosa
- deviation of the nasal septum towards the middle meatus
- the concept that **chronic rhinosinusitis** is **induced by a fungal infection** due to fungi that normally colonize the sinus, forming saprophytic crusts
- most patients with chronic rhinosinusitis show, on histology or culture, **eosinophilic infiltration and the presence of fungi**
- however, the ***presence of bacteria or fungi*** in the sinus cavity **does not prove** that ***these pathogens cause the occurrence*** of the disease

Symptoms:

- nasal blockage/congestion
- hypo/anosmia
- anterior/posterior rhinorrhea
- pain/facial pressure
- the presence of **two or more symptoms** of either:
  - nasal/facial blockage
  - obstruction/congestion
  - nasal discharge (anterior/posterior)

- +/- facial pain/pressure
- +/- hypo/anosmia is **mandatory for the diagnosis of rhinosinusitis**

Diagnosis:

- **major tests are**:
  - endoscopic examination
  - CT exam
- minor tests are:
  - nasal cytology
  - bacteriological examination
  - sinus ultrasound
  - plain sinus X-ray has low sensitivity for the diagnosis of rhinosinusitis
- **CT exam**
  - is the imaging method of choice
  - confirms the previous clinical diagnosis
  - is a guide to sinus lesions
  - provides information about **surgical anatomical characteristics**
- concha bullosa
- paradoxical middle turbinate
- Haller cells
- variations of the agger nasi cells
- CT examination in **asymptomatic patients** has an incidence of:
- sinus occupations
- cysts
- **edema of the mucous membrane greater than 3 mm**
- incidence between 30 and 60%, higher in children
- **subclinical viral infections or physiological changes of the nasal cycle may be responsible for these imaging findings**
- **Ultrasound:**
  - cheap screening
  - **high sensitivity**
  - **low specificity**
  - false-positive results:
    - Tumors
    - Cysts
    - Polyps
    - mucosal edema
- **there is no correlation** between ultrasound changes and clinical signs of sinus pathology
- assessment of the ethmoid and sphenoid sinuses is impossible
- the ethmoid sinus is the key to the pathophysiology of sinus inflammation
- quick, noninvasive test, **but not useful in diagnosis**

Microbiology studies:

- **acute rhinosinusitis**
  - Haemophilus influenzae
  - Streptococcus pneumonia
  - Moraxella catarrhalis

  - Staphylococcus
  - **chronic rhinosinusitis**
    - microbiology studies are **not conclusive**
    - infection is often polymicrobial
    - anaerobes
      - Peptostreptococcus spp.
      - Fusobacterium spp.
      - Prevotella spp.
      - Streptococcus Milleri
      - Corynebacterium
- **Pseudomonas aeruginosa** and **Proteus mirabilis** could be found in chronic rhinosinusitis in patients with ***cystic fibrosis*** or ***immunodeficiency***

Pathophysiology:

- optimal maintenance of ventilation and clearance of sinuses
- obstruction of the ostiomeatal complex contributes to endosinusal hypoxia
- edema, stagnation of secretions and increased viscosity occur
- then bacterial colonization occurs
- if conditions persist, **chronic rhinosinusitis may occur**

Treatment:

- absence of nasal polyps
- intranasal topical steroids
- chronic moderate/severe rhinosinusitis
- long-term topical steroids
- administration of macrolides
- **failure after 3 months** of treatment
- endonasal endoscopic surgery
- presence of bilateral nasal polyps
- **intranasal topical steroids** - very long term
- **failure after 3 months** of treatment
- surgery should be considered
  - short-term administration of oral steroids
  - long-term administration of macrolide antibiotics

Complications:

- periorbital edema
- exophthalmos
- diplopia or reduction of visual acuity and
- ophthalmoplegia
- severe frontal headache
- frontal edema
- meningitis
- neurological signs
  - mandatory hospitalization
  - further investigations
  - surgical procedures

SPECIFIC SINUSITIS (FUNGAL RHINOSINUSITIS)

- species of fungi involved in rhinosinusitis pathology may act as:
  - **allergen** - **all types of molds** present in the environment
  - **infectious agent** - **only some species** have the ability to develop in the human body:

Aspergillus fumigatus

- low invasive power
- opportunistic pathogens
- their pathogenicity is expressed ***only when the host's defense capacity is compromised***
- **non-invasive diseases**
  - **fungal balls**
  - allergic fungal rhinosinusitis
  - **invasive diseases**
  - **acute invasive mycotic rhinosinusitis**
  - exceptionally – indolent or chronic invasive fungal sinusitis

Fungal balls

- localized infections
- low aggressively
- without invasion of the mucosa
- immunocompetent patients
- involves the **maxillary sinus (89% of cases)**, but all sinuses may be involved, including the sphenoid
- **Epidemiology**
  - adults only
  - more common in women
- **Symptoms**
  - nonspecific
  - clinical picture of chronic rhinosinusitis
  - poor response to medical treatment
  - unilateral
  - **chronic pain** (especially when the sphenoid is involved)
  - purulent rhinorrhea
  - obstruction
  - cacosmia
  - fungal balls **are commonly incidental findings** on imaging
- **Diagnosis:**
  - inspection – imprecise information
  - rhinoscopy and nasal endoscopy
    - normal, or shows at the level of the meatus:
      - purulent secretions
      - edema, sometimes with polyposis
  - fragments of fungal balls
  - **CT exam:**
    - unilateral opacity, often heterogeneous, involving one or sometimes several sinus cavities
    - bone thickening

        - **bone lysis**
        - **pseudometallic foreign body** or one or more macrocalcifications
        - microcalcifications
        - sometimes extensive bone erosion (pseudotumoral forms)
    - **macroscopic appearance:**
        - solid, crumbly mass
        - brown-black
        - creamy secretion
        - **acid-Schiff, Grocott-Gomori staining**
        - fungal filaments
        - nonspecific inflammation of the mucosa
        - **direct microscopic examination**
        - fungal filaments - over 70% of cases
    - **culture**
        - identifies the fungal species in 30-60% of cases; Aspergillus – **A. fumigatus** – **is the most commonly involved species**
    - **additional/useful diagnostic procedures**
        - dental examination
        - devitalized teeth
        - x-ray sinuses
        - unilateral maxillary opacity
        - dentoalveolar x-ray
        - MRI: **not useful**
            - fungal balls have a hypointense signal on T1-weighted and a moderate signal on T2-weighted images
            - the mucosa has a hyperintense signal on T2-weighted and a moderate signal on post-gadolinium T1-weighted images
- **Surgical treatment**:
    - complete surgical removal of the fungal ball and secretions
    - maintaining the integrity of the sinus mucosa
    - antifungal therapies are unnecessary
    - creating a wide opening of the sinuses involved
    - restoration of drainage and ventilation of the sinus
- **Differential diagnosis:**
    - the CT examination is suggestive of the diagnosis
    - surgical intervention
    - pathological and mycological examination
    - pseudotumor forms
    - fungal allergic sinusitis
    - invasive fungal rhinosinusitis
    - tumor
- **Prognosis**
    - **healing in all patients** after complete elimination of fungal balls and restoration of ventilation and drainage of the sinuses
    - relapses are exceptional and generally occur when some fungal fragments remain after surgery

- **Surgical treatment**
- endoscopic sinonasal surgery
  - middle meatal antrotomy
  - inferior meatal antrotomy
  - minimal trephination of the canine fossa (Caldwell-Luc procedure)
  - ethmoidectomy – ethmoidal extension of the fungal balls
  - sphenoidectomy - sphenoid fungal balls
  - possible external approach and frontal sinusectomy
- **Allergic fungal rhinosinusitis**
  - patients with recurrent nasal polyposis and asthma
  - the concept was inspired by ***allergic bronchopulmonary aspergillosis***
    - pathogenicity of fungi:
      - mechanisms **dependent on IgE**
      - immunopathological mechanisms of infectious origin
      - present in adults and children
- **Symptoms:**
  - nasal obstruction / congestion
  - rhinorrhea / sneezing
  - headaches
  - parosmia
  - **unilateral - aggressive symptoms with orbital signs and symptoms:**
    - ptosis
    - diplopia
    - loss of vision
  - **Diagnosis**
    - inspection and clinical examination may reveal edema of the cheek and/or eyelids, ptosis, ophthalmoplegia
    - nasal endoscopy
      - normal
      - edema of the mucosa / nasal polyposis
    - sinus CT exam:
      - sinus opacity with or without pseudocalcification
      - bone erosion on one or both sides
      - histopathological examination
      - fungal hyphae
      - eosinophils
      - Charcot-Leyden crystals
    - mycology:
      - **identification of different fungal species**
    - **serum hypereosinophilia**
    - **type I hypersensitivity**
    - **Differential diagnosis**
      - fungal balls
      - invasive fungal sinusitis
      - tumors

- **Treatment**
  - surgical removal of large lesions
  - **systemic steroids** - 2 weeks to a few months
  - **antifungals** (local or systemic)
  - **local steroids** - ***not unanimously accepted***
  - specific immunotherapy - ***not unanimously accepted***
- **Prognosis**
  - long-term follow-up
  - relapse is commonly reported

Invasive fungal rhinosinusitis

- **immunodeficiency**
- germination of fungi -> mycelia
- invasion of surrounding tissues
- the presence of fungal tissue invasion, detected on pathological examination
- clinical forms:
  - **acute** invasive fungal rhinosinusitis
  - **indolent** invasive fungal rhinosinusitis
  - **chronic** invasive fungal rhinosinusitis

Acute invasive fungal rhinosinusitis

- **Epidemiology**
  - **rare**
- **Appears at:**
  - immunodepressed patients
  - leukemia
  - lymphoma
  - chemotherapy with aplasia and bone marrow transplantation
  - neutropenia (neutrophils ≤ 500/ml)
  - long-term corticosteroid therapy
  - graft-versus-host disease
  - AIDS
  - type I diabetes mellitus
  - radiation therapy
  - long-term antibiotic therapy
  - malnutrition
- **Symptoms:**
  - immunodepressed patient
  - eyelid or cheek edema
  - fever
  - nasal congestion
  - rhinorrhea / epistaxis
  - headache
  - **Diagnosis:**
    - host factors / clinical factors / microbiological factors
    - nasal endoscopy
      - edema

        - ulceration and/or necrosis with crusts
    - **imaging is mandatory (CT, MRI):**
        - sinus opacity
        - bone necrosis
        - orbital and/or intracranial invasion
    - **imaging is mandatory (CT, MRI)**
    - nasal biopsy and/or sinus biopsy
    - immunohistochemical and/or molecular identification techniques
    - **mycological cultures**
        - **Aspergillus** fumigatus
        - **Aspergillus** flavus
        - **Mucormycosis**
            - distinct entity
            - patients with **type I diabetes mellitus**
    - multidisciplinary approach
        - ENT / hematologist / mycologist / anatomo-pathologist
        - ophthalmologist/ neurosurgeon / anesthesiologist
    - performing biopsies
    - wide excision of invaded tissue
    - **general antifungal therapy**
        - Amphotericin B
        - Itraconazole
        - Voriconazole
    - **severe neutropenia** - GM-CSF (Granulocyte-macrophage colony-stimulating factor) may be used
    - **diabetes mellitus** - kept under control
- **Differential diagnosis**
    - other rhinosinusitis
    - **infectious diseases**
        - Pseudomonas aeruginosa
        - tuberculosis
        - syphilis
        - rhinoscleroma
    - **inflammatory or granulomatous diseases**
        - sarcoidosis
        - Wegener's disease
- **Prognosis**
    - follow-up - endoscopic and imaging
    - relapses may occur in case of neutropenia
    - mortality of 20-80% in immunocompromised patients
- **Surgical treatment**
    - surgical excision of invading tissues
    - external or endoscopic approach
        - orbital involvement - wider excisions
        - intracranial invasion – combined transcranial approach

Indolent invasive fungal rhinosinusitis

- immunocompetent patients in Africa

Chronic invasive fungal rhinosinusitis

- imaging - pseudotumor appearance
- surgery
- biopsies
- cultures
- systemic antifungal treatment

SINUSITIS COMPLICATIONS

- extension to adjacent structures
- extension to orbital structures
- intracranial extension
- potential for morbidity and mortality

Orbital complications

- **more commonly observed in children**
- ***ethmoidal sinusitis*** – responsible in most cases
- bacteria responsible for **uncomplicated sinusitis**
  - Streptococcus pneumoniae
  - Haemophilus influenzae
  - Moraxella catarrhalis
- bacteria responsible for **complicated sinusitis**
  - can change depending on the type of complication
  - Haemophilus influenzae - **young children**
  - Staphylococcus and S. pneumoniae – **in adults**

Intracranial complications:

- Staphylococcus aureus
- S. Pneumoniae
- mixed flora
- ***occasionally – no pathogen is identified (20%)***
- **Diagnosis:**
  - complete history
  - recent upper respiratory tract infection
  - surgery / dental or nasal trauma
  - ENT clinical examination:
    - decongestion of the nose
    - anterior rhinoscopy
    - nasal endoscopy
  - ophthalmologic examination
  - laboratory investigation
  - **leukocyte count** – if it does not respond to treatment, it is suggestive of a complication
  - radiological assessment
  - **plain X-ray** of the sinuses
  - limited value in complications of sinusitis
  - **high-resolution CT examination** with and without contrast

- gold standard
- important role in establishing the diagnosis and therapeutic management
- non-contrast CT may miss an abscess

– **MRI exam**
  - **essential in identifying any intracranial extension**

– **Local complications:**
  - osteomyelitis
  - extension to bone – osteomyelitis
  - possibly involving the brain or orbit
  - osteomyelitis of the frontal sinuses
  - erosion of the anterior wall – subperiosteal abscess
  - Pott's puffy tumor
  - osteomyelitis of the jaw – rare
  - sphenoid osteomyelitis – rare, but can be disastrous
  - mucoceles
  - mucopyoceles

– **Orbital complications**
  - most often caused by acute ethmoiditis
  - (1) periorbital (or preseptal) cellulitis
  - (2) subperiosteal abscess
  - (3) orbital abscess
  - (4) orbital cellulitis
  - (5) cavernous sinus thrombosis

– **Periorbital cellulitis**
  - most commonly observed in children
  - **is not** a true orbital complication
  - **oedema** of the upper eyelid
  - infection is limited to the paranasal sinuses
  - there is **no chemosis**, limitation of eye movement or loss of visual acuity
  - under antibiotics - rarely progresses

– **Subperiosteal abscess**
  - accumulation of purulent material between the periosteum and the medial (bony) wall of the orbit
    - ethmoid – the most common source of infection
    - periosteum - the progression of infection is periorbital
  - the eyeball is displaced downwards and laterally
  - ***visual acuity may be affected***

– **Orbital abscess**
  - serious complication: **severe ptosis**
  - complete ophthalmoplegia
  - visual acuity is affected
  - can progress to **blindness**

– **Cavernous sinus thrombosis**
  - retrograde spread of infection to the cavernous sinus along the **orbital veins**
  - usually bilateral, although it starts unilaterally
  - clinical signs:

- sepsis / orbital pain
- chemosis / ptosis
- ophthalmoplegia
- reserved prognosis
- evolves towards **coma** and **death**

Intracranial complications

- extension to the adjacent anterior skull base
- signs of intracranial hypertension (ICH)
- headache
- vomiting
- altered mental status
- stiffness of the neck
- contrast CT or brain MRI
- **interdisciplinary approach**

Classification:

- **epidural abscess**
- **subdural abscess**
- **meningitis**
- **cerebral abscess**

Meningitis

- the most common intracranial complication
- signs and symptoms:
- headache / fever
- stiffness of the neck
- vomiting
- behavioral changes
- seizures
- CT exam
- **lumbar puncture**

Epidural abscess

- **asymptomatic**
- clinical manifestations occur only as they increase in size
- The dura mater is a natural barrier to infections

Subdural abscess

- subdural empyema
- accompanied by meningitis
- **may evolve severely**
- **neurological deficit**
- seizures

Cerebral abscess

- clinical signs depend on:
- location
- size

- number
- the most consistent symptom:
- **headache refractory to symptomatic therapy**
- **other symptoms**
  - fever
  - vomiting
  - seizures
  - focal signs
  - impaired level of consciousness
  - asymptomatic
- **Treatment**
- **conservative** parenteral treatment with
  - antibiotics
  - nasal decongestants
  - there is **no improvement** within **48-72 hours** or especially if the progression of the disease is evident
- **surgery**
  - traditionally
  - purulent collections – external drainage
  - **morbidity and mortality** in complicated sinusitis are directly related to the delay in starting treatment
  - multidisciplinary team approach:
    - otolaryngologists / neurosurgeons / specialists in infectious diseases / radiologists / anesthesiologists
- **Medical treatment**
  - empirical administration of broad-spectrum antibiotics that can cross the blood-brain barrier
  - intensification of antibiotic therapy to achieve therapeutic concentrations
  - parenteral antibiotics for at least 14 days
  - topical decongestants are essential
  - supportive treatment
- **Surgical treatment**
  - endoscopic sinus surgery - **election treatment**
  - completion of the endoscopic approach with an open approach
  - endocranial complications - craniotomy and surgical drainage
  - osteomyelitis – surgical drainage and intravenous antibiotics

RHINOPHYMA

- chronic inflammatory, hypertrophic skin disease of the nasal pyramid
  - red-violet nasal pyramid
  - **comedones (chronic hypertrophic acne)**
  - predominantly affects men
- 50–70 years of age
- **Favoring factors:**
  - excessive lipid consumption
  - temperature variations
    - metabolic, liver, circulatory deficiencies

- excessive alcohol consumption
- histopathologically: **hypertrophy of the sebaceous glands**
- **Clinical exam:**
  - unaesthetic appearance
  - change in the shape of the nasal pyramid
  - hypertrophic appearance of the skin of the nasal lobule, pseudotumoral, irregular, multilobulated, pinkish-blue, with **expression of fetid sebum**
  - deformity of the tip of the nasal pyramid

- **Evolution:** slowly
- **Treatment:**
  - surgical - excision / skin decortication
  - skin graft

BENIGN SINONASAL NEOPLASMS

Sinonasal epithelial tumors

- papilloma
- glandular sinonasal tumors
- inverted papilloma
- pleomorphic adenoma
- myoepithelioma
- oncocytoma

Neuroectodermal tumors

- paraganglioma
- schwannoma
- neurofibroma

Neural tumors

- nasal glioma
- pituitary adenoma

Soft tissue tumors

- vascular tumors (juvenile angiofibroma / hemangiopericytoma)
- fibrous tumors (nasal fibroma)
- muscle tumors (leiomyoma / rhabdomyoma)
- bone and cartilage tumors
  - osteoma / ossifying fibroma
- osteoblastoma / aneurysmal bone cyst

Surgical treatment

- total removal of the tumor
- external approaches - successfully used
- preoperative biopsy – **except** in juvenile nasopharyngeal angiofibroma (JNA), where radiological examination is performed
- encephaloceles–**MRI exam**

Fibrous dysplasia

- benign tumor
- slow growth
- more frequently affected - the **maxillary sinus (90%)**
  - in the first two decades of life
  - no sex predilection
  - development anomaly
  - **normal mineralized bone is replaced by proliferative fibrous tissue and immature bone, with a ground-glass appearance on imaging**
  - can be **monostotic (70% of cases) or polyostotic**
  - **malignancy is not common (4%)**
  - radiation therapy increases the risk of malignant transformation
  - **Treatment**
- surgical - can accelerate tumor growth
- functional deficits may become permanent or irreversible
- radical excision
  - reconstructive plastic surgery

Inverted papilloma

- **the most frequent tumor**
- histologic particularity:
  - arises from the sinonasal mucosa
- ***histological structure with inverted growth of the epithelium into the underlying stroma***
  - the most common site of origin:
    - lateral wall of the nasal fossa
    - middle meatus, corresponding to ethmoid cells (82%)
    - extends to the sinuses
      - ***maxillary sinus (69%)***
      - ***sphenoid sinus (11–20%)***
      - ***frontal sinus (11–16%)***
      - ***nasal septum (8%)***
      - ***bilateral or multiple locations (4%)***
  - 5th–7th decades of life
  - men/women – 2/1 to 9/1
  - **cause is unknown**
- **Symptoms:**
  - nasal obstruction
  - headache
  - epistaxis
  - because the symptoms are similar to those of chronic sinusitis, **diagnosis is frequently delayed**
  - **bone damage** may be due to tumor pressure and is **not predictive of malignancy**
  - secondary sinusitis occurs due to ostial obstruction
  - **MRI differentiate:**
    - retention of secretions
    - chronic sinusitis
    - mucoceles

        - cancer
    - risk of **malignant transformation** of 2-50%, with an average of 10%
- **Treatment**
    - **surgical excision** with adequate margins
    - transnasal approach - high relapse rate (40–80%)
    - lateral rhinotomy - relapse rate of 0–50%
    - midfacial degloving, to avoid scars – relapse rate of 3–13%
    - medial maxillectomy – with a low relapse rate
    - endoscopic approach - selected cases; relapse rate is 0–17%

Juvenile nasopharyngeal angiofibroma

- benign tumor
- well vascularized
- male teens
- 0.05% of head and neck neoplasms
- **unknown etiology**
    - fibroangiomatous hyperplasia, an ectopic focus of hamartomatous or paraganglionic tissue
    - **hormonally influenced** – progesterone, androgen and glucocorticoid receptors have been found
- **Symptoms:**
    - nasal obstruction
        - epistaxis
        - headache
        - obstruction of the Eustachian tube
        - nasal voice
        - mucopurulent rhinorrhea
    - eye symptoms
    - cranial nerve paralysis
    - ***CT and MRI exam:***
    - essential for diagnosis
    - ***Biopsy:*****not necessary, even** dangerous
    - ***Extension:***
        - anteriorly - nasal cavity
        - superiorly - sphenoid sinus
        - posteriorly – nasopharynx
        - laterally - pterygopalatine and infratemporal fossa,
        - orbit
        - cranial cavity

There are multiple tumor classification systems; a commonly used one is the **Radkowski classification:**

- IA limited to **nose and/or nasopharynx**
- IB extension to **one or more sinuses**
- IIA minimal extension into the **pterygomaxillary fossa**
- IIB occupation of the **pterygomaxillary fossa**, with or without **erosion of orbital bones**
- IIC extension to the **infratemporal fossa** with or without involvement of the cheek or pterygoid process

- IIIA **skull base erosion** – **minimal** intracranial extension
- IIIB erosion of the base of the skull – **intracranial extension** with or without involvement of the cavernous sinus
- **Surgical approaches:**
  - transpalatal
  - sublabial
  - facial translocation
  - approach through the lateral infratemporal fossa
  - craniofacial
  - transantral
  - transzygomatic
  - transmandibular
  - lateral rhinotomy
  - endonasal
  - combined
- **JNA treatment:**
  - **detailed knowledge of the anatomy of the base of the skull**
  - in-depth understanding of tumor behavior
  - embolization, the endoscopic approach, navigation systems and all newly developed instruments are very useful in surgery
  - **all tumor extensions should be removed**

MALIGNANT TUMORS OF THE NOSE AND PARANASAL SINUSES

Soft tissues

- carcinoma "in situ"
- squamous cell carcinoma
- adenocarcinoma
- small cell carcinoma
- undifferentiated carcinoma
- lymphoepithelial carcinoma
- malignant melanoma
- tumors of the minor salivary glands
  - mixed malignant tumors
  - adenoid cystic carcinoma
  - mucoepidermoid carcinoma
- metastatic carcinomas

Connective tissue

- malignant histiocytoma
- lethal midline granuloma
- rhabdomyosarcoma
- embryonic
- alveolar
- pleomorphic
- fibrosarcoma / angiosarcoma
- lymphosarcoma / myosarcoma

- chondrosarcoma

Bone

- osteogenic sarcoma
- Ewing's Sarcoma
- giant cell malignancies

Lymphoepithelioma

- **rare** in the sinonasal region
- more common in nasopharynx
- poorly differentiated epithelial malignant tumors with predominant lymphoid stroma
- developing in the posterior portion of the nasal fossa, they cause **early regional metastases**
- **treatment:** radiotherapy

Malignant melanoma

- pigmented tumor
- **rarely** found in the nose and paranasal sinuses
- more frequently located in the nasal vestibule or on the septum
- extremely aggressive
- early metastasis
- biopsy is contraindicated
- complete excision of the tumor with wide peritumoral safety margins
- post-excisional histopathological examination

Malignant tumors of mesodermal origin

- rare
- approximately 5% of all malignant tumors of the nose and paranasal sinuses
- difficult to treat and to keep under control, regardless of the treatment

Lymphosarcoma

- frequently in long bones
- located in the face region
- the **maxillary sinus being most commonly involved**
- nasal bones
- great infiltrative potential
- radiotherapy
- there is no curative treatment

Extramedullary plasmocytoma

- rarely
- 5-year survival after surgical treatment and irradiation is 50–60%

Chondrosarcoma

- occurs especially in **children**
- usually in the ethmoid, but also in the septum
- very aggressive
- destroy the surrounding structures
- deforms the nasal pyramid
- rarely metastasis

- election treatment:
- surgical
- radiation therapy slows tumor development

Ewing Sarcoma

- is predominantly found in **children and adolescents**
- frequently involves **long bones**
- primary tumor in the maxillary sinus and mandible

Osteosarcoma

- bone tumor
- develops in the **ethmoid**, from which it extends into the **nasal fossa**
- any age
- men / women – 3/1
- survival is around 3 years

Staging of maxillary sinus cancer

Primary tumor (T)

- T0 – Primary tumor **not** clinically evident
- T1 – Tumor confined to the **mucosa of the infrastructure** of the sinus, without bone erosion or destruction
- T2 – Tumor confined to the **mucosa of the superstructure**, without bone destruction, or involving only the inferior or medial bony wall
- T3 – Tumor extended to the **skin of the cheek, orbit, anterior ethmoid or pterygoid muscles**
- T4 – Massive tumor with invasion of the cribriform plate, posterior ethmoid, nasopharynx and skull base

Regional lymph nodes (N)

- N0 – No clinically positive (palpable) lymph nodes
- N1 – a single clinically positive ipsilateral lymph node with a diameter of **3 cm or less**
- N2 – a single palpable ipsilateral lymph node with a diameter between **3 and 6 cm**
  - N3 – massive ipsilateral lymph nodes, bilateral or contralateral lymph nodes

Metastases (M)

- M0 – No distant metastasis
- M1 – Distant metastasis present

Staging of maxillary sinus cancer

Staging according to TNM

- Stage I: T1 N0 M0
- Stage II: T2 N0 M0
- Stage III: T1, T2 or T3 with N1 M0
- Stage IV: any T with N2 or N3 and M0, any T with any N and M1

Symptoms of sinonasal cancer

- malignant tumors of the nasal fossa and sinuses have a **particularly extensive and infiltrative potential** due to the complexity of the anatomical relationships and the proximity to the orbit, oral cavity, nasopharynx and skull base
- invasion from the nasal fossa to the sinuses or vice versa occurs so early that it is difficult to establish the actual site of origin
- **early diagnosis** of sinus malignant tumors
  - **difficult**
  - clinical picture evolving for a long time as an **asymptomatic sinusitis**
  - vague manifestations of rhinosinusitis
  - recognized only at an **advanced stage**
  - establishing the sinus or nasal origin of the advanced tumor

Signs and symptoms:

- invasion of neighboring structures:
  - **rhinological** symptoms
  - **sinus** symptoms
  - **orbito-ocular** symptoms
  - **neurological** symptoms
  - **buco-dental** symptoms
- **rhinological symptoms are unilateral:**
  - small, repeated, spontaneous **epistaxis** or prolonged **epistaxis**
  - **unilateral nasal obstruction** that progressively increases over a long period of time
  - **mucous rhinorrhea** that becomes mucopurulent, then purulent, dirty, bloody, sometimes fetid, with sloughs, due to secondary infections accompanying the tumor
  - **subjective cacosmia, hyposmia, anosmia**
  - expulsion of small tumor fragments during sneezing or nose blowing

Unilateral orbito-ocular symptoms:

- **orbital invasion:**
  - swelling of the internal angle of the orbit
  - epiphora
  - dacryocystitis, conjunctivitis
  - ocular protrusion, diplopia (paralysis of the oculomotor muscles)
  - infraorbital hypoesthesia
- **neurological symptoms:**
  - pain absent in the early stages
  - continuous or paroxysmal facial pain in the zygomatic region (maxillary nerve, ophthalmic nerve V1)
  - radiation to the upper teeth
  - unilateral frontal headache, radiating headache, orbital pain
  - secondary infection of paranasal sinuses
  - extension to the anterior cranial fossa
  - intracranial hypertension (ICH)
  - sensory disorders, paresthesia, hypoesthesia
- ***buco-dental symptoms:***
- **infrastructure cancer**

- abnormal mobility of teeth
- expulsion of teeth
- edentation
- bleeding tumor buds in place of the teeth
- gingiva-labial deformation
- double swelling, sometimes ulcer, bleeding alveolar process
- progressive bulging of the hard palate
- ***trismus – invasion of the pterygomaxillary fossa***
- deformation of the nasal pyramid
- zygomatic ipsilateral erosion of the anterior wall of the maxillary sinus with infiltration of the skin

Diagnosis of sinonasal cancers

- **difficult during the intrasinus latency period – lack of symptoms**
- objective ENT examination
- radiological examination
  - classic
  - computed tomography (CT)
  - magnetic resonance imaging (MRI)
- ultrasonography
- sinonasal endoscopy with **biopsy**
- sinus puncture with cytological examination of secretions

objective ENT clinical examination

- inspection
- oropharyngoscopy
- anterior rhinoscopy
- posterior rhinoscopy
- examination of the lymph nodes in the face and neck regions
- rigid or flexible nasal endoscopy
- decisive factor in the **correct early diagnosis** of malignant tumors
- indicated for areas where direct or indirect visual access is difficult
- **biopsy** under endoscopic control
- sinus endoscopy via the canine fossa or the inferior meatus
- sinus puncture
- location of the tumor
- tumor density
- anatomo-pathological structure
- tumor extension
- integrity and deformation of the sinus walls
- disappearance of bone contours
- presence or absence of **lymph node metastases**

Radiological examination

- conventional X-ray:
  - anterior sinuses
  - posterior sinuses
  - in different views

- haziness and intense opacity of the sinus
- loss of the bony outline
- bone rarefaction and destruction
    - **computed tomography (C.T.)**
    - **magnetic resonance imaging (MRI)**
    - **ultrasonography**

Treatment: surgery, radiotherapy, chemotherapy

CHAPTER II — PHARYNX AND ORAL CAVITY PATHOLOGY

PHARYNX ANATOMY

The nasopharynx is bounded by the following structures:

- **superior** – the base of the skull
- **inferior** – a horizontal line passing through the soft palate (its upper portion)
- The nasopharynx has a cubic shape and has 6 walls:
  - **anterior**:
    - choanae (posterior openings of the nasal fossae)
    - interchoanal septum
  - **superior:**
    - is vault-shaped and is related to the ***basilar process of the occipital bone and the atlanto-occipital ligament***
    - on this wall lies **Luschka's pharyngeal tonsil**
  - **posterior:**
    - extends to the upper edge of the anterior arch of the atlas
    - on this wall there are lymphoid formations
    - **lateral walls;** the most important element at this level: **1 cm from the tail of the turbinate lies the opening of the Eustachian tube**, which is triangular in shape, with an anterior edge and a more prominent posterior one
- **lymphoid tissue** around the tube – **tubal tonsil (Gerlach)**
- **Rosenmüller's fossa**, located posterior to the tube; lymphoid formations are found at this level

The oropharynx is bounded by the following structures:

- **superior** – a horizontal line passing through the upper edge of the soft palate
  - **inferior** – upper edge of the epiglottis
  - The oropharynx has **six walls:**
- 1 anterior wall, 1 posterior wall, 2 lateral walls, 1 superior wall and 1 inferior wall
- communicates with the nasopharynx
- during deglutition, the soft palate contracts
- inferiorly it communicates with the larynx and hypopharynx
- the **anterior wall** communicates with the oral cavity through the oropharyngeal isthmus, bounded by: the lower edge of the soft palate with the uvula, the anterior tonsillar pillars and the base of the tongue
- posterior wall:
- prevertebral aponeurosis
- axis
- is visible when the patient opens the mouth and says "a"

Hypopharynx (lower pharynx)

- delimited inferiorly by the **lower edge of the cricoid**
  - continues superiorly with the oropharynx
- continues inferiorly with the esophagus
- has six walls: anterior, posterior, 2 lateral walls, superior and inferior
- **anterior** represented by
  - base of the tongue with the lingual tonsil

  - lingual surface of the epiglottis
  - the two valleculae
  - the posterior surface of the arytenoids and cricoid cartilage
- **posterior:**
  - prevertebral aponeurosis
  - body of vertebrae C3 - C6
- **the lateral walls** correspond to the pyriform sinuses or pharyngo-epiglottic sulcus

The pharynx consists of four layers:

- mucosal layer
- pharyngeal aponeurosis
- muscle layer
- peripharyngeal adventitia

The **mucosal layer** has structures differentiated according to the 3 pharyngeal levels:

- the **nasopharynx** has a ***respiratory structure – ciliated columnar epithelium***
- a transitional epithelium between the nasopharynx and the ***oropharynx: stratified squamous epithelium***
- **Waldeyer's lymphatic ring** consists of ***subepithelial lymphoid structures*** located in the pharyngeal submucosa:
- palatine tonsil, nasopharyngeal tonsil (Luschka), tubal tonsil (Gerlach), lingual tonsil (tongue base)

Palatine tonsil

- situated on the lateral wall of the oropharynx between the **two tonsillar pillars** (anterior and posterior) and separated from the tongue by the **tonsilloglossal sulcus**
- has **two surfaces** (medial, lateral), **two edges**, an **upper pole**, where the largest crypt is located, and a **lower pole**
- the **medial surface** contains **12-15 crypts**
- is covered by a ***stratified squamous epithelium***
- the mucous glands open into the crypts
- the crypts separate lymphoid follicles made up of lymphatic tissue with a germinal center where lymphocytes are formed
- **the lateral (deep) surface** is covered by a fibrous capsule
- **is related to the pharyngeal constrictor muscles**

Nasopharyngeal tonsil (Luschka)

- consists of lymphoid tissue separated by grooves converging towards a common point located on the posterior wall of the nasopharynx
- is located on the ***upper and posterior wall of the nasopharynx***
- repeated infections cause its hypertrophy

Tubal tonsil

- made up of lymphoid tissue located around the **Eustachian tube**; this tissue also extends into **Rosenmüller's fossa**

Lingual tonsil

- extends from the lingual V to the epiglottis and forms part of the base of the tongue, its crypts being shallower than those of the palatine tonsil

Pharyngeal aponeurosis

- forms the **fibrous skeleton of the pharynx**
- **is more fibrous in the upper portion**
- continues with the corresponding layers of the esophagus

Muscle layer

- consists of the ***constrictor muscles*** and the ***levator muscles of the pharynx***
- the constrictor muscles (superior, middle and inferior) form the outer muscular layer
- **Superior Constrictor**
  - inserted at the **base of the skull** (pterygomandibular ligament, mandible - the posterior extremity of the internal oblique line of the mandible)
- **Middle constrictor**
  - inserted on the **greater and lesser horns of the hyoid bone**
- **Lower constrictor**
  - inserted on the **cricoid arch** and the posterior edge of the lateral lamina of the cricoid cartilage
- the inferior constrictor is divided into an **upper thyropharyngeal part** and a **lower cricopharyngeal part**, thus participating in the formation of **Laimer's triangle** on the posterior wall – the so-called weak area of the hypopharynx, where the **pulsion diverticulum (Zenker)** develops

Pharyngeal constrictors

- **The levator muscles of the pharynx are:**
  - **stylopharyngeus**
  - **salpingopharyngeus**
  - **palatopharyngeus**
  - **stylohyoid**
  - **styloglossus**

Peripharyngeal Adventitia

- cover the constrictor muscles
- is related to the prevertebral fascia

Pharynx vascularization

- **The arteries** are branches of the **external carotid artery**
  - ascending pharyngeal artery
  - ascending palatine artery
  - tonsillar branches of the facial artery
  - branches of the internal maxillary artery
  - branches of the lingual artery
- **The veins** form plexuses that drain into:
  - facial vein
  - pterygoid plexus
  - internal jugular vein
- **Lymphatics** drain into:
  - retropharyngeal nodes, then the deep superior cervical (jugulo-carotid) nodes (***Küttner's subdigastric node*** – most commonly affected in pharyngeal neoplasms)

Pharynx innervation:

- **motor**: IX, X, XII, VII
- **sensitive**:
  - nasopharynx: maxillary nerve (V)
  - oropharynx: glossopharyngeal (IX)
  - hypopharynx: pneumogastric or vagus (X)

PHYSIOLOGY AND PATHOPHYSIOLOGY OF THE ORAL CAVITY AND PHARYNX

Oral cavity and pharynx functions:

- respiration
- mastication
- deglutition
- taste
- phonation

Respiratory function with an important role in:

- **air heating**
- **air moistening**
- **air filtration** by incorporating various particles into mucus - filters the air of dust, bacterial agents, etc.
- conduit between the nasal fossae and the larynx, or between the oral cavity and the larynx
- **alteration of this function may occur in**
- **acute infectious processes**, when **dyspnea** may occur in
  - retropharyngeal abscess
  - peritonsillar quinsy
  - Ludwig's angina (abscess of the floor of the mouth)
  - acute adenoiditis / chronic infectious processes
- **chronic hypertrophic tonsillitis**
- **benign tumors (nasopharyngeal fibroma)**
- **malignant tumors of pharynx**

Masticatory function

- role in the **formation of the food bolus**; taking part in this masticatory function are: ***the teeth, masticatory muscles, temporomandibular joint and salivary secretions***, stimulated by taste and olfactory receptors
- may be altered by:
- pathologies of temporomandibular joint
- peritonsillar quinsy
- malignant tumors of the tonsil, tongue, etc.
- secondary gastrointestinal disorders
- decreased salivary secretion
- collagenosis
- cobalt irradiation, radiotherapy for neoplasms
- thus swallowing disorders occur
- preparation of the food bolus requires varying amounts of water

Deglutition function:

- is **the most important function** of the pharynx; through it the food bolus passes from the oral cavity into the stomach
- has **three phases:** oral, pharyngeal and esophageal
- it is **a voluntary act** up to the oropharyngeal isthmus, after which it becomes a reflex act
- Neural pathways of deglutition:
- **nerve center**: reticular formation of the medulla oblongata
- **afferent pathways**: V, IX, X
- **efferent pathways**: XII, IV, VII, IX, X
- Alteration of this function:
  - **odynophagia**, painful deglutition, is characteristic of acute infectious processes, TB, malignant tumors
  - **reflex otalgia (pain radiating to the ear)**: tonsillectomy, pyriform sinus cancer, ulcerated pharyngeal tumors, superior laryngeal neuralgia
  - strokes (brain tumors may cause swallowing difficulties) – **dysphagia**

Phonatory function

- resonance cavity, through the raising and lowering movements of the larynx
- changes the sound quality
- the tongue and soft palate play a role in the formation of consonants and vowels
  - **very important role in phonation in patients with total laryngectomy**
  - Disorders of this function cause:
- **closed rhinolalia (stomatolalia)**: adenoiditis, nasopharyngeal angiofibroma, nasopharyngeal neoplasm, tonsil hypertrophy, peritonsillar quinsy
- **open rhinolalia:** short soft palate, disorders of soft palate innervation
- **dysarthria:** palatoschisis, cleft lip and palate

Gustatory function

- **taste buds** in the mucosa of the tongue are also a site of tactile, thermal and pain sensation
- taste detects the sensations of ***sour, bitter, salty and sweet***
- the integrity of the taste receptors and the liquid state of food are indispensable to **taste sensitivity**
- loss of this function causes **ageusia**
- The **local defense function** is performed by:
  - **taste function** (control of substances that are introduced into the oral cavity)
  - the **mucus** covering the pharyngeal mucosa has an antiseptic role and neutralizes various substances
  - **specific immunological function** of Waldeyer's ring

The lymphoid tissue has a defense function by:

- control and protection from pathogens
- production of lymphocytes
- production of specific antibodies (all types of immunoglobulins are found in the lymphatic tissue of the tonsils)
- rejection of lymphocytes necessary for immune stimulation
- production and release of immune stimulating lymphocytes

Ageusia - loss of taste:

- temporary/definitive
- unilateral/bilateral
- **Etiology**
- **Central:** - tumors, hemorrhages, abscesses, multiple sclerosis
- **Peripheral:** IX; nerve filaments: chorda tympani, lingual n., nervus intermedius (Wrisberg) of VII
- influenzal **neuritis**, toxic **neuritis** (alcohol, arsenic)
- **compressions:** tumors (syphilitic gumma, acoustic neuroma, meningioma)
- trauma
- **alteration of the taste buds** (tongue), alcoholism, smoking, syphilis, malignant tumors and burns of the tongue
- Functional ageusia
  - nervous
  - surgery
  - the anterior 2/3 of the tongue is responsible for **sweet and salty taste**
  - tongue base (9-10 circumvallate papillae) – **bitter taste**

Taste examination:

- bitter (n. IX glossopharyngeal n. - V lingual)
- salty, sweet, acidic (anterior 1/3 of the tongue): lingual n., chorda tympani, nervus intermedius (Wrisberg)

PHARYNX INVESTIGATION METHODS

Anamnesis

Clinical examination of the pharynx

- ***inspection and palpation***
- ***oropharyngoscopy***
  - oral cavity examination
  - examination of the floor of the mouth and tongue
  - examination of the oropharyngeal cavity
- ***nasopharyngeal examination (cavum)***
  - posterior rhinoscopy
  - nasopharyngeal endoscopy

Radiological investigations

- Hirtz incidence
- tomography (T.C.)
- MRI
- carotid angiography

ORAL CAVITY AND PHARYNX MALFORMATIONS

Etiology:

- **complex,** most commonly caused by intrauterine **viral infection**, **embryopathies, genetic lesions, chronic intoxication** (alcoholism, lead poisoning) of the parents, **syphilis** etc.

Mouth malformations:

- congenital malformations of the **teeth** (total edentation, partial edentation, supernumerary teeth, dental malocclusion)
- congenital malformations of the **tongue** (bifid tongue, macroglossia, microglossia, aglossia, ankyloglossia – short lingual frenulum, lingual thyroid)

Pharynx malformations

- cavum atresia
- velopalatine insufficiency
- bifid uvula
- palatoschisis
- cleft lip and palate ("wolf's mouth")

Velopalatine insufficiency

- is characterized by a short soft palate, phonation disorders and reflux of fluids through the nose
- this malformation is an **absolute contraindication** to **tonsillectomy or adenoidectomy**

Cheilopalatoschisis (labio-maxillo-palatine cleft)

- Clinical forms:
  - labia cleft
  - labio-maxillary cleft
  - labio-maxillo-palatine cleft
- Symptoms:
  - feeding difficulties (reflux of food and fluids through the nose)
  - open rhinolalia
- Treatment:
  - surgical procedures aimed at closing the clefts and correcting feeding and phonation disorders

Congenital fistulas and cysts of the pharynx

- **congenital cysts** (mucous retention cysts, ranula – retention cyst of the mucous glands of the floor of the mouth, cysts and fistulas of the thyroglossal duct, branchial cysts and fistulas located on the anterior edge of the sternocleidomastoid muscle)
- **Clinical forms of fistulas**:
- **median:** the fistula of the thyroglossal duct opens at the foramen caecum
- **lateral:** derive from the remnants of the branchial arches
- **The fistulas** might be:
  - complete
  - incomplete
  - Symptoms are related to the presence of a **viscous secretion at the external orifice**

Thyroglossal duct cyst and fistulas

- occurs in the first years of life, rarely in adolescence and very rarely in adults
- **is median**
- its location and presence are explained by the **embryology of the thyroid gland:** normally the duct that starts at the base of the tongue and crosses the hyoid bone is resorbed or obliterated and becomes a tract without a lumen – failure of resorption or obliteration leads to thyroglossal duct fistulas or cysts

- **Diagnosis:**
  - small, round, mobile, painless mass located in the midline of the neck, which moves upward during swallowing
  - a **fistula** occurs if secondary infection develops, when saliva is observed during feeding
  - red, infiltrated skin
  - fistulography allows visualization of the fistula tract
- **Treatment:**
  - **surgically**, with excision of the corresponding portion of the hyoid bone to avoid relapse

Lateral fistulas

- **the external orifice** is located on the anterior edge of the sternocleidomastoid muscle
- **the internal opening** can be located
- base of the tongue
- tonsil lodge
- Rosenmüller's fossa
- treatment is **surgical**

Cervical cysts

- when superinfected, it can **fistulize**, leading to diagnostic errors
- treatment is **surgical**

PHARYNX FOREIGN BODIES

- can be located in all three segments of the pharynx
- occur more frequently in children, the mentally ill, elderly people with denture problems, etc.
- **small, sharp foreign bodies** can easily lodge in the tonsils, the base of the tongue, the valleculae or the lateral wall
  - the most commonly encountered of these are:
    - bones – especially fish bones
    - toothbrush bristles
    - shards of glass
    - pieces of wire
- at the esophageal opening and at the level of the pyriform sinuses, **larger foreign bodies** (fragments of toys, coins, dentures, etc.) may become impacted
- **Diagnosis:**
  - characteristic anamnesis and symptoms
  - **odynophagia, dysphagia, sialorrhea**
  - ENT examination (oropharyngoscopy, posterior rhinoscopy, laryngoscopy)
  - plain or contrast X-rays
- **Treatment:**
  - extraction of the foreign body directly or through a rigid tube (esophagoscope), using appropriate forceps

PHARYNX TRAUMA

- occur in both sexes and at all ages
- most commonly they are **accidental** and occur in children who fall during play while holding various objects in their mouths: pencils, sticks, etc.

- these wounds can be **superficial** and heal quickly, or **deep** and require careful exploration followed by suturing and antibiotic therapy
- broad-spectrum antibiotic therapy, tetanus prophylaxis and rigorous local hygiene are mandatory
- **wasp stings, burns from inhalation of hot vapors or ingestion of corrosive substances (bases, acids)** can produce **massive edema** accompanied by ulcers and necrosis of the pharynx that endanger the patient's life
- **required:** monitoring, corticotherapy, and sometimes tracheotomy

ACUTE INFLAMMATIONS OF THE PHARYNX

Clinical forms:

- acute non-specific angina
- specific acute angina
- anginas in some hematological conditions

Nonspecific acute anginas

- occur in both sexes and at all ages
- occur especially in spring and autumn
- **Favoring factors:**
  - low immunity
  - poor nutrition
  - alcohol, tobacco
  - metabolic diseases, diabetes, hepatopathies
  - persistent chronic infections
- **Clinical forms:**
  - erythematous catarrhal angina (red)
  - erythemato-pultaceous angina (white)
  - pseudomembranous angina
  - ulcerative angina (Moure)
  - uvulitis

Erythematous catarrhal angina (red)

- **Etiology:**
  - **viral** (adenoviruses, rhinoviruses, myxoviruses)
- **Diagnosis:**
  - **sudden onset** with fever, chills, headache, myalgia, arthralgia, fatigue, dry throat sensation followed by odynophagia and dysphagia
  - the tonsils are enlarged and congested
  - painful **subangulomandibular adenitis**
  - lasts 3-4 days
- **Complications are common:**
  - bacterial superinfection, acute otitis
- **Treatment:**
  - bed rest for 7 days, anti-inflammatory drugs, vitamin therapy, alkaline gargles and **antibiotic therapy** in case of superinfection

Erythemato-pultaceous angina (follicular or white)

- **Etiology:**

  - **Beta-hemolytic streptococcus** in the first days of the disease, after which **staphylococcus, pneumococcus,** etc. may be superadded
- **Diagnosis:**
  - **sudden onset** with fever of 39-40°C, chills, headache, agitation, convulsions, odynophagia, intense dysphagia
  - enlarged, congested tonsils, covered by a **white exudate,** purulent, creamy (pseudomembranous)
  - painful **subangulomandibular adenitis**
  - lasts 7-8 days
- **Complications:**
  - local suppuration
  - peritonsillar quinsy
  - laryngeal edema
  - general complications
    - rheumatic carditis
    - nephritis, etc.
- **Treatment:**
  - **antibiotics** active against beta-hemolytic streptococcus
  - minimum 7 days
  - when laboratory tests are abnormal (ESR, fibrinogen, ASLO), treatment with **long-acting penicillin** (moldamin, extencilin, tardocilin, retarpen) is mandatory for **2-3 months**
  - anti-inflammatory
  - alkaline gargles
  - oropharyngeal antiseptics
  - **tonsillectomy** when the patient has 6-7 episodes per year or has suppurative pharyngeal complications

Pseudomembranous angina

- **Etiology:**
  - **beta-hemolytic streptococcus, staphylococcus, pneumococcus, Haemophilus influenzae**
- **Diagnosis:**
  - onset may be insidious or sudden, with fever, chills, headache, odynophagia, dysphagia
  - enlarged, congested tonsils; yellowish-white pseudomembranes cover the palatine tonsils without extending beyond them, detach easily and leave **no ulcers**
  - bacteriological examination is mandatory for the differential diagnosis with **diphtheritic angina**
- **Treatment:**
  - **broad-spectrum antibiotics** (amoxicillin, amoxiclav, in more serious forms cephalosporins, etc.)
  - anti-inflammatory
  - vitamin therapy
  - alkaline gargles

Ulcerative angina (Moure)

- **Etiology:**
  - **banal germs**

- **Diagnosis:**
  - insidious onset with few symptoms in the first days, followed by odynophagia and severe dysphagia
  - enlarged, congested tonsils with **necrotic ulceration at the upper pole of the palatine tonsil,** covered by a **fibrous exudate**
  - painful subangulomandibular adenitis
  - mandatory bacteriological examination for the differential diagnosis with specific angina or exacerbated chronic angina
- **Treatment:**
  - applications of **5% silver nitrate solution**, 5% zinc chloride, alkaline gargles
  - broad-spectrum antibiotics
  - vitamin therapy

Specific acute anginas

Measles angina

- **red angina (viral)** preceded by acute respiratory infection (oculonasal catarrh)
- **Koplik's sign**, characterized by the presence of yellowish-white deposits located around the oral opening of Stensen's duct

Rubella angina

- red angina
- the patient has painful adenitis: cervical, retroauricular, occipital

Scarlet angina

- **erythematous angina with pus** that precedes scarlet fever

Diphtheritic angina (Löffler's bacillus)

- **pseudomembranous angina** in which the pseudomembranes are grayish, thick and adherent
- they leave **bleeding surfaces** when detached, reform quickly and extend into the pharynx, nasal fossae and larynx
- severe forms show grey-brown pseudomembranes accompanied by **multiple hemorrhages and ulcerations**
- the general condition of the patient is serious (**hypertoxic**)
- **Treatment:**
  - administration of **anti-diphtheria serum** associated with **diphtheria toxoid** and **broad-spectrum antibiotics**

Aphthous angina

- **viral angina**
- the virus is transmitted through milk or milk products
- grey-white vesicles are present on the mucosa of the oral vestibule, soft palate, uvula and tonsils
- the vesicles are followed by superficial ulcerations covered by a **fibrous exudate**
- **Treatment:**
  - alkaline gargles, local applications of 5% silver nitrate solution
  - in bacterial superinfection, **antibiotic therapy** is indicated

herpes zoster angina

- **viral** angina

- vesicles with clear fluid are polymorphic and appear on an erythematous background along the course of the maxillary nerve
- febrile state, hyperesthesia along the course of the nerve
- heals in 2 weeks, but neuralgia persists
- Treatment:
- zovirax, polydin, B group vitamins, oropharyngeal disinfectants

Fusospirillar angina (Plaut-Vincent)

- etiological agents are **fusiform bacilli and spirilla**
- begins as an erythematous angina, after which a ***thick yellowish-white pseudomembrane*** is observed on one tonsil; when removed, it leaves a ***deep ulceration with irregular, bleeding edges***
- the subangulomandibular or retroangulomandibular adenitis present is painful
- occurs in patients with dental problems
- **Treatment:**
- local applications of silver nitrate solution or 5-10% zinc chloride, alkaline gargles, vitamin therapy; dental treatment

Acute angina in hematological conditions

- are **ulcero-necrotic anginas**, with altered general condition
- complete laboratory examinations (hemogram, sternal puncture, etc.) and collaboration with a hematologist and an infectious diseases specialist are required

Leukemic angina

- **may be the first symptom** of acute leukemia or of an acute exacerbation of chronic leukemia
- presents as ulcerative tonsillitis with stomatitis and cervical adenitis (the ulceration is hemorrhagic)
- **does not respond to usual treatment** and the patient's general condition worsens under conventional treatment
- hemogram reveals **hyperleukocytosis up to 300,000/mm3** with the presence of ***myeloblasts or lymphoblasts***
- **Treatment:**
  - is that specific to leukemia, associated with broad-spectrum antibiotics, corticotherapy, oropharyngeal disinfectants, interferon

Agranulocytic angina

- **the etiology of the disease is not known**, but it occurs especially in women following the **administration of certain drugs:** arsenobenzoate, gold salts, bismuth, sulfonamides, salicylic acid, synthetic hormones, or radiation, which produce a ***blockade of granulopoiesis***
  - **is a myelotoxicosis**
- the pharynx and tonsils have ulcers and crusts with a gangrenous odor (fetor)
  - cervical adenitis is present
- hemogram shows ***leukopenia with agranulocytosis***
  - left untreated, it leads to the death of the patient
- **Treatment:**
- immunosuppressants, corticotherapy, broad-spectrum antibiotics, blood transfusions, oropharyngeal disinfectants, immunotherapeutic treatment

Monocytic angina

- occurs more frequently in **young people** with an **immune deficiency**
- lasts 3-4 weeks and is followed by a marked asthenia
- initially presents as viral, erythemato-pultaceous and ulcerative angina; several ulcero-necrotic lesions are present on the same tonsil
- patient has polyadenopathy, hepatosplenomegaly, marked asthenia
- hemogram: **leukocytosis 20,000/mm3, monocytes 40-60%**
- the course of the disease is benign
- **Diagnosis of certainty: Paul-Bunnell reaction**
- **Treatment:**
  - broad-spectrum antibiotics, corticotherapy, vitamin therapy, immunotherapy, oropharynx hygiene

ACUTE ADENOIDITIS

- **Definition:**
- acute inflammation of **Luschka's tonsil**
  - occurs in newborns and children up to the age of 6-7 years
- **Favoring factors:**
  - repeated viral infections
  - acute rhinitis, acute rhinosinusitis
  - humidity and temperature variations
  - poor nutrition
  - low immunity
- **Clinical forms:**
  - acute
  - prolonged (subacute)
- **Diagnosis:**
  - sudden onset / fever of 39-40°C / altered general condition
  - agitation, convulsions in the newborn and young child
  - nasal obstruction, mucopurulent nasal secretions
  - eating disorders
  - oropharyngoscopy reveals purulent secretions running down the posterior wall of the oropharynx
- **Differential diagnosis:**
  - acute rhinitis, acute sinusitis, acute angina
- **Complications:**
  - acute otitis, acute sinusitis, tracheobronchitis, gastroenterocolitis
- **Treatment:**
  - disinfectants and nasal decongestants / nasal aspiration
  - broad-spectrum antibiotics or cephalosporins / anti-inflammatory drugs
  - aerosols

SUPPURATIVE COMPLICATIONS OF PHARYNGITIS

Peritonsillar quinsy

- **Definition:**

  - **purulent collection** located between the tonsillar capsule and the lateral wall of the pharynx, favored by the presence of loose connective tissue in this space
  - infection spreads by contiguity or by the lymphatic and venous routes
  - occurs more frequently in adults and is unilateral
- **Favoring factors:**
  - acute non-specific angina
- **Clinical forms:**
  - anterosuperior
  - posterior
  - intra-tonsillar
- **Diagnosis:**
  - insidious or sudden onset
  - febrile or subfebrile state
- **Diagnosis:**
  - odynophagia
  - dysphagia
  - sialorrhea
  - reflex otalgia and **trismus** (anterosuperior quinsy)
  - stomatolalia
  - oropharyngoscopy: **infiltration and bulging** of the anterior or posterior tonsillar pole, depending on the clinical form; the palatine tonsil is pushed towards the midline by the purulent collection; hyperemia and hypertrophy of the tonsil, edema and congestion of the corresponding half of the soft palate
  - painful cervical adenitis
- **Differential diagnosis:**
  - difficult eruption of the lower third molar
  - suppuration of the pterygomaxillary fossa
  - superinfected tonsillar sarcoma
- **Treatment:**
  - **surgical:** puncture followed by incision and drainage (opening of the collection with Lubet-Barbon forceps under local anesthesia)
  - general antibiotic therapy
  - gargles with 12-volume hydrogen peroxide (one tablespoon per glass of water)
  - anti-inflammatory drugs
  - tonsillectomy 14 days after healing

Parapharyngeal quinsy

- **Definition:**
  - parapharyngeal purulent collections of tonsillar origin (acute angina, after tonsillectomy), pharyngeal trauma, dental infections (lower molars)
- **Clinical forms:**
  - adenophlegmon
  - cellulo or viscerophlegmon

Adenophlegmon

- cervical lymph node infection
- **Diagnosis:**
  - fever of 39-40ºC / painful local swelling

- trismus, odynophagia, dysphagia, torticollis
- local fluctuation (in the collection phase)
- oropharyngoscopy: a swelling located behind the posterior tonsillar pillar, forming a single mass with the cervical swelling

- **Treatment:**
  - external surgical approach: puncture/incision and drainage
  - systemic antibiotic and anti-inflammatory therapy

Laterocervical cellulo or viscerophlegmon

- characterized by **diffuse suppuration of the cervical lymph nodes,** accompanied by a septic state with marked edema of the cervical region that may cause **acute respiratory failure**
- **Treatment:**
- is preferably carried out by a team, in intensive care units
  - surgical: drainage through multiple cervical incisions
  - wide-spectrum antibiotic
  - anti-inflammatory

Retropharyngeal quinsy

- occurs most frequently in **infants and children up to 2 years of age**
- **Definition:**
  - purulent collection located between the posterior wall of the nasopharynx and the prevertebral fascia, due to infection of the Gillette and Morestin lymph nodes
- **Etiology:**
  - acute rhinopharyngitis, acute adenoiditis
  - Pathogen: **streptococcus, staphylococcus**
- **Diagnosis:**
  - fever 39-40ºC, agitation, convulsions
    - nasal obstruction
    - breathing disorders: dyspnea, cyanosis
    - feeding disorders with nasal reflux of fluids,
    - coughing
    - stiffness of the neck with torticollis
- oropharyngoscopy: swelling of the posterior wall of the pharynx, congestion of the mucosa, fluctuation on palpation (inconstant), laryngeal edema in more serious cases
- laterocervical adenopathy
- **Differential diagnosis:**
  - **retropharyngeal cold abscess (Pott's disease)** – X-ray of the cervical spine shows bone erosion of the vertebral structures
- **Treatment:**
  - **surgical:** puncture, incision and drainage of the collection (with the head in hyperextension), followed by aspiration of purulent secretions to prevent flooding of the tracheobronchial tree
  - **tracheotomy** if needed
  - broad-spectrum antibiotic therapy
  - anti-inflammatory

Phlegmon of the floor of the mouth

- **Definition:**

  - suppuration of the floor of the mouth
- **Etiology:**
  - **fusospirillar flora / anaerobic flora**
    - occurs in patients with **infected teeth**
    - low immunity
- **Diagnosis:**
  - insidious onset / fever / altered general condition
  - swelling and woody infiltration of the floor of the mouth
  - satellite adenitis
- **Treatment:**
- **surgical** – multiple vertical external incisions (parallel to the rami of the mandible)
- **broad-spectrum antibiotic therapy**, cephalosporins, metronidazole (for anaerobic flora)
- **prophylactic**: adequate dental treatment

CHRONIC INFLAMMATIONS OF THE PHARYNX

Chronic pharyngitis

Nonspecific chronic inflammation

Clinical forms:

- chronic pharyngitis
- pharyngeal ozena
- pharyngokeratosis
- adenoid hypertrophy
- chronic tonsillitis

Specific chronic inflammation

Clinical forms:

- syphilis of the pharynx
- pharyngeal tuberculosis
- pharyngeal lupus
- retropharyngeal cold abscess

Chronic Pharyngitis

- **Favoring factors:**
- tonsillectomized patients (with a constitutionally wide pharynx)
- chronic nasal obstruction: septal deviation, adenoiditis, nasal polyposis
- rhinopharyngitis
  - metabolic diseases / occupational noxae
  - tobacco, alcohol, dry atmosphere, dust, cold, moisture
  - prolonged vocal efforts
- **Clinical forms:**
  - **chronic catarrhal pharyngitis**
  - **chronic hyperplastic pharyngitis** (hypertrophic lymphoid formations present throughout the posterior wall of the pharynx – **diffuse form**, or located behind the posterior pillar, forming true cords – **localized form**)
  - **chronic atrophic pharyngitis** (pale, dry mucosa with viscous secretions and crusts)

  - **chronic pharyngo-esophagitis** (**Paterson-Brown-Kelly disease**) occurs exclusively in **women** after the age of 40, in the hypopharynx, due to ***deficiencies in iron, vitamins etc.***
- **Diagnosis:**
  - foreign body sensation, burning, pharyngeal pruritus
  - cough, odynophagia, dysphagia
  - pharyngeal dryness (atrophic form)
  - oropharyngoscopy: reveals the changes presented in the description of the clinical forms
- **Differential diagnosis:**
  - **specific chronic pharyngitis**
  - **oropharyngeal mycoses**
  - **hypopharyngeal cancer**
- **Treatment:**
  - vitamin A / aerosols with alpha-chymotrypsin
  - oropharyngeal disinfectants (hexoral, phenosept, codecam, orofar)
  - spa and seaside climate therapy
  - elimination of predisposing factors
  - surgical treatment only in localized forms of hypertrophy (CO2 laser)

Pharyngeal Ozena

- is a downward extension of **nasal ozena** and has the same characteristics
- generally **does not respond to treatment**

Pharyngokeratosis

- **Definition:**
  - excessive keratinization of the epithelium in the tonsillar crypts
  - this keratinization produces hard, adherent, whitish-yellow growths
- **Symptoms:**
  - is characterized by a **foreign body sensation (cancer phobia)**
- **Differential diagnosis:**
  - **mycosis with leptothrix** (growths over the entire surface of the pharyngeal mucosa, leptothrix present on bacteriological examination)
- **Treatment:**
  - tonsillectomy

CHRONIC ADENOIDITIS

- **Definition:**
  - chronic inflammation accompanied by **hypertrophy of Luschka's tonsil** (chronic hypertrophic nasopharyngeal tonsillitis or chronic adenoiditis)
  - occurs frequently in children between 3 and 6 years of age, exceptionally in adults or the elderly
- **Favoring factors:**
  - repeated rhinitis and rhinosinusitis
  - repeated acute adenoiditis
  - lymphatism
  - repeated tonsillitis
  - infectious-contagious diseases

  - low immune status
  - poor nutrition and hygiene
  - damp, cold climate
- **Clinical forms:**
  - **large** (occupies the nasopharynx entirely)
  - **medium-sized** (partially obstructs the choanae)
  - **small** (occupies the superior wall of the nasopharynx)
- **Symptoms:**
  - subfebrile / nocturnal cough / frontal headache
  - nasal respiratory insufficiency / submandibular lymph nodes
  - in the course of its evolution, chronic adenoiditis causes a **number of disorders:**
    - adenoid facies (small nose, open mouth, retracted upper lip, inert lower lip, pale, expressionless face)
    - phonation disorders (closed rhinolalia)
    - hearing disorders (conductive hearing loss)
    - disorders of the facial bones (faulty implantation of the teeth, delay in the normal development of the skeletal system)
    - respiratory disorders (catarrhal laryngitis, subglottic
    - digestive disorders (gastroenterocolitis)
    - mental and intellectual disorders (apathy, mental and intellectual retardation)
    - micturition disorders (nocturnal enuresis)
- **Diagnosis:**
  - anamnesis
  - inspection and palpation
  - oropharyngoscopy: purulent secretions running down the posterior wall of the oropharynx; secretions also present in the nasal fossae (anterior rhinoscopy)
  - posterior rhinoscopy (hypertrophic lymphoid tissue sometimes occupies the nasopharynx)
- **Differential diagnosis:**
  - TB adenoiditis
  - nasopharyngeal syphilis
  - benign and malignant tumors of the nasopharynx
- **Treatment:**
  - adenoidectomy
  - surgery performed at the optimal time prevents the disorders described above

CHRONIC TONSILLITIS

- **Definition:**
  - chronic inflammation of the palatine tonsils accompanied by hyperplasia of all elements and the presence of caseum in the crypts
- **Favoring factors:**
  - repeated acute tonsillitis in childhood
  - repeated acute adenoiditis, chronic adenoiditis
  - low immunity
  - damp, cold atmosphere with harmful factors (smoke, dust, toxic gases)
  - poor nutrition
- **Clinical forms:**

  - **atrophic**
  - **hypertrophic**
- **Diagnosis:**
  - odynophagia, dysphagia when consuming cold liquids or foods, or with changes in temperature
  - subfebrile state / nocturnal cough in children
  - caseum in the crypts – fetor
  - asthenia
  - hypertrophic or atrophic tonsils with caseum in the crypts
  - congestion of the free edge of the anterior pole
  - subangulomandibular lymph nodes (small, painful)
- **Differential diagnosis:**
  - secondary syphilis
  - simple hypertrophic tuberculosis
  - sarcoma
  - malignant lymphogranulomatosis
- **Treatment:**
  - tonsillectomy

TONSILLAR FOCAL INFECTION

BILLINGS introduced the theory of tonsillar focal infection in 1912

- **Definition:**
- a **latent, closed, anatomically and immunologically delimited infectious process** with **weak or absent local manifestations**, which may intermittently produce **functional disorders or histopathological lesions** at a distance through an **allergic-hyperergic mechanism**
- possible mechanisms of action of the tonsillar focal infection in the body:
  - **septicemia or sepsis** (pathogens from the tonsil and pharynx reach the blood and settle in organs, where they cause damage)
  - the pathogens remain in the tonsil and **their toxins** reach the body and cause complications
  - **local neurovegetative irritation** triggers remote neuroendocrine reactions (articular, renal, cardiac)
  - **Group A beta-hemolytic streptococcus produces an exotoxin** which induces specific antibodies (antihyaluronidase, antistreptodornase, antistreptokinase, etc.); on a new attack by beta-hemolytic streptococcus, the exotoxin formed reacts with the previously formed specific antibodies
- **allergic mechanism** causing an inflammatory reaction in the joint, renal parenchyma or vascular wall
- **Diagnosis:**
  - anamnesis
  - local clinical examination
  - paraclinical examinations: ESR, hemogram, fibrinogen,

ASLO, C-reactive protein, serum complement, rheumatoid factor, urine examination, etc.

- **Complications:**
  - acute polyarticular rheumatism
  - acute and chronic glomerulonephritis

- endocarditis, arteritis, thrombophlebitis
- gastroenteritis, cholangitis, appendicitis
- chronic laryngitis, asthma, tracheobronchitis
- dermatosis, psoriasis, eczema
- blepharitis, conjunctivitis, optic neuritis
- migraines, headache, neuralgia
- **Surgical treatment**
- tonsillectomy is performed under **pre- and postoperative antibiotic cover** (at least three days before and five days after the intervention), preferably with a **penicillin-series antibiotic** (penicillin, erythromycin, amoxicillin, amoxiclav)
- surgery is performed when the patient **is not in an acute episode** and laboratory examinations are normal
    - ASLO close to 250 IU
    - fibrinogen around 3.5
    - ESR up to 10 mm/h
    - normal urinalysis
- **Contraindications to tonsillectomy are**:
    - the presence of focal complications in the acute period
    - acute conditions
    - syphilis, tuberculosis
    - pregnancy
    - age under 3 years, etc.

CHRONIC SPECIFIC INFLAMMATIONS

- **Syphilis of the pharynx**
- **Congenital syphilis**
    - **early** (from the second month to 5-6 months after birth – manifestations as in acquired secondary syphilis)
    - **late** (appears in later childhood and presents simultaneous tertiary lesions in the pharynx, larynx and nose)
- **Acquired syphilis**
    - Etiology:
        - **treponema pallidum**
    - Clinical phases:
        - primary
        - secondary
        - tertiary

Primary syphilis

- anginal form (fever, odynophagia, unilaterally enlarged, red tonsil, satellite lymph node)
- erosive, ulcerative form (deep ulceration with regular, indurated edges)
- differed form
- gangrenous form
- pseudoneoplastic form
- **Diagnosis:**
    - identification of Treponema pallidum from the secretion, directly on the slide
    - BWR negative

Secondary syphilis

- Whitish patches present on the oropharyngeal mucosa
- syphilitic roseola on the skin
- BWR positive – 4-5 weeks after the onset of the primary phase
- represents the **period of maximum contagiousness**

Tertiary syphilis

- the **gumma** can be located on the soft palate, the posterior wall of the pharynx or the hard palate
- syphilitic gumma with its four stages (crudity, softening, ulceration, scarring)
- deep destructive, painless lesions
- **specific treatment of the underlying disease**

Tuberculosis of the pharynx

- most frequent between 20-40 years of age
- secondary to pulmonary tuberculosis
- **Clinical forms:**
  - acute miliary tuberculosis (Isambert)
  - ulcerative tuberculosis
  - latent form
  - pharynx lupus
  - retropharyngeal cold abscess

Acute miliary tuberculosis

- occurs in young people with cavitary pulmonary tuberculosis
- fever of 39-40
- marked asthenia
- sweating
- weight loss
- odynophagia, **marked dysphagia**, sialorrhea
- oropharyngoscopy: **small yellowish-white nodules that ulcerate** (the ulcerations are superficial, with a dirty base, surrounded by pale mucosa)

Ulcerocaseous tuberculosis

- **ulcerations** of different sizes with irregular edges, surrounded by a **pale mucosa**

Tuberculosis of the pharynx

- **Latent form**
  - hypertrophy of the three pharyngeal tonsils (palatine and Luschka)
  - TB adenitis

Pharyngeal lupus

- **secondary to nasal lupus**
- **more attenuated form of Koch's bacillus**
- **lesion polymorphism** (red-yellow nodules, superficial ulcers with irregular edges covered with a gray-yellow deposit, stellate scars)

Retropharyngeal cold abscess

- is of **lymph node and spinal origin**

- the differential diagnosis with the retropharyngeal abscess of the young child, where the lateral X-ray shows the integrity of the spine, is very important

ORAL CAVITY AND LABIAL INFLAMMATIONS

General or dermatological conditions cause changes in these mucous membranes

Rhagades of the oral commissure

- **Favoring factors:**
  - mycoses
  - diabetes
  - deficient dentures
  - **iron-deficiency anemia**
  - non-specific infections with pyogenic germs
  - the onset of a **carcinoma of the oral commissure**
- **Symptoms:**
  - ulcerative-erosive lesions
  - pain when opening the mouth
  - mild bleeding
- **Treatment:**
  - local applications of silver nitrate (1-2%)
  - investigations to establish the cause and treatment of the underlying disease

Glossitis

- **Definition:**
  - all inflammatory lesions involving the tongue
- **Etiology:**
  - intolerance to the dental materials used and to various medicinal substances
  - **anemia, diabetes, liver cirrhosis**
- **Clinical manifestations:**
  - **burning of the tongue** (on the tip and edges), with minimal objective lesions
- **Treatment:**
  - elimination or relief of the above-mentioned symptoms by **painkillers, local antiseptics and removal of the etiological factor**
  - a number of other chronic inflammatory conditions of a different kind may occur in the tongue: **mycosis, actinomycosis, scleroderma, allergy**

The superficial lesions of the tongue that most often favor inflammation are:

- **geographic tongue**: red or pale map-shaped areas
  - the etiology is **not known** and it does **not require treatment**
- **fissured tongue:** shows depressions and grooves
  - variable dimensions and shapes
  - form islands on the surface of the tongue
- **treatment: symptomatic**, with local antiseptics

Allergic glossitis

- **drugs:** antibiotics, barbiturates, sulfonamides, aspirin, local anesthetics
- **food:** fish, fruit (strawberries, etc.)
- **Symptoms:**

  - from pain and itching to swelling with marked edema
  - the presence of allergic Quincke's edema
- **Treatment:**
  - **corticotherapy, antihistamines**
  - analgesic
  - identification of allergens

Mycotic glossitis

- **Etiology:**
  - **Candida albicans**
- **Favoring factors:**
  - **low immunity**
  - long-term administration of antibiotics, chemotherapy, contraceptives, corticotherapy
- **Diagnosis:**
  - small, superficial whitish-grey deposits on the tongue and oral mucosa that are easily detached
  - after their detachment, the tongue and oral mucosa are red and irritated, and the patient experiences burning and pain
  - smear with bacteriological examination
- **Treatment:**
- local administration of **5% borax glycerin**
- **local antifungal administration** (nizoral, pimafucin, clotrimazole)
- systemic administration of antifungals, alkaline mouthwash

Stomatitis

Ulceromembranous stomatitis

- **Etiology:**
  - viral
  - fusospirillar infections
  - poor oral hygiene
  - dental treatments with various drugs
  - drug allergies
- **Diagnosis:**
  - abundant sialorrhea
  - fetidity
  - subfebrile or febrile state
  - dysphagia, odynophagia
- initially the gingivae are swollen, red and painful on palpation, then **superficial ulcers** appear on the buccal and lingual mucosa, or deep ulcers covered by fibrin deposits
- bacteriological examination shows **fuso-spirillar flora**
  - **Differential diagnosis:**
    - herpes
    - aphthous stomatitis
    - herpes zoster
    - syphilis
    - tuberculosis
    - hematological diseases (agranulocytosis, leukemia)

        - cancer
    - **Treatment:**
        - **local:** applications of 1% gentian violet solution, 1% silver nitrate, local antiseptics, antifungals if needed
        - **general:** antifungals, antibiotics according to the antibiogram
        - **local and oral hygiene**

Herpetic stomatitis and gingivitis

- **Etiology:**
    - herpes virus, Coxsackie virus
    - herpes virus is present in 90% of the population and only 1% develop the disease
    - occurs **most frequently in children**
- **Diagnosis:**
    - sialorrhea / **intense dysphagia** / fever / fetid halitosis
    - **blisters** with clear fluid that appear on the tongue and then spread
    - the **vesicles** turn into round or oval **ulcers** with a reddish halo
    - **is very contagious**, especially in children
- **Evolution:**
    - ulcers heal with crusts, but **do not leave scars**
- **Treatment:**
    - acyclovir in the early stages
    - local applications: antiseptic solutions, 1% gentian violet
    - oral hygiene
    - ***corticosteroids are not given***

Stomatitis due to metal poisoning

- stomatitis with gingival manifestations occurs most frequently after **chronic intoxication** with mercury, bismuth, lead, arsenic, zinc, nickel, chromium, chlorine, fluoride, organic substances (benzol, dimethyl sulphate, carbon tetrachloride, trichloroethylene), synthetic resins, insecticides
- **Treatment:**
    - is specific to the underlying disease

Medicinal stomatitis

- occur after **administration of** bromine, iodine, salicylate, antibiotics, sulfonamides, antidepressants, antiepileptics, barbiturates, oral contraceptives

Allergic stomatitis

- produced by a **variety of allergens**: medicines, dental products, cosmetics, chewing gum, food: fruit, fish, protein, milk
- **allergy testing** establishes the exact diagnosis
- accompanied by Quincke's edema
- **Treatment:**
    - antiallergic therapy

Mycotic stomatitis

- **Etiology:**
    - **Candida albicans** is most commonly found, and **Aspergillus** less commonly

  - occurs in individuals with low resistance, after chemotherapy or prolonged antibiotic therapy
- **Diagnosis:**
  - white or grey superficial dots on the oral mucosa, easily detached, surrounded by red mucosa
  - may cause **superficial ulceration**
  - bacteriological exam
- **Treatment:**
  - **local** hygiene, antiseptic and alkalinizing solutions, 5% borax glycerin, antifungals
  - **general:** antifungals, vitamin therapy, immunotherapy

Herpes zoster

- **Etiology:**
  - **zoster virus**, which is neurotropic and affects the **V2 and V3 branches**
  - **Symptoms:**
    - intense dysphagia
    - sialorrhea
  - **vesicles** along the nerve branches that evolve into **ulcerations** covered by **fibrinous deposits**
  - **Complications:**
    - trigeminal neuralgia
  - **Symptomatic treatment:**
    - local antiseptics
    - anti-inflammatory drugs
    - b-group vitamins

Chronic recurrent aphthous ulcers

- **Etiology:**
  - **unknown**; it is assumed that they are most commonly of **viral origin**
  - occurs more frequently in children, adolescents and young people with a **labile autonomic nervous system**, and during **menstruation**
- **Differential diagnosis:**
  - herpes simplex
  - herpes zoster
- **Treatment:**
  - antiseptic solutions
  - gentian violet 1%
  - borax glycerin 3%
  - cortisone ointments

Behçet Disease

- **Etiology:**
  - is presumed to be of **viral origin**, a **generalized vasculitis** or an **autoimmune disease**
- **Symptoms:**
  - **mouth and genital aphthous ulcers**
  - unilateral eye involvement (iritis, papilledema, retinal damage)
  - rheumatic signs and renal impairment

  - **cochleo-vestibular deafness**
- **Evolution:**
  - **fatal** over time
- **Treatment:**
  - blood transfusions
  - corticotherapy
  - immunosuppressive drugs

Hyperkeratosis and leukoplakia

- **Hyperkeratosis** – **white** granular epithelial proliferation
- **Leukoplakia** – limited or more extensive smooth epithelial proliferation, **white in color**
- **Location:**
  - tongue, buccal mucosa, floor of the mouth
- **Etiology:**
  - unknown
- **Favoring factors:**
  - chronic irritation /dental infections /prostheses
  - smoking / alcohol
  - syphilis / tuberculosis
  - lichen planus / lupus
- **classification of leukoplakia:**
  - simple, verrucous, erosive (**35% become malignant**)
  - leukoplakias are considered **precancerous stages**
- **Treatment:**
  - surgical – removal with safety margins using the CO2 laser
  - mandatory histopathological examination

Bowen's disease (erythroplasia)

- considered an intraepithelial **carcinoma in situ**
  - occurs in adults between 40 and 70 years of age
  - clinical appearance: leukoplakia, vegetative lesion, papillomatous appearance
- evolution is usually towards **squamous cell carcinoma**

ORAL CAVITY INFLAMMATIONS IN DERMATOLOGICAL DISEASES

Plan lichen

- **Etiology:**
  - **unknown**
  - the presence of **neurological disorders** is assumed
- **Diagnosis:**
  - **white nodules**, grouped or scattered, on the lingual, buccal and gingival mucosa
  - **smooth gray or whitish plaques** are found on the tongue
  - the lesions **can detach** and are firm, flat and painless
  - **2-3 mm itchy red-brown papules** located on the tongue
- **Treatment:**
  - laser vaporization, vitamin A, local corticosteroids
  - removal of external toxic agents (**smoking, alcohol, toxic substances**)
  - should be **monitored as precancerous conditions**

Polymorphic erythema

- **Etiology: unknown**
- **Favoring factors:**
  - intolerance to antibiotics, laxatives, tranquilizers
  - bacterial, fungal, viral infections
- **Diagnosis:**
  - sialorrhea, fetid halitosis, local pain
  - oral lesions: **fibrin deposits, crusts, blisters** (tongue and oral mucosa); they coexist with skin and joint lesions and signs of acute infection
  - regional adenitis
- **Evolution:** periodic; occurs frequently in male adolescents
- **Prognosis: reserved**
- **Treatment:**
  - **local:** antiseptic solutions, anesthetics, vitamin A
  - **general: corticotherapy**

Pemphigus

- **Etiology:**
  - autoimmune disease
- **Diagnosis:**
  - occurs most frequently in patients between 40 and 60 years of age
  - biopsy plus **Tzanck test (cytology)**
  - skin lesions that precede the lesions in the oral cavity (on healing they leave **pigmented spots**)
  - in the oral cavity: flat or flaccid bullae with a **tendency to evolve towards superficial epithelial erosion**
  - periodic evolution
  - fetid halitosis, intense dysphagia
- **Treatment:**
  - high-dose corticotherapy

ORAL CAVITY PATHOLOGY

Oral cavity

Anatomy

- the oral cavity is divided into **two sections:**
- **the oral vestibule** is the part of the oral cavity outside the teeth, bounded externally by the mucosa of the lips and
- **the cheeks**, and internally by the maxillary and mandibular alveolar processes
- the **facial nerve** innervates the muscles of facial expression
- the mucous membranes of the lips and cheeks contain multiple salivary glands
- **the parotid duct (Stensen)** opens into the oral cavity at the level of the second upper molar
- **the oral cavity proper** is the space medial to the maxillary and mandibular alveolar processes

Tongue

- The lingual "V" divides the tongue into **two portions**

- **the base of the tongue**, covered with lymphoid tissue belonging to Waldeyer's lymphatic ring – the **lingual tonsil**
- besides the circumvallate papillae, the **anterior, mobile part of the tongue** is covered by other types of **papillae**
- **filiform papillae** have only a mechanical role and are distributed in the posterior region of the tongue
- **foliate and circumvallate papillae** are taste receptors
- the muscles of the tongue can be divided into an **intrinsic and an extrinsic system**
- the **extrinsic muscles** contribute to the movement of the tongue – lowering, retracting and raising it
- the **intrinsic muscles** are made up of fibers arranged vertically, transversely and longitudinally – they give the tongue the ability to change its shape

The floor of the mouth

- inferior limit of the oral cavity
- consisting of multiple muscle layers
- formed mainly by the **mylohyoid muscle**

Physiology

- the functions of the oral cavity are:
  - ingestion of food
  - speech
  - recognition of taste
  - accessory respiratory function

Clinical tests

- **Inspection and palpation**
  - the opening of **Stensen's duct**
  - by massaging the cheeks, **saliva** is expressed and its consistency can be assessed
  - the function of the **cranial nerves** should also be assessed
  - the movement of the tongue provides information on the activity of the **hypoglossal nerve**
  - to examine the motor innervation of the soft palate, the **gag reflex** must be tested
  - in case of paralysis of the **glossopharyngeal nerve**, the soft palate deviates to the healthy side
  - **sensory innervation** is evaluated using a cotton swab moved over the entire surface of the tongue and cheeks, to check whether the sensations are **bilaterally equal**
- **Taste evaluation**
- impairment of taste can be tested by applying to the tongue solutions with the **four main tastes**: ***sweet, salty, sour, bitter***
- the following are used for testing:
  - **glucose** for the sweet stimulus
  - **sodium chloride** for the salty stimulus
  - **citric acid** for the sour stimulus and
  - **quinine sulphate** for the bitter stimulus
- the solutions are applied with a cotton swab to the sides of the tongue
- **normally all solutions are identified at the lowest concentration**

- these **subjective tests** are easy to perform, but have a fairly low degree of reliability and reproducibility

Congenital Diseases

- there are **multiple congenital conditions in the oral cavity**
- for appropriate treatment, the history of the disease and the patient's history should be known
- in case of ***congenital syndromes*** the patient may have multiple abnormalities; therefore the involvement of a **pediatrician** and a **geneticist** in treatment is desirable
  - clinical examination
  - X-rays
  - deglutition function testing and
  - hearing function examination

Congenital Epulis

- **Etiology/Epidemiology**
  - this is a **myoblastoma** consisting of granular cells, a benign tumor, generally located at the boundary ***between the oral mucosa and the maxillary and mandibular alveolar processes***
  - is common in female patients
- **Symptoms:**
  - red tumor of the oral mucosa, which may cause difficulties in ingesting food
  - X-rays show **erosion of the bony alveolar process**
- **Treatment**
  - **exclusively surgical**, by excision of the tumor, including the periosteum

Palatal and mandibular torus

- **Etiology/Epidemiology**
  - **bone exostoses**, located on the surface of the hard palate or on the lingual surface of the mandible
  - covered by mucous membranes
- **Treatment**
  - only if the formation is massive, dentures no longer fit, or the exostoses undergo recurrent infections, ulcerations or neoplastic change

Ankyloglossia

- **Etiology/Epidemiology**
  - shortened lingual frenulum, which fixes the entire tongue to the floor of the mouth, limiting the mobility of the tongue
- **Symptoms**
  - difficulty in speaking, chewing and swallowing
- **Treatment**
  - surgical, consisting of division of the lingual frenulum
  - in severe cases (a very thick lingual frenulum) a Z-plasty is required

Lingual Thyroid

- **Etiology/Epidemiology**
  - thyroid tissue originates at the **foramen caecum**, in the middle of the base of the tongue, and descends to its anatomical position anterior to the trachea
  - **between 7-12 weeks of embryogenesis**

  - the thyroglossal duct is obliterated
  - if the thyroid fails to descend, it persists at the base of the tongue
- **Symptoms**
  - dysphagia, breathlessness and impaired speech, symptoms of **hypothyroidism**
- **Diagnosis**
  - mass in the lymphoid tissue at the **base of the tongue**
  - in case of lingual thyroid, the normal thyroid tissue is assessed by clinical tests, cervical ultrasound and thyroid scintigraphy
- **Treatment**
  - **thyroid hormone suppression** to relieve the lingual thyroid
  - radioactive iodine ablation
  - surgery

Hypoplastic mandible

- usually associated with **first arch syndrome**

Pierre Robin Anomaly

- **the characteristic triad** of this anomaly is:
- cleft palate
- **micrognathia and** glossoptosis
- first, **development of the first arch is arrested**; the characteristic triad appears secondarily
- during childhood **airway obstruction** may occur – a life-threatening situation that requires tracheotomy and glossopexy
- in some cases **mandibular growth reaches a normal stage by the age of 6 years**

Treacher Collins Syndrome

Franceschetti-Treacher Collins Syndrome

MandibuloFacial dysostosis

- a craniofacial dysmorphism syndrome
- typically occurs in combination with
  - hypoplastic mandible
  - low-set ears
  - oral atresia
  - hypoplastic mastoid and epitympanum
  - cleft palate
  - **autosomal dominant transmission**

Crouzon Syndrome and Apert Syndrome

- the main factor in both syndromes is **craniofacial dysostosis**
  - premature abnormal synostosis of multiple suture lines of the skull
- **Crouzon syndrome** - maxillary hypoplasia, mental retardation, congenital oral atresia and mandibular prognathism
- **Apert syndrome** – early fusion of the cranial sutures causes brachycephaly, in combination with cleft palate, syndactyly of the hands and feet, and mental retardation

Tongue anomalies

Fissured tongue

- **Synonyms:** Lingua plicata
- **Etiology/Epidemiology**
  - the incidence of fissured tongue is 10%
  - **autosomal dominant transmission**
- **Symptoms:**
  - there are **no clinical signs**
  - fissures may be lateral, medial or randomly spread over the surface of the tongue
  - may occur in the context of ***Melkersson-Rosenthal syndrome, Down syndrome or acromegaly***
- **Treatment: not necessary**

Geographic tongue

- **Etiology /Epidemiology**
  - desquamation of filiform papillae in different regions of the tongue
  - characteristic irregular shape with **white and red spots**
  - the areas change their position, color and pattern, giving the characteristic "map" appearance
- **Symptoms**
  - there are **no clinical symptoms**
  - except for burning of the tongue and decreased or lost taste
  - histopathological examination shows **inflammatory signs**
- **Treatment**
  - local application of **vitamin A and zinc**
  - should help re-epithelialization of the mucosa

Hairy tongue

- **Etiology/Epidemiology**
- **filamentous hyperkeratosis** of **the filiform papillae**, which looks like hair on the surface of the tongue
- elongation of the papillae is caused either by lack of desquamation or by **increased production of keratin**
- hyperkeratosis is triggered by
  - antibiotic treatments
  - nicotine abuse
  - chronic irritation of the mucosa
  - metabolic imbalance
- **Symptoms:**
  - the surface of the tongue has a **hairy appearance** caused by **elongation of the filiform papillae**
  - there is **no** pain or burning of the tongue
- **Treatment:**
  - treatment of the causative disease

Hunter glossitis

- **Etiology/Epidemiology**

  - nutritional anemia, or **pernicious anemia** in the case of malabsorption with intrinsic factor deficiency
- **Symptoms:**
  - tongue is inflamed, atrophic
  - burning of the tongue, paresthesia and dry mouth
  - the surface of the tongue has an atrophic and glossy appearance
- **Diagnosis/Treatment:**
  - measuring serum levels of **vitamin B12**
  - in case of low levels, **folic acid should be administered**

PHARYNX BENIGN TUMORS

Benign tumors of the pharynx

- these tumors involve the three segments of the pharynx:
  - nasopharynx
  - oropharynx
  - hypopharynx

are specific to each level:

- fibromyxoma
- papillomas
- angiomas
- fibromas
- lipomas
- chondromas
- thyroid tumors arising from the thyroglossal duct
- teratomas
- chordomas

Nasopharyngeal fibroma

- occurs predominantly in males – **tumor of male puberty**
- the commonly incriminated **etiology** is **endocrine** (17-ketosteroid assay shows a deficiency; the tumor no longer develops after the age of 20)
- **Diagnosis:**
  - repeated epistaxis in a male patient at puberty
  - progressive nasal obstruction, headache, hearing loss
  - deformity of the face (frog face)
  - posterior rhinoscopy reveals a sessile, hard, fibrous, bleeding, **pale or pink tumor**
  - grows by extension, **invading** the nasal fossa, orbit, sinuses, zygomatic and temporal fossae and the base of the skull
  - **palpation or biopsy is not recommended because of bleeding**
  - **17-ketosteroid assay** shows hormonal deficiency
- **Differential diagnosis:**
  - adenoid hypertrophy
  - other benign tumors
  - malignancies
- **Treatment:**

- **surgical** (tumor removal) with controlled hypotension to avoid bleeding; preoperative preparation, ICU, including blood grouping

PHARYNX MALIGNANT TUMORS

Malignant tumors of the nasopharynx

- **Etiology:**
  - the **viral origin hypothesis** is increasingly accepted (Epstein-Barr virus)
- **Frequency:**
  - represent 2% of upper aerodigestive tract cancers and 0.8-1% of all cancers
  - age: most commonly **between 45 and 60 years**, but can also be found at the extremes of age
- **Anatomopathological forms:**
  - epitheliomas 85-95% (undifferentiated, poorly differentiated, differentiated)
  - lymphoepithelioma
  - cylindroma
  - sarcoma, lymphosarcoma, reticulosarcoma
  - plasmocytoma
  - dysembryoplastic malignant tumors (chordoma, craniopharyngioma, rhabdomyosarcoma)
- **Symptoms:**
  - **phases of evolution: onset, established disease, extension and terminal**
  - determine the patient's symptoms (the symptoms are characteristic of each phase)
  - **Debut phase:**
    - in this phase the patient has discreet symptoms with minimal discomfort: retronasal foreign body sensation, nasal obstruction, hearing loss
  - **The established phase** has 5 forms:
    - rhinologic
    - otic
    - neurologic
    - ocular
    - lymph node
- ***The symptoms*** are **characteristic** of each form:
  - **rhinologic**
    - nasal obstruction
    - micro-epistaxis
    - muco-purulent nasal secretions
  - **otic**
    - tinnitus
    - autophony
    - conductive hearing loss
    - chronic catarrhal otitis unresponsive to treatment, sometimes acute otitis
  - **neurologic** (manifested by damage to one or more **cranial nerves**)
  - **anterior syndromes:**
    - superior orbital fissure syndrome (III, IV, VI plus ophthalmic branch of V)
    - orbital apex syndrome (II, III, IV plus ophthalmic branch of V)
    - petrosphenoidal syndrome (V, VI, subsequently II, IV)

        - paratrigeminal syndrome (V,VI)
    - **posterior syndromes:**
        - jugular foramen syndrome (IX, X, XI)
        - condylar and jugular foramen syndrome (IX, X, XI, XII)
        - syndrome of the jugular foramen, anterior condylar canal and superior cervical sympathetic (IX, X, XI, XII plus superior cervical sympathetic)
        - **Trotter syndrome** (invasion of the tumor into the maxillopharyngeal space): hemiparalysis of the soft palate, trigeminal neuralgia (V), unilateral conductive hearing loss
        - **Garcin syndrome** (cranial nerves from I to XII)
- **neurologic**
    - anterior syndromes
    - posterior syndromes
- **ophthalmic** (the **VI nerve** is more commonly affected):
    - the patient has diplopia and limited lateral movement of the eyeball
- **lymph node**
    - high cervical lymph nodes, or nodes in the space between the mastoid tip and the ramus of the mandible: firm lymph node, fixed to deep structures, painful on palpation
- **Diagnosis:**
    - anamnesis
    - complete ENT clinical examination
    - neurological examination / ophthalmological examination
    - radiological examination: skull base X-ray (Hirtz view), CT
    - **biopsy** of the nasopharyngeal tumor or cervical lymph nodes
- **Differential diagnosis:**
    - adenoid hypertrophy
    - benign tumors
    - TB, syphilis
- **Treatment:**
    - radiation therapy to the tumor and lymph node areas
    - chemotherapy / combination of radio-chemotherapy
    - immunotherapy
    - the prognosis is **generally unfavorable** when the tumor has invaded the base of the skull

Malignant tumors of the oropharynx

Cancer of the palatine tonsil

- the most common tumor of the pharynx; unilateral; occurs at the age of 45-60 years
- anatomopathological forms:
    - **epithelial** (most common epithelioma)
    - **connective tissue** (sarcomas: lymphosarcomas, reticulosarcomas, etc.)
- clinical forms: vegetative, ulcerative, infiltrating
- **Diagnosis:**
    - odynophagia, dysphagia
    - sialorrhea, fetid halitosis
    - reflex otalgia, trismus

- weight loss
- lymph nodes are present in 20% of patients at the first consultation

Cancer of the palatine tonsil

- **Diagnosis:**
  - oropharyngoscopy reveals the tumor:
    - deep, crateriform ulceration
    - tonsil infiltration
    - vegetative tumor
    - infiltrative-ulcerative or ulcerative tumor
- **biopsy** - histopathological examination
- **Treatment:**
  - depending on the TNM stage
  - **surgical only in stage I**, when the tonsil is mobile and the tonsillar pillars are not infiltrated
  - radiotherapy (6000-8000 r)
  - chemotherapy
  - immunotherapy
  - a combination of radio-chemo-immunotherapy is commonly used

Cancer of the floor of the mouth

- **Diagnosis:**
- is **frequently asymptomatic** for a long time
  - minimal subjective and functional symptoms
  - satellite lymph nodes
  - oropharyngoscopy: infiltration or ulceration of the floor
- **biopsy** and histopathological examination
- **Treatment:**
  - surgical / radiotherapy / chemotherapy

Cancer of the anterior 2/3 of the tongue

- develops in front of the lingual V, on the dorsal surface or the edges of the tongue
  - minimal symptomatology
  - **Treatment:**
    - surgical/radiotherapy

Malignant tumors of the hypopharynx (cancer of the pyriform sinus)

- **the most common location** of hypopharyngeal cancers
  - age 45-60
- **Diagnosis:**
  - early lateral cervical lymph node
  - foreign body sensation
  - odynophagia, dysphagia
  - reflex otalgia
  - hypopharyngoscopy / indirect laryngoscopy / fibroscopy / esophagoscopy
  - contrast X-ray and CT
  - **biopsy**
- **Differential diagnosis:**

  - laryngeal cancer
  - pharyngeal foreign bodies
- **Treatment:**
  - surgery, radiotherapy, chemotherapy

BACTERIAL INFECTIONS

Chronic gingivitis

- **Etiology/Epidemiology**
  - the **most common form of gingivitis**; the incidence is 90% in middle age
  - poor hygiene in combination with bacterial factors and food particles causes irritation of the gingival mucosa
- **Symptoms**
  - diffuse and painful erythema of the gums; ulcerations sometimes occur
- **Treatment**
  - dental hygiene and dental treatment

Acute gingivitis

- **Etiology/Epidemiology**
  - the most important etiological factors are poor dental hygiene associated with immune and nutritional deficiency
  - acute gingivitis is caused by **beta-hemolytic streptococcus** and **staphylococcus**
- **Symptoms**
  - the gingiva is erythematous and inflamed, bleeds slightly and hurts during mastication
  - oral fetor
- **Differential diagnosis**
  - acute ulcero-necrotic gingivitis and herpetic gingivitis
- **Treatment**
  - management of acute bacterial infection with **oral antibiotics** (penicillin or ampicillin) and improvement of **oral hygiene**
  - local anesthesia may be useful

Acute ulcero-necrotic gingivitis

- **Synonyms: Vincent Stomatitis, Gangrenous pharyngitis**
- **Etiology/Epidemiology**
  - older people or people with a weak immune system
  - necrosis and gingival retraction through **overgrowth of bacteria and Borrelia vincentii**
  - poor oral hygiene and immune deficiency
- **Symptoms**
  - hyperemia and swelling of the gingiva, then a pseudomembrane appears, covering the necrotic tissue at the gingival margins
  - crater-shaped lesions on the alveolar margins and throughout the gingiva
  - slight gingival bleeding
  - very painful, sometimes associated with lymphadenitis
  - **metallic taste**
- **Differential diagnosis**
  - erythemato-pultaceous tonsillitis, herpes infections, agranulocytosis, syphilis

- **Treatment**
  - aggressive oral hygiene against **fusiform bacteria**
  - antibiotic treatment with penicillin or ampicillin, local antiseptic treatment

Syphilis (Syphilis)

- **Etiology/Epidemiology**
  - sexually transmitted disease
- **Symptoms**
- **primary syphilis**: a **solitary chancre** occurs after an **incubation period** of 2-3 weeks – extragenital sites are the **lips, tongue, oral mucosa, soft and hard palate**
- **secondary syphilis**: a rash and patches appear on the mucosa of the lips and inside the mouth – this follows a **latency period** of 4-6 months – the lesions are **very painful**
- **tertiary syphilis**: **syphilitic gumma** occurs after 3-10 years – the gumma is a well-delimited, painless ulceration
- **Diagnosis**
- detection of Treponema pallidum by **serological tests**
- **Differential diagnosis**
  - neoplasm
  - specific angina of mononucleosis
  - diphtheria and
  - Vincent stomatitis
- **Treatment**
- 2.4x10 mil. IU of penicillin G twice a week, intramuscularly, or
- 1.2x10 mil. IU of penicillin daily intramuscularly for 15 days in primary, secondary and latent syphilis (duration of infection of less than one year)
- after treatment it is necessary to **repeat serological tests**

Abscess of the submandibular space (Ludwig's angina)

- **Etiology / Epidemiology**
  - bacterial proliferation and invasion from **dental caries** cause necrosis of the bone tissue around the root of the tooth
  - patients require dental treatment, with incision and drainage of the dental abscess
  - infections spread especially from the mandibular teeth, sometimes from mucosal lesions
  - such abscesses can also form as a result of **trauma, lingual tonsillitis** or pathologies of the **submandibular glands**
  - infections can extend to the **deep cervical tissues and the submandibular, parapharyngeal, retropharyngeal or prevertebral spaces**
  - **Ludwig's angina** is diagnosed when **all three primary submandibular spaces** (bilateral, sublingual, submandibular) are affected
- **Symptoms**
  - high fever, pain, trismus and hypersalivation are the most common symptoms
  - the skin is indurated and edematous; the edema may extend down to the **anterior cervical region and the chest wall**
- **Diagnostic**
  - complete blood tests
  - culture
  - CT and ultrasound

- **Treatment**
  - **conservative treatment**
    - antibiotics
  - **surgical treatment**
    - necessary when the abscess has collected, when anaerobic bacteria are involved and if no improvement is observed after conservative treatment

Actinomycosis

- **Etiology / Epidemiology**
  - **bacteria Actinomycetes**
- **Symptoms**
- it starts with a **painless nodule** located in the mucosa or submucosa
- the nodule often ruptures into the mouth or onto the skin surface
- swelling and edema of the skin or mucosa
- **chronic fistula**, especially when the submandibular or parotid gland is involved
- **Treatment**
  - identification of Actinomycetes
  - long-term treatment with penicillin:
  - 10x10 ml U - 20x10 ml U penicillin G / day intravenously for 4- 6 weeks, followed by
  - 2-4 g penicillin V / day or 1.5g amoxicillin / oral for 6 -12 months

FUNGAL INFECTIONS

Oral candidiasis

- **Etiology / Epidemiology**
- **Candida albicans** is found in 15-20% of normal oral mucosa
  - it is normally a **saprophytic organism**
  - often patients with **immune deficiency** develop oral candidiasis
- **Diagnostic**
  - dysphagia, odynophagia, inflammation of the oral cavity and pharynx
    - diagnosis
    - the mucosa of the oral cavity presents with irregular, red lesions
  - **whitish plaques with red edges, which bleed slightly on touch**
  - culture
- **Treatment**
  - **local application** of antifungal drugs such as nystatin or clotrimazole, 5 times daily, 7-14 days
  - in severe cases, **systemic treatment** with 100-200 mg of fluconazole daily for 14-21 days is recommended

VIRAL INFECTIONS

Herpetic gingivostomatitis

- **Etiology / Epidemiology**
  - **Herpes simplex infections** of the oral cavity are most often caused by **type 1**
  - **type 2** Herpes virus infections are most often localized in the **genital area**
  - incubation lasts 5-7 days
- **Symptoms**

  - gingivitis and erythema of the oral mucosa, subfebrile state, general condition slightly affected
  - lymphadenitis
  - vesicles on the oral mucosa that rupture within 24 hours, leaving gray ulcerated lesions
  - fetor, hypersalivation, and odynophagia
  - the eruption heals within a week without scarring
- **Complications**
  - in patients with **immune deficiencies**, the infection may progress, a condition known as **"Pospischill-Feyrter's disease"**
  - the vesicles extend to the skin of the face
  - life-threatening herpes meningitis
- **Diagnosis**
  - based on clinical symptoms
  - **paraclinical investigations – costly, not normally required**
- **Treatment**
  - the virus cannot be eliminated; early treatment with **antivirals** is necessary to **lower viremia**
  - **primary infection** – acyclovir 5x200 mg daily, orally, for 10-14 days
  - **recurrence** – local applications of penciclovir or acyclovir are required
  - local application of antiseptics to prevent **bacterial superinfection**

Herpes Zoster

- **Etiology / Epidemiology**
  - the first contact with the varicella-zoster virus causes **varicella**, mainly during childhood
  - the **latent virus persists in ganglion cells** (trigeminal ganglia) and can be reactivated
  - predisposing factors are immune deficiencies in HIV and immunosuppressive therapy
- **Symptoms**
  - **vesicular lesions** similar to those of varicella, located unilaterally, in the areas innervated by the ***second and third branches of the trigeminal nerve***
  - the patient has **neuralgic pain** before the appearance of the vesicles
- **Treatment**
  - useful if started within the first 3 days after the onset of the lesions; consists of acyclovir 5x800 mg/day orally for 7-10 days
  - analgesics / amitriptyline / carbamazepine

Herpangina

- **Etiology / Epidemiology**
  - **Coxsackie A** virus; **Coxsackie B** infection is sometimes possible
  - children up to 7 years of age are the most affected
- **Symptoms**
  - fever, headache and pharyngitis
  - **intense odynophagia, hypersalivation and nausea**
  - small blisters surrounded by red edges on the soft palate, which rupture early
- **Differential diagnosis**
  - the herpes virus
  - the main difference is the involvement of the entire gingiva in **herpes simplex** infection

  - stomatitis, bacterial pharyngitis and the varicella rash are often confused with herpangina
- **Treatment**
  - symptomatic
  - local anesthetics and silver nitrate

Oral mucosal disorders associated with HIV infection

- **Etiology / Epidemiology**
  - involvement of the oral mucosa or lips is often associated with HIV infection
  - the lesions are **not caused by HIV itself but are a consequence of immune deficiency**
- **Symptoms**
  - the course of the disease is much more severe than in patients who are not infected with HIV
  - viral infections promote HIV progression
- **Treatment**
  - **systemic therapy** with **acyclovir and ganciclovir** – in herpes simplex, varicella and cytomegalovirus infections; the dose depends on the severity of the infection and the immunological status of the patient
  - **interdisciplinary approach** to the treatment of these opportunistic infections

Hairy leukoplakia

- immunocompromised patients, most often in HIV infections
- the lateral part of the base of the tongue has a **hairy appearance**, with colors ranging from white and gray to black; **there is no treatment** other than symptomatic

Kaposi's sarcoma

- since the introduction of antiretroviral therapy and protease inhibitors, Kaposi's sarcoma has become **rare**
- in 90% of patients with Kaposi's sarcoma associated with HIV infection, human **herpes virus type 8** is found
- Kaposi's sarcoma may occur before infection
- presents **two phases:**
- an **early phase** with skin manifestations and lymphatic involvement and
- an **advanced phase** with gastrointestinal and/or pulmonary involvement, sometimes with extensive oral involvement
- **Treatment:** optimization of antiretroviral therapy
- **limited skin sarcoma**: radiotherapy, laser therapy, excision
- **disseminated sarcoma**: systemic therapy with interferon alpha

ORAL CAVITY NEOPLASMS

Benign tumors

- the oral cavity contains multiple structures in which tumors may develop
- these include **epithelial tumors** with mesenchymal and ectodermal elements, as well as tumors of the **nerve structures** of the oral cavity
- treatment consists of **surgical excision**

Epidermoid cyst

- mass of firm consistency, well delimited by a capsule
- treatment consists of **surgical excision**
- **relapse is rare**
- if **adnexal structures** (e.g. hair follicles, sebaceous glands) are found, the diagnosis of dermoid cyst is established

Haemangioma

- well-delimited, large tumor with blood content, appearing in the submucosa of the **floor of the mouth, the oral vestibule or the lips**
  - present at birth or appearing immediately after
- hemangiomas frequently **involve the tongue**, and **airway obstruction** may occur
- it is worth mentioning that the **true size of the hemangioma is assessed with the patient lying down**; the blood content then expands, revealing the true extent of the tumor, which is often underestimated
- the natural evolution of hemangiomas is **spontaneous regression** over the next few years
- this is **not always the case**: upper respiratory tract hemangiomas that cause obstruction, or enlarging hemangiomas, require **surgery**

Lymphangiomas

- appear after birth
- unlike hemangiomas, **spontaneous regression is not possible**
- they can reach a considerable size and are in close contact with the cervical structures, which makes **surgical excision of these tumors very difficult**
- this tumor naturally tends to **recur after excision**
- at the initial operation, total excision of the tumor must be performed
- the use of **steroids is controversial**

Ranula

- **mucocele** of the sublingual glands
- well-delimited **blue mass** under the mucosa of the floor of the mouth, lateral to the lingual frenulum
- when the ranula is located **above the mylohyoid muscle**, excision is performed through a **transoral approach**
- it can herniate laterally through the mylohyoid muscle; in this case it appears as a soft mass in the submandibular region, requiring a **transoral approach combined** with an **external approach**
- **the submandibular gland should be excised to prevent tumor recurrence**

Malignant tumors of the oral cavity and lips

- **Epidemiology**
  - > 95% of malignant tumors of the oral cavity are **squamous cell carcinomas**
  - alcohol and smoking are risk factors
  - the incidence is higher in **males**
- **Diagnosis**
  - for all tumors of the oral cavity the diagnostic procedures are as follows:
    - inspection and palpation
    - endoscopy
    - **biopsy** for histopathological examination

- imaging procedures (MRI, CT)

Anatomoclinical and histopathological classification:

- **floor of the mouth carcinoma**
- **carcinoma of the tongue**
- **retromolar trigone carcinoma**
- **carcinoma of the lip**
- **non-squamous cell carcinoma of the oral cavity**
- **melanoma**
  - melanoma of the oral cavity **is rare**
  - <1% of all melanomas
  - **wide excision** and **dissection of cervical lymph nodes** in case of clinically detectable laterocervical lymph nodes

Malignant tumors of the oral cavity and lips

- many lesions remain **asymptomatic** for a long time and are discovered by dentists at a routine check-up
  - evaluation of the primary tumor and regional metastases
  - **tumor biopsy**
  - **Treatment**
- three therapeutic options:
- primary tumor **surgery**
- **surgery combined with radiotherapy or radiochemotherapy**
- primary radio-chemotherapy
- these therapeutic options should be discussed with the patient, depending on the individual situation
- the **best cure rate** is obtained through combined therapy consisting of surgery associated with radio- and/or chemotherapy

SALIVARY GLAND PATHOLOGY

Introduction

- three pairs of major salivary glands
  - parotids
  - submandibular glands
  - sublingual glands
- several hundred minor salivary glands distributed in the mucosa of several anatomical regions of the head and neck (most in the oral cavity)

The parotid glands

- the largest major salivary glands
- exocrine, tubuloacinar glands, with serous acini
- from superficial to deep, the parotid is crossed by the **following anatomical structures**: the facial nerve, the auriculotemporal nerve, the retromandibular vein and the external carotid artery
- the **facial nerve** divides into five branches: ***temporal, zygomatic, buccal, mandibular and cervical***

Parapharyngeal space

- located bilaterally in the upper cervical region
- **is not accessible** to ENT clinical examination; it becomes relevant when **pathological processes** appear (infections, tumors)
- it is shaped **like a pyramid** with the base up, at the base of the skull, and the tip down, at the level of the greater horn of the hyoid bone
  - **prestyloid space** (anterior compartment of the parapharyngeal space) and
  - **retrostyloid space**, containing the carotid sheath and extending down to the upper mediastinum

Submaxillary gland

- consisting of a **large, superficial lobe**, located in the submandibular (digastric) triangle, and a **smaller, deep lobe**, located posteriorly in the floor of the mouth
- the two lobes are continuous around the posterior margin of the mylohyoid muscle
- the two lobes are not true lobes, because embryologically the gland – of mixed seromucous type – is an epithelial invagination

Sublingual glands

- develops from several epithelial thickenings of the gingivolingual groove, on its outer edge, when the embryo is 20 mm long
- its main duct and many of the ducts of the sublingual glands open directly into the sublingual sulcus

Inflammatory and non-inflammatory disorders of the salivary glands

infectious sialadenitis is an acute or chronic inflammation of the gland manifested by painful or painless enlargement, with or without clinical manifestations, affecting the major and minor salivary glands

the most common etiological agents are **viruses** and **bacteria**

Viral etiology

- **Epidemic parotitis (mumps)**
  - the most common viral infection of the salivary glands; manifests as unilateral or bilateral enlargement of the parotid gland
  - **in 85% of cases it occurs in children under the age of 15, but it can also occur in adults (especially the elderly)**
  - the **prodromal period** lasts **1-3 days**
  - general malaise, discomfort, chills, headache, fever and odynophagia; the enlargement persists for several days up to a week, without expression of purulent secretions
  - **leukopenia with reactive lymphocytosis**

Other infections

- **influenza and parainfluenza viruses (types 1 and 3), Coxsackie viruses (A and B), echovirus, lymphocytic choriomeningitis virus, cytomegalovirus and adenoviruses**, especially in HIV patients
- the symptoms are identical to those of mumps (viral parotitis)
- **enlargement of the gland** as a result of multiple lymphoepithelial cysts
- these cysts can be evaluated by ultrasound or by fine-needle aspiration, which will reveal a serous fluid with a **rich infiltrate of lymphocytes and macrophages**
- intraparotid lymphadenopathy may occur

- **differential diagnosis:** solid tumors of the parotid gland with a high incidence in HIV patients

Acute bacterial infections

- **Clinical evolution**
  - the onset is acute and consists of **enlargement** of the salivary gland, which becomes **painful**
  - the gland is **tender on palpation** and pus is expressed at the papilla of Stensen's duct
  - ***massage of the gland with expression of the pus, although very painful, reduces the ductal pressure and therefore reduces the pain***
- **Treatment**
  - broad-spectrum **antibiotics** (based on etiology) and **anti-inflammatory drugs**
  - **corticosteroids** to reduce inflammation
  - **secretagogue** medication offers **no substantial benefit**

Cytomegalovirus infections

- found in newborns, infants and children **under 2 years**
- **congenital infection** across the placental barrier is **benign**
- **severe forms** are accompanied by
  - jaundice
  - petechial
  - hepatosplenomegaly
  - thrombocytopenia
  - hemolytic anemia
  - chorioretinitis
  - psychomotor and mental retardation
- **Evolution** - **high mortality in newborns**
- **Diagnosis** - **serology**, which reveals **antibodies**
- **Treatment** – **symptomatic**

Chronic sclerosing sialadenitis of the submandibular gland

- **Etiology and pathological anatomy**
  - chronic inflammation of the gland with destruction of the serous acini, lymphocytic infiltration of the interstitial tissue and periductal sclerosis
  - appearance of "**salivary cirrhosis**", through metaplasia of the glandular parenchyma and connective tissue
  - the etiology is **presumed autoimmune**
- **Diagnosis**
  - **histopathological examination**
- **Treatment**
  - surgical excision

Chronic recurrent parotitis

- **Etiology**
  - **congenital ductal dilation** is assumed to be a predisposing factor
- **Symptoms**
  - unilateral or alternating clinical signs
  - swelling that can be bilateral and sometimes painful
  - the saliva is milky or purulent, with a salty taste

  - trismus
- **Diagnostic**
  - history and clinical evolution
  - on sialography it looks like a leafy tree
  - **histopathological examination**
- **Treatment:**
  - oral hygiene / gland massage / sialagogues
  - **parotidectomy**, with preservation of the facial nerve
- **Evolution**
  - **after a long time**, **salivary secretion stops** through **fibrous scarring and destruction of the glandular parenchyma**, and the symptoms disappear

Gougerot-Sjögren Syndrome (Myoepithelial Sialadenitis)

- **Etiology**
  - secondary to an **autoimmune disease** (collagenoses, rheumatoid arthritis
  - the autoimmune reaction results in **atrophy of the glandular parenchyma**, **interstitial lymphocytic infiltration** and **myoepithelial proliferation**
- **Symptoms**
  - xerostomia / recurrent chronic arthropathy
  - atrophy of the lacrimal glands causes keratoconjunctivitis sicca
  - rheumatoid purpura, nodular periarteritis, scleroderma
- **Diagnostic**
  - **Dysproteinemia**
  - increasing ESR
  - signs of **lupus erythematosus**
- **Treatment**
  - difficult, the **etiology being unknown**
  - symptomatic treatment – saliva and tear substitutes (**pilocarpine hydrochloride**)
  - corticosteroids
  - immunosuppressant
  - surgical treatment - **submaxillectomy / parotidectomy**

Sialadenitis with epithelioid cells (Heerfordt syndrome, uveoparotid fever)

- **Etiology**
  - **extrapulmonary manifestation of sarcoidosis** (**Besnier-Boeck-Schaumann disease**) or **benign lymphogranulomatosis**
- **Symptoms**
  - symmetrical, sometimes painful swelling of the parotids and lacrimal glands
  - uveitis
  - **facial paralysis**
  - meningoencephalitis, sensorineural hearing loss
  - the **triad conjunctivitis / parotitis / facial paralysis** constitutes **Heerfordt syndrome**
- **Diagnosis:** histopathological – **manifestations of sarcoidosis** (tuberculoid follicles, but ***without caseation*** and ***without Koch bacilli***)
- **Differential diagnosis:** TB
- **Treatment:** corticosteroids

Salivary lithiasis

- **Etiology**
  - glandular infections of **dental origin**
  - **saliva alkalinity** causes the precipitation of calcium phosphates and carbonates
  - in 85% of cases the stones are located in the submandibular gland and in only 15% in the parotid
- **Symptoms**
  - swelling of the gland
  - painful
  - **salivary colic is pathognomonic: intense pain during meals; the salivary duct is turgid**
- **Differential diagnosis**
  - calcified TB lymph node
  - intraglandular tumor
- **Surgical treatment**
  - calculus extraction
  - submandibular gland removal

Benign tumors

Classification:

1. Epithelial tumors

- adenomas:
  - pleomorphic (mixed tumors)
  - monomorphic - adenolymphoma cyst (Warthin tumor)
- oncocytoma
- other types

2. Benign tumors of other origin, from non-parenchymal elements of the gland:

- vascular tumors
- lipomas
- nerve tumors and
- tumors of the lymphoreticular system

Mixed tumors (pleomorphic adenomas)

- **the most common** salivary gland tumors
- 65% of these are pleomorphic adenomas
- about 60% of tumors of the parotid gland are pleomorphic adenomas
- **in most cases, the tumor is surrounded by a capsule**
- they are **often multicentric**, recurrent tumors, revealed after inadequate, incomplete surgery (simple enucleation)
- **Symptoms**
  - unilateral
  - it develops slowly over several years
  - **women** develop the tumor more often
  - hard, pseudocartilaginous consistency, painless, quite mobile in the parotid region
  - the **facial nerve remains unaffected**, even in the presence of a very large tumor

- **Diagnosis**
  - clinical appearance, evolution
  - sialography
  - puncture biopsy
- **Treatment**
  - surgical
    - superficial subtotal parotidectomy
    - preservation of the facial nerve
    - total parotidectomy with nerve preservation
- **Evolution**
  - in **3-5%** of cases **malignant transformation** occurs
  - this frequency is higher in recurrences after inadequate surgery
- **Prognosis**
  - **good** in benign cases treated by superficial parotidectomy

Cystadenolymphoma (Warthin tumor)

- cystic tumor that generally develops at the **lower pole of the parotid gland**
- derives from segments of the salivary ducts included, during the embryonic period, in **intra- or extraglandular lymph nodes**
- histopathological examination shows a dense lymphoreticular stroma, with lymphoid follicles between the glandular epithelial areas
- **Symptoms**
- usually unilateral swelling, bilateral in 10%, recurrent, mobile, painless
- more common in **men** between 60 and 70 years of age
  - **Diagnostic**
    - ultrasound
    - uptake of technetium-99 on scintigraphy
    - histopathological examination
  - **Treatment**
- **surgery** – **superficial parotidectomy** with facial nerve preservation, or excision of the submandibular gland
- **Prognosis** – **good**; malignant transformation is rare

Malignant tumors

- 25-30% of salivary tumors are malignant
- **the main signs of malignancy:**
- **rapid growth or growth in spurts, except for adenoid cystic carcinoma, which grows very slowly**
- severe infiltration, sometimes skin or mucosal ulceration, reduced mobility of the tumor on the deep planes
- **cervical lymph node metastases**
- **facial paralysis** in parotid tumors

Malignant epithelial tumors

- Classification:
  - **tumors with acinar cells**
  - **mucoepidermoid tumors**
  - **carcinomas:**

    - adenoid cystic carcinoma (formerly cylindroma)
    - adenocarcinomas
    - Malpighian carcinoma
    - carcinoma on pleomorphic adenoma
- **the most common** are **mucoepidermoid tumors** and **adenoid cystic carcinoma**

Tumors with acinar cells

- **Diagnosis**
  - based on histopathological examination
- **Treatment**
  - **total parotidectomy**; the recurrence rate after limited surgery is very high
  - lymph node dissection – in the case of palpable lymph nodes
- **Evolution - Prognosis**
  - prognosis is **better than in carcinomas**
  - **metastases are rare**

Mucoepidermoid tumors

- **Etiology**
  - well-differentiated tumors with **low-grade malignancy**
  - frequent between 40 and 50 years of age, in the parotid and the palatal minor salivary glands
- **Diagnosis**
  - is based on histopathological examination
- **Treatment**
  - surgical, **total parotidectomy**, with lymph node dissection

Adenoid cystic carcinoma (Cylindroma)

- **Etiology**
  - arises from the **epithelium of the ducts and myoepithelial cells**, giving rise to cystic glandular cell islands and dense trabecular formations
  - the former name, **cylindroma**, led to **underestimation of its severity** and malignancy and should no longer be used
  - evolves through significant **local, diffuse, perivascular and perineural infiltration**
  - it also develops in the **aberrant minor salivary glands**, especially in the palate, less often in the submandibular and parotid glands
  - age: **35-50 years**
- **Symptoms**
  - **growth** is often **very slow**
  - pain – locoregional paresthesia
  - **facial paralysis – 25%** of cases in parotid tumors
  - extension to the base of the skull
  - **regional lymph node metastases**
  - distant, blood-borne metastases (lung, skeleton) in 20% of cases
- **Diagnosis**
  - symptoms
  - evolution
  - CT, sialography
  - histopathological exam

- **Treatment**
  - **radical surgery** – in the absence of lung or bone metastases there is hope of cure
  - **the facial nerve must be sacrificed** when the parotid is involved
  - **radiation therapy is controversial**
- **Evolution - Prognosis**
  - they generally have a **long evolution, over 10 years**
  - prognosis may be **aggravated** by
    - locoregional infiltration
    - early locoregional metastases and
    - distant metastases (lung, brain, bone)

Adenocarcinomas

- papillary and mucus-secreting carcinomas, deriving from the salivary duct system – infiltrating and destructive
  - pain
  - **facial paralysis**
- **cervical lymph node involvement** is common

Malpighian carcinoma

- infiltrative tumors with **rapid growth**

Carcinoma on pleomorphic adenoma

- **malignant transformation** occurs after a long latency period
- around **3-5%** of all pleomorphic adenomas
  - **facial paralysis**
  - regional lymph node metastases
  - preoperative extemporaneous histopathological exam
- **total parotidectomy** is performed, with cervical lymph node dissection

CHAPTER III — LARYNX PATHOLOGY

LARYNX ANATOMY AND PHYSIOLOGY

The larynx

- part of the respiratory system
- located in the upper part of the airways
- organ of phonation, able to produce voice
- **physiologically**, it is a **valve or sphincter** with a **triple function**:
- open valve in **breathing**
- partially closed valve, which produces **phonation** by modulating its opening
- closed valve that **protects** the tracheobronchial tree during deglutition

Laryngeal cavity:

- extends from the tip of the epiglottis to the lower edge of the cricoid cartilage
- continues with the trachea
- located in the visceral compartment of the neck
- corresponds to the anterior cervical triangle
- bounded superiorly by the hyoid bone, inferiorly by the sternal notch and laterally by the medial edges of the sternocleidomastoid muscles

Supraglottic region

- portion of the larynx above the glottis
- **Includes:**
- laryngeal aditus (opening between the larynx and pharynx)
- laryngeal ventricle (the space between the false and true vocal cords)
- false vocal cords
- laryngeal (or posterior) surface of the epiglottis
- arytenoid cartilages and laryngeal (or medial) surfaces of the aryepiglottic folds

Glottis

- Includes:
- true vocal cords
- anterior and posterior commissure
- glottic space, located between them

Glottis space

- space between the vocal cords anteriorly and the arytenoid cartilages posteriorly
- two regions:
- **intermembranous portion**, anteriorly
- **intercartilaginous portion**, posteriorly
- **is the narrowest portion** of the upper airways
- the vocal cords form the anterolateral edges of the glottic space and produce the voice
- **Reinke's space** extends along the free edge of the vocal ligaments and slightly superiorly on the surface of the vocal cords
- the vocal cords meet anteriorly at the anterior commissure
- the fibers of the vocal ligament pass through the thyroid cartilage at this level, piercing the underlying perichondrium and forming **Broyles' ligament**

Subglottic region

- extends **from 1 cm below the free edge of the vocal cords** to the lower edge of the cricoid cartilage
- walls – covered by respiratory mucosa, formed by the cricothyroid ligament superiorly and the cricoid cartilage inferiorly
- prolonged intubation, trauma or inadequate endoscopic maneuvers may cause subglottic and/or tracheal **stenosis**

Cartilaginous structures:

- epiglottis
- thyroid cartilage
- cricoid cartilage
- arytenoid cartilage
- the thyroid cartilage, cricoid cartilage and epiglottis are **unpaired**
- while the arytenoid cartilages are **paired**
- the larynx is suspended from the U-shaped hyoid bone by the thyrohyoid membrane (and ligaments) and the thyrohyoid muscles
- the **hyoid bone** (the only bone in the human body that does not articulate with other bones) is U-shaped, open posteriorly, and is attached to the mandible and tongue by the suprahyoid muscles

Laryngeal joints

- the most important joints of the larynx are:
- **cricoarytenoid**
- **cricothyroid**

Soft tissues

- The skeletal structures of the larynx are interconnected by **ligaments and fibrous membranes**
- The most significant **membranes** are:
- thyrohyoid
- cricothyroid
- cricotracheal
- quadrilateral
- cricovocal (or conus elasticus)

The laryngeal muscles are divided into intrinsic and extrinsic

- the **intrinsic muscles** are responsible for changing the length, tension, shape and position of the vocal cords
- as a result of these movements, the glottis opens during inspiration and closes during phonation and deglutition

Intrinsic muscles are:

- cricothyroid muscles
- lateral and posterior cricoarytenoid
- transverse and oblique interarytenoid
- aryepiglottic
- thyroarytenoid
- vocal

- thyroepiglottic
- Depending on their **main action**, the **intrinsic** laryngeal muscles can be divided **into 3 groups**:
- three main vocal cord **adductors** (thyroarytenoid, lateral cricoarytenoid and interarytenoid muscles)
- an **abductor** (posterior cricoarytenoid muscle)
- **tensor** (cricothyroid muscle)

Extrinsic muscles

- connect the larynx to the neighboring structures
- responsible for its **vertical displacement** during phonation and deglutition
- include the **infrahyoid muscles** (thyrohyoid, sternothyroid, sternohyoid and omohyoid) and the inferior constrictor muscle of the pharynx

Vascularization

- the **arterial** supply comes mainly from
- **superior laryngeal artery**, from the superior thyroid artery (branch of the external carotid artery)
- and
- **inferior laryngeal artery**, from the inferior thyroid artery (branch of the thyrocervical trunk, subclavian artery)
- **venous** drainage is carried out
- **superior thyroid vein**, to the internal jugular vein and
- **inferior thyroid vein**, to the brachiocephalic vein

Lymphatic drainage

Supraglottic region

- **has a rich, multilayered lymphatic network**
- responsible for bilateral and/or contralateral **lymph node metastases** commonly observed in supraglottic cancer
- drainage to the deep cervical lymph nodes **(levels IIA-III)** and then to the mediastinum **(level VI)**

Vocal cords

- is basically **a barrier** separating the two lymphatic regions, although posteriorly communication between them is practically free
- the glottis is **very poor in lymphatics**, so 95% of cancers at this level cause voice changes and airway obstruction, but **without lymph node metastases** at presentation

Subglottic region

- the capillary network **is not as rich as the supraglottic network, but denser than the glottic network**
- below the vocal cords, drainage is towards the lower deep cervical lymph nodes (**levels III-IV**) and partly through the nodes anterior to the larynx and trachea (**level VI**), one of which is almost constant – the **Delphian node**, located anterior to the cricothyroid or cricotracheal membrane

Innervation

- superior and recurrent laryngeal nerves, branches of the vagus nerve (X).
- **Superior laryngeal nerve**
- arises directly from the vagus nerve
- its internal branch (**sensory**) innervates the mucosa of the larynx
- its external branch (**motor**) innervates the **cricothyroid muscle**
- **Recurrent laryngeal nerve**
- ***on the right***
- arises from the vagus nerve and loops under the **subclavian artery**
- enters the larynx posterior to the cricothyroid joint
- ***on the left***
- arises from the vagus nerve and loops under the **aortic arch**
- runs in the tracheoesophageal groove, similarly to the right nerve
- **motor** innervation of ***all ipsilateral intrinsic laryngeal muscles***, **except** the **cricothyroid**, and **sensory** innervation of the mucosa below the level of the vocal cords

Physiology of the Larynx

- ***Breathing at rest***
- the glottic space is open (by **abduction** of the vocal cords)
- ***Deep breathing***
- the glottis is in **active and tense abduction** by contraction of the cricothyroid and cricoarytenoid muscles
- the glottic space widens considerably
- ***Sphincter function of the larynx***
- is intended to **protect** the airway
- participates in **important reflexes** that protect the airway against external stimuli and foreign bodies (glottic closure reflex):
- **laryngospasm** (in response to mechanical stimuli), **cough, apnea**
- **bradycardia, hypotension, glottic closure reflex**
- ***Phonation***
- requires several **complex mechanisms**, primarily adequate breathing to achieve optimal subglottic pressure
- is a complex phenomenon, **under cortical control**, which also depends on lung capacity, chest wall compliance, pharyngeal, nasal and oral anatomy and mental status

LARYNX CLINICAL EXAM

Examination

- laryngeal symptoms require a **full examination** of the head and neck, which must include:
- otoscopy, rhinoscopy, inspection of the oral cavity, oropharynx and nasopharynx
- palpation of the neck

Laryngoscopy

- **mirror** examination – introduced by Manuel Garcia in 1855
- **endoscopic** examination
- **clinical** examination and laryngoscopy – the initial assessment of the patient with laryngeal symptoms; it can visualize:
- the base of the tongue, the lingual and laryngeal surface of the epiglottis, the aryepiglottic folds

- arytenoids, true and false vocal cords

Laryngeal electromyography

- a set of **electrophysiological procedures** with a useful diagnostic, prognostic and therapeutic role in:
- laryngeal dysphonia
- paralysis of the vocal cords
- other neuro-laryngological disorders

LARYNX INFLAMMATORY DISEASES

- **Definition**
  - the term 'laryngitis' **does not refer** to a specific condition, but to **inflammation of the larynx, regardless of the cause**
- **Etiology**
  - the most common causes are:
  - infection
  - voice abuse
  - gastroesophageal reflux
  - laryngitis is usually **multifactorial**, and several causes can be identified
  - other etiological factors may be:
    - allergies
    - acute or chronic sinonasal infections
    - vomiting / aspiration
    - inhalation of irritants

Infection

In adults, infection might be:

- **Viral**
- **Bacterial**
  - streptococcus
  - staphylococci
  - pneumococcus
  - Haemophilus influenzae type B
- **Mycotic**
  - **candidiasis**, caused by: steroids, broad-spectrum antibiotics, diabetes, alcohol, prolonged intubation, caustic or infectious irritation of the larynx, immunosuppression
  - **aspergillosis**: severity depends on the immunocompetence of the patient
  - can mimic laryngeal cancer
  - **other rare mycoses**: blastomycosis, histoplasmosis, coccidioidomycosis and sporotrichosis
- **Tuberculosis**
  - the clinical aspect is varied:
  - simple hyperemia
  - monocorditis
  - exudative edema of the vocal cords
  - ulceration in different regions of the larynx
  - **can mimic laryngeal cancer**

- **Syphilis**
  - laryngeal involvement is rare and polymorphic
  - caused by Treponema pallidum
- **AIDS**
  - specific infections of the larynx, including tuberculosis and syphilis

In children, infection might be:

Viral

Acute edematous subglottic laryngitis

- is the ***most common form of laryngitis associated with dyspnea***
- occurs in children aged **1 to 3 years**
- is caused by ***myxoviruses, parainfluenza virus types 1 and 3, rhinovirus, adenovirus and echovirus***
- in infants it may be caused by ***respiratory syncytial virus***
- **bacterial etiology is rare**
- is more common in **autumn and winter**, although it can occur in any season, sometimes as a local epidemic
- **diagnosis is clinical**
- dyspnea sets in especially at night, with stridor and a barking cough
- **voice and deglutition are normal**
- fever is moderate, around 38°C
- treatment should be started as soon as possible: **oral corticotherapy**
- if symptoms persist, **emergency intubation** or **tracheotomy** may be required
- for prophylaxis, esophageal reflux, allergies and adenoiditis should be treated

Stridulous or spasmodic laryngitis

- is **the least severe form**
- probably caused by **reflux, allergy, inflammation, psychological factors**
- manifests as short attacks of dyspneic laryngitis, especially at night
- may be associated with a suffocating cough and cyanosis
- **remits spontaneously within minutes, up to 1 hour**
- relapses are common
- **oral corticotherapy** for a few days with air humidification in the bedroom
- may accompany **eruptive fevers**

Bacterial

Epiglottitis and subglottic laryngitis

- **in adults** it is caused by ***Haemophilus influenzae type B*** and is characterized ***not only by inflammation of the glottis, but also of the subglottic region***
- the situation improved with the **introduction of vaccination in 1992**
- affects children **between 3 and 6 years of age** and is preceded by rhinopharyngitis
- **dyspnea** sets in and the child takes a characteristic position, sitting and leaning forward, with the mouth open, and **cannot swallow saliva**
- the voice is muffled
- the general condition is altered and associated with anxiety
- emergency hospitalization is required

- **the effect of steroids is limited**
- **antibiotic therapy effective** against Haemophilus influenzae, e.g. cefotaxime, 200 mg/kg/day

Bacterial laryngotracheobronchitis, also called "pseudomembranous"

- **is rare, but severe**
- is caused by ***staphylococcus, Haemophilus influenzae, streptococcus and Branhamella catarrhalis***
- may complicate a subglottic laryngitis, with signs of sepsis and dyspnea due to obstruction of the larynx and trachea by pseudomembranes
- diagnosis is completed by endoscopy
- ***it is sometimes necessary to remove the pseudomembranes, which may be complicated by bleeding or toxic shock***
- relapse is usually present
- **intubation and targeted antibiotic therapy**, based on secretions sampled during endoscopy, are required

Specific infectious laryngitis

Diphtheric laryngitis

- is **extremely rare** today, but should nevertheless be considered in **migrants**
- caused by **Corynebacterium diphtheriae**
- the clinical picture includes:
- altered general condition
- tachycardia
- rough cough
- low voice

Pseudomembranous laryngitis

- is associated with **measles**
- dyspnea caused by **ulcero-necrotic or pseudomembranous laryngitis**

Allergy

- vasodilation and increased capillary permeability due to **histamine** cause swelling of the laryngeal mucosa
- **angioneurotic edema or Quincke's edema** is characterized by sudden swelling of the face and throat, which may also involve the larynx
- treatment includes:
- oxygen therapy
- epinephrine
- steroids
- antihistamines
- androgen derivatives

Toxic inhalations

Esophageal reflux

Vocal abuse (muscular tension dysphonia)

CHRONIC LARYNGITIS

Systemic disorders

Rheumatoid arthritis

- affects the larynx in 30% of cases
- in the **acute phase** it causes ***laryngeal congestion and edema***
- **chronic arthritis** causes ***reduced mobility through fibrosis and, occasionally, rheumatoid nodules***
- treatment is medical
- ankylosis with fixed vocal cords may require ***tracheotomy*** or ***endoscopic widening*** of the posterior region of the larynx

Lupus erythematous

- is an **autoimmune condition**, in which circulating immune complexes alter the vessels, connective tissue and mucosa
- the clinical signs are: **arthritis, skin rash, nephritis**

Recurrent polychondritis

- chronic painful inflammation affecting the **external ear, nose and larynx**
- progressive cartilage damage with **progressive laryngeal stenosis**
- treated with ***dapsone and steroids***

Pemphigus and pemphigoid

- are two distinct **autoimmune conditions** with the formation of **blisters and ulcers** in the skin and/or mucous membranes
- ***pemphigus affects the epithelium, and pemphigoid the subepithelial tissue***
- chronic pemphigoid may cause severe **laryngeal stenosis and ocular complications**
- treated with ***steroids, methotrexate and cyclophosphamide, or even plasmapheresis***

Sarcoidosis

- **is an idiopathic granulomatous disease**
- **dysphonia** occurs with the development of nodules in the submucosa
- ***local steroid injections*** may be useful in case of failure of systemic treatment

Wegener Granulomatosis

- is a **necrotizing granulomatosis** that affects the airway (8.5% in the trachea and subglottic region) and induces glomerulonephritis

Amyloidosis

- occurs through ***extracellular deposition of fibrillar proteins*** in the tissues
- ***primary and idiopathic or secondary to a systemic condition***, such as rheumatoid arthritis or tuberculosis
- is usually asymptomatic, although it can sometimes induce ***dysphonia or dyspnea*** requiring surgical treatment

- ***complete removal is difficult*** and ***local recurrence is common***
- a distinction is classically made between **"red" laryngitis**, in which inflammatory phenomena dominate, and **"white" laryngitis or keratosis**

Pseudomyxomatous laryngitis or Reinke's edema

- occurs particularly in **smokers** and is associated with **voice abuse**
- the lesions are located on the upper surface of the vocal cords, from the anterior commissure to the vocal process, sparing the lower surface and the ventricle
- vocal cords are **whitish, translucent**
- lesions are generally **bilateral**
- can be associated with eversion of the ventricle
- **dysplasia is generally rare**

White laryngitis has three types:

- **leukoplakia**
- **white pachydermia**
- **horny papilloma in adults**
- the **white color** is due to **surface keratinization**

Leukoplakia:

- is a flat, wax-like, ***opalescent or grey patch with poorly defined contours***
- can be single or multiple, localized or diffuse
- **adjacent mucosa is usually inflamed**

White Pachydermia:

- the lesion has a **pseudotumoral appearance**
- is gray, with clearly defined limits
- the surface is irregular and hard on palpation, and the suppleness of the vocal cords is altered

The following aspects are suggestive of malignant transformation:

- **irregular surface**
- **vocal cord induration**
- **ulceration, hemorrhage**
- **significant inflammation of the surrounding area**
- **microcapillary abnormalities**

Etiological factors

- **Smoking:**
- is an important causal factor in their development, especially in the case of keratinization of the mucosa
- the risk of dysplasia is **7 times higher** in a smoker
- **Asbestos:** studies are contradictory; however, the influence of asbestos is combined with that of tobacco
- **Other factors:**
- **nickel, sawdust, chromium, arsenic, sulphuric acid**
- **Alcohol:**
- acts **synergistically and enhances the effect of tobacco**
- **can act as a solvent, favoring penetration of tobacco carcinogens into the epithelium**

- in case of massive and prolonged ingestion, alcohol favors **nutritional deficiencies, especially of vitamin A,** which has a protective role for the mucosa
- **Esophageal reflux:** its role is still **controversial**
- gastroesophageal reflux causes inflammatory reactions, which may induce cellular changes (metaplasia, keratinization)
- There are **several classifications** that grade these changes, based on the **alteration of tissue architecture and cellular morphology**

The most frequently used is the three-grade classification:

- **Grade 1**: **mild dysplasia** with/without hyperplasia and/or keratosis
- **Grade 2**: **moderate dysplasia**
- **Grade 3**: **severe dysplasia** or **carcinoma in situ**

Carcinoma in situ:

- can be defined as an entity that has the **morphological characteristics of a cancer**
- its extension is limited to the site of origin
- the **distinction between carcinoma in situ and microinvasive carcinoma is difficult**
- the most widely used criterion is involvement of the basement membrane

Hypertrophic laryngitis

- **subepithelial cordectomy (type 1) or decortication** is indicated, according to the classification of the European Laryngological Society
- depending on the degree of severity, the entire epithelium may be affected, thereby requiring complete resection to avoid leaving a dysplastic or cancerous area
- subepithelial cordectomy allows examination of the entire epithelium, its role being mainly diagnostic
- ***if histopathological examination confirms the presence of hyperplasia or dysplasia, its role is also therapeutic***
- **invasive carcinoma** – the technique is **not sufficient**
- **carcinoma in situ** (where microinvasion is excluded) – this type of cordectomy **may be sufficient**, with clinical and endoscopic follow-up

GLOTTIC MUCOSA DISEASES

Glottis symptoms:

- the most common symptom of laryngeal diseases is **dysphonia**
- voice abuse
- cough
- inhalation of irritants
- smoking
- crying
- **inappropriate use of the voice** refers to **speaking at high intensity or with an altered pitch**
- clinical examination:
- vocal nodules
- voice polyps
- Reinke's edema
- cysts
- treatment should include **voice training of the patient**

Vocal nodules

- are **bilateral, chronic thickenings of the epithelium of the vocal cords**
- usually located at the junction of the anterior and middle thirds
- develop more frequently in women, boys and people with professions requiring sustained vocal effort (**teachers, singers, vendors,** etc.)
- also appear in **children with cleft palate**
- nodules are linked in particular to **improper use of the voice**
- the main symptom is **permanent or intermittent dysphonia**, sometimes associated with non-specific pharyngeal symptoms
- singers have **difficulty producing high notes**
- need to cough
- on laryngoscopy, the vocal nodules are:
- **bilateral, white, broad-based spicules**
- **at the junction of the anterior and middle thirds of the vocal cord**
- **Treatment**
- rehabilitation and voice hygiene (hydration)
- surgery

Vocal polyps

- **benign lesions**, usually located on the free edge of the vocal cord, preventing proper apposition of the vocal cords during adduction
- **larger than nodules,** with a **broad or pedunculated base**; may be associated with a contact lesion on the contralateral vocal cord
- occur mainly in adult males, the risk factors being voice abuse and a noisy work environment
- other etiological factors are aspirin consumption, anxiety, extroverted behavior and excessive use of the sphincter function of the larynx (weight lifting, glass blowing, etc.)
- the consequence of a short, violent trauma caused by high voice intensity, with rupture of the capillaries in the submucosa and subsequent hemorrhage
- if voice abuse persists, the initial lesion evolves into a **hemorrhagic polyp** and, over time, a **fibrous polyp**
- dysphonia usually has a sudden onset, coinciding with voice abuse
- **Treatment** is:
- by sectioning the pedicle and sparing the vocal ligament
- endoscopic laser ablation
- **medical treatment is usually ineffective**
- in case of gastroesophageal reflux, anti-reflux measures will be taken

Reinke's edema

- accumulation of **unilateral or bilateral edema** in Reinke's space
- the mobility of the vocal cords is preserved
- patients have a **deepened voice**
- chronic irritation of the larynx causes edema, vascular congestion and capillary stasis in the submucosa
- other causes are ***chronic sinusitis, hypothyroidism and exposure to environmental or industrial irritants***
- patients with Reinke's edema are usually **smokers**, with **vocal abuse** and **a long history of dysphonia**

- the laryngoscopic appearance is typical, with fusiform edema of both vocal cords; a yellow fluid can sometimes be seen through their mucosa
- voice improvement is slow and should be monitored by indirect laryngoscopy
- Surgical treatment consists of:
- incision on the upper surface of the vocal cord
- aspiration of the fluid from Reinke's space
- excess mucosa should be removed

Vocal cord cysts

- **white, subepithelial, spherical, unilateral**
- located in the middle third of the vocal cord
- present in approximately 5% of patients with voice alterations
- vocal cord cysts may be **congenital** or **acquired**
- the symptoms are similar to those produced by vocal nodules
- **dysphonia** is the most constant clinical symptom and improves after spontaneous emptying of the cyst
- **epidermoid cysts** are:
- larger, ovoid, yellow, sometimes bilateral, with more marked dysphonia
- **Treatment:**
- surgery is the treatment of choice: **superior cordotomy** and **cyst dissection**
- **corticosteroid injections** into the vocal muscle are recommended to avoid postoperative inflammation and adhesions between the epithelium and the vocal muscle
- **phoniatric therapy**

LARYNX BENIGN TUMORS

Histopathological classification of benign laryngeal tumors

Epithelial

- squamous epithelium
- recurrent respiratory papillomatosis
- keratinized papilloma
- glandular
- pleomorphic adenoma
- oncocytic tumor

Non-Epithelial

- **Vascular**
  - hemangioma
  - lymphangioma
- **Cartilage and bone**
  - chondroma
  - osteoma
  - giant cell tumor
- **Muscle**
  - leiomyoma
  - rhabdomyoma
  - angiomyoma

  - epithelioid leiomyoma
- **Fat:** lipoma
- **Nerve**
  - neuroma
  - neurofibroma
  - schwannoma
  - paraganglioma
- **Pseudotumors**
  - fibroid
  - fibroblastic inflammation
  - amyloid
- **Laryngeal cysts**

Papilloma

Keratinized papilloma (keratosis papilloma)

- occurs mainly in **adults**, as a **solitary lesion** arising from the true vocal cord
- most keratinized papillomas are **not related to viral infection**; they are associated with **smoking** and have potential for **malignant transformation**
- recurrence after simple excision is variable

Recurrent respiratory papillomatosis (RRP) or non-keratinized papilloma

- is the **most common benign laryngeal tumor**
- the incidence is estimated at 4 per 100,000 in children and 2 per 100,000 in adults
- its origin is **viral**, being caused by **human papillomavirus (HPV)**
- more than 90 HPV subtypes have been identified, the most common being **HPV-6 and HPV-11**
- RRP occurs in response to HPV infection of the mucosa and may occur at any level of the upper aerodigestive tract
- **malignant transformation occurs in 1-7%** of patients and is associated with:
- infection with HPV-11, HPV-16 and HPV-18 subtypes
- anterior irradiation
- **Diagnosis**
- **in adults**, the most common symptoms are **dysphonia** and sometimes mild **dyspnea**
- **in children** the following appear:
- progressive dyspnea
- stridor
- respiratory failure
- macroscopically, papillomas are:
- **pink or whitish**
- pedunculated or sessile, with a broad base
- **Surgical techniques**
- surgical options in the treatment of RRP include:
- microsurgery with cold instruments
- CO2 laser / microdebriders / angiolytic laser treatment in the outpatient setting
- adjuvant therapy
- cidofovir
- α-interferon
- indol-3-carbinol

Hemangioma

- **rare but important tumor**, with possible obstructive manifestations in the airways
- hemangiomas are of **two types**: **pediatric (10%)** and **adult (90%)**
- **the pediatric form** is typically **subglottic**. Its pathogenesis is controversial; it is considered a congenital anomaly, although its aggressive behavior sometimes suggests a malignant neoplastic lesion
- **in the adult form**, the lesions are located at or **above the vocal cords**, and patients present with dysphonia and occasionally dysphagia
- these tumors are more common in males (60-70%); most are cavernous or capillary
- multiple tumors may occur in syndromes such as **Rendu-Osler-Weber and Sturge-Weber disease**; in this case **the term hemangiomatosis** is used
- **Treatment:**
- surgical excision, with the risk of hemorrhage during biopsy or excision
- laser therapy for vascular lesions – Nd:YAG laser photocoagulation, CO2 lasers
- **temporary tracheotomy**

Chondromas

- laryngeal chondromas **are rare**
- occur mainly in the **6th and 7th decades of life**
- the **cricoid cartilage** is most commonly involved, then the **thyroid cartilage** and sometimes the **epiglottis**
- the treatment and prognosis of slow-growing chondrosarcomas are practically the same
- most authors recommend **conservative techniques** for slow-growing tumors
- **radiation therapy** for these conditions is controversial

Amyloidosis

- **isolated amyloidosis (without plasmacytoma)** occurs frequently along the false vocal cords, although any part of the larynx may be affected
- the term "amyloidosis" denotes an ***extracellular accumulation of homogeneous, eosinophilic, fibrillar protein-derived material with well-defined histochemical characteristics***
- the amyloid tumor is firm, waxy, translucent on cross-section, ranging from **brown-yellow to red-gray**
- it is important that patients diagnosed with amyloidosis are investigated, because they may have **amyloid tumors**:
- **primary systemic amyloidosis**
- **secondary amyloidosis**
- **amyloid tumors:**
- **primary systemic amyloidosis**
- diagnosed by serum and urine immunoelectrophoresis
- rectal biopsy
- **secondary amyloidosis**
- associated with other predisposing conditions such as **plasmacytoma, solitary or part of multiple myeloma**
- **Surgery**
- partial laryngectomy
- microlaryngoscopic excision of the lesion
- CO2 laser excision

Laryngeal cysts

- their location depends mostly on the **site of origin and the structural type** (saccular, retention/inclusion, ductal, vascular or traumatic)
- can be considered **internal or external**, depending on the degree of compression or their extension in the larynx

Laryngoceles

- air-filled herniation or saccular dilation, **external or internal**

Saccular cysts (anterior or lateral)

- submucosal; contains mucus deposits and inflammatory elements
- may appear anywhere in the larynx
- classified as: **congenital, retention, inclusion**

LARYNX MALIGNANT TUMORS

Definition:

- **95% are squamous cell carcinomas**
- other **primary** malignancies are
  - tumors of the salivary glands
  - neurogenic tumors
  - chondromas
- other malignancies may **metastasize** to the larynx (**secondary**)
  - malignant melanoma
  - hypernephroma
  - general systemic tumors
  - non-Hodgkin lymphoma

Etiology/epidemiology

- **laryngeal cancer is common**
- currently accounts for 3% of male cancer cases in Europe
- incidence is higher in men than in women **(6:1)**
- the incidence in women is increasing due to the increased prevalence of smoking and alcohol consumption
- appears in the **second half of life,** with a peak incidence in the fifth and sixth decades (on average, 55 years for men and 60 years for women)
- **genetic predisposition** continues to be investigated
- genotypic and phenotypic deficiencies in the metabolism of tobacco and other carcinogens, as well as abnormal DNA repair mechanisms, may also play an etiological role

Symptoms

- Patients with laryngeal cancer frequently have a history of:
  - cervical pain (odynophagia)
  - dysphonia
  - dysphagia
  - reflex otalgia
  - a cervical tumor mass
- ***dysphonia*** is a common symptom of glottic carcinoma
- ***cervical foreign body sensation*** occurs in patients with supraglottic and/or glottic cancer

- ***progressive dysphagia***, initially for solid foods and later for liquids, is commonly observed in patients with supraglottic tumors
- ***weight loss*** may occur due to pharyngeal invasion in more advanced stages
- ***palpable lymph nodes*** are common at the time of diagnosis, more so in supraglottic than in glottic cancers

Diagnosis

Clinical exam

- ***indirect laryngoscopy*** remains the basic diagnostic procedure and, when necessary, is supplemented by examination with a rigid endoscope or fiberscope
- ***fiberoptic endoscopy*** is especially useful in patients with exaggerated reflexes
- patients should be evaluated both **during breathing** and **during phonation**
- particular attention should be paid to ***indirect signs*** such as edema or mucosal erythema, which may suggest deeper hidden processes; in this case examination by ***direct laryngoscopy*** and possibly ***biopsy*** is recommended
- ***cervical palpation*** is essential in determining lymph node involvement
- the size, site and mobility of the lymph nodes must be well documented
- pain on mobilization of the ***thyroid cartilage*** suggests tumor extension into the surrounding structures
- ***Direct pharyngolaryngoscopy***
- direct endoscopic examination of the pharynx, larynx and esophagus is carried out **under general anesthesia**
- is an indispensable method of evaluation and has several objectives:
- **multiple biopsies**
- accurate assessment of superficial tumor extension
- systematic examination of the entire upper aerodigestive tract and esophagus for the detection of **synchronous cancers**

Imaging Diagnosis

- imaging techniques are increasingly accurate and are now commonly used to complement the clinical and endoscopic assessment of patients with laryngeal carcinomas
- **CT is currently the most useful imaging technique**, especially helical CT, which is simple and fast and also allows multiplanar reconstruction

Treatment

Surgery

- **in the early stages** of supraglottic and glottic cancers, endoscopic surgery may be used with good results, with both the CO2 laser and classical approaches
- **in more advanced stages**, the decision and the surgical approach, either endoscopic or external, will be based on the expertise of the practitioners
- some patients may be referred directly for ***radiochemotherapy***
- for the **treatment of glottic cancers:**
  - cordectomy
  - frontolateral laryngectomy
  - frontal anterior laryngectomy with epiglottoplasty
- for the **treatment of supraglottic cancers:**
  - supraglottic horizontal laryngectomy

  - partial supracricoid laryngectomy with cricoidopexy
- tumors that **massively involve the larynx**
  - total laryngectomy

Radiotherapy

- the radiotherapy sources are either cobalt-60 or 4-6 MV linear accelerators with electron beams
- conventional treatment consists of one **fraction** of 2 Gy per day, 5 days a week, for a total of 65-70 Gy
- it may also be **hyperfractionated** (2 fractions per day) for a total of 70-80 Gy

Chemotherapy

- the most commonly used cytostatics are **cisplatin** (or carboplatin as a substitute), **5-fluorouracil** or, for palliation, **methotrexate**
  - there are three treatment options:
    - **induction chemotherapy**
    - **adjuvant chemotherapy**
    - **concomitant chemotherapy**

Laryngeal preservation

- There are three ways to **avoid total laryngectomy**:
- **1. Induction chemotherapy,** then CT examination, followed by **radiotherapy** in patients who respond well
- randomized studies conducted on this basis have shown that ***survival is not affected and that about 60% of cases*** might be treated in this way
- **2. Simultaneous chemoradiotherapy**
- randomized trials (RTOG) show a higher rate of larynx preservation with this strategy, but also ***higher mucosal toxicity***, while ***survival remained unchanged***
- **3. Induction chemotherapy followed by concurrent chemoradiotherapy** in patients who responded well
- ***the strategy is being evaluated***; the results are promising

TNM Staging

- after clinical, pathological and imaging examination, the tumor is staged, which is important for the treatment decision and prognosis
- in laryngeal cancer the TNM staging also takes each laryngeal level into account

Primary tumor (T):

- Tx – primary tumor **cannot be evaluated**
- T0 – primary tumor **cannot be identified**
  - Tis – carcinoma in situ

Supraglottic:

- T1 – tumor limited to one subsite of the supraglottis, **normal mobility of the vocal cords**
  - T2 – the tumor invades the mucosa of more than one supraglottic subsite, with **decreased mobility of the vocal cords**
    - T3 – the tumor is limited to the larynx, **with fixation of the vocal cords**, and may invade the surrounding structures (retrocricoid area, pre-epiglottic space, paraglottic space) or the thyroid cartilage

    - T4a – **moderately advanced local tumor**: invades surrounding tissues such as the trachea, extrinsic muscles of the tongue, esophagus, thyroid gland
- T4b – **very advanced local tumor**: invasion of the prevertebral space, mediastinum or carotid artery

Glottic:

- T1 – tumor limited to the vocal cords, with or without involvement of the commissure, **normal mobility of the vocal cords**
    - T1a – a single vocal cord
    - T1b – both vocal cords
- T2 – **decreased mobility of the vocal cords**; local extension to the supraglottic or subglottic levels
- T3 – the tumor is limited to the larynx, with **fixation of the vocal cords**; it may invade the thyroid cartilage or paraglottic space
- T4a, T4b – **moderately and very advanced local disease, respectively** (as for the supraglottic level)

Subglottic:

- T1 – tumor located exclusively in the subglottic area, **normal mobility of the vocal cords**
- T2 – extension to the vocal cords, **decreased mobility of the vocal cords**
- T3 – **tumor with fixation of the vocal cords**, limited to the larynx
- T4a, T4b – similar to the lesions **described above**

Lymph nodes (N):

- Nx – Regional lymph nodes **cannot be assessed**
- N0 – **no regional** lymph node metastases detected
- N1 – one involved lymph node, on the same side as the tumor (**ipsilateral**), with a maximum diameter of **<3 cm**
- N2a – metastasis in a single **ipsilateral** lymph node, with a diameter of **>3 cm, but <6 cm**
- N2b – metastases in **several ipsilateral** lymph nodes, all with a maximum diameter of **<6 cm**
- N2c – **bilateral or contralateral** metastases, in lymph nodes with a maximum diameter of **<6 cm**
- N3 – an involved lymph node with a diameter **> 6 cm**

Metastases (M):

- M0 – **absent**
- M1 – **present**

Stage	T, N, M
0	Tis N0 M0
I	T1 N0 M0
II	T2 N0 M0
III	T3 N0 M0; T1/T2/T3 N1 M0
IVA	T4a N0/N1/N2 M0; T1/T2/T3 N2 M0
IVB	T4b any N M0; any T N3 M0
IVC	any T, any N, M1

Prognosis

- **1. T1-T2**
    - local control of **more than 85%**

  - five-year survival for **supraglottic cancers** is around **70%** and for **glottic cancers** about **80%**
- **2. T3**
  - local control **over 80%**
  - five-year survival for supraglottic cancers is around **60%** and for glottic cancers about **70%**
- **3. Resectable T4**
  - local control **over 66%**
  - five-year survival for supraglottic cancers is around **40%**, and for glottic cancers about **50%**
- **4. Non-resectable T4:**
  - local control and 5-year survival **below 30%**

LARYNGOTRACHEAL TRAUMA

Definition

- trauma to the larynx and trachea is the heterogeneous consequence of perforating or endoluminal injuries affecting the head, neck and mediastinum

Etiology/epidemiology

- laryngeal or tracheal trauma may result from internal (endoluminal) action or from external forces
- ***endolaryngeal and endotracheal (endoluminal) lesions*** are caused by **intubation**, most commonly producing:
  - soft tissue hematoma
  - lacerations, perforations and ruptures
  - dislocation of arytenoid cartilages
- **Trauma** caused by **endotracheal intubation for anesthesia** or by treatment of the respiratory tract includes:
  - intubation granulomas
  - post-intubation cysts
  - inoculation of pigment into the mucosa
  - synechiae between the true or false vocal cords
  - stenosis/scarring of the posterior wall of the larynx
  - subglottic stenotic web
  - some form of laryngotracheal or tracheal stenosis
  - partial or total destruction of the larynx and/or trachea
- ***additional causes*** of endoluminal trauma include:
  - ingestion of xenobiotics (e.g. caustics, acids, gases)
  - endolaryngeal surgery
  - other surgical and diagnostic procedures
  - **external trauma** is heterogeneous in terms of mechanism of action and consequences
    - endoluminal lesions of soft tissues
    - fractures of the thyroid cartilage / cricoid / trachea
    - ruptures of the laryngeal, laryngotracheal and tracheal ligaments
    - combined fractures and ruptures with soft tissue damage
  - **external perforating trauma** can lead to:

        - laceration of the larynx, trachea and laryngeal nerves, as well as of other cervical structures
        - aspiration, gas embolism and pneumothorax
        - bleeding from major vessels
        - fistula - tracheoarterial and tracheoesophageal
    - **other causes** of external laryngeal and tracheal trauma are:
        - coniotomy
        - tracheostomy
        - dilative tracheotomy
        - partial resections
        - surgical and diagnostic procedures

The symptoms of laryngotracheal trauma vary greatly depending on the cause and severity

- common indicators of laryngeal and/or tracheal trauma may be:
    - hoarseness
    - dyspnea, cyanosis
    - stridor
    - crepitation
    - hemoptysis and cough attacks
    - soft tissue emphysema
    - hematoma
    - strangulation marks
    - pain, fear, shock
    - open ruptures and fractures, sprains, dislocations
    - bleeding, aspiration, hypoxia
    - cardiorespiratory arrest
- obviously, the history of trauma and the onset of symptoms can clarify the diagnosis in cases of endoluminal trauma

Complications

- scarring and stenosis
- impairment of voice, swallowing and breathing

Diagnosis

- **standard diagnostic steps:**
    - detailed history
    - inspection
    - palpation
    - endoscopy
    - microlaryngoscopy and tracheobronchoscopy
- **additional/useful diagnostic procedures:**
    - clinical examination – auscultation and percussion of the chest
    - X-ray of the chest and neck
    - CT scan - thorax and neck
    - high-resolution CT scan
    - monitoring blood oxygen saturation
    - pulmonary function testing
    - MRI in children

- fluorescence MRI for functional evaluations

Treatment

- **conservative treatment**
  - therapeutic stages:
    - ensuring respiratory and circulatory functions
    - anti-inflammatory drugs (steroidal and non-steroidal)
    - painkillers
- **additional measures / useful therapeutic strategies**
  - locally applied cold
  - inhalations
  - voice and swallowing therapy
- **surgical treatment**
  - **proposed surgical procedures:**
    - endolaryngeal microsurgery
    - endoscopic surgery of the trachea
    - endo-extraluminal repositioning and fixation of fragments
  - **additional useful surgical strategies:**
    - procedures for enlarging the glottic space
    - procedures for increasing subglottic space
    - resections of tracheal and tracheolaryngeal segments
    - temporary tracheostomy
    - tracheal and/or tracheolaryngeal stents

Prognosis

- in **severe laryngotracheal trauma**, the large cervical blood vessels, intrathoracic organs, and respiratory and circulatory functions may be compromised, and in these major emergencies death can occur rapidly
- in the management of these situations, **maintenance of vital functions** is the first step
- for patients who **survive the initial shock**, an exact diagnosis, based on an understanding of the action of the causative agent, and promptly applied treatment are the only solution
- **long-term therapy** will focus on:
- effective breathing
- decannulation
  - the quality of the voice
  - they depend very much on the initial and long-term treatment

LARYNGEAL AND TRACHEAL STENOSIS

Supraglottic laryngeal stenosis

- **Ethology**
  - 1. external trauma (accident)
  - 2. toxic agents - chemicals (bases, acids, insecticides)
  - 3. infections
    - tuberculosis
    - scleroma
    - diphtheria
    - syphilis

        - typhus
        - mycoses
        - scarlet fever
    - 4. Boeck sarcoidosis
    - 5. pemphigus
    - 6. Wegener granulomatosis
    - 7. amyloidosis

Symptoms

- in the **onset phase** (or in mild cases) the symptoms are ***dysphonia and dyspnea***; after progressive narrowing of the lumen there is loss of voice (***aphonia***) or severe respiratory difficulty (***dyspnea***) at rest

Treatment

- **1. Conservative treatment**
- respiratory and circulatory functions may be maintained by:
    - oxygen / • humidification / • steroids
    - inhalations / • analgesics
- **2. Surgical treatment**
- a) external trauma
    - **acute:** depending on severity, tracheotomy or thyrotomy / debridement should be performed
    - **chronic:** in the case of scarring with destruction of the laryngeal inlet, supraglottic laryngectomy or laryngeal reconstruction
- b) internal trauma
    - local damage leading to severe edema with exudate – **emergency tracheotomy**

Glottic laryngeal stenosis

Anterior commissure synechia

- **Definition:**
- **Congenital:**
    - anterior commissure synechia is ***one of the mildest forms*** of congenital malformation
    - consists of a thin membrane that extends into the subglottic area
    - the intervention required depends on the extent of the membrane
    - due to ***failure of resorption of the proliferating epithelium in the intrauterine period, at 7-8 months***
- **Acquired:**
    - tuberculosis / syphilis / scleroma / diphtheria / typhus / scarlet fever
    - mycoses / pemphigoid / sarcoidosis Boeck / amyloidosis
    - different types of external trauma to the thyroid cartilage
    - surgery (partial laryngectomy), endolaryngeal microsurgery
    - prolonged intubation
    - **Symptoms:**
    - **dysphonia and dyspnea**
- **Treatment:**
    - **1. Conservative treatment**
        - acute – fibrin coating of de-epithelialized areas to prevent adhesions

        - mitomycin C
    - **2. Surgical treatment**
        - division with the laser or microscalpel
        - **tracheotomy** is only necessary in severe cases, when dyspnea is severe

Interarytenoid scar bridge

- **Definition:**
- lesion that fixes the **vocal cords in the paramedian position**
    - **congenital:** rarely occurs
    - **acquired:** in almost all cases after prolonged intubation or as a consequence of laryngeal surgery
- treatment without tracheotomy is possible by division of the scar bridge using the laser or microscalpel

Bilateral cricoarytenoid ankylosis

- **Definition:**
- ankylosis of the cricoarytenoid joints causes the **vocal cords to be fixed, usually in the paramedian position**
- due to prolonged intubation, or chronic in common synovial diseases
    - **dyspnea** if both joints are involved
    - **dysphonia** if one joint is involved or there is dislocation
- **1. conservative treatment**
- **2. surgical treatment**

Subglottic laryngeal stenosis (Cricotracheal stenosis)

- **Definition**
    - subglottic laryngeal stenosis (cricotracheal stenosis) is a pathological condition that causes narrowing of the laryngeal lumen in the subglottic or cricotracheal portion, manifested clinically by **moderate to severe dyspnea**
- **Etiology**
- **congenital:** malformations of the cricoid cartilage
- **acquired:** in adults, most often after prolonged intubation
- **Treatment**
- **1. Conservatory:**
- conservative treatment is **NOT successful**
- mitomycin C
- **2. Surgical**
- **congenital:** laser surgery and dilation
- **acquired:** laser surgery / open procedure

NEUROLOGICAL DISEASES OF THE LARYNX

- the larynx has two major functions
    - **sphincter** to close the lower airways during swallowing, preventing the aspiration of food and saliva
    - **phonate organ**
- laryngeal function is based on the corresponding motor efferents and sensory afferents
- the X nerve provides innervation of the larynx and pharynx

- the X nerve leaves the skull through the jugular foramen, then gives off the pharyngeal, superior laryngeal and recurrent laryngeal nerves
- the **pharyngeal nerves** enter the pharynx and form the **pharyngeal plexus**, which innervates the striated muscles of the pharynx and soft palate, **except** the tensor veli palatini and stylopharyngeus muscles, which are innervated by nerves V and IX
- the **superior laryngeal nerves** innervate the **cricothyroid muscles** and the **inferior constrictor muscle**
- the **recurrent laryngeal nerves** innervate **all the intrinsic muscles** of the larynx **except the cricothyroid muscles**
- the **parasympathetic fibers** of the X nerve are distributed to the smooth muscles of the airways, contributing to breathing, producing bronchoconstriction or regulating the peristalsis of the esophagus
- *sensation of the vocal cords* and of the *lower laryngeal level* (below the vocal cords) is provided by the **sensory nerve fibers of the recurrent laryngeal nerves**
- *sensation of the larynx above the vocal cords* is provided by **sensory fibers from the superior laryngeal nerves**

Paralysis – **complete loss of voluntary motor function** due to neural or muscle problems

Paresis – incomplete loss of motor function (reduced function)

- Nerve lesions are classified by:
  - morphological characteristics
  - functionality
  - possibility of spontaneous regeneration
- EMG (electromyography) makes a special contribution to the classification

Etiology / epidemiology

- vocal cord paralysis may be the result of:
  - **central nerve damage**
    - CNS – nucleus ambiguus or supranuclear tract
  - **peripheral**
    - vagus nerve trunk or its branches: superior laryngeal nerves or recurrent laryngeal nerves
  - **thyroid surgery** is **the most common cause** of laryngeal nerve paralysis
  - **surgery of the cervical spine** by an anterior approach, and surgery of the larynx, pharynx, cervical esophagus, upper mediastinum and carotid artery may result in damage to the laryngeal or vagus nerves

Symptoms

- loss of mobility of the vocal cords (abduction – adduction)
- disturbance (incoordination) of food bolus transport during swallowing
- **phonation and swallowing are affected; aspiration of food, fluids and saliva into the trachea is possible**
- **unilateral paralysis**, due to a lesion of the vagus nerve or its recurrent branch, is manifested by ***impaired vocal cord abduction***, or even ***complete immobility in the paramedian position***
  - **hoarseness**
  - **weak cough**
- **bilateral recurrent paralysis** fixes the vocal cords in the paramedian position, causing a ***functional laryngeal stenosis***
  - **dyspnea (sometimes mild, sometimes severe)**

- **inspiring stridor**
- **respiratory distress even without effort**
- **dysphagia**

Diagnostic steps

- detailed history (previous surgical interventions?)
- inspection
- palpation
- rigid and/or flexible endoscopy
- thyroid gland investigations
- ultrasound examination of the cervical region

Additional/useful diagnostic procedures:

- stroboscopy
- microlaryngoscopy, pharyngo-esophagoscopy and tracheobronchoscopy
- CT examination of the thorax and skull base
- pulmonary function tests
- voice analysis
- laryngeal electromyography
- barium transit with fluoroscopy
- MRI studies of the brain
- laboratory tests

Treatment

- **conservative treatment**
  - corticosteroids
  - nonsteroidal anti-inflammatory drugs
  - antiviral
  - antibiotics (oral) if an infection is suspected
  - logopedic support
- **surgical treatment**
  - unilateral vocal cord paralysis very rarely requires surgical treatment to improve the voice
  - arytenoidectomy
  - cordectomy
  - posterior cordotomy
  - temporary lateral fixation of the vocal cords, described by Lichtenberger
  - endoscopic laser procedures

Bilateral vocal cord paralysis

- **Definition**
- vocal cord paralysis is a pathological condition that causes the vocal cords to be fixed in a **paramedian position**
- **Etiology**
- congenital
- **surgery** (cervical, thoracic, cardiac, brain stem, spinal cord, etc.)
- **infections**, mostly viral infections
  - **degenerative processes**

- **tumors** (brain, mediastinum, thoracic, pulmonary, esophageal)

- **Treatment**
- 1. **Conservative treatment:** as a rule it is **NOT effective**
  - botulinum toxin injection into the cricothyroid muscles reduces the tension of the vocal cords; their relaxation increases the glottic lumen
- 2. **Surgical treatment:**
  - arytenoidectomy
  - submucosal cordectomy
  - posterior cordotomy
  - submucosal resection of the anterior third of the arytenoid cartilage and partial submucosal resection of the medial and lateral thyroarytenoid muscles

LARYNGOTRACHEAL FOREIGN BODIES

Clinical symptoms

- penetration of a foreign body into the respiratory tract may produce **severe acute respiratory failure**, which, if prolonged beyond **6 minutes**, causes **irreversible** damage to the **central nervous system**
- once it has entered the airways, the foreign body will cause **disturbances of air passage**, as well as a series of immediate and late bronchial reactions, depending on the nature of the foreign body, its size and its ability to swell or release toxic substances
  - the clinical picture is characteristic and has three phases:
    - **acute onset phase**
    - **quiet phase**
    - **late phase**

Acute stage

- **the most characteristic**, and the one with the greatest clinical significance in establishing the diagnosis
- ***noisy symptoms, revealing the penetration of the foreign body into the airways***
  - sudden feeling of suffocation
  - explosive cough
  - fear / agitated patient
  - the face is congested at first, then becomes cyanotic
  - breathing is difficult
- asphyxia seems imminent
- if the foreign body is large and completely obstructs the airway, **sudden death by asphyxia** may occur
- after this onset phase, a **silent phase** follows after about **30 min.**

The most characteristic

- 1. laryngeal foreign bodies
- large foreign bodies, which cannot pass through the glottis
- sharp foreign bodies, which become lodged in the mucosa of the larynx (fish bones)
- indirect laryngoscopy can reveal the endolaryngeal foreign body, especially those lodged in the laryngeal mucosa
- after a period of time, local edema and inflammation may set in
- 2. tracheal foreign bodies

- tracheal foreign bodies are located in the trachea, where they can be **mobile or fixed**
- after the initial period, the clinical symptoms improve; they may recur as paroxysmal cough and dyspnea when the foreign body moves
- tracheal foreign bodies, especially mobile ones, carry a risk of subglottic or tracheal impaction leading to asphyxia
- on interscapular auscultation – a characteristic sound – the **"flag-flapping sound"**
- **3. bronchial foreign bodies**
- are located in the bronchi
- usually in the **main bronchi**, and can be fixed or mobile
- obstruction of a main bronchus leads to **massive atelectasis** (abolished vesicular murmur, dullness, mediastinal shift, immobility of the hemithorax)
- obstruction of a **lobar bronchus** leads to **partial atelectasis**
- impaction acting as a **valve** causes **pulmonary emphysema**
- bronchial foreign bodies can sometimes be **well tolerated** in the tracheobronchial tree and discovered late, when **bronchopulmonary complications** occur
- bronchiectasis
- bronchial asthma
- pulmonary abscess, etc.

Diagnosis

- the diagnosis of tracheobronchial foreign bodies is based on the clinical signs described above, radiological examination and tracheobronchoscopy
- radiological examination plays an important role in the diagnosis, including through **indirect signs** of bronchial and pulmonary obstruction

Treatment

- **rigid** bronchoscopes with special **magnifying optics**
- in some medical centers **flexible** bronchoscopy is used

CHAPTER IV — EAR PATHOLOGY

EAR CLINICAL ANATOMY

External ear

- ***Sensitive innervation:***
  - 1. great auricular nerve (C3),
  - 2. lesser occipital nerve (C2 and C3),
  - 3. auricular branch (Arnold) of the vagus nerve (X)
  - 4. auriculotemporal nerve (V)

Middle ear and mastoid

- ***Innervation:***
  - the anterior part – the auriculotemporal branch of the V nerve
  - the posterior part – the auricular branch (Arnold) of the X nerve
  - internal wall – the tympanic (Jacobson) branch of the IX nerve
- ***Middle ear muscles***
  - Tensor tympani muscle, innervated by a branch of the mandibular nerve (V)
  - Stapedius muscle, innervated by a branch of the facial nerve (VII)

Inner ear

The bony labyrinth (otic capsule, the densest bone in the body):

- vestibule
- three semicircular canals
- cochlea
- two aqueducts:
  - **vestibular aqueduct** containing the endolymphatic duct
  - **cochlear aqueduct**, which connects the scala tympani with the subarachnoid space
- the cochlea makes 2.5 turns around the modiolus and contains the cochlear duct or scala media
- the three semicircular canals are perpendicular to each other
- the lateral (external) semicircular canal forms a 30-degree angle with the horizontal plane

Membranous Labyrinth

is located within the bony labyrinth and consists of:

- **cochlear duct**, which communicates with the saccule via the ductus reuniens
- saccule
- utricle
- membranous semicircular canals
- the **utricular duct and the saccular duct** form the endolymphatic duct, which ends with the endolymphatic sac in the posterior cranial fossa
- cochlear duct, which communicates with the saccule via the ductus reuniens
- saccule
- utricle
- membranous semicircular canals: utricular and saccular duct
- the ampullae of the semicircular canals contain sensory organs called **ampullary crests**
- the sensory organs contained in the utricle and saccule are called **maculae**

- the ampullary crests and maculae contain **vestibular hair cells and supporting cells**
- the cochlear duct is separated from the scala tympani by the **basilar membrane** and from the scala vestibuli by **Reissner's membrane**
- the **organ of Corti**, located on the basilar membrane, consists of ***sensory hair cells (inner, outer) and supporting cells (Deiters' cells, Hensen's cells)***

Peripheral innervation

- vestibular sensory organs
- **the superior vestibular nerve** consists of:
  - utricular nerve
  - superior and lateral ampullary nerves and
  - saccular branch (Voit's)
- **the inferior vestibular nerve** consists of
  - the saccular nerve and
  - posterior ampullary nerve
- **cochlea**
  - **afferent innervation:**
    - 35,000 first-order neurons – spiral ganglion located in Rosenthal's canal in the modiolus
  - **efferent innervation:**
    - much sparser (about 1,600 neurons) – transmit signals from the central nervous system to the cochlea

Central innervation

- **E.C.O.L.I.**
- **E. Eighth nerve** – cochlear nerve fibers (first-order neurons)
  - internal auditory canal
  - pontocerebellar angle
- **C. Cochlear nucleus** – the fibers enter the ventral or the dorsal cochlear nucleus
  - synapse with the second-order neuron
- **O. Superior olivary complex** on the same side
  - most fibers cross to the opposite side via the trapezoid body
- **L. Lateral Lemniscus**
  - the third neuron is located in the superior olivary complex and the lateral lemniscus
- **I. Inferior Colliculus**
  - neurons at this level may synapse in the inferior colliculus or end in the medial geniculate body of the thalamus
- **The medial geniculate body of the thalamus** is the **subcortical relay** of the auditory pathway – nerve fibers from this level ascend along the **acoustic radiation** to the auditory cortex – **tonotopic organization**

EAR PHYSIOLOGY

External ear and auditory canal

- collects, amplifies and transmits sound to the tympanic membrane

The tympanic membrane and the ossicular chain

- **efficiently transfer sound energy** from the gaseous environment to the liquid environment of the inner ear through two mechanisms:

- **hydraulic effect:** the surface of the tympanic membrane is **17 times** that of the stapes footplate
- **lever effect:** the handle of the malleus is **1.3 times** longer than the long process of the incus
- the total gain is about **27.5 dB**

Oval and round window

- sound waves reach the **two windows at different times**
- the round window is protected by the intact tympanic membrane and the edges of the round window niche

Inner and outer hair cells

- ***the inner hair cells*** are the mechanoelectrical transducers of the inner ear
- ***the outer hair cells***, with cilia connected to the **tectorial membrane**, receive neural signals from the olivocochlear efferent tract
- the contractile activity of the outer hair cells is responsible for the cochlea's ability to produce sounds (**otoacoustic emissions**)

Semicircular canals, Utricle and Saccule

- ***the semicircular canals*** and their ampullae provide information on ***angular acceleration***
- the receptor organ contained in each ampulla is called the '***crista***'. It contains vestibular hair cells, supporting cells and a gelatinous mass called the "***cupula***". The movements of the cupula are caused by the endolymphatic current
- ***the maculae of the utricle and saccule*** are covered by the otolithic membrane, on the surface of which there are calcium carbonate crystals called ***otoliths.*** They provide information about ***linear acceleration*** and ***head position in space***
- the ***endolymphatic sac*** plays a role in regulating endolymphatic pressure through its ability to absorb water. Its contents are ***hyperosmolar*** compared to the endolymph

TEMPORAL BONE IMAGING

- **Conventional X-ray**
  - the **Schüller view** and the **transorbital view** are still useful in identifying pre- and postoperative details
- **Computed tomography**
  - **method of choice** in the evaluation of most pathologies of the temporal bone
  - may assess:
    - anatomy of the temporal bone
    - tympanic membrane and tensor tympani muscle
    - the ossicular chain, the bony labyrinth, congenital malformations, ossification of the cochlear turns due to meningitis, labyrinthine fistula
    - round and oval windows, vestibular aqueduct
    - intratemporal facial nerve (labyrinthine, tympanic and mastoid segments)
    - defects in the temporal bone, fractures
- **Magnetic resonance imaging**
  - **an excellent way to visualize:**
    - cranial nerves
    - vestibulocochlear system
    - soft tissue lesions in the temporal bone and cerebellopontine angle

EAR MALFORMATIONS

Congenital ear malformations are present at birth

- may be caused by:
- **genetic factors** (chromosomal abnormalities, spontaneous mutation of a gene)
- **acquired factors** affecting the embryological development of the ear
- acquired factors may be:
- **extrinsic origin** (medicines, nutritional deficiencies, viral infections, hormonal disorders)
- **intrinsic origin**, such as autoimmune reactions or metabolic disorders
- malformations of the outer ear and middle ear are often combined, while **malformations of the inner ear occur independently**
- **minor:** small auricle malformations
- **moderate:** microtia, stenosis or atresia of the external auditory canal, associated with reduced or normal pneumatization and malformation or fixation of the ossicular chain
- **severe:** aplasia of the auricle, atresia of the external auditory canal. It is associated with markedly reduced or absent pneumatization, absent or undifferentiated ossicles, absence of one or both windows, aberrant facial nerve (20%)
- malformations or dysplasia of the cochleo-vestibular system may be associated with other **craniofacial abnormalities** and **hereditary syndromes**

EXTERNAL EAR DISEASES

Ear trauma

- because of its position, the external ear is at risk of trauma: puncture and crush wounds, lacerations, bites, burns and frostbite

Ear hematoma

- **Definition**: accumulation of serous fluid or blood between the perichondrium and the underlying cartilage (subperichondrial hematoma)
- **Etiology/Epidemiology**
- the cause is blunt trauma (contact sports, assault) to the external ear
- small blood vessels of the perichondrium rupture, leading to accumulation of blood between the perichondrium and the cartilage
- **Symptoms**
- fluctuant mass on the lateral surface of the auricle
- tenderness, heat, pain
- **Treatment**
- aseptic drainage of the blood or serous fluid and prevention of its reaccumulation
- minimal retroauricular incision with fenestration of the cartilage and drainage of the blood
- application of a compressive dressing soaked in ointment and fixed in place with sutures
- **Complications**
- relapse requires incision and drainage
- complications are: cartilage necrosis, organization of the hematoma, infection (perichondritis); the end result is the “cauliflower” ear

Frostbite

- are the consequence of prolonged exposure to very low temperatures
- the upper portion of the auricle is the most affected

Burns

- if the skin is still intact, antibiotic ointment
- surgical treatment if cartilage has been exposed

Infections of the auricle and external auditory canal

- **Etiology**
- infections of the auricle are produced by ***Staphylococcus aureus or Streptococcus pyogenes; Pseudomonas aeruginosa*** is the main agent responsible for infection of the perichondrium
- **Symptoms**
- edema, erythema, induration; painful (bacterial infection) or itchy (fungal infection, allergy)
- the skin produces a serous or purulent exudate
- **Complications:** extension of the infection to the perichondrium and cartilage (chondritis)
- **Treatment**
- **bacterial infection:** local disinfection, anti-inflammatory drugs, oral antibiotics
- **erysipelas:** antibiotic treatment targeting Streptococcus pyogenes (penicillin) for 10-14 days

External otitis

- **Definition:**
- bacterial/fungal infection of the EAC, without damage to the tympanic membrane
- **Etiology/Epidemiology:**
- occurs after **microtrauma** (cotton swabs, foreign objects, fingernails)
- **Favoring factors:**
- exostoses
- chronic otitis media
- swimming in a warm and humid environment

Acute, diffuse external otitis (swimmer's ear)

- **Infectious agents** most commonly found in diffuse external otitis are:
  - Pseudomonas aeruginosa
  - Staphylococcus aureus
  - Proteus vulgaris
  - Streptococcus species
  - Candida albicans, Aspergillus niger
- **Symptoms**
  - **itching**, ear fullness
  - **localized pain** in both the EAC and the auricle
  - the pain is exacerbated by mastication and by moving the tragus
  - there may be ear **discharge** and **hearing loss**
- **Diagnosis**
  - Inspection:
  - diffuse edema of the EAC with accumulation of cellular debris; the secretion is clear to purulent (yellow-greenish)
  - **culture** – in treatment-resistant cases, to identify the pathogenic organisms
  - laboratory tests: blood sugar, to exclude **diabetes**
- **Differential diagnosis**
  - furunculosis
  - chronic otitis media
  - malignant tumor

  - benign osteitis of the EAC
  - malignant external otitis
  - keratosis obturans
  - inflammation of the temporomandibular joint
- Treatment
  - **thorough cleaning** of the external auditory canal under the microscope
  - avoiding **wetting**
  - oral antibiotics
  - oral analgesics

External auditory canal furuncle

- acute, **localized** infection of the outer, cartilaginous portion of the EAC, arising from a ***hair follicle and its sebaceous and ceruminous glands***
- the pathogen is usually **Staphylococcus aureus**
- the symptoms are **pain, pruritus and edema**
- treatment depends on the stage of the infection:
  - oral antibiotics such as amoxicillin + clavulanic acid or a cephalosporin
  - drainage

Otomycosis (fungal external otitis)

- fungal infections of the EAC are **rarely primary**
- they commonly occur after treatment of external otitis with antibiotic and corticosteroid drops
- the most common pathogenic organisms are: ***Aspergillus niger, Aspergillus fumigatus, Aspergillus flavus and Candida albicans***
- the symptoms are ***pain, pruritus and thick secretion***
- treatment involves thorough aspiration of the EAC followed by application of a topical antifungal lotion (**clotrimazole, miconazole**)
- ear washing is not recommended

Chronic external otitis

- **Definition:** mild chronic inflammatory condition of the skin of the EAC
- **Etiology:**
  - incomplete treatment of acute diffuse external otitis
  - repeated contamination with water
  - allergens, purulent secretions from a chronic otitis media
- **Symptoms:**
  - pruritus and ear fullness
  - lack of cerumen
- **Differential diagnosis:**
  - **eczematous external otitis (chronic dermatitis, eczema):**
    - patients with a history of atopic dermatitis, psoriasis or eczema
    - pruritus, skin peeling, crusts
    - fissures and discharge on the auricle and in the canal
- **Treatment:**
  - the underlying cause must be removed
  - thorough, repeated cleaning of the canal with 70% alcohol solution
  - topic
- **Complications:**

  - the most common complication is **EAC stenosis**
- **Prevention of external otitis:**
  - excessive cleaning and microtrauma destroy the protective barrier of the EAC
  - avoid scratching with foreign objects, cotton swabs or fingernails

Malignant external otitis

- **Etiology:** infection caused by **Pseudomonas aeruginosa** in elderly, diabetic and immunodeficient patients
  - **Symptoms:**
    - intermittent otalgia, especially at night
    - headache and temporomandibular joint pain (spontaneous and on chewing)
    - scanty, fetid otorrhea
    - hearing loss
    - inflammation of the parotid gland
  - **Complications:**
    - paralysis of cranial nerves:
- **nerve VII**, due to spread towards the parotid gland and stylomastoid foramen
- **nerves IX, X, XI**, due to spread towards the jugular foramen
- **nerve XII**, at the hypoglossal canal
  - meningitis
  - epidural abscess
  - thrombophlebitis of the lateral sinus and internal jugular vein
  - **Diagnosis:**
    - clinical examination
    - external otitis unresponsive to conventional treatment
    - granulation tissue on the floor of the EAC
    - ulceration at the junction between cartilage and bone
    - diffuse inflammation of soft tissues
    - Laboratory: **biopsy** (to exclude squamous cell carcinoma)
    - glucose
    - ESR (commonly >80 mm/h)
    - culture: to confirm the pathogenic organism
    - **CT** is most useful in defining the extent of the disease and in assessing the integrity of the bone
  - **Treatment:**
    - proper treatment of diabetes
    - ear drops with topical antibiotics are of **little value**
    - long-term systemic treatment (6-8 weeks) with high-dose antibiotics:
    - quinolones: ciprofloxacin 750 mg per day orally
    - 3rd-generation cephalosporins: ceftriaxone 1-2 g/day i.v. or ceftazidime 1-2 g/day i.v., associated with an aminoglycoside
    - **additional therapies**
    - analgesic/anti-inflammatory: e.g. diclofenac 75-150 mg orally or as a suppository
    - **surgery:** limited to **biopsies** and **removal of necrotic bone fragments**
- **Differential diagnosis:**
  - EAC squamous cell carcinoma – **biopsy**

Exostoses

- **Definition:** new bone growth in the bony part of the EAC, usually **bilateral**
- **Etiology/Epidemiology:**
- patients with a history of **aquatic activities** (swimming, diving, surfing)
- may be caused by chronic asymptomatic periostitis
- **Diagnosis:**
- **sessile, ovoid** bony tumor, covered with normal, very thin skin
- asymptomatic
- **Surgical treatment** in:
  - rare cases of subtotal occlusion of the EAC with cerumen retention
  - cases with water retention between the exostoses and the eardrum

Osteoma

- **Definition:** benign bone tumors of the EAC extending into the mastoid, usually **unilateral**
  - **Symptoms:**
    - conductive hearing loss due to the accumulation of keratin and cerumen
    - recurrent external otitis
- **Diagnosis: solitary bone mass** covered by normal-looking skin
  - **Treatment:**
    - surgical removal
    - it is recommended to preserve as much skin as possible

Keratosis obturans

- **Definition:**
- rare disease, characterized by **excessive accumulation of keratin** in the external auditory canal with progressive erosion of its walls
- Etiology:
- altered mechanism of epithelial migration in the EAC
- Diagnosis:
- bulky keratin plug that obstructs the EAC
- should be **soaked with olive oil** and removed under the microscope
- **Differential diagnosis:**
- cholesteatoma of the external auditory canal:
- invasion of squamous epithelial tissue into a localized area of bone erosion
- intermittent otorrhea and mild, chronic otalgia
- CT reveals the extension into the EAC
- **Treatment:**
  - periodic cleaning under the microscope
  - avoiding self-cleaning

Pavilion tumors

- **Benign tumors**
  - keloid
  - chronic nodular chondrodermatitis (Winkler node)
  - keratoacanthoma
  - epidermal inclusion cysts
- **Malignancies**

- **basal cell** carcinoma
- **squamous cell** carcinoma
- malignant melanoma

EAC tumors

- **Benign lesions**
- osteoma
- adenoma of the ceruminous glands
- squamous papilloma
- **rare benign lesions**: neurofibromas, paragangliomas, angiomas
- **Malignant lesions**
- **squamous cell** carcinoma
- **basal cell** carcinoma
- **rare** malignancies:
  - adenocarcinoma
  - adenoid cystic carcinoma
  - rhabdomyosarcoma
  - melanoma

TRAUMATIC PERFORATIONS OF THE TYMPANIC MEMBRANE

- **Definition:**
  - tympanic membrane perforations caused by **direct injury or indirect trauma**
- **Etiology:**
  - **direct trauma:** cotton swab, hairpin, match, etc.
  - **indirect trauma:** sudden change of pressure in the EAC caused by an explosion, a hard slap, diving, barotrauma, etc.
- **Symptoms:**
  - otalgia, otorrhagia, ear fullness, tinnitus, hearing loss
- **Diagnosis:**
  - microscope otoscopy
  - examination of hearing:
    - mild conductive hearing loss (10-30 dB)
    - severe mixed hearing loss in case of explosion
  - vestibular evidence
- **Treatment:**
  - **avoiding water entering the EAC**
  - small perforations that do not heal spontaneously: **fibrin or fat myringoplasty**, left in place for 3-4 weeks
  - mesotympanic perforations: **standard myringoplasty** is performed
- **Prognosis:**
  - the prognosis is generally very good, with spontaneous healing **within 3 months**

MYRINGITIS

- **Definition:**
- limited to the tympanic membrane
- ***viral myringitis (bullous):***

  - acute painful inflammation of the epidermal layer of the tympanic membrane caused by **influenza virus**
  - **red vesicles with serohemorrhagic content** under tension, between the epidermis and the fibrous layer of the tympanic membrane
- ***granular myringitis (chronic myringitis):***
  - small pyogenic granulations on the external surface of the tympanic membrane following local trauma (frequently with a cotton swab)
- ***external myringitis:***
  - bacterial inflammation involving the epidermal and fibrous layers of the tympanic membrane
- **Diagnosis:**
  - **otoscopy:**
    - specific red vesicles in viral otitis
    - bulging and pulsation of the tympanic membrane with serohemorrhagic discharge are specific to acute otitis media
    - pyogenic granulations specific to chronic myringitis
  - **audiogram:**
    - mild conductive hearing loss or hearing within the normal range
    - in case of damage to the inner ear (viral myringitis) – sensorineural hearing loss
- **Treatment:**
- ***Viral myringitis:***
  - local anesthesia and puncture of the blisters: the pressure decreases and the pain diminishes
  - **wick soaked in antibiotic and cortisone**
  - analgesic
  - in case of inner ear involvement, treatment is **similar to that of sudden idiopathic hearing loss**
- ***Granular myringitis:***
  - local anesthesia and removal of the infected granulations
  - wick soaked in **acetic acid** (1.5% solution)
  - untreated, granular myringitis can lead to lateralization of the tympanic membrane
- ***External myringitis:***
  - treatment for external otitis

ACUTE OTITIS MEDIA

- **Definition:**
  - acute inflammation of the mucosa of the middle ear caused by **viruses and/or bacteria** from the nasopharynx that reach the middle ear through the Eustachian tube
- **Etiology/Epidemiology:**
  - main causes:
    - **acute** upper respiratory tract infection
    - **chronic** infection of nasopharynx
  - **80% viral**
  - **20% secondary bacterial** infection
    - ***Streptococcus pneumoniae (40-50%)***
    - ***Haemophilus influenzae (30-40%)***

        - ***Moraxella catarrhalis (10-20%)***
        - ***Staphylococcus pyogenes and Staphylococcus aureus***
    - in children the main cause is **chronically inflamed adenoids**
- **Symptoms:**
    - intense, throbbing **otalgia**
    - temperature can reach **40 °C**
    - irritability and abdominal pain
    - conductive hearing loss is always present
    - adults may have **tinnitus**
- **Complications:**
    - mastoiditis
    - acute labyrinthitis (dizziness, vertigo, deafness)
    - facial paralysis
    - sigmoid sinus thrombosis
    - meningitis
    - epidural abscess
    - **Gradenigo syndrome:**
        - **abducens nerve paralysis** and **trigeminal neuralgia (V1 and V2)**
        - the inflammatory process spreads to the **apex of the temporal bone**
- **Diagnosis:**
    - **inspection: red auricle**, pushed anteriorly; edematous skin over the mastoid in case of mastoid reaction
    - **palpation: pain** on touching or pressing the mastoid
    - **otoscopy:**
        - accentuated vascular pattern along the handle of the malleus
        - congestion and thickening of the tympanic membrane
        - bulging of the tympanic membrane with loss of landmarks
        - perforation with otorrhea
    - **audiometric:** tuning fork tests, audiogram
    - **nystagmus:** to exclude vestibular damage
    - **microbiology:** cultures taken from nose/nasopharynx
- **Treatment:**
    - ***conservative treatment:***
        - antibiotics
        - analgesic/anti-inflammatory
        - nasal vasoconstrictors
    - ***additional therapies:***
        - **local heat** applications twice daily for 5-10 min
        - **antibiotic ear drops** are of **no value** when the tympanic membrane is intact
    - ***surgical treatment:***
        - **myringotomy (paracentesis)**: indicated if bulging of the tympanic membrane persists; it helps relieve pain
        - **adenoidectomy**
        - **antrotomy/mastoidectomy**

SEROUS OTITIS MEDIA

- **Definition:**

- accumulation of **non-purulent fluid** in the middle ear due to **Eustachian tube dysfunction**

- **Etiology/Epidemiology:**
  - recurrent upper respiratory tract infections (adenoid vegetations)
  - Eustachian tube dysfunction
  - allergy
  - chronic rhinosinusitis
  - posterior packing
  - radiotherapy in the ear/nasopharynx area
  - **the highest incidence of serous otitis is in the 2-5 year age group**
- **Pathophysiology:**
  - inflammation or allergy causes **Eustachian tube dysfunction** and reduced or absent aeration of the middle ear
  - **negative pressure** in the middle ear, with the formation of a ***serous transudate***, metaplasia of the middle ear mucosa into an active secretory epithelium and the formation of an ***exudate (mucotympanum)***
- **Symptoms:**
  - sensation of pressure, short episodes of pain
  - conductive hearing loss
  - tinnitus
  - **commonly asymptomatic**
- **Complications:**
  - acute otitis media, caused by bacterial infection of the effusion
  - chronic or subacute masked mastoiditis
  - retraction pockets, atelectatic otitis, cholesteatoma
  - perforation of the tympanic membrane
  - tympanosclerosis
- **Diagnosis:**
  - **endoscopic examination of the oropharynx, nose and nasopharynx – mandatory!**
    - **otoscopy:**
      - fluid level or air bubbles behind the eardrum
      - the tympanic membrane is bluish, gray or brownish in mucous effusions
      - reduced mobility of the tympanic membrane on pneumatic otoscopy
    - **hearing tests** – tuning fork, audiogram, **conductive deficit < 40 dB.**
    - **tympanometry:** flat curve type B
    - **allergy tests** (prick test, IgE, RAST – radioallergosorbent test)
    - **biopsy** from nasopharyngeal hypertrophic tissue
- **Treatment:**
  - **conservative treatment:**
    - **antibiotic therapy IS NOT** usually necessary
    - vasoconstrictor nasal drops in combination with antihistamines
    - exercises for ventilation of the Eustachian tube **(Valsalva maneuver)**
  - **surgical treatment:**
    - ***myringotomy*** with aspiration of the fluid
    - insertion of ***transtympanic ventilation tubes*** if:
      - hearing loss **>30 dB,** unresponsive to treatment for **over 3 months**

        - retraction pockets form in the tympanic membrane
    - ***adenoidectomy***
    - ***antrotomy or mastoidectomy:*** persistence or recurrence of the middle ear effusion and chronic discharge through the ventilation tubes
- **Differential diagnosis:**
    - **eosinophilic otitis media:** extensive accumulation of eosinophils in the middle ear exudate
    - **otoliquorrhea**
    - haemotympanum
    - Wegener's granulomatosis of the middle ear

ACUTE OTOMASTOIDITIS

Definition:

- bacterial infection of the mastoid cell system during, or as a complication of, acute, subacute or chronic otitis media

Etiology/Epidemiology:

- in most cases acute mastoiditis occurs in **young children**
- during otitis media, bacteria spread through the antrum into the mastoid cell system, causing **edema and hyperemia of its mucosa**
- a **serous effusion** appears, followed by a **purulent effusion**, **demineralization** of the bony cell walls and **bone necrosis**
- finally a cavity forms, which may break through into neighboring structures
- the **bacteria** responsible for acute mastoiditis are similar to those of ***acute otitis media***

Types:

Masked mastoiditis:

- chronic inflammation of the mucosa lining the cells **without abscess formation** (in children it is associated with chronic or mucous otitis)
- there are **non-specific symptoms**, rarely pain or fever

Acute mastoiditis:

- occurs a few days or 1-2 weeks after the onset of untreated acute otitis media
- **redness, swelling, tenderness and fluctuation in the mastoid area**
- **the auricle is displaced laterally and downwards**
- exacerbation of ear pain, fever and creamy otorrhea
- the pain tends to be persistent and throbbing
- leukocytosis, subfebrile state or septic fever and sometimes chills
- a ***subperiosteal retroauricular abscess*** may develop

Symptoms:

- otalgia, with spontaneous pain or pain on palpation of the mastoid
- fever
- hearing loss
- **these symptoms may be masked by insufficient antibiotic therapy**
- the tympanic membrane does not return to its normal appearance

Complications:

- facial paralysis
- meningitis
- labyrinthine
- sigmoid sinus thrombosis
- Bezold abscess (perforation of the mastoid tip and abscess in the neck muscles)
- petrositis (temporal bone osteomyelitis)

Diagnosis:

- Inspection:
  - ***retroauricular erythema***
  - ***swelling***
  - ***auricle pushed anteriorly and downwards***
- otoscopy:
  - congested, bulging or dull tympanic membrane due to effusion
  - central perforation of the tympanic membrane with purulent secretion
  - **pathognomonic – sagging of the posterosuperior wall of the EAC**
- rhinoscopy:
  - **signs of rhinopharyngitis**
  - **purulent nasal secretion**
- tuning fork tests, audiogram – **conductive hearing loss**
- Frenzel glasses: **spontaneous nystagmus**
- X-ray – Schüller view:
  - reduced pneumatization
  - resorption of the mastoid cells
  - formation of cavities
- CT:
  - the mastoid cells are **opaque** because they are **filled with fluid**
  - information on mastoid pneumatization and the ossicles of the tympanic cavity

Additional diagnostic procedures:

- nasal and nasopharyngeal exudate
- hemogram, ESR
- paranasal sinus X-ray

Differential diagnosis:

- acute or subacute external otitis
- furunculosis of the external auditory canal
- suppuration of retroauricular lymph nodes
- auricular erysipelas

Treatment:

- **conservative treatment:**
  - **the antibiotic** given initially should cover the main organisms involved and penetrate the CNS if a complication appears imminent
  - a sample of the otorrhea is taken for **culture and antibiogram**
  - antibiotic therapy should be continued **for at least 2 weeks**
- **surgical treatment:**

- **myringotomy** with or without the insertion of ventilation tubes
- **cortical mastoidectomy** is performed as an emergency if there is no improvement after the initial antibiotic therapy or in the event of a complication

Prognosis:

- acute mastoiditis usually heals **without sequelae** if treatment is correct

CHRONIC OTITIS MEDIA (WITHOUT CHOLESTEATOMA)

Definition:

- chronic inflammation of the mucosa of the middle ear characterized by the **persistence of a central eardrum perforation** with **permanent or intermittent otorrhea**

Etiology / Epidemiology:

- **persistent central perforations** occur in patients with repeated infections due to Eustachian tube dysfunction
- the most common pathogenic organisms detected in cultures of chronic otorrhea are:
  - **Pseudomonas aeruginosa (60-80%)**
  - **Staphylococcus aureus (10-25%)**
  - **Proteus (10-20%)**
  - **mycotic superinfection is possible**

Symptoms:

- recurrent episodes of **otorrhea**
- in the **active period** the discharge is odorless and abundant
- **reactivation** episodes are generated by:
  - upper respiratory tract infections
  - manipulation of the EAC
  - water in the ear
- during the **inactive period** the patient has:
  - usually mild conductive hearing loss
  - dry central perforation

Complications:

- ossicular resorption - especially the **long process of the incus**
- ossicular fixation - especially the **head of the malleus**
- **tympanosclerosis:**
  - **hyalinization** of the middle ear mucosa, limited to the tympanic membrane (with or without perforation) and/or extending to the ossicles, with their fixation
  - **small plaques** of tympanosclerosis limited to the tympanic membrane ***do not affect hearing***
  - in **extensive forms** (involving the stapes and promontory) there is ***severe conductive or mixed hearing loss***
- **conductive or mixed hearing loss** (toxic involvement of the inner ear)
- **external otitis**
- **otic eczema**

Diagnosis:

- **Otoscopy:**

  - from a **small central perforation** to a **subtotal perforation**
  - **calcifications**
  - in the **inactive stage** the ear can be dry, with the edges of the perforation covered by healed epithelium
  - in the **active stage** the central perforation is surrounded by a ring of granulation tissue and a purulent yellowish exudate
- **Hearing examination:**
  - tuning fork tests and audiometry: **conductive hearing loss**
  - **sensorineural impairment** is not common, but it is possible

Additional diagnostic procedures:

- X-ray (Schüller view) to assess the degree of mastoid pneumatization
- high-resolution CT (**cholesteatoma**)
- X-ray of paranasal sinuses
- nasopharyngeal fibroscopy

Differential diagnosis:

- **middle ear atelectasis and retraction pockets:**
  - ***partial (retraction pocket) or total retractions*** of the tympanic membrane are caused by **Eustachian tube dysfunction**
  - feeling of ***aural fullness***
  - ***hearing loss***
  - otoscopy: the eardrum appears ***retracted***, sometimes with ***adhesion*** to the incus, stapes and promontory
  - ***whether the tympanic membrane is perforated or intact is determined by the Valsalva maneuver***, which, if successfully performed, pushes the atelectatic membrane outwards
  - ***retraction pockets*** are commonly located in the **posterosuperior portion of the tympanic membrane**
  - they may become "non-self-cleaning" and potentially dangerous, with the formation of a **cholesteatoma** when keratin debris accumulates

Treatment:

- **Conservative treatment:**
- periodic ear cleaning under the microscope
- topical treatment: quinolone drops applied twice a day for 7-10 days
- oral or parenteral antibiotic therapy
- in the case of an eardrum perforation, a **conventional hearing aid** is **NOT recommended**, even if the perforation is dry, because wearing the device will cause ear discharge
- the tympanic membrane perforation must be closed first
- **Additional therapeutic options:**
- treatment of disorders of the nose, paranasal sinuses or nasopharynx
- elimination of allergic factors
- **Surgical treatment**
- ***1. Myringoplasty/tympanoplasty without mastoidectomy:*** if the perforation is dry, the eardrum (myringoplasty) and possibly the ossicular chain (ossiculoplasty) are repaired
- **Myringoplasty/tympanoplasty without mastoidectomy** may be performed by either an **endaural or a retroauricular** approach

- ***2. Tympanoplasty with mastoidectomy:*** if otorrhea persists and the mastoid mucosa is involved
- **Tympanoplasty with mastoidectomy** is performed by a **retroauricular** approach

MIDDLE EAR CHOLESTEATOMA

Definition:

- cholesteatoma involves the presence of **keratinized squamous epithelium** in the **tympanic cavity**
- it consists of **a matrix which desquamates** continuously
- **exfoliated keratin debris** accumulates and forms the cholesteatoma

Etiology/epidemiology:

- the middle ear cholesteatoma is **relatively common** and may appear throughout life
- it is **rare as a primary lesion** (congenital cholesteatoma), originating from epithelial rests of ectodermal origin (in the petrous portion of the temporal bone)
- in the **vast majority** of cases cholesteatoma is **secondary to infection** of the middle ear (**acquired cholesteatoma**)
- **acquired forms** occur due to **Eustachian tube dysfunction**, which causes negative pressure in the middle ear with retraction of the tympanic membrane, which can perforate
- tympanic membrane retractions are usually localized in:
- the upper portion of the tympanic membrane (**pars flaccida cholesteatoma**)
- the posterosuperior portion (**pars tensa cholesteatoma**)
- local inflammatory processes, especially those caused by ***Pseudomonas aeruginosa*** and ***Proteus***, stimulate **keratin production and exfoliation**, and thus the cholesteatoma grows
- **acquired cholesteatoma** may also occur with a **marginal perforation** located in the posterosuperior portion of the tympanic membrane, with destruction of the fibrous annulus
- this allows the keratinized epithelium of the EAC to migrate into the tympanic cavity and grow in the middle ear

Symptoms:

- **asymptomatic** for years, due to slow accumulation of keratin
- infections increase the rate of exfoliation and the mass of keratin; an infected **cholesteatoma may grow rapidly**
- **otorrhea:** usually scanty, but **foul-smelling**
- **hemorrhage:** from **granulations** or **polyps**; may alarm the patient
- **otalgia:** due to inflammation of the EAC, or may indicate a complication
- **hearing loss:** may range from a normal hearing threshold to severe sensorineural hearing loss
- tinnitus
- **vertigo:** may indicate a **labyrinthine fistula**
- **headache:** suggestive of **intracranial extension**

Extracranial, intratemporal complications:

- labyrinth fistula
- acute labyrinthitis
- facial paralysis

Intracranial complications:

- extradural abscess

- subdural abscess
- localized or diffuse meningitis
- lateral sinus thrombosis
- temporal lobe abscess
- cerebellar abscess

Diagnosis:

- **otoscopy:** the cholesteatoma can usually be seen through the perforation
- **audiometry:** performed to assess the degree and type of hearing loss
- **high-resolution CT** reveals:
- location and size of cholesteatoma
- information about the integrity of the ossicular chain or any complications

Special types:

- **congenital cholesteatoma** (**rare**): white spherical cyst seen through an **intact tympanic membrane** in young children with no history of ear disease
- **post-traumatic cholesteatoma:** following a fracture of the temporal bone, EAC epithelium may remain trapped in the fracture line
- **iatrogenic cholesteatoma**: after transtympanic ventilation tube insertion

Differential diagnosis:

- **active chronic otitis media** with a polyp in the EAC
- **tuberculous otitis media**: sensorineural hearing loss, facial paralysis
- cholesterol granuloma

Prognosis:

- long-term clinical follow-up (**minimum 10 years**) is required to detect residual cholesteatoma and recurrences

Treatment:

- Conservative **treatment:**
- **periodic observation** indicated in:
- limited cases of dry cholesteatoma
- in the case of an only hearing ear
- elderly patients
- patients with poor health
- **Surgical treatment:**
- cholesteatoma requires surgical treatment with mastoid exploration
- the purpose of the operation is to ***eradicate the disease*** and ***close the tympanic cavity***
- ***reconstruction of the ossicular chain*** can be carried out immediately or at a later stage
- the cholesteatoma can be removed by **closed or open techniques**:
- **closed techniques**: indicated in children or cases with a well- pneumatized mastoid
- **open techniques**: indicated in the case of a sclerotic mastoid and in recurrences

INTRATEMPORAL AND INTRACRANIAL COMPLICATIONS OF OTITIS MEDIA

Definition:

- the infection spreads to adjacent structures outside the middle ear and mastoid

- **coalescent mastoiditis** is **a stage in the evolution** of acute otitis media and **not a complication**

Etiology/ Epidemiology:

- **complications** usually occur in **association with cholesteatoma**, and less frequently with chronic otitis media without cholesteatoma or with acute otitis media
- **their incidence has decreased substantially with the availability of antibiotics**
- Routes of spread:
- **thrombophlebitis of small veins**, in acute otitis media or in acute exacerbations of chronic otitis media
- **bone erosion** as a result of coalescent mastoiditis or a cholesteatoma
- preformed paths:
- oval window and round window
- IAC
- endolymphatic sac and duct
- congenital dehiscence of the tegmen and hypotympanum
- fracture line
- previous ear surgery

Complications:

- **1. Extracranial - Intratemporal**
- **labyrinthitis:** serous or purulent (**most common**)
- a dead labyrinth is usually the consequence of purulent labyrinthitis
- **labyrinthine fistula** (with or without simultaneous labyrinthitis)
- petrositis
- **2. Intracranial**
- **extradural abscess**: purulent collection between the dura and the bone
- **meningitis**, with two clinical types:
- localized
- diffuse
- lateral sinus thrombophlebitis
- cerebral abscess

Diffuse meningitis and brain abscess are the most important complications

- **Symptoms:**
- The signs and symptoms of **complications** are as follows:
- early signs:
- fetid otorrhea
- constant and deep pain
- headache and drowsiness
- **visual field changes (early sign – brain abscess)**
- **late signs:**
- **high fever**, which indicates meningitis or lateral sinus thrombosis
- rare symptoms:
- balance disorders
- hearing shearing
- cervical stiffness
- cranial nerve involvement

- seizures

Diagnosis of specific complications:

Labyrinth fistula

- the patient frequently complains of **dizziness** when **pressure** is applied to the tragus or when the auricle is manipulated (**Hennebert's sign**)

Inflammatory serous labyrinthitis

- vestibular symptoms:
- **spontaneous (irritative) nystagmus**, initially towards the affected ear
- then **(destructive) nystagmus** towards the opposite ear
- vertigo, nausea, vomiting, ataxia
- **cochlear symptoms:**
- hearing impairment predominantly at **high frequencies**
- **diplacusis** (the same sound perceived differently by the two ears)

Inflammatory suppurative labyrinthitis:

- the signs are similar to those of serous labyrinthitis, but with a **more sudden onset**

Petrositis

- **frontal or retro-orbital pain** in **anterior** petrositis
- **occipital, parietal or temporal pain** in **posterior** petrositis
- **persistent otorrhea** following a classical mastoidectomy
- **Gradenigo syndrome:**
- persistent otorrhea
- periocular pain in the region of the trigeminal nerve (V1 and V2) and
- diplopia due to paralysis of nerve VI

Facial palsy

- may be a complication of **acute otitis media** (dehiscence of the Fallopian canal) or
- of **chronic otitis media** (the onset of symptoms is slow and progressive)

Subperiosteal abscess

- usually associated with the **onset of acute mastoiditis** after an episode of acute otitis media

Extradural and perisinusal abscess

- risk of extension, leading to lateral sinus thrombophlebitis, subdural abscess, meningitis or brain abscess
- **persistent headache** on the side of the affected ear
- **abundant or pulsating otorrhea**, accentuated by compression of the internal jugular vein
- **signs of meningism** (Kernig, Brudzinski)
- **lumbar puncture**: pleocytosis, usually with a predominance of lymphocytes – normal glucose values, no microorganisms
- **MRI: epidural collection** with intense peripheral signal
- **most perisinus and extradural abscesses do not produce symptoms and are discovered by chance during surgery**
- Localized meningitis
- Symptoms:
- headache, fever, irritability or drowsiness, convulsions (in infants)

- Signs:
- neck stiffness
- **Kernig's sign** (inability to extend the leg on the thigh when the thigh is flexed on the abdomen)
- **Brudzinski's sign** (flexion of the thigh and knee when the neck is flexed)
- **Babinski's sign** (extension of the toes instead of flexion on stimulation of the sole)
- **CSF is clear or only slightly turbid**, 10-1000 cells/mm3, predominantly lymphocytes, glucose within normal limits, **without microorganisms** on culture

Generalized meningitis:

- pyogenic infection of the **arachnoid membrane** of the entire brain and spinal cord, with viable organisms in the CSF
- occurs **in adults** with **chronic otitis media**
- occurs **in children** with **acute otitis media**
- **the symptoms are similar to those of the localized form, but more intense**
- a **chill** may accompany the initial rise in temperature
- the pulse is rapid
- the temperature is high **(41-42°C)**

Lateral sinus thrombophlebitis:

- characteristically caused by infection (in order of frequency) with:
- hemolytic streptococcus
- pneumococcus type III
- Staphylococcus
- **septic fever** is rarely seen nowadays, owing to aggressive antibiotic therapy
- between bouts of fever the patient **feels well**, **as opposed to the apathy of brain abscess and the prostration of meningitis**
- headache
- progressive, rapid and pronounced **anemia**, especially with hemolytic streptococcus
- **progressive emaciation**, not as rapid and pronounced as in cerebellar abscess
- fundus changes – **papilledema**
- **edema over the mastoid process**, due to thrombosis of the mastoid emissary vein (**Griesinger's sign**)
- the definitive diagnosis is made by surgical exploration

Brain abscess

- **First stage (encephalitis):**
- moderate, transient but **characteristic symptoms**:
- a slight/moderate rise in temperature (a few days)
- headache and nausea, sometimes with vomiting, **but not projectile**
- the patient may seem apathetic, drowsy or irritable
- **convulsions especially in children**
- possible mild neck stiffness, with a moderate increase in cells and proteins but normal glucose in the CSF
- Second stage (latent):
- lasts from 10 days to a few weeks
- symptoms may be minimal or absent:
- general malaise

- inappetence
- intermittent headache
- slight increases in temperature
- drowsiness, apathy, irritability due to ongoing encephalitis
- moderate, transient symptoms
- **Second stage (latent):**
- symptoms may be minimal or absent
- **Third stage (manifestation of the expanding abscess)**
- encephalitis and edema fluctuate from day to day
- **severe, continuous headache**, refractory to symptomatic treatment
- projectile vomiting – pathognomonic
- **bradycardia** due to **compression of the vagal center**
- slightly increased, normal or below normal temperature
- apathy, drowsiness, disorientation, **Cheyne-Stokes breathing,** increased blood pressure
- **Jacksonian seizures** (30-50%) and ocular palsies with pupillary changes
- **CSF is rarely normal**; it usually shows slight increases in cell count and protein
- Focal symptoms:
- **aphasia** (left temporal abscess in a right-handed person)
- **facial paresis** on the **opposite side**, of central origin (sparing the frontal muscles)
- visual field disturbances
- symptoms of **intracranial hypertension** are more constant and defined in a cerebellar abscess than in a temporal lobe abscess
- In cerebellar abscesses the following are also found:
- **ataxia** on the same side (finger-to-nose test)
- **adiadochokinesia** (inability to perform rapidly alternating movements)
- intentional tremor
- **ataxic gait** with a tendency to fall towards the affected side
- **emaciation** despite increased appetite
- **CT:** a hypodense area surrounded by a well-defined ring (ring sign)
- **MRI:** better contrast between peripheral edema and surrounding brain tissue

Treatment of specific complications:

Serous labyrinthitis:

- if it is caused by acute otitis media, **antibiotics and myringotomy** are indicated
- if it is caused by perilabyrinthine osteitis or cholesteatoma, **mastoidectomy and antibiotics** are indicated

Suppurated labyrinthitis:

- bed rest and antibiotic treatment
- examination of the CSF and **emergency surgical drainage** of the labyrinth
- suppurative labyrinthitis secondary to meningitis **does not require drainage of the labyrinth**

Petrositis:

- cortical mastoidectomy
- if there are **no signs of meningitis**, wait 8-10 days before approaching the apex
- if **symptoms of petrositis** persist, the cells should be surgically opened as soon as possible

Facial paralysis:

- **immediate surgery** in the case of chronic otitis media
- **myringotomy** in acute otitis media

Subperiosteal abscess:

- **abscess drainage** and a **cortical mastoidectomy**

Extradural and perisinusal abscess:

- **simple or radical mastoidectomy** with exploration of the pathway to the abscess
- **antibiotic therapy** to prevent the spread of infection

Localized meningitis of otic origin:

- **immediate surgical exploration** with wide exposure of the dura

Generalized meningitis of otic origin:

- any febrile patient should be examined for neck stiffness and Kernig's sign
- **lumbar puncture** (the centrifuged sediment should be Gram-stained)
- **antibiotic treatment:** third-generation cephalosporin 2 g intravenously every 12 hours for 7-10 days, associated with dexamethasone

Lateral sinus thrombophlebitis

- intravenous **antibiotic therapy**
- **simple or radical mastoidectomy** in case of cholesteatoma
- inspection and palpation of the exposed sinus wall
- **ligation of the internal jugular vein** in cases with persistent sepsis after mastoidectomy and sinus exploration
- the use of **anticoagulants** is **controversial**

Otitic hydrocephalus:

- **repeated lumbar or ventricular puncture** or placement of lumbar drainage
- diuretics, hyperosmolar agents
- steroids
- **subtemporal decompression** to **prevent optic nerve atrophy** due to prolonged papilledema

Otoliquorrhea (Otic CSF leak):

- **surgical exploration** to prevent meningitis

Brain abscess:

- intravenous **antibiotic** treatment (third-generation cephalosporin and metronidazole)
- intravenous **dexamethasone**
- mannitol
- **craniotomy** with abscess drainage
- **mastoidectomy** 3-4 days after craniotomy

OTOSCLEROSIS

Definition

- the condition is characterized by a primary, focal bone **dystrophy** localized to the **labyrinthine capsule**, which causes fixation of **the stapes**
- **otosclerosis, otospongiosis or stapedovestibular ankylosis** is a **common cause of hearing loss** in otological practice

Historical aspects

- the term “otosclerosis” was first used by **Joseph Toynbee**, more than 100 years ago, for an ear disease characterized by early onset and progressive evolution, associated with treatment-refractory hearing loss and structural pathology of the temporal bone
- stapedovestibular ankylosis had been **described** long before by **Valsalva, in 1704, at an autopsy**

Pathological anatomy

- the otosclerotic focus arises through a **bone resorption process**, similar to inflammation
- the **avascular, ivory-like enchondral bone** of the labyrinthine capsule is **replaced by new spongy, soft and well-vascularized bone (otospongiosis)**
- characteristic of the active, immature otosclerotic focus, which consists of irregularly arranged bony lamellae separated by wide vascular spaces and containing numerous histiocytes, osteoblasts and a few osteoclasts
- **a process of sclerosis follows**, leading to mature, inactive bone with a denser structure, reduced vascular spaces and decreased cellularity (otosclerosis)
- the active otospongiotic bone is covered by a thickened, vascular mucosa, giving the promontory a pinkish, congested appearance, which is sometimes visible through a more transparent eardrum and is known as **Schwartze’s sign**
- the mature bone has a **white, ivory-like color**, in contrast to the rest of the labyrinthine capsule

Localization:

- the disease has a predilection for the **oval window**, in the **anterior portion of the stapedovestibular joint**, causing fixation of the stapes (**80–90%**)
- much less often, it is located at the **round window** or causes enchondral ossification in **other areas of the cochlea**, leading to **sensorineural hearing loss** as the only manifestation of the disease
- a less favorable situation, encountered intraoperatively, is **obliterative otosclerosis**, which **fills the oval window niche**. It must be removed with the microdrill in order to open the oval window
- in about **50% of cases** there are **two or more otosclerotic foci**
- in **70–80% of cases** the disease is **bilateral**

Incidence

- varies widely, between **3 and 10 per 100,000 white adults**
- Caucasians are more commonly affected; the disease is a common cause of hearing loss in Europe, the Middle East, the USA, Israel and India
- otosclerosis is rarely found in Africa and in East Asian populations (1%)
- the condition is more common in women than in men
- **may start at any age**, but most diagnoses are made in adulthood

Etiology

- is not precisely known
- some consider it to have a **genetic cause**, which is why termination of pregnancy on request is allowed
- it is considered by some to be a **localized form of Paget’s disease**, due to the similarity between the two conditions
- a **significant correlation** has also been found between **HLA antigens** (human leukocyte antigens) and otosclerosis

- some consider otosclerosis to be an **autoimmune disease** – in fact a **reaction of the labyrinthine capsule to cartilage tissue** (cartilaginous islands in the capsule), as demonstrated by experimental studies
- Symptoms:
- characteristic of otosclerosis is a **progressive hearing loss** of insidious onset, without a history of ear disease
- although the condition is frequently **bilateral**, in most cases the two ears evolve differently over time
- at the beginning the hearing loss is **conductive** and is due to stapedovestibular ankylosis (immobilization of the stapes footplate in the oval window)
- during the course of the disease a cochlear component may be added, which worsens hearing, resulting in a **mixed or sensorineural hearing loss**
- some patients understand speech better in noisy environments (train, tram, crowds), a phenomenon known as **paracusis of Willis**
- the **explanation** of this phenomenon would be that the person speaking to the patient instinctively raises their voice to overcome the masking effect of the noise – noise that hardly bothers the patient at all
- **tinnitus** frequently accompanies the hearing loss; it is **low-pitched** and refractory to treatment
- otosclerosis is not painful

Objective examination:

- **otoscopy DOES NOT** reveal significant pathological changes, Schwartze's sign being inconstant (15%)
- the Eustachian tube is functional and, if Schüller mastoid X-rays are performed (which is not strictly necessary), they show a very well-pneumatized mastoid

Positive diagnosis:

- the diagnosis of otosclerosis is based on the **symptoms** described above and several methods of **examining hearing and middle ear function**
- **pure-tone threshold audiometry** shows, at first, a purely conductive hearing loss (75%), which may in time become mixed (20%) or even predominantly sensorineural, through involvement of the labyrinth by the otosclerotic focus
- **in 5%** of cases, the hearing loss is sensorineural from the onset, due to capsular foci that do not affect the mobility of the ossicular chain
- the diagnosis of otosclerosis is based on the **symptoms** described above and **several methods of examining hearing and middle ear function**
- pure-tone threshold audiogram
- in conductive hearing loss there is frequently a slight dip (15-20 dB) in the bone conduction curve between 1000 and 4000 Hz, called the **Carhart notch**
- this appears to be related to the transmission of the sound wave and disappears after surgery
- **tuning fork tests** show, in most cases, the Bezold triad of conductive hearing loss: Weber lateralized to the more affected ear, prolonged Schwabach and negative Rinne
- the **Gellé test** is also important; it is **negative** in otosclerosis and indicates fixation of the stapes footplate
- the diagnosis of otosclerosis is based on the **symptoms** described above and **several methods of examining hearing and middle ear function**
- testing of the stapedial reflex

- **impedance audiometry** is very important in the diagnosis of otosclerosis; the tympanograms obtained are type As, due to fixation of the stapes
- in some cases, the diagnosis is established **intraoperatively**, by inspection of the tympanic cavity and testing of the ossicular chain

Differential diagnosis:

- is made, first of all, with other causes of **conductive hearing loss with an intact eardrum**
- fixation of the head of the malleus
- aseptic necrosis of the long process of the incus
- ***minor malformations of the ossicular chain*** may sometimes have a clinical picture similar to otosclerosis
- ***post-traumatic dislocation of the chain or its fixation*** by an adhesive or tympanosclerotic process (anamnesis and otoscopic appearance)
- ***ventilation disorders of the tympanic cavity*** (serous or seromucous otitis, atelectasis) are excluded on the basis of the tympanogram and local appearance
- ***post-otitic sequelae and cholesteatoma*** have a history of ear suppuration and a well-defined otoscopic appearance
- there are also systemic bone disorders that lead to ***fixation of the ossicular chain***, such as
- osteogenesis imperfecta (van der Hoeve syndrome)
- Paget disease
- **sometimes the diagnosis is difficult and is established intraoperatively by exploring the tympanic cavity**

Treatment:

- the treatment of choice is **surgical**
- surgery is indicated when the conductive component of the hearing loss is **at least 20 dB.**
- the surgical techniques currently practiced worldwide are
- stapedectomy
- posterior partial stapedectomy
- stapedotomy
- over time they have replaced fenestration and footplate mobilization techniques, which were frequently followed by refixation
- **stapedectomy** consists of complete removal of the fixed stapes
- the oval window is usually sealed with vein, perichondrium or fascia
- the stapes may be replaced with a prosthesis of **steel wire and connective tissue** or with various types of pistons made of different materials (Teflon, platinum, steel, gold)
- in **stapedotomy** only the stapes superstructure is removed, and a hole is made in the fixed footplate with the drill or a manual perforator (some surgeons also use the laser, especially the argon laser)
- a piston is hung from the long process of the incus and inserted into the footplate hole; it moves freely, transmitting the sound vibrations
- **posterior partial stapedectomy** is an **intermediate technique between the first two**, in which only the posterior third of the footplate is removed
- **the ear with the greater hearing loss** is always operated on **first**
- the interval between the first and second intervention must be **at least one year**, in order to assess the outcome and avoid compromising hearing in both ears
- in the case of **an only hearing ear**, **surgery is avoided,** a hearing aid being preferred

- surgery carried out in time **may prevent progression of the disease to sensorineural hearing loss**
- conservative treatment has uncertain results and is used only exceptionally
- **sodium fluoride** may be given in **6–12 month** courses in forms with rapidly progressive sensorineural hearing loss, or for **4 months** preoperatively in patients with severe forms of active otosclerosis
- **hearing aids** are **NOT useful** in operable cases
- **hearing aids** remain **an alternative** in patients
- who have not received effective surgical treatment
- in whom the operation is contraindicated, or who refuse it
- and cases of cochlear otosclerosis

Evolution and prognosis:

- if **not operated** in the conductive phase, the disease usually evolves with worsening hearing loss over time, which becomes mixed and ultimately predominantly sensorineural
- the younger the age of onset, the faster the evolution and the less favorable the functional prognosis
- the results of surgery are remarkable
- in the hands of an experienced surgeon and in the presence of normal cochlear function, significant and long-term improvement of hearing can be achieved in **80–90% of cases**
- the progression of the disease to marked **hearing loss and the tinnitus are favorably influenced in 80% of cases**
- postoperative sensorineural hearing loss, up to profound hearing loss, occurs in **1%** of cases
- **postoperative vertigo (20%)** usually lasts 1–2 days; cases with prolonged vertigo are rare

HERPES ZOSTER OTICUS

Definition:

- **severe otalgia** and **varicelliform vesicles** affecting the auricle and external auditory canal, associated with **peripheral facial paralysis** of sudden onset

Etiology / Epidemiology:

- **reactivation** of the varicella-zoster virus in the ganglion cells of cranial nerves VII and VIII
- otic herpes zoster is responsible for **10-15% of cases of facial paralysis**

Symptoms:

- altered general condition accompanied by **fever**
- **otalgia** with burning sensation
- **vesicular rash** involving the external auditory canal and the auricle
- within a few days the blue-red blisters form **crusts**

Symptoms:

- altered general condition accompanied by **fever**
- **otalgia** with burning sensation
- facial paresis
- sensorineural hearing loss and dizziness (n. VIII)
- dysphagia (involvement of cranial nerves IX and X)
- **facial pain** (involvement of nerve V)
- headache

- photophobia

Complications:

- complete hearing loss
- persistent dizziness
- persistent pain
- persistent facial paralysis

Diagnostic:

- inspection:
- **varicella-zoster vesicles** located on the lateral surface of the auricle, the concha and the entrance to the auditory canal
- vesicles can appear on the face, neck and oral mucosa
- **crusts** - after vesicles disappear
- otomicroscopy:
- **vesicles** or remnants of vesicles in the external auditory canal and on the tympanic membrane
- audiometric evaluation:
- sensorineural hearing loss
- vestibular tests:
- **spontaneous nystagmus** towards the affected side at the beginning, in the opposite direction later

Differential diagnosis:

- **Bell's palsy** (idiopathic facial paralysis)
- external otitis
- perichondrium
- erysipelas

Treatment:

- **Medical treatment:**
- local treatment of the vesicles with **acyclovir** cream
- antiviral treatment with **acyclovir** (800 mg orally, five times daily)
- **prednisolone** 100 mg/day for 3-5 days, along with proton pump inhibitors, or prednisone 1 mg/kg/day orally for 7-10 days, tapered to zero over the next 10 days
- Surgical treatment:
- if facial paralysis persists, **hypoglossal-facial anastomosis** is required
- Prognosis
- the prognosis for facial function is **less favorable** than in Bell's palsy
- **partial recovery** is common (<50% of patients recover completely)
- the prognosis is **worse in elderly patients**
- the neuralgia often persists for a long time

LABYRINTHITIS

Definition:

- acute or chronic, serous or purulent **inflammatory reaction** within the spaces of the cochleovestibular labyrinth, caused by **bacteria, viruses, spirochetes or fungi**
- the route of infection can be **otogenic, meningogenic or haematogenic**

Viral labyrinthitis:

- **Symptoms:**
- otologic:
- occur during viral infections of the upper respiratory tract: influenza virus, parainfluenza virus, respiratory syncytial virus, coronavirus, adenovirus
- the serous effusion of the middle ear
- vestibular disorders
- sensorineural hearing loss with tinnitus
- without otalgia
- meningeal:
- occur during mumps, measles or parainfectious meningitis
- **the route of infection is the internal auditory canal and the cochlear aqueduct**
- **classic clinical signs of meningitis** (**vomiting, fatigue, neck stiffness, fever, and unilateral or bilateral deafness**)

Bacterial or purulent labyrinthitis

- **Etiology / Epidemiology:**
- bacterial purulent labyrinthitis may be **secondary** to acute otitis media
- purulent labyrinthitis is frequently **followed by meningitis**
- **the most common bacteria** are:
- pneumococcus
- Haemophilus influenzae
- Streptococcus group A
- Escherichia coli
- Klebsiella pneumoniae
- Symptoms:
- otogenic labyrinthitis:
- severe dizziness with nystagmus, vomiting, high fever, severe / profound sensorineural hearing loss and facial paralysis
- meningogenic labyrinthitis:
- classic symptoms of meningitis, vertigo with nystagmus, vomiting, fluctuating hearing loss or complete, bilateral deafness
- Complications of otogenic bacterial labyrinthitis
- meningitis, encephalitis, brain abscess, deafness, death

Hematogenic labyrinthitis:

- ***Treponema pallidum:***
  - **congenital or acquired syphilis** (third stage)
  - fluctuating hearing loss due to endolymphatic hydrops, tinnitus
- ***Mycobacterium tuberculosis:***
  - **tuberculous granulomas** along the spaces of the labyrinth
    - progressive sensorineural hearing loss
- **Diagnosis:**
  - **otoscopy:**
    - serous or purulent otitis media
    - cholesteatoma
  - **hearing exam:**

        - mixed or sensorineural hearing loss
        - deafness
    - **vestibular exam:**
    - **nystagmus** towards the **affected ear** at the beginning, later in the **opposite direction**
    - **imaging exam:**
        - high-resolution CT
    - **microbiology:**
        - **culture** of the purulent ear secretion or of the nasopharynx
        - **CSF examination:** cellularity, protein, glucose level, culture
    - **additional diagnostic procedures:**
        - serology:
        - rubella virus
        - paramyxovirus
        - influenza virus
        - adenovirus
        - syphilis
- **Treatment:**
    - **viral labyrinthitis**
    - **bacterial labyrinthitis**
- **1. Viral labyrinthitis:**
    - **supportive treatment**
    - **corticosteroids** – 500 mg intravenously for 3 days,
    - then oral **prednisolone**, 120 mg daily, with dose reduction by 10 mg daily
    - **antiemetic** drugs and vestibular suppressants
    - **intravenous antibiotics** to prevent bacterial superinfection
- **2. Bacterial labyrinthitis**
    - **supportive treatment**
    - **antibiotics:**
    - cephalosporin (third generation)
    - chloramphenicol and aminoglycoside, depending on the results of the culture
    - **vestibular suppressant and antiemetic** drugs
        - in tertiary syphilis, penicillin G or tetracycline
        - in tuberculosis, tuberculostatic treatment
        - **antipyretics**
    - fluid and electrolyte rebalancing
- **Surgical therapy**
    - any bacterial otogenic labyrinthitis **requires surgery**
    - **myringotomy, insertion of ventilation tubes**
    - **radical or modified radical mastoidectomy** with **labyrinthectomy**

Prognosis:

- **early treatment** is **mandatory** in this infection
- in case of complete **bilateral** deafness following bacterial **meningitis**, **early cochlear implantation** is recommended **before cochlear ossification occurs**

LABYRINTH CONTUSION

Definition:

- **microlesions of the cochleovestibular organ** (bleeding, rupture of membranes, microfractures), caused by a blunt blow to the skull
- it can be associated with fractures of the skull base

Symptoms:

- **high-frequency sensorineural hearing loss** – above 3 kHz
- dizziness and tinnitus
- in **lateral trauma**, the **opposite ear** is **mainly affected**

Complications:

- ossicular dislocations
- perilymphatic fistula
- progressive sensorineural hearing loss, up to deafness
- long-term dizziness

Diagnosis:

- **anamnesis**: detailed history (forensic importance)
- skull inspection: skin lesions, hematoma
- examination of the nasal and oral cavities and the nasopharynx
- **otoscopy:** signs of fracture, hemotympanum, perforation of the tympanic membrane
- hearing examination: tuning forks, audiogram, tympanogram, stapedial reflex
- vestibular examination: to determine irritation or loss of vestibular function

Additional diagnostic procedures:

- high-resolution CT: to exclude fractures
- examination of vestibular function (electronystagmography, video-oculography)
  - **smell tests**

Treatment:

- supportive treatment:
- **anti-inflammatory** and anti-edema medication
- surgical treatment:
- if the middle ear structures are injured, **reconstruction of the ossicular chain** is recommended a few weeks after the injury

Prognosis:

- the prognosis is uncertain:
- **complete recovery** of the cochleovestibular deficit is **possible**, **but not always**

SUDDEN HEARING LOSS

Definition:

- sudden, sensorineural, **unilateral** hearing loss, of **unknown pathogenesis and origin**
- hearing loss can be moderate, profound, or complete
- the disease is accompanied by **tinnitus in 85%** of cases and **vertigo in 30%** of cases

Epidemiology:

- incidence of 11 to 20 cases / 100,000 inhabitants in industrialized countries
- women are affected as often as men, most commonly at the **age of 50 ± 5 years**

Etiology / Classification:

- the cause is **unknown**
- the following mechanisms are involved:
- insufficient cochlear blood flow due to **vasospasm**
- insufficient cochlear blood flow due to **systemic rheological disorders**
- dysfunction of the **ion channels** of the hair cells
- **synaptic dysfunction** due to insufficiency or toxicity of the neurotransmitters involved
- **inflammatory changes** in the endolymphatic sac

The following types of sudden sensorineural hearing loss can be distinguished:

- **idiopathic sensorineural hearing loss** at **high frequencies**, caused by dysfunction of the **outer** (below 50 dB HL) and **inner** (over 60 dB HL) **hair cells**
- **idiopathic sensorineural hearing loss** at **low frequencies**, caused by **endolymphatic hydrops**, possibly as a result of impaired blood flow in the stria vascularis
- **idiopathic sensorineural hearing loss** at **middle frequencies**, caused by **impaired blood flow** in the spiral lamina and hypoxic cellular damage to the organ of Corti
- **idiopathic sensorineural hearing loss** at **all frequencies**, due to **insufficient blood flow** in the modiolar artery, leading to hypoxic damage to the cochlear tissues
- **profound idiopathic sensorineural hearing loss**, due to obstruction of blood flow by **thrombi or emboli** in the common cochlear artery and the modiolar artery

Symptoms:

- patients have the following symptoms:
- acute **unilateral hearing loss** (may go unnoticed if mild)
- **tinnitus** (in about 85% of cases)
- sensation of pressure in the ear
- **vertigo** (in about 30% of cases)
- hearing disturbances: diplacusis, hyperacusis
- secondary **psychoemotional impairment:**
- anxiety
- fear
- stress
- restlessness
- agitation

Diagnosis:

- **1. detailed history**
  - side, onset and duration of the hearing loss
  - **associated symptoms** (tinnitus, dizziness, sensation of pressure in the ear, periauricular dysesthesia)
  - previous sudden sensorineural hearing loss, previous loss caused by exposure to noise or other diseases (previous audiograms).
  - **history** of ENT diseases, head trauma and/or other accidents
  - history of internal, neurological, psychiatric or orthopedic diseases
- **2. complete ENT examination**

- **3. tympanometry and stapedial reflex measurements**
- **4. pure-tone audiogram**
- **5. transient evoked otoacoustic emissions**
- **6. vestibular tests**
- **7. auditory brainstem responses (ABR)**
- **8. brain MRI**
- **9. blood pressure measurement**

Additional diagnostic procedures:

- electrocochleography
- serological tests for Borrelia, syphilis, herpes virus type 1, herpes zoster virus, HIV

Treatment:

- **It's an emergency!**
- spontaneous recovery rates range from 31 to 68%
- patients **CANNOT** be considered fully recovered while the **tinnitus** persists

Differential diagnosis:

- sudden sensorineural hearing loss can be seen in the following diseases:
- perilymphatic fistula
- perilymphatic fistula of the **oval window** is rare
- may occur during head trauma, spontaneously, or following middle ear surgery
- the perilymphatic fistula in the area of the **annular ligament of the stapes** causes sensorineural hearing loss and dizziness
- appears after stapes surgery
- perilymphatic fistula of the **round window** membrane is rare
- has no characteristic clinical symptoms
- can occur after strenuous physical effort
- **hematological diseases** (polycythemia, polyglobulia, leukemia)
- cardiovascular diseases
- **bacterial labyrinthitis** (otitis media, Borrelia, syphilis)
- **viral infection** (adenovirus, herpes virus type 1, herpes zoster, mumps, HIV)
- **disseminated encephalitis** (multiple sclerosis)
- **autoimmune vasculitis** (Cogan syndrome)

Supportive therapy:

- **1. Idiopathic sensorineural hearing loss at high frequencies:**
- daily dose of 250-500 mg intravenous **prednisone** for three days
- prednisone should be accompanied by gastric proton pump inhibitors (40 mg omeprazole / day or 150 mg ranitidine / day).
- intravenous **pentoxifylline** (100 mg) may be used for 5-10 days
- additional administration of **antioxidants - α-lipoic acid** (600 mg / day, oral)
- **2. Low and medium frequency idiopathic sensorineural hearing loss**
- daily dose of 500 mg intravenous **prednisone** for three consecutive days
- in case of partial or no remission, treatment should be continued with oral prednisone for 16 days, together with gastric proton pump inhibitors.

- an **osmotic infusion** can be added, using **mannitol** (15 g in 100 ml solution) and **acetazolamide** (500 mg) intravenously for three consecutive days; acetazolamide (250 mg / day) may be continued orally for 10 days
- in patients with **contraindications to prednisone** (diabetes, severe chronic gastritis, gastric or duodenal ulcer), **initial hemodilution therapy is recommended**
- **3. Idiopathic sensorineural hearing loss at all frequencies**
- daily dose of 250-500 mg intravenous **prednisolone** for three consecutive days
- in the case of partial or no remission, the treatment is identical to that of idiopathic sensorineural hearing loss at high frequencies
- **4. Profound idiopathic sensorineural hearing loss and deafness**
- daily dose of 500 mg intravenous **prednisolone** for three consecutive days
- hemodilution infusion and **α-lipoic acid**
- **prednisone** should be continued orally for 16 days, together with gastric proton pump inhibitors
- **hemodilution infusion therapy and α-lipoic acid** should be continued for another 7 days

MÉNIÈRE'S DISEASE

Definition:

- the syndrome is characterized by a **characteristic triad**:
- spontaneous recurrent attack of **vertigo**
- **fluctuating hearing loss** (initially at low frequencies)
- **tinnitus** and
- aural fullness
- to confirm the diagnosis, there must be **at least two attacks of rotatory vertigo, each lasting at least 20 minutes**
- Ménière's disease is always a **diagnosis of exclusion**, because other diseases of the cochleovestibular system may mimic its symptoms

Etiology / Epidemiology:

- histopathologically, there are **dilations (hydrops)** or **ruptures of the membranes that separate the endolymph from the perilymph**
- the hydrops can develop simultaneously in the **cochlear and vestibular compartments**, but **also in isolation** in a **single compartment**
- the cause of endolymphatic hydrops seems to be a dysfunction of the spiral ligament and/or the endolymphatic sac
- There are several **known causes** of endolymphatic hydrops:
- viral labyrinthitis
- autoimmune diseases of the inner ear (Cogan syndrome)
- otosclerosis
- syphilis
- leukemia
- surgical trauma to the inner ear
- temporal bone trauma etc.
- **when the cause cannot be identified, the term "Ménière's disease" is used**

Symptoms:

- recurrent attacks of **rotatory vertigo**, which can last from minutes to several hours, usually associated with nausea and vomiting

- **at first, unilateral fluctuating sensorineural hearing loss** at **low frequencies**
- **in advanced stages** of the disease, the **hearing loss curves are flat** (threshold of 50-60 dB, speech discrimination 50-60%)
- during the attacks **tinnitus** and a feeling of **ear fullness** appear
- **between attacks, patients are not dizzy, but tinnitus may persist**
- vertigo attacks vary in frequency, intensity and duration, and increase in severity and frequency as the disease progresses
- **the frequency and intensity of vertigo attacks will decrease after about 5-10 years**

Diagnosis:

- clinical records are extremely important
- **physical examination**: between attacks, patients show spontaneous horizontal nystagmus beating towards the opposite side
- **electrocochleography**: a summating potential to action potential ratio greater than 30% is found in 60% of patients with Ménière's disease
- **contrast-enhanced MRI** excludes a retrocochlear lesion (vestibular schwannoma) in a patient with unilateral symptoms

Additional diagnostic procedures:

- ESR, antinuclear antibodies for autoimmune ear disorders
- fluorescent treponemal antibody absorption test **for syphilis**
- **thyroid function tests**, for possible associated hypothyroidism
- **glucose tolerance test**, because diabetes may also be associated
- **caloric test** (with electronystagmography): reduced response on the affected side

Differential diagnosis:

- vestibular schwannoma
- sudden sensorineural hearing loss
- hearing loss due to autoimmune diseases
- migraine

Treatment:

- **Supportive treatment:**
- in **acute attacks** the following are recommended:
- bed immobilization
- vestibular suppressant drugs (diazepam)
- antiemetic (transdermal scopolamine, thiethylperazine, prochlorperazine)
- to **prevent attacks**, the amount of fluid in the inner ear is reduced:
- diet: **low salt intake** (less than 3 g per day) and reduced water intake
- **diuretics**: acetazolamide, hydrochlorothiazide, furosemide
- **potassium** supplementation if necessary
- Additional:
- **vasoactive drugs** – betahistine (improves blood flow to the inner ear)
- **steroids**, to suppress inflammation and/or allergic tissue reactions in the inner ear and endolymphatic sac
- avoid caffeine, alcohol, tobacco
- **Antiviral treatment:**
- oral **acyclovir** (800 mg 3 times) for 3 weeks

- Surgical treatment:
- **endolymphatic sac decompression** controls vertigo attacks in 70% of cases
- **vestibular neurectomy** (through the middle fossa or retrosigmoid approach) has a 98% success rate in improving vertigo, but does not improve hearing
- **labyrinthectomy** results in total hearing loss, but has a high success rate in eliminating vertigo attacks
- **vestibulotomy** – high success rate: vertigo control up to 90%, reduction of tinnitus intensity up to 60%, but with a **risk of hearing loss**

NOISE-INDUCED HEARING LOSS

Definition:

- hearing loss caused by **acute or chronic** exposure to high-intensity sounds
- noise exposure with a peak intensity of **more than 150 dB** and a duration of peak acoustic pressure **greater than 3 ms.**

Etiology / Epidemiology:

- **blast** injuries are seen in the army, in the chemical industry and in car accidents
- they can also result from an injury to the ear such as a **blow to** the side of the head

Symptoms:

- **blast** trauma to the ear causes concomitant damage to the tympanic membrane and possibly disruption of the ossicular chain
- the result is **unilateral or bilateral hearing loss**, moderate or severe, mixed (conductive and/or sensorineural), with **otalgia, tinnitus and vestibular symptoms**

Complications:

- the most feared complications are **profound hearing loss** and **tinnitus**
- other complications are long-term **dizziness** and rupture of the round window membrane

Diagnostic:

- the detailed history
- complete ENT examination
- otoscopy
- audiological test
- vestibular test
- electronystagmography (ENG)
- caloric test

Treatment:

- **Supportive treatment:**
- as in **sudden hearing loss**
- **Surgical treatment:**
- in case of complete hearing loss and suspected rupture of the round or oval window: **tympanotomy and sealing of the perilymphatic fistula**
- in case of tympanic membrane perforation: **myringoplasty, tympanoplasty**

Differential diagnosis:

- rupture of the round window
- sudden hearing loss

- damage to the inner ear by toxic substances
- cranio-cerebral injury

INNER EAR TRAUMA DUE TO INTERMITTENT NOISE — ACUTE ACOUSTIC TRAUMA

Definition:

- exposure to sounds with an intensity **greater than 100 dB** for a few minutes to hours

Etiology:

- exposure to loud sounds causes **metabolic and mechanical damage** to the organ of Corti
- increased oxygen demand and progressive ischemia of the cochlea lead to the production of free radicals, depletion of endogenous cellular antioxidants and cell death

Epidemiology:

- **discos:** average exposure of over 15 minutes – the sound pressure level of the music is **100-110 dB**
- **music concerts:** sound pressures **above 120 dB**
- **headphones:** average sound pressure level of **85-89 dB**
- **musical instruments:** drums and wind instruments

Symptoms:

- acute, bilateral, mild or moderate **sensorineural hearing loss** at high frequencies, with **good recovery**
- **tinnitus** - frequent
- otalgia or vestibular symptoms occur very rarely

Complications:

- permanent hearing loss and / or tinnitus

Diagnosis:

- the patient's history is very important from a medico-legal point of view
- full ENT examination:
- **otoscopy**, without pathological findings
- **hearing tests**: pure-tone audiogram, speech audiogram

Additional diagnostic procedures:

- vestibular testing
- videonystagmography / ENG with caloric vestibular testing
- OAE, ABR

Treatment:

- supportive treatment
- as in the case of **sudden hearing loss**

Differential diagnosis:

- trauma caused by an explosion / gunshot
- sudden idiopathic hearing loss
- labyrinthine contusion

Prognosis:

- the prognosis is **unfavorable**

- recovery is possible

OCCUPATIONAL HEARING LOSS

Definition:

- the hearing loss is **bilateral** and symmetrical and follows **intermittent exposure to noise above 80-85 dB daily** for **6-8 hours** over **several years**

Etiopathogeny:

- **chronic exposure** to noise causes mechanical and metabolic damage to the organ of Corti
- **initially** it causes **loss of the outer hair cells**, which eventually leads to **neuronal damage**
- typically, the initial sensorineural hearing loss is maximal at **4 kHz.**
- the speech frequencies are considerably affected later
- rehabilitation is always possible
- in about **70%** of cases there is **bilateral tinnitus**
- the extent and progression of the hearing loss depend on the intensity and duration of exposure, the frequency composition of the sound and individual susceptibility

Etiology / Epidemiology:

- occupational noise-induced hearing loss occurs mainly in metallurgy, mining, airport workers, etc.

Symptoms:

- **progressive hearing loss** leading to communication problems
- **tinnitus** occurs in 70% of patients
- Complications:
- social isolation as a result of hearing loss
- psychological impairment caused by tinnitus

Diagnosis:

- detailed **patient history**, including occupational exposure
- full ENT examination:
- **otoscopy:** no pathological findings
- **hearing tests:** tuning fork, pure-tone and speech audiogram, tympanometry
- **tinnitus analysis**, with assessment of subjective intensity and masking possibilities

Additional diagnostic procedures:

- OAE
- ABR
- VOG / ENG
- vestibular testing: evaluation of spontaneous and provoked nystagmus with Frenzel glasses

Differential diagnosis:

- presbycusis
- progressive idiopathic hearing loss
- sensorineural hearing loss caused by toxic drugs
- post-traumatic sensorineural hearing loss

Treatment:

- **Supportive treatment:**

- there is **NO** effective **drug treatment** for hearing loss caused by chronic noise
- **glucocorticoids** can be used to treat tinnitus
- hearing loss can be compensated by **hearing aids**
- Additional therapeutic strategies:
- noise avoidance
- use of **hearing protection** when working in a noisy environment

Prognosis:

- after **cessation** of activity in a noisy environment, the **hearing loss does NOT progress**
- if a sensorineural hearing loss **progresses** despite cessation of noise exposure, **other causes** should be investigated

OTOTOXICITY

Definition:

- adverse effects on the **cochlear or vestibular** component of the ear caused by ***pharmaceutical agents***

Etiology / Epidemiology:

- the **most ototoxic** compounds in clinical practice are:
- aminoglycoside antibiotics (streptomycin, neomycin, kanamycin, gentamicin)
- diuretics
- quinine
- chemotherapy agents (cisplatin)
- the incidence has not been accurately determined
- risk factors are:
- impaired renal function
- use of high doses daily
- prolonged administration
- co-administration of more than one ototoxic drug

Symptoms:

- the following **symptoms** may be **temporary or permanent**:
- loud, acute tinnitus (first sign of cochlear damage)
- hearing loss (with or without dizziness)
- nausea
- dizziness
- initially the hearing loss affects high frequencies; low frequencies are involved later
- Audiometric monitoring of patients receiving ototoxic drugs:
- **establishing the baseline hearing level** (air conduction thresholds at 0.5, 1, 2, 4, 6 and 8 kHz)
- **repeating the test** during treatment (every two days or at least every week)
- ototoxicity **is defined as a decrease** of **at least 15 dB at 6 and 8 kHz,** unilaterally or bilaterally, evaluated 5-7 weeks after starting treatment

Prevention:

- discontinuation or change of medication
- **antioxidant therapy** (iron chelators, vitamin E, ascorbic acid)
- **prophylactic** treatment (aspirin)

- in patients with **renal impairment**, the dose should be adjusted

Rehabilitation:

- if there is residual hearing, amplification with a **hearing aid** should be used

PRESBYCUSIS

Definition:

- presbycusis is **progressive sensorineural hearing loss starting in the 5th decade of life**
- the loss may or may not be **symmetrical** and **begins at the highest frequencies**, with or without tinnitus

Etiopathogeny:

- **degeneration of** the hair cells, cochlear neurons, stria vascularis, cochlear nerve and central auditory pathways

Etiology / Epidemiology:

- hearing loss is **caused by**:
- the **physiological** processes of aging
- **exogenous degeneration** of parts of the inner ear and central auditory pathway
- the consequence of **environmental influences, dietary habits, drug toxicity, noise exposure**, etc.

Symptoms:

- the hearing loss is **progressive**, with the greatest loss at high frequencies
- **speech comprehension** is reduced, especially in noise, with loss of discrimination
- **tinnitus is** present

Complications:

- **psychosocial isolation** caused by the loss of the ability to communicate
- **depression** that can be triggered by tinnitus

Diagnosis:

- detailed patient history, including occupational noise exposure and family history
- complete ENT examination
- otoscopy, hearing tests: tuning fork, pure-tone audiogram, speech audiogram, tympanogram
- the sensorineural hearing loss is symmetrical, at **frequencies above 1 kHz.**
- speech comprehension is poor

Differential diagnosis:

- hereditary sensorineural hearing loss
- otosclerosis with sensorineural hearing loss
- sensorineural hearing loss in neurological or autoimmune diseases
- noise-induced hearing loss
- hearing loss induced by ototoxic drugs
- traumatic sensorineural hearing loss

Treatment:

- management includes the following:
- **hearing aids:** must be prescribed as soon as possible

- **socialization**, possibly with concomitant psychotherapy
- **medical therapy** only when the progression of presbycusis is rapid: **α-lipoic acid or antioxidants (vitamin C or E)**

Prognosis

- normally, gradual loss of hearing is inevitable

CHILDHOOD HEARING LOSS

Definition:

- hearing impairment in childhood refers to any hearing loss occurring **between birth and late childhood (16 years)**
- it can be unilateral or bilateral
- it affects **language acquisition** and can also be classified in relation to it:
- prelingual (0-2 years)
- perilingual (2-4 years) and
- post-lingual (over 4 years old)

hearing loss can be classified according to severity as:

- **mild** (hearing level between 10 and 39 dB.)
- **moderate** (40-69 dB.)
- **severe** (70-94dB.)
- **profound** (hearing loss is greater than 95 dB.)

Definition:

- hearing loss can be:
- transmission
- sensorineural
- mixed
- according to evolution:
- temporary
- fluctuating
- progressive
- permanent

Etiology / Epidemiology:

- **Conductive hearing loss**
- the most common cause of hearing loss in childhood is ***serous otitis media***
- the hearing loss is usually transient; if not, it can be treated by ***myringotomy, placement of ventilation tubes and adenoidectomy***
- **other causes** of conductive hearing loss:
- chronic otitis media (mesotympanic)
- cholesteatoma
- congenital malformations of the external auditory canal or middle ear
- **Sensorineural hearing loss** can be caused by:
- dysfunction of the organ of Corti (**sensory hearing loss**)
- dysfunction of the auditory nerve (**neural hearing loss**) – less common
- dysfunction at the interface between the sensory hair cells and the spiral ganglion cells (**auditory neuropathy**)

- permanent hearing loss has an incidence of about 1-2 / 1000 children

Etiology / Epidemiology:

Congenital hearing impairments:

- in **> 50% of cases** a **genetic cause** can be identified
- ¾ of these cases are non-syndromic
- ¼ are associated with hereditary syndromes that have other manifestations in addition to deafness
- most commonly, they are **autosomal recessive**
- most commonly, mutations are found in the **GJB2 gene**, encoding **connexin 26,** an inner ear protein that ***maintains the endocochlear potential***
- other hereditary causes are **autosomal dominant**, with varied phenotypes, or X-linked

Acquired hearing impairments

- **during** the **perinatal period**, they are due to:
- hypoxemia
- severe infections
- prolonged jaundice of the newborn and others
- **after the perinatal period**:
- **bacterial meningitis - the most common cause**
- 2% of children affected by meningitis may develop permanent, profound bilateral hearing loss
- **vaccines against Haemophilus influenzae type B and Streptococcus pneumoniae** reduce the risk of meningitis
- **other causes** include:
- infections (measles, mumps, varicella)
- trauma
- diseases of the inner ear
- administration of ototoxic drugs
- **Progressive hearing impairment:** may be associated with **genetic mutations** that cause syndromes such as:

Pendred syndrome

- genetic syndromes associated with an enlarged vestibular aqueduct

Usher syndrome

- pigmented retinitis
- progressive hearing loss

Alport syndrome

- other **genetic forms** of progressive hearing loss occur in adolescence; most often the cause of progressive hearing loss remains unknown

Symptoms:

- delayed or absent normal **language development**
- the diagnosis of even severe hearing loss is often **delayed** until around the age of 2-4 years
- **the risk factors** associated with permanent hearing loss should raise suspicion:
- ***familial history*** of hearing loss
- infections during ***pregnancy*** (rubella, toxoplasmosis, cytomegalovirus)
- ***prematurity***, low birth weight

- prolonged newborn ***jaundice***
- administration of ***ototoxic*** drugs (aminoglycosides)
- ***craniofacial malformations*** and ***hereditary syndromes***
- deficiencies in language development involve both the **ability to understand language and the ability to speak**
- it is important to monitor the child's language development, as some children may develop **progressive hearing loss**

Diagnosis:

- Neonatal screening for hearing impairments
- **universal neonatal screening** is recommended, or at least screening in the presence of risk factors
- **all newborns should be tested during the first days of life:**
- automated measurement of **otoacoustic emissions (OAE)**, such as transient evoked OAE (TEOAE) and distortion product OAE (DPOAE)
- brainstem evoked potentials (BERA)
- if the screening shows abnormal results it is repeated, and then **comprehensive hearing tests** are carried out to confirm or exclude hearing loss

Audiological tests:

- There are **subjective** and **objective** hearing tests
- **subjective tests** require some form of response from the subject tested, while **objective tests** can be performed without active feedback
- the diagnosis must be based on a combination of subjective and objective tests
- the diagnosis should be re-evaluated as the child develops in order to **distinguish** between **transient, permanent or progressive hearing loss**

The subjective tests are:

- **Behavioral audiometry** (0-2 years old):
- spontaneous responses to sounds are observed, such as the baby becoming quiet, rapid blinking or widening of the eyes
- testing is bilateral, in free field
- the hearing thresholds obtained are generally ***20-30 dB above the actual hearing thresholds***
- **Play audiometry** (**2-5 years old**):
- requires the subject to be engaged in an activity
- the test uses a conditioned response (stacking cubes)
- testing is in free field and bilateral
- **Pure-tone threshold audiometry** (**from 3.5-4 years of age**):
- the child must be able to ***wear headphones and cooperate***
- Vocal audiometry:
- speech tests with ***age-appropriate language material***

The objective tests are:

- **Screening tests:**
  - designed to detect **hearing loss greater than 30-40 dB.**
- **Tympanometry:**
  - to detect middle ear problems
  - stapedial reflex measurements – estimate the hearing threshold
- **OAE:**

  - reflect the normal activity of the **outer hair cells**
  - are **present** when the **hearing loss does not exceed 30-40 dB.**
  - they **do NOT reflect the function** of the inner hair cells and the auditory nerve
  - may be present in the case of auditory neuropathy
- **Auditory responses of the brainstem:**
  - the synchronized **neural activity** of the auditory pathway (spiral ganglion and cochlear nerve, cochlear nucleus, lateral lemniscus, superior olive and inferior colliculus) is recorded **in response to click stimuli** (broad frequency) **or tone bursts** (limited frequency)
  - the method **requires sedation**
- **Evaluation of language and speech development**
- **Ophthalmological and neurological examination**
- **Imaging diagnosis:**
  - **high-resolution CT** is the method of choice to evaluate the bone structure of the ear
  - **MRI** is indicated to visualize the **contents of the inner ear** and the **auditory, vestibular and facial nerves** in the internal acoustic meatus
- **Genetic testing and counseling:**
  - **specific genes** associated with Waardenburg, Pendred and Usher syndromes have been identified
  - the **GJB2 gene** encoding **connexin 26** has been identified
  - the number of **mutant genes** associated with deafness continues to increase
  - **over 100 mutations** have been described; therefore negative findings on genetic testing do not exclude a genetic origin of the hearing loss
- **Conservative therapy:**
  - **early intervention** is a key factor in preventing the sequelae of hearing loss
  - even children **3-6 months old** can **wear hearing aids**
  - the first step in the treatment of **permanent hearing loss** is the provision of amplification by appropriate means (**the hearing aid**)
  - in **ear malformations**, a **bone conduction hearing aid** is used
  - **monitoring of the child's development**: reactions to sound and speech, as well as language development in children fitted with a hearing aid, should be monitored by parents, audiologists, teachers and therapists
  - if **speech and language development remain insufficient** in patients with severe or profound hearing impairment despite optimal fitting of the hearing aid, a **cochlear implant** is required

Surgical treatment:

Conductive hearing loss

- in **otitis media with effusion**, if conservative treatment fails, **myringotomy** and placement of **transtympanic ventilation tubes** should be performed
- **surgical treatment** is indicated in **chronic mesotympanic otitis media and chronic otitis media with cholesteatoma**
- in **ear malformations**, the options are reconstruction of the external auditory canal and the ossicular chain, implantation of active middle ear devices, or fitting of a bone conduction hearing device

Sensorineural hearing loss

- cochlear implant

Prognosis:

- early identification and treatment of hearing loss, together with adequate rehabilitation and educational support, can yield excellent results, even in children with profound deafness
- **negative** prognostic factors include:
- advanced age at the time of diagnosis
- the presence of other cognitive disabilities
- improper communication strategies
- inadequate rehabilitation and educational support and
- poor socio-economic situation

VESTIBULAR NEURITIS

Definition:

- **unilateral** loss of vestibular function, which might be partial or total
- frequently caused by inflammation of **viral etiology**

Etiology/Epidemiology:

- **viral** origin **(herpes simplex virus)**, similar to facial nerve paralysis
- owing to **immunologic deficiencies**, herpes simplex viruses, which were already present in the patient as a result of an earlier infection, are reactivated and destroy vestibular sensory fibers
- one of the **most frequent peripheral vestibular disorders** (**25% of vertigo patients**)

Symptoms:

- marked by the acute appearance of severe vertigo, mostly purely rotatory, sometimes accompanied by vomiting, nausea and ataxia
- in the acute state a **violent horizontal-rotatory nystagmus**, beating towards the intact side, is always present
- hearing impairment and tinnitus are not part of vestibular neuritis
- in the **acute state** a tendency to fall is obvious, so **prevention of falls is necessary** to avoid orthopedic sequelae
- not infrequently, benign paroxysmal positional vertigo follows vestibular neuritis after a short interval; this is called ***Lindsay-Hemenway syndrome***.

Diagnosis:

- sudden onset of the vertiginous complaints, which decrease over a period of several days
- the only objective sign is a strong horizontal-rotatory nystagmus, beating to the intact side
- **the caloric test** shows hypofunction of the affected side, which at the beginning is not compensated in the rotatory test
- **vestibulospinal tests** show a marked deviation and a tendency to fall towards the affected side.

Additional diagnostic procedures:

- **Hearing tests** to rule out other lesions
- **MRI** to rule out **vestibular schwannoma,** which very rarely presents with the same symptoms

Differential Diagnosis:

- the differential diagnosis of vestibular neuritis is **simple**, because vertigo lasting several days is very characteristic; therefore:
- benign paroxysmal positional vertigo

- vestibular paroxysmal
- Ménière's disease
- vestibular migraine can be excluded by careful history-taking
- although **vestibular schwannoma** only very rarely presents with vertigo

Treatment:

- exclusively conservatory:
- in the early stages **H1 antagonists** are recommended, but **not for more than 2 days**; **steroids** – in the first **14 days** (i.v. 500 mg, tapered to 0 over 10 days)
- vestibular habituation training should be started to induce a rapid vestibular compensation
- to accelerate vestibular compensation active movements (**sports**), stimulating agents such as **caffeine** and **avoidance of calming procedures** such as bed rest seem useful

Prognosis:

- the prognosis is generally **favorable**
- factors which **inhibit vestibular compensation** cause the symptoms to persist for a long time:
- ageing
- sedatives
- CNS lesions

BENIGN PAROXYSMAL POSITIONAL VERTIGO (BPPV)

Definition:

- benign paroxysmal positional vertigo (BPPV) is a **mechanically induced vertigo** caused by **canalolithiasis or cupulolithiasis**

Etiology/Epidemiology:

- normally **otoliths are fixed in the otolithic membrane**
- after head trauma, in old age or idiopathically, otoliths can become loose and travel into one or more of the semicircular canals
- **BPPV is one of the most frequent kinds of vertigo**, mainly in the elderly
- the semicircular canals are not affected equally: in **96% of cases the posterior vertical canal** is involved, the horizontal canal in 3% of cases and the anterior vertical semicircular canal in only 1% of cases.

Symptoms:

- short duration, only a few seconds
- triggered by head movement
- BPPV does not appear when the head is not moving

Complications:

- disequilibrium and fall

Diagnosis:

- Vertigo appears when **the head is moved and lasts for a few seconds (<60)**
  - apart from the characteristic complaints, the diagnosis is made by **observation of nystagmus with Frenzel glasses**
  - by specific positioning of the head in the plane of one of the semicircular canals (**Hallpike maneuver**), BPPV can be demonstrated if a typical nystagmus appears

- the involved semicircular canal can be identified by analysis of the nystagmus, because the **pattern of eye movements for each semicircular canal is known**

Treatment:

- because of the mechanical etiology of BPPV, **only mechanical treatment is adequate,** by the **Epley or Semont maneuvers**
- both have essentially the same aim, namely to bring the dislodged otoliths, by specific movements of the head, to a "neutral point" in the vestibular apparatus near the utricle
- surgical treatment is rarely indicated

Prognosis:

- BPPV prognosis is **favorable**
- after **one liberatory maneuver** about **70% of patients no longer have complaints**
- in the **remaining 30%**, **repeated liberatory maneuvers** lead to a complete cure
- if necessary, Brandt–Daroff exercises must be continued
- **the recurrence rate is relatively high**

MOTION SICKNESS

According to the environment: car, sea or space sickness

Definition:

- motion sickness is a **special kind of physiological vertigo**, induced by an **unusual stimulation of the multisensory system**, which normally guarantees adequate orientation in space.

Etiology/Epidemiology:

- motion sickness develops when there is a **sensory conflict between the different sensory systems responsible for orientation in space**
- both **intrasensory mismatch** (within the **vestibular system**) and **intersensory mismatch** (between the **vestibular and visual system** for example) can trigger the symptoms of motion sickness
- the **crucial condition** is that **different sensory receptors** give **different signals** about the **passive motion** of an individual in space.

Symptoms:

- motion sickness is characterized by:
- **nausea, pallor, yawning, vomiting and, above all, a feeling of severe discomfort**
- these symptoms last not only during the conflicting stimulation but also for a certain period afterwards, when the motion stimulation has stopped

Complications:

- **there are no complications**
- falls and aspiration caused by vomiting are sequelae of the symptoms themselves

Diagnosis:

- motion sickness is diagnosed **very simply**, because the **coincidence of the inducing motion and the typical symptoms is pathognomonic**
- further diagnostic procedures **are not necessary**
- **Treatment:**

- **prophylactic therapy** can be useful **if it is predictable** that a sensory conflict inducing motion sickness will occur
- **the best prevention** consists of vestibular habituation training, intended to prepare the orientation system for conflicting stimulation
- as **prophylactic drug treatment**, **scopolamine** in the form of a transdermal patch can be recommended
- when the symptoms of motion sickness appear, one can try to break free from the sensory conflict situation
- in the case of seasickness, the affected person should leave the cabin and go on deck, then fixate on an object not too far away. In this way the correspondence between visual and vestibular information increases
- drug treatment consists of an **H1 histamine antagonist** such as **dimenhydrinate or meclizine**
- it must be pointed out that all H1 antagonists have sedative side effects.

Prognosis:

- is favorable
- after a few hours the symptoms disappear

FACIAL NERVE — IDIOPATHIC FACIAL NERVE PARALYSIS

Synonyms:

- Bell's palsy, *a frigore* facial palsy

Definition:

- Bell's palsy is characterized by the **sudden onset** of incomplete or complete paralysis of facial nerve function **without detectable cause**
- it is usually **unilateral**

Etiology / Epidemiology:

- Bell's palsy affects about two in 10,000 people
- the cause remains unclear
- the most probable cause is a **mononeuritis** triggered by reactivation of **herpes simplex virus type 1** in the geniculate ganglion
- this increases the nerve diameter and leads to compression in its bony canal as it courses through the temporal bone
- **other infections** known to be associated with facial palsy are early-summer **meningoencephalitis virus, Borrelia burgdorferi, herpes zoster, influenza virus, HIV and HTLV-1**

Symptoms:

- it is a **diagnosis of exclusion:**
- the palsy usually **starts suddenly** and ranges from weakness of facial movements to total facial paralysis
- it may also include **difficulty in eye closure** on the affected side
- face twitching
- **drooling of saliva** due to the drooping corner of the mouth
- dry eye or mouth
- loss or alteration of the sense of taste
- and sometimes hyperacusis and dysaesthesia on the affected side

Complications:

- **disfigurement** from loss of facial movement
- **difficulty** with eating, drinking and speaking
- **dry** eye with corneal ulcers and keratitis due to lagophthalmos
- **chronic spasm** of the facial muscles or eyelids
- **chronic taste** abnormalities

Diagnosis:

Recommended European Standard

- *History*: time and type of onset (slow, rapid), first time or recurrent, additional symptoms (neurological, hearing, vertigo, taste, pain)
- *Inspection*:
- usually **unilateral facial weakness** [classification according to House and Brackmann (HB)
- **vesicles** in or around the auricle and external auditory canal (Ramsay Hunt syndrome similar to herpes zoster oticus)
- *Palpation*: parotid gland (tumor), dysaesthesia of the auricle (Ramsay Hunt syndrome similar to herpes zoster oticus)
- *Complete ENT examination*
- *Micro-otoscopy*: *effusions, cholesteatoma, blisters of herpes zoster*
- normal auditory canal and tympanic membrane in Bell's palsy.
- *Hearing tests*: tuning fork, audiogram, tympanogram, **stapedial reflexes** (topography of facial nerve palsy)

Other diagnostic tools:

- **parotid gland sonography**: to rule out a parotid gland tumor

Topographic diagnosis:

- **Schirmer test**: reduced tear production if palsy is **at or proximal to the geniculate ganglion** (greater petrosal nerve)
- **Stapedial reflex:** elevated threshold or absence if the site of the lesion is located **in or proximal to the tympanic segment** of the facial nerve
- **Taste evaluation:** loss of taste on the anterior two thirds of the tongue on the affected side shows loss of function of the chorda tympani and indicates **damage proximal to the second genu**
- Note that if all of these tests result in normal findings, the lesion must be distal to the tympanic segment of the facial nerve (e. g. mastoid or extracranial segment)

House-Brackmann classification

GRADE 1 (normal):

- **normal functions** in all areas

GRADE 2 (mild dysfunction):

- At rest:
- **slight weakness** noticeable only on close inspection
- normal symmetry of forehead
- Motion:
- **ability to close eye** with minimal effort and **slight asymmetry**
- ability to move corners of mouth with maximal effort and **slight asymmetry**

- no synkinesis, contracture or hemifacial spasm

GRADE 3 (moderate dysfunction):

- At rest:
- **obvious, but not disfiguring** difference between two sides
- no functional impairment
- noticeable but not severe synkinesis, contracture and/or hemifacial spasm
- normal symmetry and tone
- Motion:
- slight to no movement of forehead
- ability to close eye with maximal effort and obvious asymmetry
- ability to move corners of mouth with maximal effort and obvious asymmetry
- Patients who have **obvious, but not disfiguring synkinesis**, contracture and/or hemifacial spasm are **grade 3**, regardless of the degree of motor activity

GRADE 4 (moderate-severe dysfunction):

- At rest:
- obvious weakness and/or disfiguring asymmetry
- Motion:
- no movement of forehead
- **inability to close eye completely** with maximal effort

GRADE 5 (severe dysfunction):

- At rest:
- only barely perceptible motion
- possible asymmetry with drop of corner of mouth and decreased or absent nasal labial fold
- Motion:
- no movement of forehead
- incomplete closure of eye and only slight movement of lid with maximal effort, slight movement of corner of mouth
- synkinesis, contracture and hemifacial spasm are usually absent

GRADE 6 (total paralysis)

- loss of muscle tone
- asymmetry
- no movement
- no synkinesis, contracture or facial spasms

Treatment:

- if the patient is referred **early,** during the **first week**, **medical treatment** is the initial form of therapy
- the HB (House and Brackmann) grading system is used to assess clinical outcome
- **three different situations** are to be differentiated:
- 1. If the palsy is **incomplete and not worsening**, appearing benign, **medical treatment** is optional and the patient is observed clinically until total recovery (HB 1)
- 2. If the palsy is **complete** but the **EMG pattern is favorable** (suggesting neurapraxia), or if daily electroneuronograms show less than an 85% denervation rate, **medical treatment** is applied and the patient is closely followed up clinically and, if possible, electrophysiologically

- 3. If the palsy is **complete** and the **ENOG pattern is unfavorable** (90% denervation rate), or if follow-up of an initially benign-looking Bell's palsy shows clear clinical and electrical deterioration, **surgical decompression** of the nerve can be discussed during the first 3 weeks after onset
- confirmation of the **poor prognosis by EMG**, which is more reliable than ENOG alone, and an MRI scan are two prerequisites
- the aim of **surgery is to decompress** the probably swollen nerve by opening the Fallopian canal at the level of the geniculate ganglion and the labyrinthine portion up to the meatal foramen, thus avoiding total denervation and its unavoidable sequelae (synkinesis, hemifacial spasm; HB 3–4)
- if the patient is referred late **(after 3 weeks)**, the usefulness of any medication is debatable
- great attention must be paid to these patients, with careful monitoring of their outcome
- if no recovery at all is observed **6 months after onset**, contrary to the natural course of the disease, the diagnosis of Bell's palsy must be reconsidered
- a full MRI and CT scan workup, searching for intrinsic tumors of the nerve, must be performed and surgical exploration of the nerve could be considered

Conservatory treatment:

- **corticosteroids:**
- i.v. 500 mg/day for 3 days
- might be followed by oral administration for a few days
- may reduce facial nerve edema and pressure
- the earlier they are introduced in the treatment scheme, the more effective they are
- **antiviral drugs** - acyclovir:
- i.v. 3x10 mg/kg/day or orally 5x800 mg/day
- **combined therapy** leads to faster recovery than monotherapy
- if **eye closure is not complete**, the eye should be protected with ointment or eye drops

Additional therapeutic options:

- pentoxifylline
- myofacial training
- electrotherapy
- if **Lyme disease** is suspected, **tetracycline, macrolides or a 3rd-generation cephalosporin** should be administered for 14-21 days

Surgical treatment:

- **normally** surgical treatment **is not indicated**
- surgery is **indicated** when **EMG shows a severe or complete nerve lesion**
- to be effective, it should be performed **within 3 weeks of the onset of facial palsy**

Differential diagnosis:

- ***Complications*** of acute otitis media, cholesteatoma, middle ear tuberculosis
- ***Post-traumatic facial palsy*** (temporal bone fracture, stab or gunshot wound), iatrogenic after middle ear or parotid surgery
- ***Tumor of the parotid gland***, ***vestibular schwannoma, cerebral tumor, facial schwannoma***. All patients with Bell's palsy lasting more than 4 weeks and all patients with recurrent facial palsy should be investigated for the presence of a tumor
- ***Central facial palsy*** (e.g. following ischemic stroke): the frontal branch of the facial nerve is functional!

- ***Ramsay Hunt syndrome*** (zoster oticus): vesicles
- ***Lyme disease***
- ***Melkersson-Rosenthal syndrome***: recurrent bilateral or alternating facial palsy with cheilitis, fissured tongue and facial edema (familial, sometimes with rheumatologic symptoms)
- Congenital hemifacial dysplasias
- ***Dissection of the internal carotid artery***: subintimal or adventitial bleeding occurring secondary to sports, chiropractic manipulation, trauma (distortion of the cervical spine) or spontaneously. Partial interruption of perfusion of the internal carotid artery leads to compression of the cranial nerves. Functional deficits can also be caused by ischemia of the cranial nerve nuclei (VI, VII, IX, X, XI). Similar spontaneous or post-traumatic conditions are also possible along the vertebral artery
- ***Bilateral simultaneous or recurrent alternating facial palsy***
- ***Neoplastic:*** leukemia, pontine gliomas, brain metastasis, carcinomatous meningitis
- ***Neurologic:*** multiple sclerosis, polyneuropathies, Guillain-Barré syndrome, bulbar paralysis, Parkinson's disease, pontine hemorrhage, bulbospinal neuropathy, Bannwarth syndrome
- ***Autoimmune diseases:*** systemic lupus erythematosus, Wegener's granulomatosis

Prognosis:

- **60-80% of patients recover fully** within a few weeks or months
- incomplete palsy has an excellent prognosis
- factors of **unfavorable prognosis**:
- complete facial nerve palsy
- rapid onset facial nerve palsy
- pain
- neurootologic symptoms
- delayed treatment