Ear, Nose, and Throat Diseases

With Head and Neck Surgery

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Oliver Kaschke
Tadeus Nawka
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Foreword by
Heinz Stammberger

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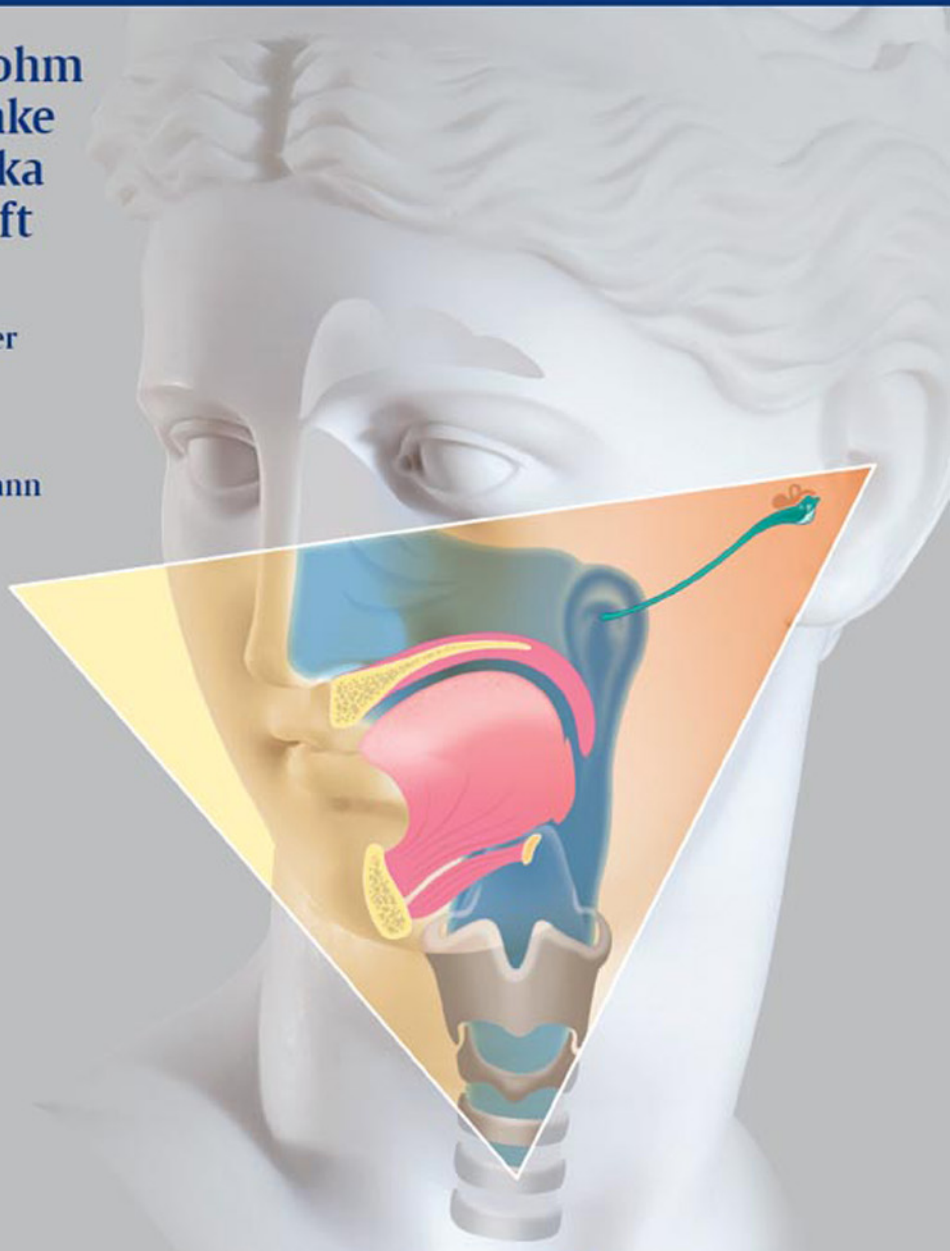
Walter Becker
Hans Heinz Naumann
Carl Rudolf Pfaltz

3rd edition



Thieme

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3rd edition

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Foreword

After two decades of ever-increasing subspecialization, it is very encouraging and rewarding to see that otorhinolaryngology, head and neck surgery still can and should be seen as an entity. In the new edition of this marvelous textbook, Professor Hans Behrbohm and his colleagues, who have taken over authorship from Professors Walter Becker, Hans Heinz Naumann, and Carl Rudolf Pfaltz, provide all the anatomical, physiological, diagnostic, and therapeutic evidence for this, should anyone be in doubt.

This brilliantly illustrated textbook addresses advanced medical students and doctors alike, and provides a modern overview of the complexity of our fascinating specialty. As such, it may serve as both a learning and a teaching reference, summarizing our present knowledge of the different aspects of all components contributing to otorhinolaryngology, head and neck surgery.

It adds to the value and beauty of this book that not only the connections between and interdependence of various subspecialties are displayed, but the interdisciplinarity with neighboring specialties is also addressed, e.g., with neurosurgery and neurology for skull base and intracranial structures, with chest medicine specialties for tracheal and pulmonary disorders, or with gastroenterology for disorders of the upper digestive tract.

Thieme Publishers have to be congratulated for providing the plentiful and outstanding colored anatomical and schematic illustrations, in addition to the photographic images.This is a truly international textbook that will be of great benefit to all of its readers!

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Preface

The first edition of *Ear, Nose, and Throat Diseases* was written by Professors Walter Becker (1920–1990), Hans Heinz Naumann (1919–2001), and Carl R. Pfaltz (1922–2003) and published in 1988. Since then, and over several English and German editions, this book has presented the essential knowledge of otorhinolaryngology, head and neck surgery in a concise and highly accessible format. In addition to this, it also provided advanced information to facilitate a better understanding of the diagnostic and therapeutic issues and challenges of the specialty. The scope and didactic presentation of its content made this book attractive to medical students, specialist trainees, residents, interested practitioners and specialists, both as a textbook and reference source. Because of its continuing success, a new edition of *Ear, Nose, and Throat Diseases* was very much in demand, and we are grateful to Thieme Medical Publishers for the opportunity to prepare this new volume.

We have been sensitive to the fact that the book has been important on an international scale and we have strived to maintain this status, recognizing that the range of presenting conditions and their management will vary between various countries.

As a surgical specialty, otorhinolaryngology–head and neck surgery has shown a fast-paced, progressive development with regard to diagnosis and treatment and the increased understanding of pathophysiological principles. It was therefore necessary to restructure some chapters completely and to revise and update the remaining ones in order to bring the new edition up to the present standard of scientific and technical knowledge and practice in this specialty.

As in the previous editions, basic information is presented in normal type; supplementary and advanced information is given in a smaller type.

The figures supplementing the text have received particular attention, as they are immensely important and key to understanding the text. Numerous new drawings have been prepared; all of the existing drawings have been recreated in full color as well as revised and updated where necessary. Visual findings are often the key to diagnosis for the otorhinolaryngologist, so all of the previous clinical photographs have been replaced with new images, and the number of images has been greatly increased.

Our medical illustrator, Ms Katja Dalkowski MD, has made a substantial contribution to the visual appearance of the book, and we would like to express our gratitude for her invaluable assistance and commitment to the project. Our constant contact with Mr Stephan Konnry, our editor at Thieme Medical Publishers, his thorough editorial work and clearing up of many, many details and questions made an extremely valuable and beneficial contribution to the book.

The book project was generously supported by Dr h. c. mult. Sybill Storz, to whom we are extremely grateful. We are also grateful to the patients who were willing to allow photographs of their sometimes serious diseases to be published for the purpose of medical education.

It is our hope that this new edition will continue to be a valuable practical guide to the vast range of otorhinolaryngology, head and neck surgery for its readers—the students of medicine and dental medicine, as well as physicians and surgeons who are either in training or practicing as established specialists in various disciplines within the field.

Hans Behrbohm
Oliver Kaschke
Tadeus Nawka
Andrew Swift

We would like to express our warm thanks here to our former head of department and teacher, H.-J. Gerhardt—now Professor Emeritus at the Department of Otorhinolaryngology at the Charité Hospital in Berlin (where he held the chair from 1973 to 1994)—for providing rare pictures of disease conditions from his image archive.

The *Berlin Diagnostic Workshops* have provided an important stimulus for the work on this book. We have been involved in obtaining and assessing visual and auditory organ findings for more than 10 years and have had the opportunity to discuss these discoveries with many physicians.

Hans Behrbohm
Oliver Kaschke
Tadeus Nawka

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1 Ear

Applied Anatomy and Physiology

Embryology

Inner ear. The sensory organs for hearing and balance develop from ectoderm. The *membranous labyrinth* develops from the ectodermal otic placode. *Embryonic mesenchymal tissue* surrounding the *membranous labyrinth* is converted into cartilage and also, by a process of vacuolization, into a fine reticular network that forms the inner layer of the *perilymphatic space.* The outer layer of the cartilage forms the *labyrinthine capsule.*

Middle ear. The eustachian tube and the mucosa of the middle ear arise from a diverticulum of the first pharyngeal pouch (endoderm).

The malleus and incus develop from Meckel cartilage, which emerges from the first branchial arch and is supplied by the trigeminal nerve. The stapes develops from the second branchial arch and is supplied by the facial nerve.

Myxomatous embryonic connective tissue lies between the ectodermal and endodermal ingrowths and makes a preformed middle ear cavity. If this myxomatous tissue does not involute properly after birth, the epitympanic recess remains as a narrow cleft. This is easily occluded by inflammation and creates a predisposition for chronic ear disease to develop.

External ear. The external meatus and the tympanic membrane develop from an ectodermal diverticulum between the first and second branchial arches. Developmental disorders may therefore cause deformities of both the external and middle ears. Bilateral lesions causing severe conductive deafness or a psychologically unacceptable deformity should be corrected, for both esthetic and functional reasons (see pp. 49 and 94) (Figs. 1.1, 1.2).

Basic Anatomy

The hearing and balance systems consist of the *peripheral receptor apparatus* (i.e., the ear in the strict sense), *neurological pathways,* and *centers in the central nervous system.* Two main subdivisions can therefore be distinguished:

Peripheral part:

- The external, middle, and inner ear.
- Vestibulocochlear nerve with its two parts, the cochlear and the vestibular divisions.

Central part:

- Central auditory pathways.
- Subcortical and cortical auditory centers.
- Central balance mechanism.

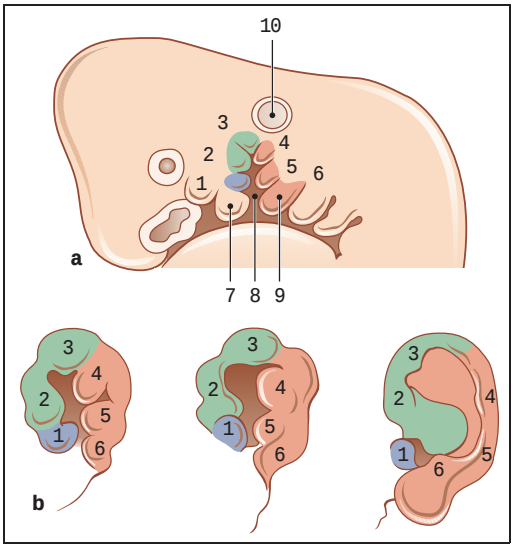


Fig. 1.1a, b Development of the external ear.
a An 11-mm embryo, lateral view.
b Development of the outer ear from six hillocks arising from the first and second branchial arches.
1, Tragus; 2, crus helicis; 3, helix; 4, crus anthelicis; 5, antihelix; 6, antitragus; 7, first branchial arch; 8, branchial cleft; 9, second branchial arch; 10, auricular plate.

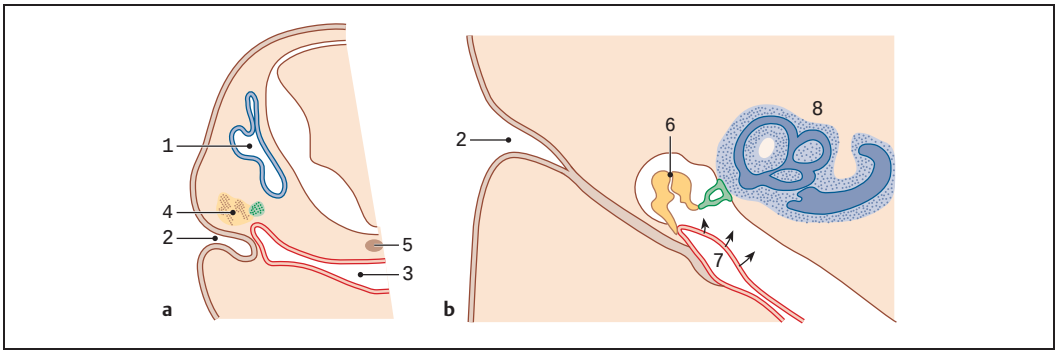


Fig. 1.2a, b Developmental stages of the external auditory canal, middle ear, and labyrinth. The epithelial auditory canal pouch with the tympanic plate opens through epithelial necrolysis (apoptosis) in the seventh month. The mesenchyme of the stapes develops from the second visceral arch; the remaining structures of the middle ear develop from the first visceral arch.
a Approx. 8th week. 1, Otic vesicle; 2, primary auditory canal; 3, tubotympanic recess; 4, mesenchymal condensation; 5, acousticofacial ganglion.
b Approx. 7th month. 2, Primary auditory canal; 6, primordium of the auditory ossicles; 7 tympanic cavity; 8, primordium for the labyrinth.

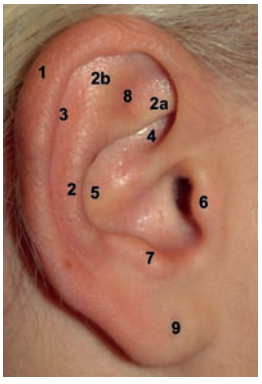


Fig. 1.3 Topography of the external ear structures. 1, Helix; 2, antihelix (a: inferior crus, b: superior crus); 3, scaphoid fossa; 4, cymba conchae; 5, cavum conchae; 6, tragus; 7, antitragus; 8, triangular fossa; 9 earlobe.

The *anatomic boundary* between the *peripheral* and *central* parts is the point of entry of the eighth cranial nerve into the brain stem (the cerebellopontine angle), at which point the peripheral part of the vestibulocochlear nerve passes into the central part, interspersed with glial cells. In functional terms, however, the peripheral neurons end in the primary centers.

External Ear

The *auricle* consists of a framework of elastic cartilage covered by skin (Fig. 1.3), located between the temporomandibular joint anteriorly and the mastoid process posteriorly. The skin adheres tightly to the perichondrium on the anterior surface, but is more loosely attached posteriorly. For this reason, contusions of the anterior surface often lead to detachment of the skin–perichondrial layer and to the formation of a hematoma (see p. 55).

The *external meatus* is ≈ 3 cm long, consisting of an outer cartilaginous part and an inner bony part. The cartilaginous meatus is curved and lies at an angle to the bony part. The tympanic membrane and the middle ear lying beyond it are thus protected from direct trauma.

> Note: For an otoscope to be introduced accurately, the curved cartilaginous mobile part of the external auditory meatus has to be drawn upward and posteriorly to bring it into the same axis as the bony part.

The cartilaginous part is attached firmly to the rim of the *bony meatus* by connective tissue. The bony canal is covered by a thin layer of skin that adheres to the periosteum. It contains no accessory structures, in contrast to the cartilaginous part of the meatus, which has numerous hair follicles and ceruminous glands that form wax (epidermis scale, sebaceous matter, pigment) (see p. 55).

The external meatus narrows medially. *Foreign bodies* may therefore become impacted at the junction of the cartilaginous and bony meatus. The meatal cartilage does not form a closed tube, but

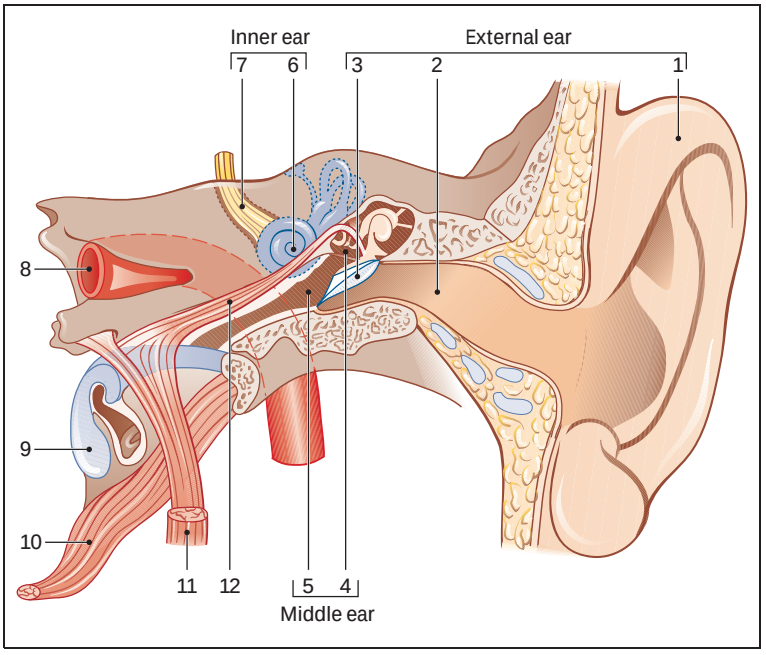


Fig. 1.4 Overview of the three sections of the ear. *External ear:* 1, auricle; 2, external ear canal; 3, tympanic membrane. *Middle ear:* 4, tympanic cavity; 5, eustachian tube. *Inner ear:* 6 & 7, labyrinth with internal ear canal and vestibulocochlear nerve; 8, internal carotid artery; 9, cartilage of eustachian tube; 10, levator veli palatini muscle; 11, tensor veli palati muscle; 12, tensor tympani muscle (Toynbee muscle).

rather a channel closed superiorly by fibrous tissue. The cartilage contains several fissures (Santorini fissures), which provide a pathway for the spread of severe bacterial infection to the parotid space, the infratemporal fossa, and the base of the skull.

The auricle and the cartilaginous meatus have very rich *lymphatic drainage* to an extensive regional lymphatic network consisting of parotid, retroauricular, infra-auricular, and superior deep cervical nodes. Infections of the external meatus with regional lymphadenitis can thus cause extensive swelling in these areas.

The *sensory innervation* is supplied by the trigeminal, great auricular, and vagus nerves and the sensory fibers of the facial nerve. Irritation of the posterior meatal wall stimulates the vagus and induces the cough reflex. Hypoesthesia of the posterosuperior meatal wall occurs with facial nerve impingement from a vestibular schwannoma (see the discussion of Hitselberger sign, p. 13 and Table 1.13, p. 94).

Relations (Fig. 1.4): The cartilaginous meatus abuts anteriorly onto on the parotid gland, allowing the spread of infection or malignant tumors.

The posterosuperior wall of the bony meatus forms part of the *lateral attic wall* (the partition between the external auditory meatus and the attic), the mastoid antrum, and the adjacent pneumatic system of the mastoid process. A middle ear infection can thus break through into the external auditory meatus, causing swelling of the posterosuperior wall or a fistula in acute mastoiditis. Destruction of the lateral attic wall by *cholesteatoma* may also lead to an open communication between the external auditory meatus and the attic or mastoid antrum. The anterior wall of the bony meatus forms part of the temporomandibular joint. There is therefore a risk of *fracture* resulting from a blow to the chin.

Middle Ear and Pneumatic System

The *middle ear cavity* consists of an extensive *pneumatic system* aerated by the eustachian tube. It has the following components:

- Eustachian tube.
- Tympanic cavity.
- Mastoid antrum.

The *eustachian tube* consists of a mobile, cartilaginous portion (two-thirds) suspended from the skull base, and a bony portion (one-third). The bony portion, together with the tensor tympani muscle, forms the musculotubal canal in the temporal bone.

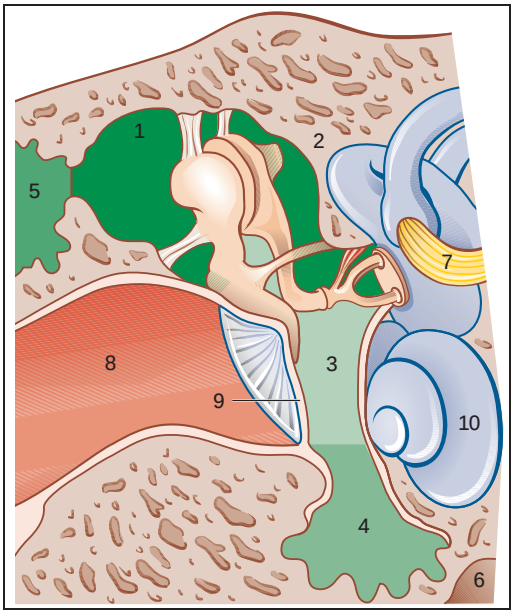


Fig. 1.5 Anatomy of the middle ear cavity. 1 & 2, Epitympanum; 3, mesotympanum; 4, hypotympanum; 5, mastoid antrum; 6, internal jugular vein. The lower part of the attic (2) is markedly narrowed by the facial nerve (7) and the horizontal semicircular canal. 8, External meatus; 9, tympanic membrane; 10, cochlea.

This canal lies adjacent to the internal carotid artery. The funnel-shaped pharyngeal ostium of the cartilaginous part (the torus tubarius) lies in the nasopharynx. The bony end opens into the middle ear.

The junction between the two parts of the tube is very narrow. This *isthmus* is the site of predilection for inflammatory stenosis of the tube. The tube serves to equalize the pressure between the middle ear and the nasopharynx, and thus to equalize the pressure on each side of the tympanic membrane (see pp. 6, 37). An increase in pressure in the tympanic cavity is usually compensated for passively via the eustachian tube to the nasopharynx, whereas a decrease in pressure usually requires active ventilation from the nasopharynx along the tube to the middle ear cavity. The tube opens and closes in response to movements of the neighboring muscles and by differences of air pressure between the nasopharynx and the middle ear cavity that tend to equalize spontaneously. The principal closing mechanism is elastic recoil of the cartilage of the tube and the valvular action of the pharyngeal ostium of the tube. The tube opens by contraction of the tensor palati and levator palati muscles. The mechanism is partially under the control of voluntary muscle, but the reflex movements on yawning and swallowing and the muscle tone are under autonomic control. Tension opposing the opening muscles is provided by the elastic recoil of the tubal cartilage and the pressure of the peritubal tissues—i. e., the pterygoid muscles, Ostmann's fatty bodies, the venous and lymphatic plexus of the tubal mucosa, and the pterygoid venous plexus.

The *middle ear cavity* is an air-containing space lying between the external ear and the inner ear. It is divided into three parts (Fig. 1.5):

- Epitympanic recess or attic.
- Mesotympanum.
- Hypotympanic recess.

There are two narrow zones within the middle ear cleft. Firstly, there is an anatomic constriction between the epitympanum and mesotympanum that can lead to retention of secretions in inflammation and to deficient aeration of the attic. This is due to the considerable narrowing of this area caused by the head of the malleus, the body of the incus, numerous ligaments, nerves (the chorda tympani), and mucosal folds and pockets. This is one of the causes of chronic inflammation of the epitympanum (chronic epitympanitis), which is one of the causative factors for epitympanic cholesteatoma (see p. 68). A second narrow zone lies at the junction of the attic and the mastoid antrum (the aditus ad antrum). This may be blocked by granulation tissue in chronic inflammation, leading to deficient aeration or drainage of the mastoid cell system.

The hypotympanum is closely related to the bulb of the internal jugular vein.

The *tympanic membrane*: The lateral wall of the middle ear cavity is formed by the tympanic membrane. The tympanic membrane consists of the pars tensa and the pars flaccida. The *pars tensa* forms the stiff vibrating surface of the membrane and is attached to a fibrous ring (the *anulus fibrosus*), lying in the tympanic sulcus of the tympanic part of the temporal bone. The *pars flaccida* is the superior part of the membrane in the area of the tympanic notch (Rivinus notch) where the anulus fibrosus ends (Fig. 1.6a,b).

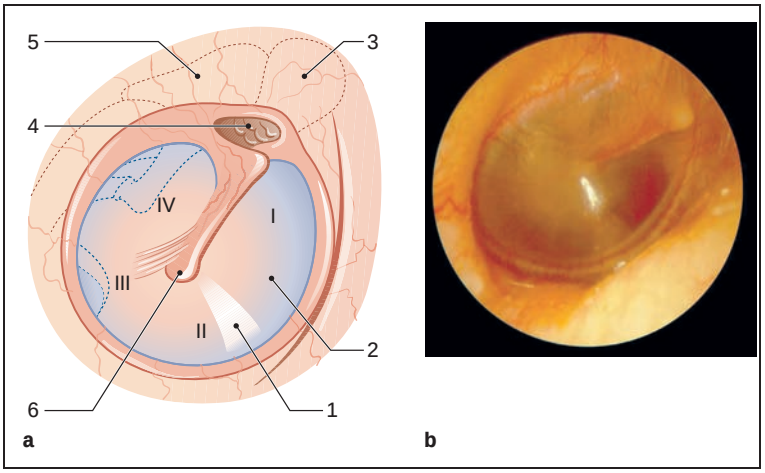


Fig. 1.6a, b a The macroscopic appearance of the right tympanic membrane. 1, Light reflex; 2, pars tensa; 3, malleus head; 4, pars flaccida; 5, incus; 6, umbo; 7, anulus fibrosus. The visible part of the surface of the tympanic membrane is divided into four quadrants, in order of investigation: I, anterosuperior; II, anteroinferior; III, posteroinferior; and IV, posterosuperior. b The oto-endoscopic appearance of a normal, transparent tympanic membrane. The tympanic ring, the handle of the malleus, and the short process of the malleus are visible. The light reflex is visible in the usual position—starting from the umbo across the anterior inferior quadrant.

The microscopic cross-sectional appearance of the tympanic membrane is shown in Fig. 1.7. The epithelial or cuticular layer (the stratum corneum) is similar in structure to the skin of the external auditory meatus. Close to the tympanic annulus is the marginal zone of the tympanic membrane. This section shows extremely active proliferation due to papillary ingrowths into the stratum germinativum. This is another important factor in the genesis of cholesteatoma (see pp. 68–76).

The keratinizing squamous epithelium regenerates through migration of the epidermis from the center of the tympanic membrane to the periphery—in contrast to superficial desquamation, as occurs in normal skin. Migration of the outer epidermal layer forms an important part of the self-cleansing mechanism of the external meatus; this can be observed clinically in the movement of a blood clot from the tympanic membrane to the external meatus.

The *lamina propria* has an external radial layer of fibers and an internal circular layer: this is evident during myringotomy. The anulus fibrosus forms a thickening of the edge of the tympanic membrane and is formed by both layers of fiber. A lamina propria can also be seen in the pars flaccida, but it lacks the characteristic radial and circular structure described above, which provides the normal pars tensa with the necessary functional tension.

The *middle ear*, or *tympanic cavity*, is empty except for air. Only the epitympanic recess contains solid structures—the ossicular chain and the chorda tympani. The ossicular chain consists of three separate bones connecting the lateral and medial wall of the middle ear. The medial wall of the middle ear also forms the lateral wall of the labyrinthine capsule. The malleus is the most lateral of the ossicles. Its inferior portion, or handle, is incorporated into the eardrum, while the superior portion, or head, is located in the anterior portion of the attic. The incus is connected to the head of the malleus by a genuine articulation surrounded by a joint capsule. The long process of the incus ends in the lenticular process, which bends medially to articulate with the head of the stapes. The lenticular process is covered by cartilage to form the incudostapedial joint (Figs. 1.8, 1.9).

The *mucosa* that lines the middle ear space consists of stratified cuboidal epithelium, which changes to pseudostratified ciliated epithelium around the mouth of the eustachian tube. A few goblet cells and submucosal glands are normally present. The submucosa is very thin, so that the mucosa lies directly on the periosteum, forming a tightly bound unit called the *mucoperiosteum.* In pathologic conditions such as tubal occlusion or chronic otitis media, the structure of the mucosa changes considerably to show hyperplasia of the glands, proliferation of the goblet cells, edema of the submucosa, vascular buds, and transformation of the flattened cuboidal epithelium to columnar epithelium.

The middle ear mucosa forms several pouches and folds *(Prussak space, Tröltsch pouch),* which narrow the junction

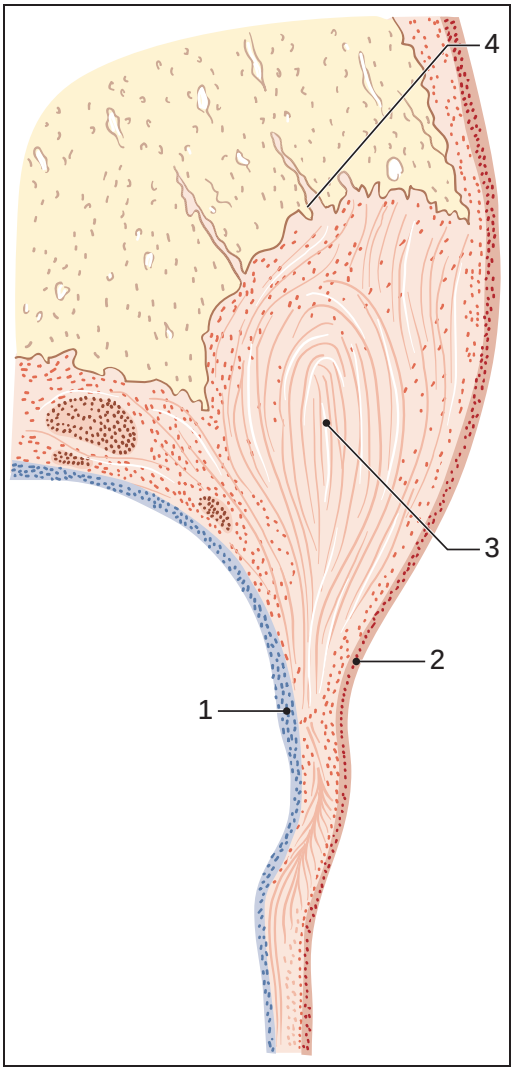


Fig. 1.7 The microscopic appearance of a sagittal section through the posterosuperior quadrant of the tympanic membrane. 1, Epidermis layer, similar to the meatal skin bordering the tympanic membrane; 2, middle ear mucosa; 3, anulus fibrosus; 4, bony sulcus of the fibrocartilaginous ring.

between the attic and the rest of the middle ear and between the attic and the antrum. The epitympanic recess may remain as a narrow cleft with development, and if chronic hyperplastic inflammation follows an infection, the "mesenchyme" can completely obliterate the epitympanum. Ventilation and drainage of the attic is then impeded by thickened masses of inflammatory tissue, despite normal tubal function. Deficient aeration and drainage of this small space

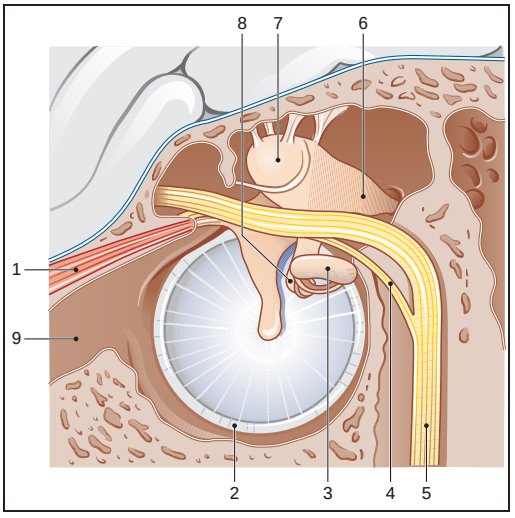


Fig. 1.8 Medial view of the middle ear, with the ossicular chain and facial nerve. 1, Tensor tympani muscle. The pars tensa is anchored by the anulus fibrosus (2) in the bony niche of the tympanic ring. 3, Stapes footplate. The handle and short process of the malleus lie lateral to the chorda tympani (4), as part of the facial nerve (5). The long process of the incus forms a joint (8) at its lenticular process with the head of the stapes. The body of the incus (6) forms the joint surface for the head of the malleus (7). The malleus and incus vibrate as one body in the middle part of the frequency range. The middle ear cavity is aerated via the eustachian tube (9).

favors the development of chronic *epitympanitis* and plays a considerable role in the pathogenesis of *chronic otitis media* (see p. 58–61), especially attic cholesteatoma.

The *arterial blood supply* originates from the basilar artery (the labyrinthine artery), the maxillary artery (the middle meningeal and tympanic arteries), and the stylomastoid artery. Venous drainage is via the middle meningeal veins, the venous plexus of the internal carotid artery and pharynx, and venous connections into the bulb of the internal jugular vein.

The *nerve supply* of the mucosa is provided from two sources: the tympanic branch of the glossopharyngeal nerve (cranial nerve IX) and the auriculotemporal branch of the trigeminal nerve (cranial nerve V).

Note: The shared sensory supply of the ear and upper respiratory tract explains why pain is referred to the ear in diseases of the teeth and the jaws, as well as of the larynx and pharynx.

Pneumatic System of the Temporal Bones

The air-containing cells of the mastoid process are continuous with the air in the middle ear. These multiple interconnecting spaces arise from the mastoid antrum, and the extent to which they are pneumatized is extremely variable. On the one hand, *pneumatization* may be well developed, extending to the temporal and occipital bones and the origin of the zygomatic arch. Acute infections of the mastoid may cause inflammatory swellings in these regions. At the other extreme, in a poorly pneumatized mastoid, the mastoid process may consist exclusively of compact bone, with the pneumatized cells lying in the immediate vicinity of the antrum.

The mastoid process begins to develop after birth as a small tuberosity, which is pneumatized synchronously with the growth of the mastoid antrum. In the first year of life it consists of cancellous bone, so that true mastoiditis cannot occur. Between the second and fifth years of life, as pneumatization proceeds, it consists of mixed cancellous and pneumatic bone. Pneumatization is complete between the sixth and twelfth years of life (Figs. 1.10, 1.11).

Principle of pneumatization (the concept of biological mucosal competence). Bone is destroyed by an enzymatic lacunar osteoclastic process. The resulting bony spaces are lined by continuous ingrowth of mucoperiosteum from the antrum. A system of hollow cavities results, consisting of numerous spaces lined by mucosa and communicating with each other.

Normal tubal function is a prerequisite for biologically active, healthy middle ear mucosa, and thus for the normal process of pneumatization. The process of pneumatization can be related to the biological competence of the middle ear mucosa. The mucosa may be described as *biologically normal* or as *inferior*, depending on the degree of pneumatization. *Good pneumatization* indicates biologically competent middle ear mucosa, whereas *restricted pneumatization* indicates biological incompetence of the middle ear mucosa. Biologically incompetent middle ear mucosa may be due to two possible mechanisms—a defective enzyme system that is impairing normal pneumatization, and/or a deficient local immune system in the respiratory mucosa and middle ear mucoperiosteum that predisposes to chronic or recurrent otitis media.

Note: Characteristically, pneumatization of the temporal bone is absent or restricted in chronic otitis media.

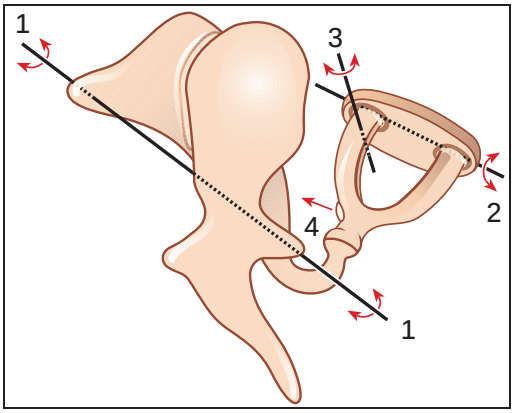


Fig. 1.9 The axis of ossicular chain movement. The malleoincudal joint can turn at a 90° angle according to the position of the footplate (1). The footplate itself can move from anterior to posterior (2) and in a lateral direction (3). The incudostapedial joint (4) moves only in a slight lateral bend.

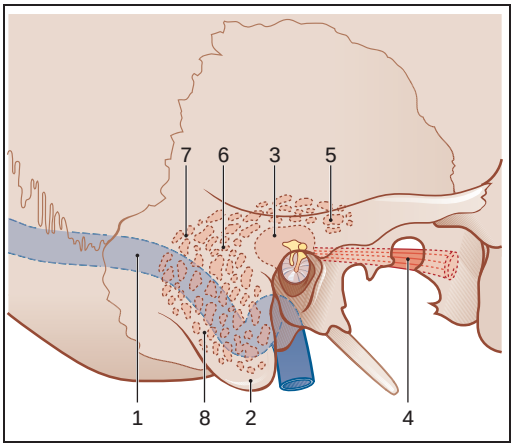


Fig. 1.10 The pneumatic system of the temporal bone. 1, Transverse sinus; 2, mastoid process with tip cells; 3, mastoid antrum; 4, eustachian tube; 5, zygomatic cells; 6, cells of the squamous part of the temporal bone; 7, sinodural angle; 8, retrosinus cells.

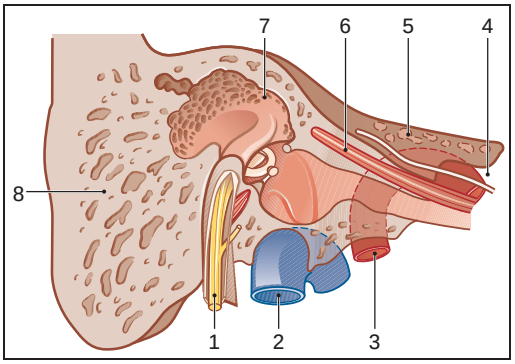


Fig. 1.11 Topographic relationships in the middle ear cavity. 1, Facial nerve—inflammation and trauma often affect the mastoid segment; 2, the bulb of the internal jugular vein, which is the site of predilection for extension of a glomus tumor into the middle ear cavity; 3, the internal carotid artery—in petrositis, the inflammation can extend into the venous plexus around the carotid artery to create a cavernous sinus thrombosis; 4, cavernous sinus; 5, apical cells—purulent infection of the cells in petrositis (see p. 80) causes Gradenigo syndrome; 6, the tensor tympani muscle; 7, the tegmen tympani, which is the site of predilection for mastoiditis to penetrate into the middle cranial fossa; 8, the pneumatic system of the mastoid process—purulent infection of the cells causes subperiosteal abscess and sigmoid sinus thrombosis.

The better pneumatized the temporal bone is, the easier it is for infection to break through the thin cortical bone. When there is poor pneumatization (known as a *dangerous mastoid process*), the inflammatory process may be concealed in the depths and lead to unexpected complications.

Inner Ear, Peripheral Hearing, and Balance Organs

The inner ear, or labyrinth, is embedded in the temporal bone and is divided into two functionally separate receptor mechanisms:

- The vestibule and semicircular canals (the vestibular end organ).
- The cochlea (the acoustic end organ).

The labyrinth can also be divided morphologically into *bony* and *membranous* parts.

The bony labyrinth. This is formed by the *labyrinthine capsule,* which develops by periosteal and enchondral ossification. In systemic bone diseases (e. g., Paget disease and osteodystrophy) and in localized bone disease (e. g., otosclerosis), the bony labyrinth shows characteristic histopathological and chemical abnormalities. These conditions demonstrate continuous bone remodelling.

The oval and round windows form the bony and membranous openings to the labyrinth from the middle ear cavity, and are closed by the stapes footplate and round window membrane, respectively (see p. 5).

Membranous labyrinth and inner ear fluids (Fig. 1.12a, b). The *membranous labyrinth* develops from the ectodermal otic placode. It encloses a hollow system filled with *endolymph.* This passes via the endolymphatic duct to end in a blind sac, the *endolymphatic sac,* in the posterior cranial fossa. The sac lies in the epidural space on the posterior surface of the petrous pyramid, close to the sigmoid sinus.

The *perilymphatic system* forms a hollow space consisting of the scala tympani and the scala vestibuli. The system communicates directly with the subarachnoid space in the jugular foramen via the cochlear aqueduct. Perilymph separates the membranous labyrinth from the internal layer of the labyrinthine capsule. Perilymph is the immediate substrate of the cochlear and vestibular sensory cells. The origin of perilymph is a matter of controversy; it may form from filtration of perilymphatic capillary blood and/or through diffusion of cerebrospinal fluid.

Endolymph is a filtrate of perilymph that has completely different concentrations of sodium and potassium, which are kept constant by the epithelium of the *stria vascularis* (see Fig. 1.18a). The electrolyte composition of the endolymph regulates the volume of the fluid circulating in the endolymphatic system. The basis of the electrolyte exchange system, which maintains a constant ion concentration, is the cellular *potassium–sodium exchange pump* found in the stria vascularis, the utricle, and the saccule. There is also passive diffusion between the endolymphatic and perilymphatic spaces, with potassium–sodium ion exchange in the endolymphatic sac. Functional disturbances of this electrolyte regulation system lead to a disorder of the middle ear known as *Ménière disease* (see p. 97).

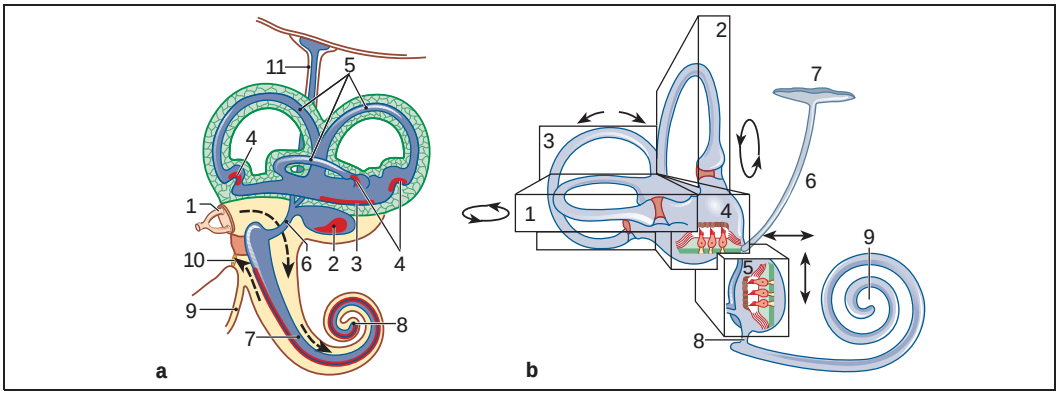


Fig. 1.12a, b a The inner ear. 1, Oval window with stapes; 2, saccule; 3, utricle; 4, ampulla of the semicircular canals, with cupula; 5, membranous semicircular canals (horizontal, superior, and posterior); 6, ductus reuniens; 7, cochlear duct; 8, helicotrema; 9, the perilymphatic duct, which passes through the cochlear aqueduct; 10, round window; 11, endolymphatic sac on the posterior surface of the pyramid. b The vestibular apparatus. 1, Lateral semicircular canal; 2, vertical semicircular canal; 3, posterior semicircular canal; 4, utricle; 5, saccule; 6, endolymphatic duct; 7, endolymphatic sac; 8, ductus reuniens; 9, cochlea. Arrows mark the direction of velocity forces.

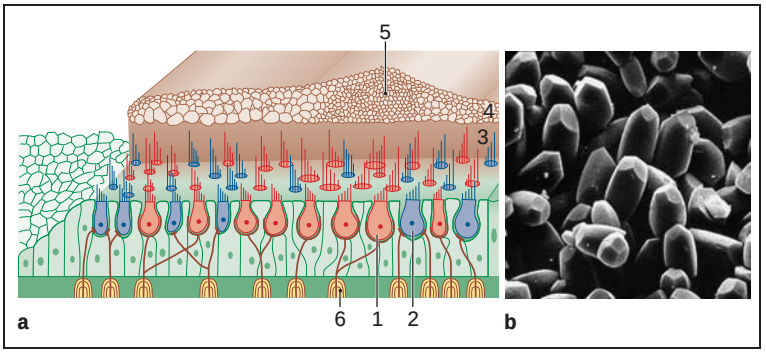


Fig. 1.13a, b a Static macula. A change in the polarity of the hair cells occurs below the striola. 1, type I hair cell; 2, type II hair cell; 3, gelatinous layer; 4, statolith membrane; 5, statoliths, striola; 6, afferent nerve fibers.
b Scanning electron-microscopic image of calcium carbonate crystals in the gelatinous layer of the utricle—otoliths.

Vestibular–Semicircular Canal System

The anatomical fine structure of the balance mechanism system is shown in Figs. 1.13a, 1.14, 1.15. It consists of the *utricle* and *saccule* enclosing the static *maculae* with the sensory end organs for the reception of linear acceleratory stimulation. These consist of *supporting cells* and *hair cells*, which have *cilia* embedded in a gelatinous mass consisting of sulfomucopolysaccharides. On their surface lie the *otoliths* (or statoconia), which consist of rhomboid calcium carbonate crystals (Fig. 1.13b). Linear acceleration changes the otolith pressure, deflecting the sensory hairs. This stimulates the sensory cell by altering the *resting potential.*

The three semicircular canals arise from the utricle and have a pear-shaped expansion at one end called the *pars ampullaris,* enclosing the sensory cells, which are stimulated by angular acceleration (Fig. 1.16). The sense organs consist of an *ampullary crest* (crista ampullaris), on which *sensory hair cells* are arranged in such a way that their cilia extend to the *cupula,* which reaches to the roof of the ampulla. The cupula acts as a mobile partition that closes off the pars ampullaris and is relatively impervious to endolymph (Fig. 1.15).

> Note: The hair cells of the maculae and ampullary crests have similar structural principles. They are mechanoreceptors that respond to tangential bending of their cilia.

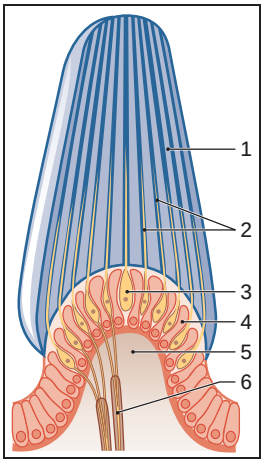


Fig. 1.14 A receptor in the semicircular canal. 1, Cupula; 2, cilia; 3, sensory cells; 4, supporting cells; 5, crista ampullaris; 6, afferent nerve fibers.

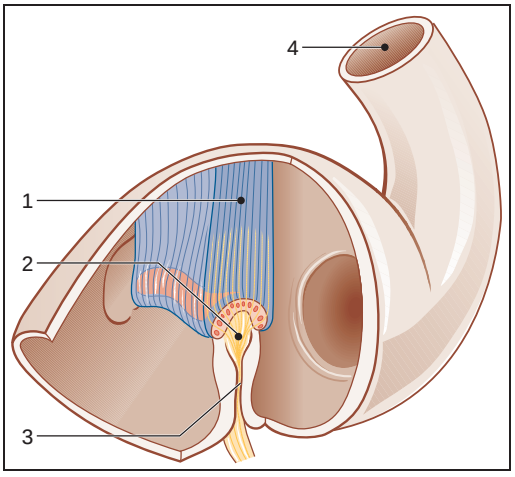


Fig. 1.15 The ampulla of a semicircular canal. 1, Cupula; 2, crista ampullaris; 3, afferent nerve fibers; 4, membranous semicircular canal.

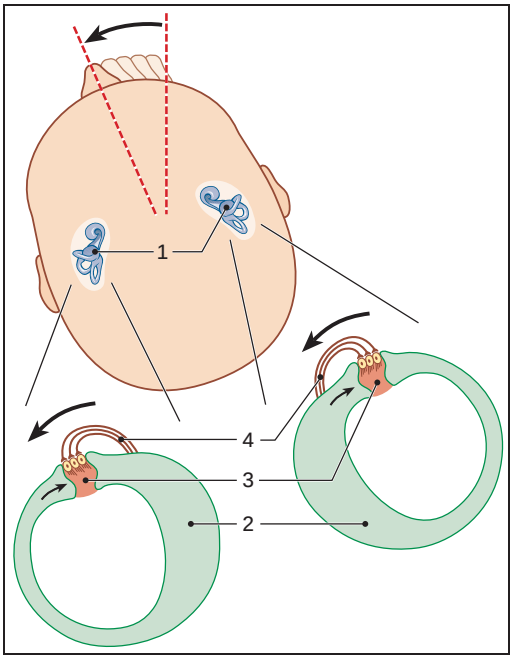


Fig. 1.16 Oscillation of the cupula. When the head is rotated (arrow), the semicircular canals rotate as well. Owing to its viscosity, the endolymph initially remains motionless and directs the cupula in the opposite direction. This causes the cilia to bend. 1, Labyrinth; 2, membranous canal of semicircular canal; 3, cupula; 4, vestibular nerve.

Cochlea (Acoustic End Organ)

The macroscopic and microscopic structure of the bony and membranous cochlea are shown in Figs. 1.17a,b, 1.18a–c.

Functional structure of the organ of Corti. The basilar membrane supports the sensory apparatus of the organ of Corti. It stretches between the bony spiral lamina and the lateral cochlear wall and forms the border to the scala tympani. Surrounded by supporting cells, there are two types of receptor cells: one row of inner and three rows of outer hair cells, totaling ≈ 16 000 sensory cells. The hair cells have fine cilia on their free surfaces, with approximately 80 cilia per cell. So-called *tip links*, ≈ 10 µm thick, extend from the tips of the small cilia to the longer, very fine protein strings. There are ion channels where the tip links connect to the cilia, providing the basis for transduction of the sound stimulus to a receptor potential. Lying on top of the organ of Corti is the gelatinous tectorial membrane. The cilia of the outer hair cells lie below the tectorial membrane, while the cilia of the inner hairs cells do not insert into the tectorial membrane. The hair cells are secondary sensory cells and have no nerve cell processes. They receive fibers from the spiral ganglion. Approximately 90 % of the nerve fibers extend to the inner hair cells, and each inner hair cell is connected to many afferent fibers, each of which undividedly connects to an individual hair cell. The remaining 10 % of the nerve fibers are widely dendritic and innervate the outer hair cells. There are

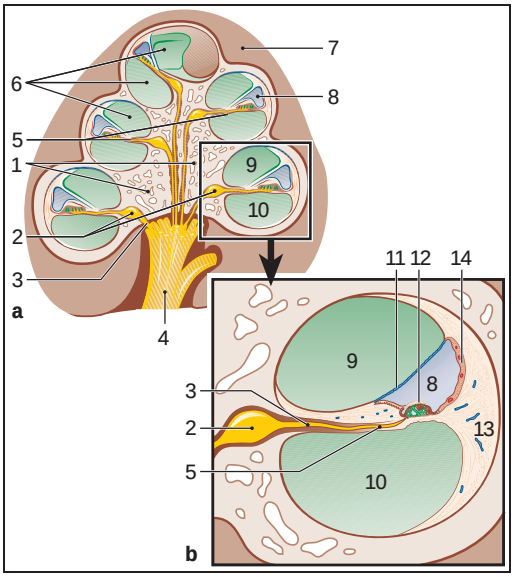


Fig. 1.17a, b Axial cross-section through the cochlea (a) and cochlear canal (spiral canal) (b). The cochlea is arranged spirally (with two and a half turns) around the central modiolus (1) lying horizontally. Its base lies against the lateral end of the internal acoustic meatus, and its apex is directed anterolaterally toward the medial wall of the middle ear. The spiral ganglion—i. e., the ganglion of the cochlear nerve (2)—is located within the modiolus, and its nerve fibers (3) join to form the stem of the cochlear nerve, the pars cochlearis of the vestibulocochlear nerve (4). The osseous spiral lamina or spiral plate (5) is a bony plate that runs spirally from the base to the apex (7). Nerve fibers pass through the channels of the spiral lamina to the spiral organ of Corti (12). The cochlear duct (scala media) (b, 8), filled with endolymph, lies between the scala vestibuli (9) above and the scala tympani (10) below, both of which contain perilymph (6). The osseous spiral lamina (5) and the basilar membrane form the separating wall between the scala tympani, on the one hand, and the scala vestibuli and cochlear duct on the other. The Reissner membrane (11) separates the scala vestibuli and the cochlear duct. The tectorial membrane (12) covers the sensory cells of the organ of Corti. The stria vascularis (14) forms the lateral wall of the cochlear duct and has numerous vessels. This layer of fibrous vascular tissue is the site of production of the endolymph. Laterally, it borders on the spiral ligament of the cochlea (13). The perilymphatic spaces of the cochlea, the scala tympani and scala vestibuli, communicate with each other at the apex of the cochlea (a, 7), at the helicotrema, (see Fig. 1.12a, 8) and are also connected with the perilymphatic space of the membranous labyrinth of the vestibule, containing both the utricle and the saccule (see Fig. 1.12a, 2, 3).

≈ 30 000–40 000 axons that lead from the spiral ganglion to form the vestibulocochlear nerve (Fig. 1.19).

Note: The entire frequency spectrum of 18–20 000 Hz is represented in the hair cells of the organ of Corti over the entire basilar membrane. The highest frequencies are localized to the most basal segment of the cochlea and the lowest frequencies near the helicotrema in the apical turn. This arrangement forms the morphologic basis of the "tonotopic" organization of the cochlea—i. e., the point-to-point connection between the sound wave receptors and the signal-converting central neurons of the auditory system.

Central Connections of the Organ of Corti

The cochlear division of the eighth cranial nerve (pars cochlearis) is formed by the bipolar neurons of the spiral cochlear ganglion. It runs through the internal auditory meatus, unites with the vestibular division, crosses the cerebellopontine angle, and enters the brain stem at the lower border of the pons, at which point the central auditory pathway begins (Fig. 1.20).

The central auditory radiation incorporates the strict tonotopic arrangement, as does the *auditory cortex.* The cochlea is thus represented unrolled, as it were, from the basal turn to the helicotrema. The *auditory cortex* is considerably larger than the area of Heschl's transverse striations, since these represent only the *primary auditory field* (AI) in which the auditory radiation ends. The secondary acoustic field (AII) and the posterior ectosylvian gyrus, like the visual cortex, include secondary integration areas such as the *Wernicke speech center.* Numerous commissural systems allow fibers to be exchanged between the two halves of the brain. These are very important for directional hearing.

Central Connections of the Balance Mechanism

The bipolar neurons of the vestibular ganglion send out their peripheral processes as two divided neural bundles—a superior division to the sensory cells in the macula of the utricle, the lateral and superior semicircular canals; and an inferior division to the posterior semicircular canal and the macula of the saccule (Fig. 1.21).

The central processes combine to form the vestibular division of the eighth cranial nerve, which

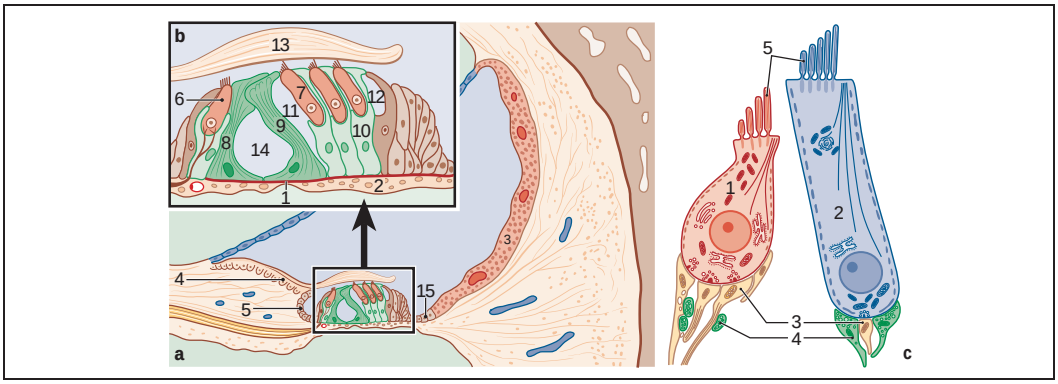


Fig. 1.18a–c a, b The cochlear duct (a) and spiral organ of Corti (b). The spiral organ of Corti (b) rests on the basilar membrane (1, 2) in the cochlear duct. Medially, at the free edge of the osseous spiral lamina, lies the limbus of the spiral lamina (4), with two labia enclosing the internal spiral sulcus (5). The highly vascularized stria vascularis (3) with intraepithelial capillaries lies laterally. The spiral organ of Corti (b) consists of inner hair cells (6) and outer hair cells (7) supported by pillar cells (8, 9), constituting the borders of the inner tunnel (perilymph or cortilymph, 14). Between the outer pillars (9) and external phalangeal cells of Deiters (10), which act as supporting cells for the spiral organ of Corti, lies the Nuel space, with perilymph (11). In the extreme lateral position, there is the outer tunnel (12), which borders on the external spiral sulcus (15) and the stria vascularis (3), respectively. Above the hair cells (inner and outer, 6, 7) is the tectorial membrane (13), a gelatinous mass extending from the limbus of the spiral lamina (4). The intercellular spaces of the spiral organ (11, 12, 14) contain perilymph, also known as cortilymph.
c The ultrastructure of the inner and outer hair cells. 1, Inner hair cells; 2, outer hair cells; 3, afferent nerve endings; 4, efferent nerve endings; 5, cilia.

Fig. 1.19 Scanning electron-microscopic image of the spiral organ of Corti, with a view of the surface of the basilar membrane. There are three rows of outer hair cells in the lower part of the picture and one row of inner hair cells in the upper left corner of the picture.

unites in the internal auditory meatus with the cochlear division to form the vestibulocochlear nerve which has a common nerve sheath. The vestibular division sends ascending fibers to the vestibular centers after it has entered the medulla oblongata. The *secondary vestibular pathway* is connected to the spinal cord by the *vestibulospinal tract.* Its fibers end at the spinal intermediate neurons and activate the alpha and gamma motor neurons of the extensor muscles. They are therefore the antagonists of the pyramidal pathway and mainly produce flexor inhibition and activation of extensors. They form part of a phylogenetically old antigravity system that serves to maintain balance. In addition, there are important ascending pathways to the cerebellum, the reticular formation (a multisensory integration center), and the centers for the eye muscles (where the oculomotor muscles are coordinated), via the *medial longitudinal bundle.*

A vestibulocortical connection is provided via the thalamus. Vestibular stimulation is projected to a small area in the *ventral postcentral somatosensory region,* near the visual area. This region represents a primary vestibular cortical area.

Note: Connections between the vestibular centers, the centers for the ocular muscles, and the cervical musculature, together with the cerebellum, form the morphologic basis for the extremely precise coordination of the three functional systems. This allows objects to be visually fixed even when the head is moving. Synchronized coordination of the ocular and cervical muscles is controlled through the vestibular apparatus via the gamma neurons.

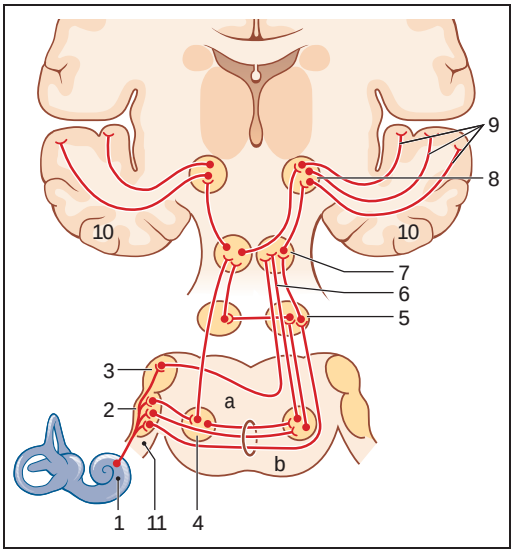


Fig. 1.20 The afferent auditory pathways. For the sake of simplicity, the pathways for only one cochlea are shown. a, Direct auditory pathway; b, indirect auditory pathway; 1, cochlea; 2, ventral cochlear nucleus; 3, posterior cochlear nucleus; 4, superior olivary nucleus; 5, nuclei of the lateral lemniscus; 6, lateral lemniscus; 7, inferior colliculus; 8, medial geniculate body; 9, acoustic radiation; 10, auditory cortex; 11, vestibulocochlear nerve.

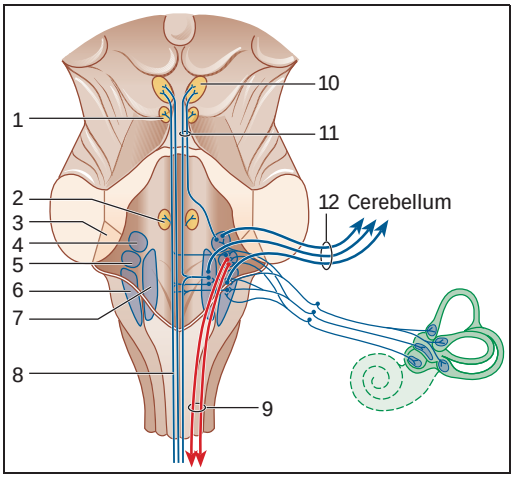


Fig. 1.21 The central vestibular connections in the brain stem. 1, Trochlear nucleus; 2, abducent nucleus; 3, inferior cerebellar peduncle; 4, superior vestibular nucleus (Bekhterev nucleus); 5, lateral vestibular nucleus (Deiters nucleus); 6, inferior vestibular nucleus; 7, medial vestibular nucleus; 8, 11, medial longitudinal bundle; 9, lateral vestibulospinal tract; 10, oculomotor nucleus; 12, vestibulocerebellar nerve fibers.

Facial Nerve

The seventh cranial nerve carries *motor fibers* for the mimetic muscles of the face, *afferent sensory taste fibers* and *visceroefferent secretory neurons* in a separate nerve bundle, the intermediate nerve. The nerve also contains the sensory fibers that supply the posterior wall of the external auditory meatus. This explains the reduced sensation of this area of skin in patients who have a vestibular schwannoma *(Hitselberger sign)* (Fig. 1.22).

The motor fibers originate from the facial motor nucleus in the floor of the fourth ventricle, run round the abducens nucleus (the internal "genu"), and exit at the lower border of the pons, together with the *visceroefferent fibers* of the intermediate nerve arising from the superior salivatory nucleus. The *gustatory fibers* insert into the subcortical taste centers in the nucleus of the solitary tract. All of these branches form the *nervus intermediofacialis,* which runs first in the internal auditory meatus (the meatal segment). It enters the bony canal immediately adjacent to the labyrinth (the labyrinthine segment) and runs to the hiatus in the canal for the facial nerve. At this point, the greater superficial petrosal nerve divides off from the main

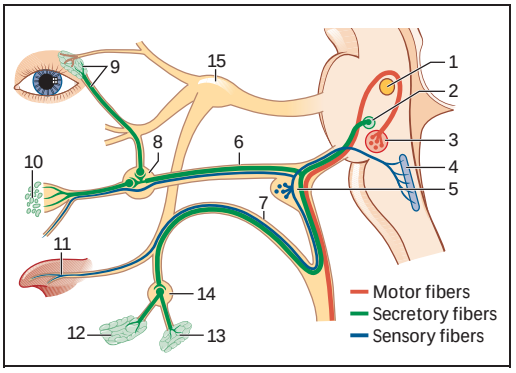


Fig. 1.22 Course of fibers in the facial nerve. 1, Abducent nucleus; 2, secretory nucleus of the nervus intermedius; 3, motor nuclei of the facial nerve; 4, nucleus of the solitary tract; 5, geniculate ganglion; 6, greater superficial petrosal nerve; 7, chorda tympani; 8, pterygopalatine ganglion with the lacrimal anastomosis; 9, lacrimal gland with greater superficial petrosal nerve; 10, nasal glands; 11, taste fibers to the anterior two thirds of the tongue; 12, sublingual gland; 13, submandibular gland; 14, submandibular ganglion; 15, trigeminal ganglion.

trunk. This branch goes to the lacrimal gland and also supplies fibers to the glands of the nasal mucosa. The first "genu" of the facial nerve lies at the level of the geniculate ganglion. The nerve then turns into the horizontal *tympanic segment* before it passes at the level of the entrance to the mastoid antrum, the second "genu," into the vertical *mastoid segment*. In this area, it branches to the stapedius muscle and the chorda tympani, which contains taste fibers for the anterior two-thirds of the tongue and carries visceroefferent fibers for the sublingual and submandibular glands. After leaving the mastoid process through the stylomastoid foramen, it divides into five extratemporal branches—the temporal, zygomatic, buccal, marginal mandibular, and cervical—to the platysma. These branches are highly variable (see Fig. 1.114).

The facial nerve is surrounded by a tough fibrous sheath in its course through the temporal bone. Its individual fascicles are embedded in a well-developed *epineurium* of loose connective tissue that encloses the vessels and nerves. The fiber bundles are enclosed in a *perineurium*. When injuries to the nerve are being repaired, the epineurium has to be resected from the stump, and a perineural suture has to be used so that the site of anastomosis can be adapted precisely, to prevent the formation of a scar tissue neuroma due to connective-tissue infiltration of the anastomosis (see p. 114).

! Note: Familiarity with the details of the regional anatomy of the facial nerve is a prerequisite for understanding the neurologic diagnosis of facial paralysis (the differential diagnosis of central and peripheral paralyses and the topographic diagnosis of the lesion; see p. 47).

Physiology and Pathophysiology of Hearing and Balance

Physiology of Hearing: Middle and Internal Ear

The functions of the various parts of the ear are as follows:

- The external and middle ear transport the stimulus.
- The cochlea distributes the stimulus.
- The function of the outer hair cells is mechanoelectric transduction.
- The inner hair cells transform the stimulus.

Stimulus Transport

In the *external auditory meatus*, the resonance effect lowers the hearing threshold to between 2000 and 3000 Hz, the main range of speech frequencies.

The *tympanic membrane* is a sound pressure receptor and transformer.

The *ossicular chain* is responsible for impedance adaptation between the middle ear, in which the medium is air, and the inner ear in a fluid medium, as well as *pressure transformation*. The pressure enhancement is 1 : 17, due to the ratio between the surface of the tympanic membrane and the stapes footplate. The ratio due to the mechanical advantage of the incudomalleolar joint is 1 : 1.3. The total pressure on the stapes footplate is therefore increased 22 times (see Fig. 1.9).

The physical movements of molecules that we perceive as sound set the tympanic membrane in motion. The frequency of the motion is the same as that of the vibrations of the air, and its amplitude is proportional. The transmission of sound waves from the air medium to the fluid medium in the perilymphatic and endolymphatic space requires a relative increase in power, due to the increase in density—i. e., impedance adaptation through sound pressure transformation (impedance = acoustic resistance).

For normal transmission of sound to the inner ear, the tympanic membrane has to be in a normal position and have normal mobility, and the air pressure in the outer and middle ears has to be similar. Measuring the impedance at the tympanic membrane can provide information about the functioning of the sound transmission apparatus, and this method—known as *impedance audiometry*—is used for clinical investigations (see p. 36). Sound energy reaches the cochlea firstly via the sound transmission apparatus of the middle ear *(air conduction)* and secondly through the bone of the skull, which is set in motion in a sound field. The sound energy is thus transmitted directly to the cochlea via the labyrinthine capsule *(bone conduction)*.

Audiometry is used to measure the hearing threshold for both air and bone conduction (see p. 27).

Stimulus Distribution

The main function of the cochlea is *mechanical frequency analysis,* which depends on its *hydrodynamics*. Periodic movements at the stapes are converted into aperiodic movements to produce a trav-

eling wave on the basilar membrane (Fig. 1.23). Since the inner ear fluids are not compressible, volume displacement at the stapes footplate leads to an equal volume displacement at the round window, and this produces a bulging of the round window membrane that is equal in extent to the depression of the stapes footplate. This volume displacement, produced by *periodic vibrations* of the stapes footplate, leads to displacement of the *cochlear duct* (scala media, Löwenberg scala; the space surrounded by the basilar membrane and Reissner membrane, between the scala vestibuli and scala tympani) (see Fig. 1.17a,b). This initial displacement forms a wave motion that proceeds along the partition to the helicotrema. This is an *aperiodic vibration,* or traveling wave. The wavelength becomes shorter as the wave approaches the helicotrema, but the amplitude becomes greater. The amplitude reaches a maximum at one specific point and then immediately begins to fall sharply, before dying away toward the helicotrema. The traveling wave causes a displacement between the tectorial membrane and the basilar membrane at its point of maximal amplitude, so that the cilia of the hair cells are displaced at this point, forming the sensory stimulus for these mechanoreceptors (see Figs. 1.18c, 1.25b).

The frequency-dependent development of the maximal amplitude on the traveling wave induces a corresponding *frequency-dependent localized stimulus* on the basilar membrane in the sensory cells of the organ of Corti that lie at the point of maximal amplitude. An initial analysis of the sound is thus achieved in accurately defined frequency stimulus patterns (Békésy's dispersion or traveling wave theory).

The maximum displacement of the traveling wave lies at a different point for each frequency: it is nearer the helicotrema for the lower frequencies and nearer the stapes footplate for the higher. The tonotopic arrangement of the cochlea means that every frequency is thus represented at a particular point on the basilar membrane (Fig. 1.24). Since the distribution of the maximal amplitude across the basilar membrane determines the point of excitation of the organ of Corti and thus the activity of the afferent nerve fibers in the cochlear nerve, the traveling wave hypothesis is also a "one-point" hypothesis, as suggested by Helmholtz. Each point on the basilar membrane therefore corresponds to a specific frequency.

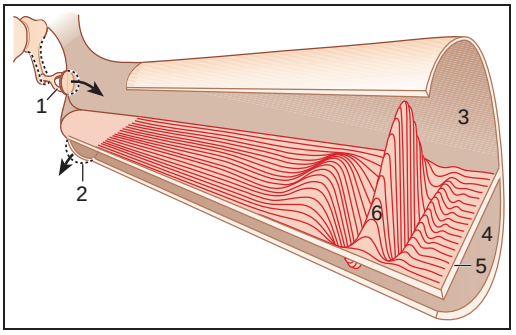


Fig. 1.23 Three-dimensional representation of the vibration of the basilar membrane. The traveling wave runs from the stapes along the basilar membrane, the tectorial membrane, and the Reissner membrane to the apex of the cochlea. The location of the maximum elongation of the basilar membrane is similar to the formation of a frequency-dependent maximum amplitude. 1, Stapes in the oval window; 2, round window; 3, scala vestibuli; 4, scala tympani; 5, basilar membrane with spiral organ of Corti; 6, maximum amplitude of the traveling wave.

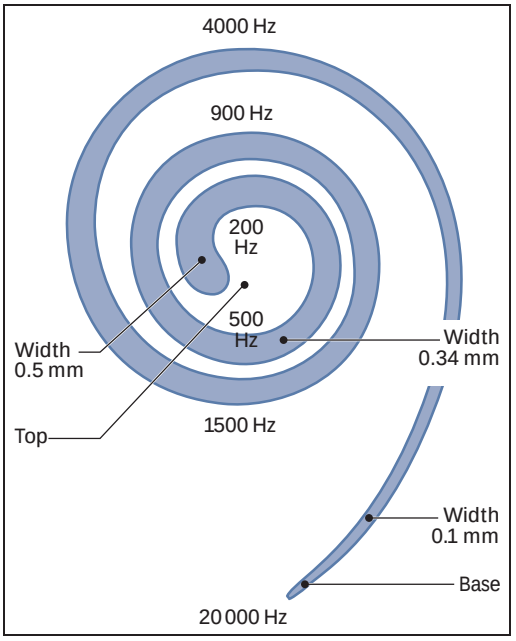
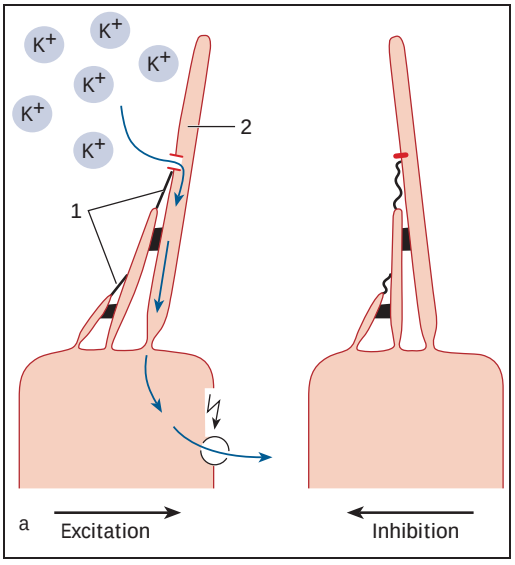


Fig. 1.24 The human basilar membrane, showing the frequency-dependent locations of sound receptors and analyzing receptors.

Mechanoelectric Stimulus Transduction

The cilia of the outer hair cells are bent to the greatest extent when the wave motion approaches the maximum range. A force pushing on the tip links causes the ion channels to open and changes the receptor potential. The outer hair cells carry out active, oscillating extension and thus locally intensify the travelling wave (Fig. 1.25a).



Stimulus Transformation

The actively intensified vibrations of the inner hair cells also cause the cilia of the inner hair cells to bend with the resulting opening of the ion channels. An influx of Ca^{2+} causes a basal discharge of glutamate as a transmitter, and the afferent nerve fibers of the vestibulocochlear nerve are consequently stimulated (Fig. 1.25b).

Otoacoustic Emissions

Active contractions of outer hair cells have natural modes of vibration and are subject to distortion. In this phenomenon of normal hearing, sounds emitted by the cochlea occur at certain frequencies as *spontaneous otoacoustic emissions* (SOAEs). *Evoked otoacoustic emissions* (EOAEs) can be recorded in the external auditory canal after induction by external acoustic stimuli (see p. 39).

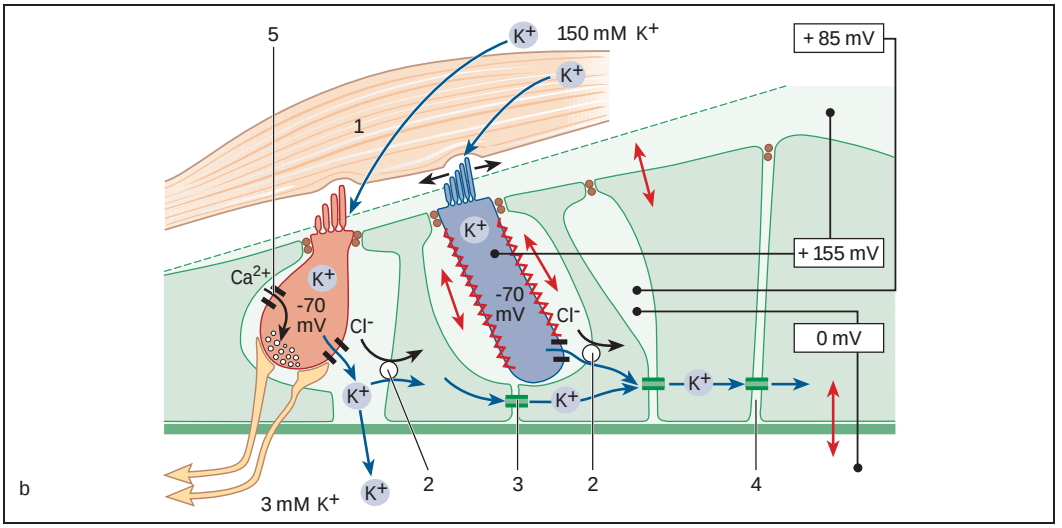


Fig. 1.25a, b a Depolarization (excitation) of the sensory hair cells by deflection of the cilia (2) and opening of stretch-sensitive potassium ion channels. Stretching of the channels is induced by tension to the tip links (1). K^+ ions escape the hair cell at the base through stretch-sensitive channels, leading to repolarization of the cells.

b The spiral organ of Corti, showing the electromotility of the outer hair cells acting as a cochlear amplifier. The frequency-dependent length changes in the outer hair cells (blue) vibrate the spiral organ of Corti and thereby stimulate the inner hair cells (red), which are normally not in contact with the tectorial membrane (1). The influx of K^+ into the hair cells is necessary for depolarization. This occurs through the high K^+ concentration of the endolymph and the endocochlear potential (+ 85 mV), which amounts to as much as 155 mV between the hair cell (resting potential –70 mV) and endolymph. Potassium ions leave the cells basolaterally by means of excitation-dependent K^+ channels (2) and are led through the cortilymph by means of K^+Cl^- cotransporters (3) and nexus channels (4) in the support cells into the spiral ligament. Excitation-dependent Ca^{2+} channels (5) regulate transmitter release during depolarization through the influx of Ca^{2+} ions.

Physiology of Hearing: Retrocochlear Analysis of Acoustic Information

The electrical stimulus pattern of sensory cells in the organ of Corti is converted in the peripheral cochlear neuron into the action potential pattern of the vestibulocochlear nerve. The sound stimulus—which has many parameters, such as frequency, intensity, temporal pattern, and the periodicity of the action potentials—has to be encoded to allow the information to be analyzed in the central nervous system.

Sound frequency and *sound intensity coding* play a very important role in the central analysis of the acoustic signal.

- *Sound intensity coding* occurs through frequency modulation. With increasing sound intensity, the number of spikes in the sensory cell discharge increases.
- In *sound frequency coding*, specific sensory cell groups in the organ of Corti are stimulated depending on the sound frequency. Tonotopicity (see below) allows these locally circumscribed stimulus patterns, produced on the basilar membrane, to be conducted by the vestibulocochlear nerve to the higher centers without distortion.

Tonotopy is a point-to-point connection between the sound receptors and the neurons analyzing the signal. Each cochlear neuron what is known as a *best frequency*—i. e., it responds only to an acoustic stimulus that has a frequency identical to the frequency assigned to it.

The acoustic system can process the duration, intensity, and frequency parameters of the acoustic signal in the following ways:

- With increasing intensity and constant frequency, the action potential rate in the nerve fibers increases, and the number of stimulated afferent neurons also increases, corresponding to the extent of the deflected area of the basilar membrane.
- At constant intensity and variable frequency, the deflected area of the basilar membrane is displaced into the appropriate segment of the organ of Corti within the cochlea, so that frequency is determined by point analysis. In addition, changes occur in the periodicity of the action potential series within the individual nerve fibers, which are analyzed by means of periodicity analysis. This provides another means of frequency determination.

Frequency analysis by means of local pattern scanning, *intensity perception* by frequency modulation, and *time-periodicity analysis* by combined evaluation of the time and place pattern also provide information that passes to the higher auditory centers as a result of tonotopicity (Fig. 1.26a, b).

Pathophysiologic Basis of Hearing Disorders

Conductive or middle ear hearing loss is caused by lesions of the stimulus transport organ. A characteristic symptom of this type of hearing loss is that bone conduction functions better than air conduction. The depression of the hearing threshold for air conduction is associated with an increase in acoustic impedance, as seen with stapes fixation due to otosclerosis.

Sensory hearing loss is caused by lesions in the stimulus transformation organ and/or in the vestibulocochlear nerves, and is therefore better known as *sensorineural hearing loss. Noise-induced hearing loss* and *age-related hearing loss (presbyacusis)* are caused mainly by mechanical overloading of the cochlear amplifier system of the outer and inner hair cells.

Disorders of sound perception are caused by lesions in the subcortical or cortical auditory centers and by pathologic processes involving the central auditory pathway. As a result, the acoustic signals are falsely coded, stimulus patterns are wrongly analyzed, and acoustic information can no longer be integrated. The patient can then hear but not understand.

Central hearing disorders are characterized by a loss of the integrative functions of the auditory centers. Differences in level of tone, differences in loudness, and temporal differences of acoustic stimulus pattern can no longer be analyzed. Redundancy is also reduced—i. e., the information content is reduced due to loss of secondary and tertiary cochlear neurons. These disorders affect the understanding of speech (whereas hearing of pure tones may be preserved), directional hearing, and speech intelligibility.

Recruitment: In certain forms of unilateral sensorineural deafness, the loudness perception rises quickly with increasing loudness intensity, so that despite different hearing thresholds both ears hear

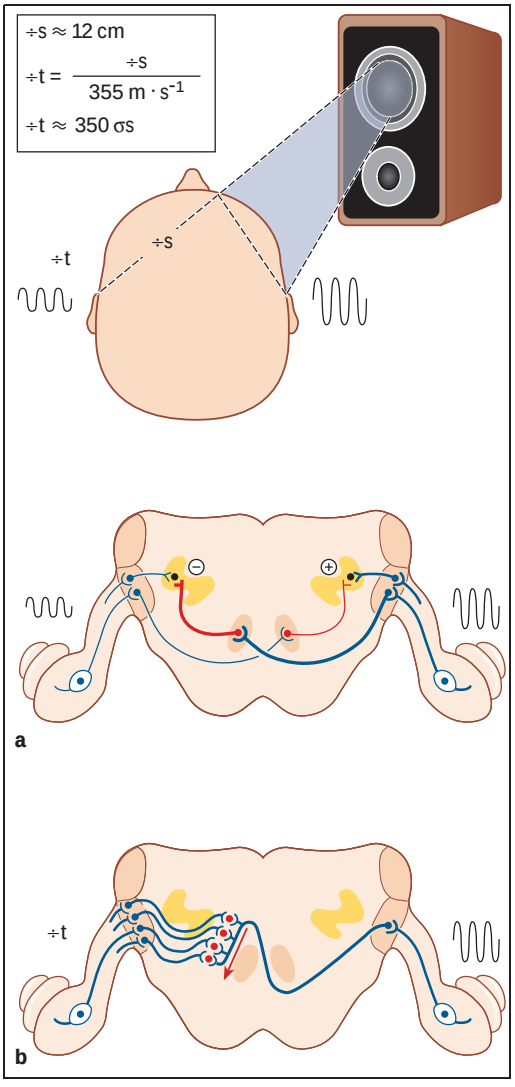


Fig. 1.26a, b Important connections for directional hearing. When level differences are being determined to identify the sound source (a), the highly stimulated neurons on the lateral superior olivary nucleus are on the sound side (low inhibition, intense excitation). With the interaural time difference Δt (b), simultaneous maximal excitation only takes place in the neurons of the medial superior olivary nucleus on the side turned away from the sound source.

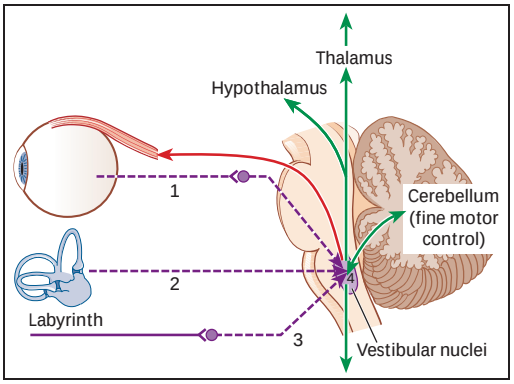


Fig. 1.27 The input and output of the vestibular nuclei. 1, Visual information; 2, vestibular information from the semicircular canals and otolith apparatus; 3, kinesthetic information from the superficial and deep receptors in the skin, muscles, tendon, and joints, which react to pressure and traction forces caused by the force of gravity and inertia; 4, vestibular nuclei.

the tone at the same loudness once a certain threshold is reached. This phenomenon is called recruitment. The pathophysiologic basis for recruitment is loss of the cochlear amplifier mechanism, with abnormal sound processing dynamics of. *Positive recruitment* can generally be regarded as a sign of a cochlear lesion, whereas *absent recruitment* indicates a retrocochlear lesion localized to the first or second neuron.

Physiology of the Balance System

Balance is maintained by coordination of visual kinesthetic and vestibular regulatory mechanisms. These serve for *spatial orientation, upright posture,* and *gait.* Control of all the static and motor muscle groups allows the body to counteract the influence of weight and centrifugal forces (Fig. 1.27).

The main functions of the vestibular system are:

- To send information to the central nervous system about the action of linear and angular acceleratory forces.
- Coordination. Movement is coordinated by continuous control of the tone of the skeletal muscles. Information from the vestibular sensory receptors is coordinated and integrated with information from the visual system. Spatial orientation is also ensured.

The potential difference between the sensory cells and the extracellular fluid forms the physiologic basis for normal functioning of the vestibular sense organ. A constant discharge of action potentials passes along the vestibular nerve fibers, even

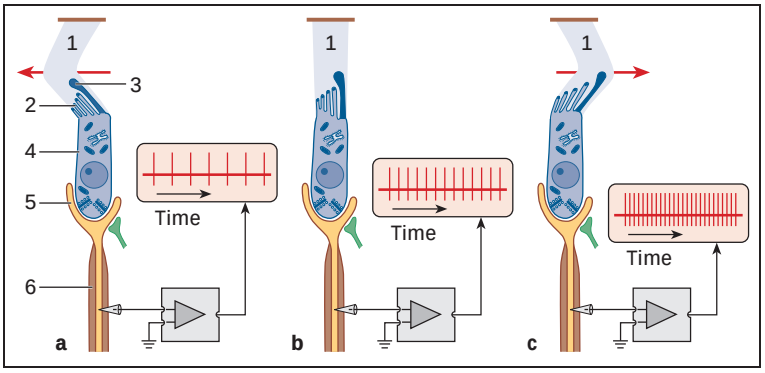


Fig. 1.28a–c The bioelectrical activity of the vestibular sensory cells at rest and in response to stimulation. Bending of the sensory hair cells away from the kinocilium (a) causes hyperpolarization and inhibition of the resting activity (b). Deflection in the opposite direction, toward the kinocilium (c), causes depolarization and an increase in the discharge frequency of the action potential. 1, Gelatinous layer; 2, cilia; 3, kinocilium; 4, sensory cell; 5, synapse of the afferent nerve; 6, afferent nerve fiber.

when the end organs are at rest *(resting activity).* As in the cochlea, a transduction channel in the vestibular hair cells is opened by a force pushing on the tip links, allowing an influx of ions and causing the receptor potential to change. Depending on the direction of the ciliary deflection of the sensory hair cells, the resting activity is altered by an increase in the discharge frequency *(depolarization)* or by inhibition *(hyperpolarization)* (Fig. 1.28a–c). Modulation of resting activity thus allows the body to sense movement both in one direction and also in the opposite direction using a single receptor.

Function of the Otolith Organ: Linear Acceleration Measurement

Linear acceleration is the sensory stimulus for the horizontally orientated macula of the utricle and the vertical macula of the saccule. Shearing forces occur during linear acceleration that shift the otoliths from their base, causing shearing of the hair cells (see Fig. 1.12b) and providing an adequate stimulus for the sensory cells. The resulting neuronal impulses release the *maculo-ocular reflex,* producing compensatory eye movements that ensure optimal static positioning of the eyes during linear movement. The *maculospinal reflex* is also evoked, which influences the musculature of the trunk and limbs via the motor anterior horn cells in the spinal cord to ensure that the position of the body remains stable during linear movement. The otolith apparatus also has another important function: due to the continuous effect of gravity, the otoliths exert constant pressure on the underlying sensory cells, even at rest. This pressure influences the resting activity of these mechanoreceptors. Linear acceleration—e.g., a fall, rapid lowering of the head, air travel, or fast movement in an elevator—changes this resting activity, thus guaranteeing continuous spatial orientation during vertical movement.

Function of the Semicircular Canals: Angular Acceleration Measurement

Positive or negative angular acceleration causes endolymphatic movement within the semicircular canals lying in the plane of the centrifugal force. The stimulus always affects the semicircular canals on both sides; the cupula is displaced toward the utricle on one side *(ampullopetal stimulation)* and in the opposite direction on the other side *(ampullofugal stimulation).* As a result, resting activity increases in the semicircular canal in which the cupula is deflected in an ampullopetal direction (depolarization effect), whereas activity decreases in the contralateral canal (hyperpolarization effect). This rule applies only to the horizontal canals, since ampullofugal deflection causes depolarization in the vertical semicircular canals. This is the neurophysiologic basis for the stimulating mechanism of the vestibuloocular reflex.

The *vestibuloocular reflex* also serves for spatial orientation. It addition, it assists in stabilizing the retinal image of the visual environment and induces vestibular nystagmus. Every movement of the head causes slow, conjugated movement of the eyes in the opposite direction, to stabilize the field of vision on the retina for as long as possible during the movement. Two modifiable parameters determine the progress of the vestibuloocular reflex: the

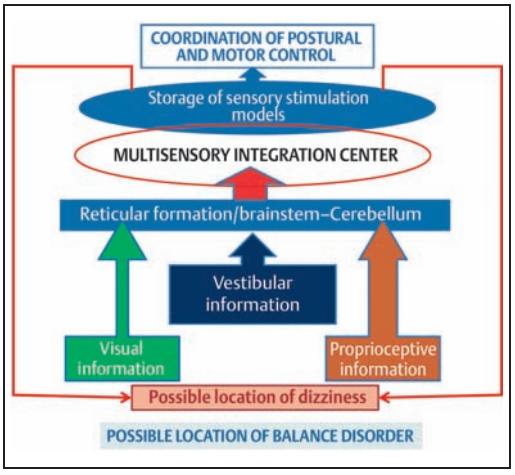


Fig. 1.29 Pathogenesis of disorders of orientation and balance. *Disorders of proprioceptive information:* loss of control over the ability to stand upright and walk straight causes a balance disorder. *Disorders of visual information:* loss of optical control of the visual field occasionally leads to *dizziness* due to a discrepancy between visual and vestibular information, causing disorientation. *Disorders of vestibular information* are due to involvement of the spatial orientation and stabilization of the gaze axis, leading to contradictory vestibular visual and kinesthetic information and *dizziness.* If central compensation of the loss of vestibular function is also absent, there is an additional balance disorder.

position of the head and the position of the eyes. The difference between these is the angle of vision (see Fig. 1.16).

Note: The vestibuloocular reflex coordinates the speed of reflex eye movements (the slow component of nystagmus) with the speed of head movement. This ensures clear visual control of the environment during movement. Fast return of the eyes is achieved by a reflex, the fast component of nystagmus.

Conjugated eye movements due to the vestibuloocular reflex, with typical slow and fast components, are classified as vestibular nystagmus (see p. 42).

The intervertebral joints of the cervical spine and the deep muscles of the neck contain mechanoreceptors, which are connected to the reticular formation by afferent fibers and from there to the vestibular and oculomotor centers. The function of these receptors is to provide continuous information about the position and movement of the head and to allow coordination of eye movements through the *cervicoocular pathway.*

The central *vestibular system* includes the cerebellum and the reticular formation of the brain stem—i. e., it is integrated into the centers for multisensory data analysis. This allows multisensory control and coordination of posture, movement, and oculomotor functions.

Pathophysiologic Basis of Functional Vestibular Disorders

Vestibular disorders become manifest through:

- *Vertigo:* partial or complete loss of spatial orientation—e. g., apparent movement of the environment as a result of spontaneous vestibular nystagmus; and/or
- *Disturbed balance,* with an inability to maintain balance, stand upright, or walk properly (ataxia) (Fig. 1.29).

Vestibular disorders may be *peripheral,* caused by sudden unilateral failure of one labyrinth, or by a unilateral lesion of the vestibular nerve. They may also be *central,* caused by a lesion in the vestibular centers or their central connections to the cerebellum and reticular formation.

Every functional disturbance in a vestibular end organ causes unequal activity in the higher vestibular centers. This central imbalance initially produces a disturbance of vestibular information. The multisensory spatial orientation is therefore no longer capable of functioning, since vestibular information on the one hand and visual somatosensory information on the other hand contradict each other. This causes a disturbance of orientation, which in turn causes dizziness. If the central imbalance in the two vestibular centers influences the main neighboring coordination centers for eye movements in the reticular formation of the brain stem, spontaneous abnormal eye movements occur that have the characteristics of nystagmus (Fig. 1.30).

Peripheral functional failure is compensated centrally by adjustment of the difference in neuronal activity in the vestibular centers and by substitution of visual and somatosensory regulatory mechanisms for the loss of peripheral vestibular function. This process is called *central vestibular compensation.* Central vestibular disorders are only incompletely compensated by the above mechanisms (or not at all), since the multisensory connections to the vestibular centers are damaged.

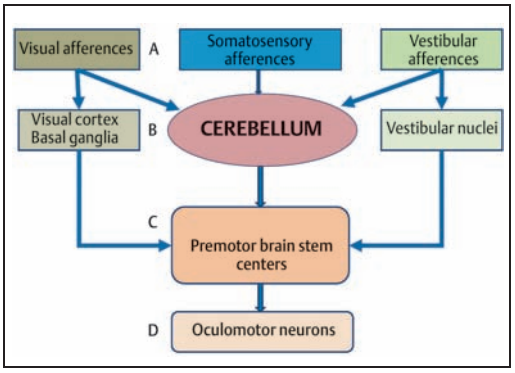


Fig. 1.30 Oculomotor system. All three sensory systems (A) send afferent signals via relay stations (B) in the premotor centers of the reticular formation of the brain stem (C). The motor neurons (D) that innervate the eye muscles begin at this point. The cerebellum is the key to coordination: visual, somatosensory, and vestibular signals are continually being compared with one another. If this structure receives contradictory information that could lead to disorientation and dizziness, the vestibular signal is modified or, if necessary, completely suppressed.

Methods of Investigation

Inspection, Palpation, Otoscopy, Microscopy

Inspection of the External Ear

The physician should look for redness, swelling, ulceration, tumors, malformations, fistula, or retroauricular scars.

Palpation

The mastoid process should be palpated with both hands to search for swelling and for sensitivity to pressure on the surface of the mastoid process and at its apex. The auricle is examined for pain when pressure is applied to the tragus or when the auricle is pulled. Finally, the regional lymph nodes in the preauricular and postauricular areas and the upper deep cervical chain are examined.

Otoscopy

The external auditory meatus and the tympanic membrane are examined, and if a perforation is present, the middle ear is also examined.

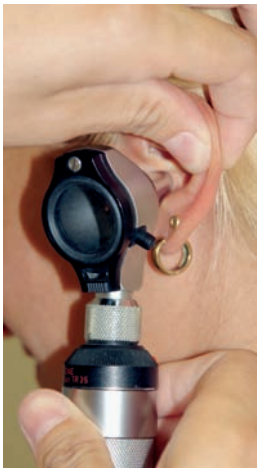


Fig. 1.31 Otoscopy with an illuminated otoscope, which consists of a disposable ear speculum, a light source, and a magnification attachment.

Indirect illumination with a head mirror is a difficult method of investigation for the nonspecialist, as correct adjustment of the light source and head mirror require time and practice, especially when patients are being examined in bed (see Fig. 2.16).

The electrical *otoscope* is more widely used, as it is easier to handle. It consists of a combination of an interchangeable *ear speculum* with a small, but strong, built-in low-voltage *light source* and a *magnification attachment* providing a magnification of 1.5–2 × (Fig. 1.31).

The *otomicroscope* provides a magnification of 6–12 × and is indispensable for accurate examination of the meatus, tympanic membrane, and parts of the middle ear in cases of perforation.

The *oto-endoscope* provides a wide-angled and magnified view over the tympanic membrane, allowing complete investigation of the anulus and anterior tympanomeatal angle. Rigid scopes with 0° and 30° views are used.

Technique of otoscopy. The cartilaginous part of the external meatus is stretched by pulling the auricle upward and backward. The speculum is then introduced into the long axis of the bony meatus. The instrument is held with one hand, so that the other hand remains free for handling instruments such as cotton-wool probes, hooks, an aspirator, and aural forceps (Fig. 1.32). The speculum has to be introduced carefully, and the end of it should not be moved abruptly, as its opening has relatively sharp edges. The wall of the bony meatus is particularly sensitive and easy to injure, and contact with it should therefore be avoided.

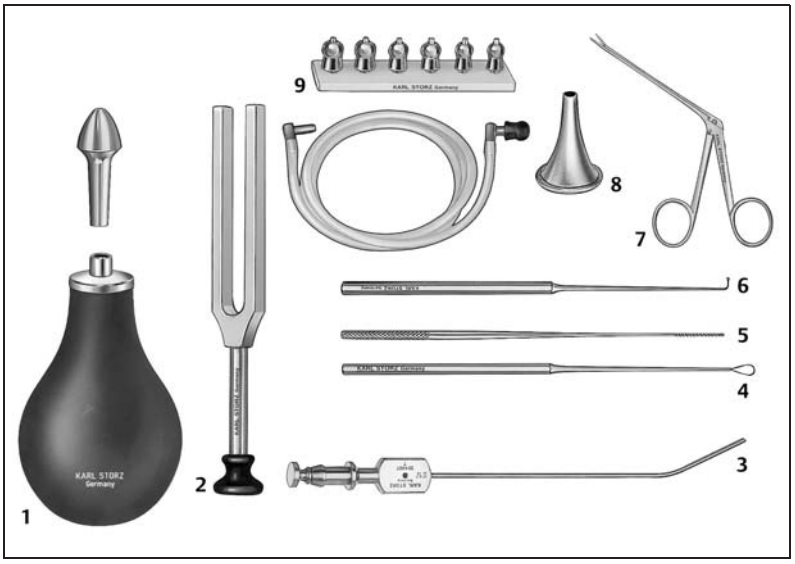


Fig. 1.32 Important ear instruments. 1, Politzer bag with olive; 2, tuning fork; 3, suction tube; 4, curette; 5, driller for cotton pads; 6, hook; 7, microforceps; 8, ear speculum; 9, hearing tube with olives (of various sizes).

In *infants* and *young children,* the auricle is pulled downward and backward to allow the speculum to be introduced. The short cartilaginous part of the external meatus is reduced to a cleft, which can only be entered with a narrow speculum with a small lumen, making otoscopy difficult. The head has to be immobilized, either by an assistant or using a headrest on the patient's chair, to prevent unnecessary movements that can cause pain.

Wax and other material obstructing the view into the external auditory meatus has to be removed using the following methods:

- By syringing for foreign bodies, wax, and exudate.
- With the hook or curette, for hard wax.
- With the aural aspirator, for exudate or fluid wax.
- With a cotton-wool probe for exudate.

The ear is syringed with tap water at body temperature. Hard wax is softened beforehand with softening drops such as 3% hydrogen peroxide, 5% sodium bicarbonate, soft soap, olive oil, or a commercial preparation.

Note: Syringing the ear is contraindicated in:

- Dry perforations of the tympanic membrane
- Fresh injuries to the tympanic membrane and meatus
- Longitudinal and transverse fractures of the petrous pyramid, with meatal trauma

It is important to obtain a history of any previous perforation, as syringing may rupture a thin scar. In the United States, failure to take a history can result in a malpractice suit.

Mistakes to be avoided:

- A speculum that is too narrow and that penetrates too deeply into the sensitive bony meatus.
- Introducing the speculum in the wrong direction—e. g., from above downward.
- Not introducing the speculum far enough, causing its opening to be blocked by otic hairs.
- Unsatisfactory cleaning of the external meatus, so that a proper view of the tympanic membrane is not obtained.

Otomicroscopy. This is performed with a speculum under the operating microscope, with a magnification of 6–40×, in all cases in which routine otoscopic examination does not allow reliable assessment of the tympanic membrane (Fig. 1.33).

Oto-endoscopy. This is performed with a tele-otoscope (0° and 30° view). This allows examination of the whole tympanic membrane, meatus, and anulus, as well as assessment of perforations and pockets in the tympanic membrane, and anterior angles and open cavities after surgery (Fig. 1.34).

Normal Otoscopic Appearance

Characteristics of the tympanic membrane: The pars tensa is grayish-yellow. The cutis layer is often slightly injected. The surface is smooth and without any relieving features, apart from the handle of the malleus. The membrane is moderately translucent and is only transparent in scarred areas. A tympanic membrane showing the properties described above is described as *normal*. The mobility of the tympanic membrane can be assessed using a pneumatic otoscope (Fig. 1.35).

The tympanic membrane is moved back and forth with positive and negative pressure while it is in the field of vision. Atrophic parts flutter, and the movement of the pars tensa may be limited by scar tissue. In the presence of a perforation, the remnants of the tympanic membrane are completely immobile.

Appearance of a Pathologic Tympanic Membrane

- Injection of the vessels and inflammation are seen in otitis externa (occasionally), myringitis, and otitis media.
- Hemorrhage is red if fresh, or brownish if old. Blood vesicles are seen in influenzal otitis, and the hemotympanum is dark blue.
- Serous exudate: A fluid level can be seen, and there are air bubbles in the fluid. The tympanic membrane looks like oiled silk when there is a complete middle ear effusion. A blue tympanic membrane or "blue drum" is seen in advanced stages.
- Retraction of the tympanic membrane as a result of decreased pressure in the middle ear: The short process of the malleus protrudes externally, and there is displacement of the manubrium of the malleus posteriorly and superiorly, causing an apparent shortening of the malleus handle. The triangular light reflex is fragmented, or disappears entirely.
- Bulging due to the formation of exudate behind the tympanic membrane, at times with an irregular surface, which may be papillary, with an opaque surface.
- Atrophy of the tympanic membrane with retraction pockets results from chronic inflammation and reduced pressure. The site of predilection is the posterosuperior quadrant.
- Thickening of the tympanic membrane, as a result of degenerative changes or as the result of inflammation, produces a surface that is dark and lacking in luster.

Fig. 1.33 Ear microscopy: variable magnification between 6 × and 12 ×, with a focused light supply.

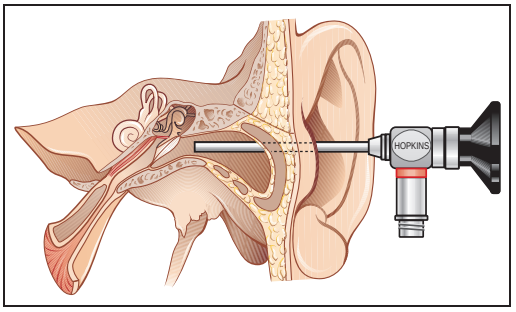


Fig. 1.34 A rigid telescope with a straight or 30° angled view and a diameter of 2.7–4.0 mm is used for oto-endoscopy. The external meatus should be stretched by pulling the auricle upward and backward, as in other otoscopy procedures.

Fig. 1.35 Pneumatic otoscope with loupe (Welch Allyn, Skaneateles Falls, New York, USA).

- Scars of the tympanic membrane: These may be thickened areas, with or without calcium deposits or atrophic areas.
- Tympanic membrane perforations: These may be either central or peripheral, mesotympanic or epitympanic. Central or mesotympanic defects are the result of chronic mucosal inflammation (see p. 67), whereas peripheral or epitympanic perforations are usually associated with a cholesteatoma (p. 68).

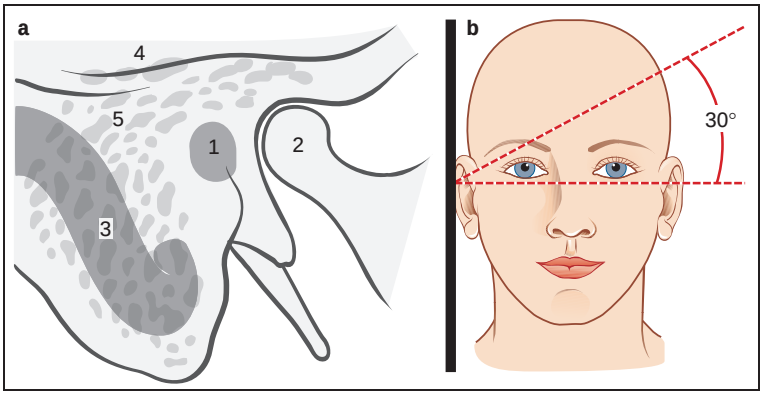


Fig. 1.36a, b Radiographs in the Schüller view. 1, The external and internal meatus superimposed; 2, the head of the mandible; 3, the sigmoid sinus; 4, the border to the middle cranial fossa; 5, the sinodural angle.

Note: A tympanic membrane that has a surface with an opaque and dull appearance—as a result of inflammatory infiltration of the pars tensa, with hyperemia, edema, formation of bullae, desquamation of the epidermal layer, and distortion of the characteristic appearance of the handle of the malleus—is designated as abnormal or pathologic.

Diagnostic Imaging

The position in regional anatomy of the petrosal bone inside the skull base generates overlapping artifacts during radiographic examinations. Special radiographic images of the temporal bone, or of both sides for comparison, are therefore essential.

Conventional imaging of the petrosal bone using the Stenvers and Schüller techniques has now to a large extent been replaced by computed tomography (CT). In selected cases, however, the Schüller radiographic view is still valuable for clinical evidence.

Conventional Radiography

Schüller technique: This provides information about the degree of mastoid pneumatization and demonstrates the intercellular space and septal bone, the course of the sigmoid sinus, the tympanic roof (tegmen tympani), and the maxillary joint. These images are useful for diagnosing otitis media, mastoiditis, and fractures of the petrosal bone (Fig. 1.36a, b).

Stenvers technique: Demonstration of the inner auditory canal, and the width of the canal in particular. The horizontal and superior arches of the equilibrium/vestibular organ, as well as the petrosal apex, are also well displayed. These images are useful in cases of acoustic neuroma, destructive processes in the internal ear, and transverse fractures of the petrosal bone (Fig. 1.37a–c).

Computed Tomography

High-resolution CT images of the petrosal bone in axial and coronal views, with slice thicknesses of 1 and 2 mm, have largely replaced conventional radiographic imaging in image-guided diagnosis. In addition to clear depiction of anatomical structures, CT images precisely show inflammatory changes such as soft-tissue enlargement (mucosal swelling, cholesteatoma, or tumor masses) and fluid retention, as well as osseous destruction or fractures (Fig. 1.38a, b). A new CT technique known as *multislice spiral CT* also allows three-dimensional interpretation of the middle and inner ear structures.

Angiography

The following supplementary *neuroradiologic investigations* are indicated for suspected vascular neoplasms or space-occupying lesions in the middle or posterior cranial fossa and cerebellopontine angle (CPA):

- Carotid angiography.
- Vertebral angiography.

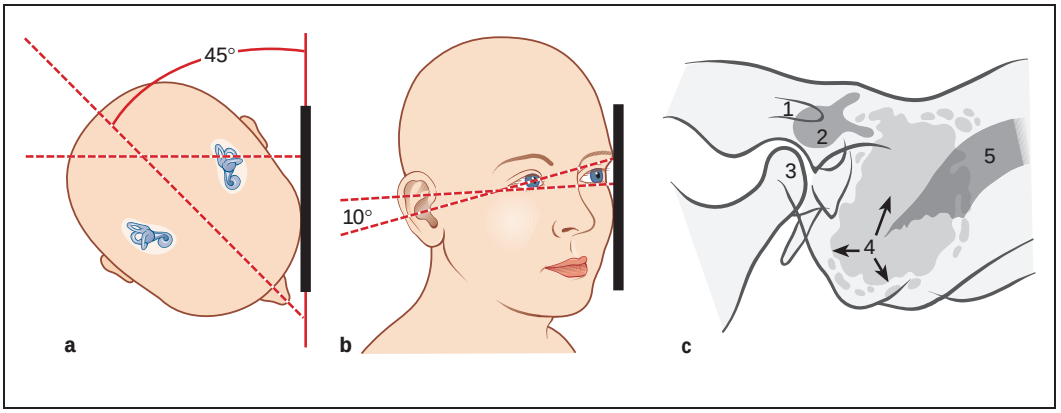


Fig. 1.37a–c Radiographs in the Stenver view. 1, Internal auditory meatus; 2, vestibule, with the superior and horizontal semicircular canals; 3, mandibular condyle; 4, pneumatic system; 5, sigmoid sinus.

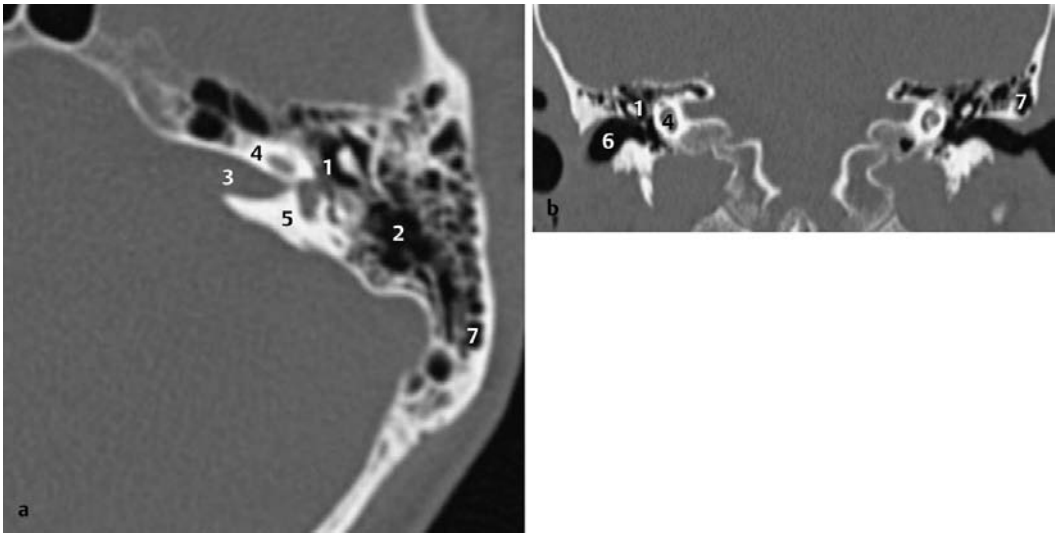


Fig. 1.38a, b Axial (a) and coronal (b) computed tomography scans of the temporal bone. 1, Tympanic cavity with ossicles; 2, antrum; 3, internal acoustic canal; 4, cochlea; 5, labyrinth; 6, external acoustic canal; 7, pneumatized mastoid cells.

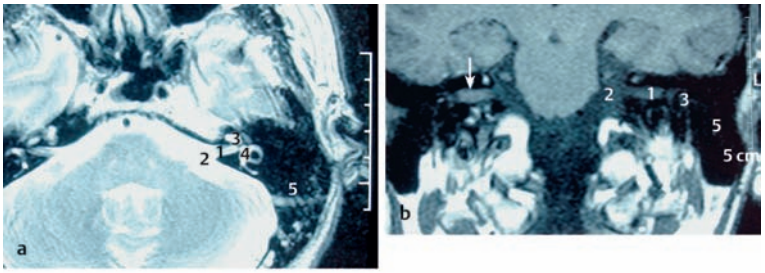


Fig. 1.39a, b Axial (a) and coronal (b) magnetic resonance images of the temporal bone area. 1, vestibulocochlear nerve in the internal acoustic canal; 2 cerebellopontine angle (CPA); 3, cochlea; 4, labyrinth; 5, pneumatized mastoid. The arrow in b shows an intrameatal vestibular schwannoma (see p. 92).

Magnetic Resonance Imaging

Magnetic resonance imaging (MRI) is an optimal method for detecting inflammatory, traumatic, or neoplastic pathology in the temporal bone and skull base. It is routinely used with T1-weighted and T2-weighted spin-echo sequences, producing thin-slice images, sometimes with contrast administration (gadolinium diethylenetriamine pentaacetic acid, Gd-DTPA). Very small vestibular neuromas in particular (acoustic neuromas) can be optimally visualized (Fig. 1.39a, b). The enhanced soft-tissue display is very helpful for assessing the extent of any neoplasms, complementing CT images.

Using high-resolution T2-weighted sequences such as the *constructive interference in steady state* (CISS) sequence—a three-dimensional magnetic resonance sequence that displays cerebrospinal fluid spaces—even details of the membranous labyrinth and neuronal structures of the inner ear canal and cerebellopontine angle become visible. The *volume-rendering* technique can even produce three-dimensional images of the entire inner ear, as well as of the topological relationships in the entire temporal bone. In addition, it provides virtual endoscopic visualization of the inner ear region.

Views can be taken in all three planes without secondary reconstruction using a computer and without the need for special positioning of the patient. Pathologic formations (e. g., tumors, ischemic lesions) can be differentiated from normal structures using contrast administration (Gd-DTPA). Other specific MRI techniques include *functional MRI* (f-MRI), which makes localized perfusion changes in the brain visible, and *magnetic resonance angiography*, which can reveal intracranial tumors (glomus tumors) and vascular structures.

Functional Assessment of the Eustachian Tube

Tests of tubal function are always necessary in all patients with middle ear hearing loss, particularly before an operation to improve hearing.

Qualitative Assessment of Tubal Function

Valsalva test. This test is used to demonstrate normal tubal patency without the need for any external aids. Failure of the test does not prove pathologic occlusion of the tube, but further functional tests may be required.

After taking a deep breath, the patient pinches his nose and closes his mouth in an attempt to blow air into his ears.

Otoscopy shows bulging of the tympanic membrane, and auscultation reveals crackling.

> **!** Note: In patients with infection of the nose and nasopharynx, inflation of air involves a risk of transmitting infected secretions into the middle ear, causing tubogenic otitis media. In patients with an atrophic scar of the pars tensa, rupture of the tympanic membrane is also possible, especially during air insufflation or catheterization of the tube.

Toynbee test. This test is used to confirm normal tubal air patency with a simple and safe method. During swallowing, pressure in the middle ear falls if the nose is closed off. This can be seen on otoscopy as a drawing-in of the tympanic membrane.

Hearing Investigations

Testing Hearing without an Audiometer

Hearing Threshold for Whispered Voice and Conversational Speech

Two-syllable words are articulated at a decreasing distance from the patient until the test words can be correctly repeated. The distance is recorded in meters. Alternatively, the examiner can say numbers or words at a fixed distance with decreasing loudness. When severe unilateral deafness is being assessed, and also when the hearing distance for conversational speech is being measured, it is necessary to mask the contralateral ear. Each ear is tested separately, with the better ear being tested first. The contralateral ear canal is closed with a finger.

Requirements:

- A sufficiently large, quiet room (6 m long).
- Good acoustic properties (no smooth walls with distorting echoes).

Tuning Fork Tests

A C^1 fork with a frequency of 512 Hz is used.

Weber test. This test is based on binaural comparison of bone conduction. The tuning fork is placed in the center of the skull at the hairline. A patient with normal hearing or with symmetrical hearing loss localizes the tone either in the center of the head or equally in both ears. A patient with unilateral conductive hearing loss (middle ear) localizes the tone in the affected ear, whereas a patient with unilateral inner ear deafness localizes the sound in the healthy ear.

Theoretical explanations:

- In middle ear disorders, the mobility of the ossicular chain is reduced and it thus transmits less sound energy than it does in normal physiologic conditions (Mach's sound wastage theory).
- Pathologic processes in the middle ear cause an increase in the mass of the sound conduction apparatus, so that increased forces are exerted at the oval window, due to inertia. This leads to greater stimulation of the inner ear (inertia theory) (Fig. 1.40a–c).

Rinne test. This test is based on monaural comparison of air conduction with bone conduction. If air conduction is better than bone conduction, Rinne's test is *positive.* This is the finding in normal hearing or sensorineural hearing loss (inner ear). If bone conduction is better than air conduction, Rinne's test is *negative.* This is found in conductive or middle ear hearing loss.

The patient is asked whether the tuning fork placed in front of the ear is heard better than when it is placed behind the ear, on the mastoid process, without striking it again. If the patient cannot decide with certainty, the decay period of the tuning fork should be determined precisely for both air and bone conduction separately (Fig. 1.41a–c).

Gellé test. This can be used to test the mobility of the ossicular chain in cases of otosclerosis (see p. 81) and fixation of the incus. The test has now been replaced by impedance audiometry (see p. 36) (Fig. 1.42a, b).

Note: Assessment of the hearing distance for whispered and conversational speech, along with tuning fork tests, provides valuable information about the site of a hearing disorder. These tests are still the basic diagnostic methods in otologic examinations (Table 1.1).

Audiometry: Fundamental Physical and Acoustic Concepts

See Tables 1.2, 1.3, 1.4, 1.5, 1.6, 1.7.

Pure-tone Audiometry

An *audiometer* is an electric tone generator used to determine the hearing threshold for pure tones—i.e., tones free of harmonics within a frequency range from 125 to 12 000 Hz.

The *hearing threshold* is measured for both air and bone conduction in decibel steps. The *normal hearing threshold is* indicated by a straight line at 0 dB. Hearing loss is measured in decibels relative to this threshold for all frequencies and is recorded on an audiogram (Fig. 1.43).

The decibel (dB) is a relative value that compares one sound pressure to another. The reference point in audiometry is the human hearing threshold of 1000 Hz. The sound pressure necessary to produce the subjective impression of hearing at a threshold of 1000 Hz is 20 µPa (2×10^{-4} µbar) (see Table 1.3). This is the average value for young individuals with normal hearing and is the reference point for the physical or absolute measurement of the hearing threshold

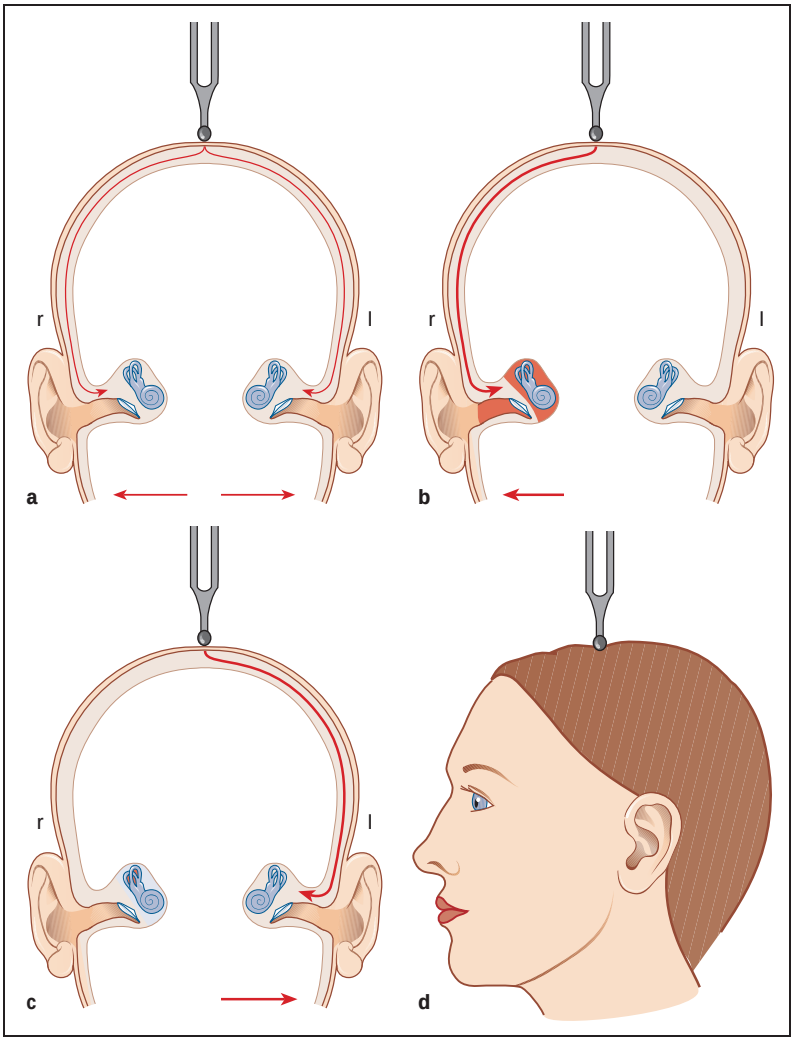


Fig. 1.40a–d The Weber test. A vibrating tuning fork is placed on the midline of the skull.
a Equal loudness perceived in both ears means symmetrical hearing.
b Lateralization of sound to the affected ear (right) is present in cases of conductive hearing loss.
c In cases of sensorineural hearing loss, the sound is lateralized to the better ear (left).
d Correct orientation of the tuning fork.

in decibels (sound pressure level, SPL). The *relative hearing threshold* for pure tones is a simpler method of demonstrating and describing the hearing threshold. The reference point is no longer the absolute sound pressure, but the just-audible threshold of hearing, measured in dB (hearing level, HL). This makes it possible to use a coordinate system with a horizontal zero line. The *absolute hearing threshold* is curved in comparison with the relative hearing threshold. The reason for this is that greater sound pressure is needed at high and low tones to produce a similar sensation of sound near the threshold than for the central part of the frequency range around 1000 Hz (see Fig. 1.44).

A disorder of sound conduction can be identified by assessing the difference between the hearing threshold for air and bone conduction, in the same way as with tuning fork tests.

Relationship between Air Conduction and Bone Conduction

Normal conduction of sound to the inner ear via the sound-conducting apparatus is defined as *air conduction* (conduction via earphones). Sound is also conducted via the bones of the skull to the inner ear, either via the middle ear *(osteotympanic* or *craniotympanic bone conduction)* or by direct transmission via the labyrinthine capsule *(osteal* or *cranial bone conduction)* (conduction via a vibrator).

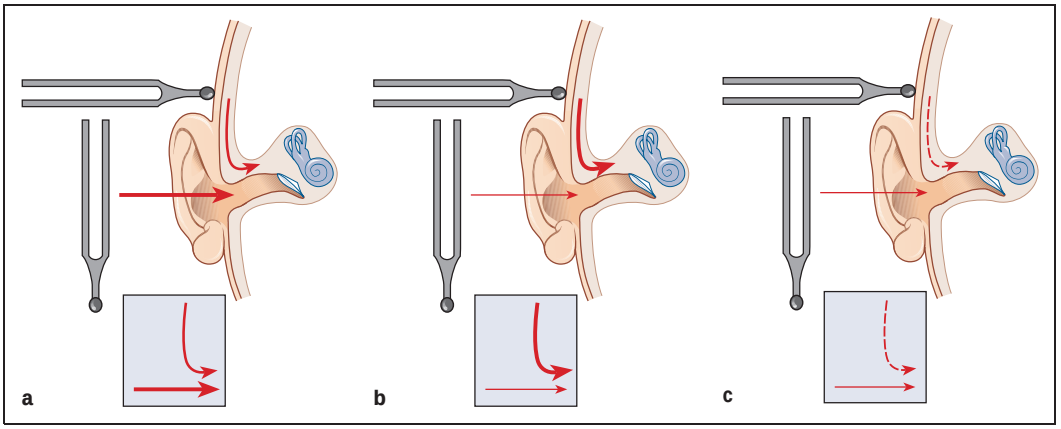


Fig. 1.41a–c The Rinne test. Air and bone conduction are compared in the same ear.
a Rinne positive, normally hearing ear. Air conduction is perceived louder or longer than bone conduction in the test ear.
b Rinne negative, conductive hearing loss. Bone conduction is perceived louder or longer than air conduction.
c Rinne positive, sensorineural hearing loss. Air conduction is perceived louder or longer than bone conduction, but the duration is shorter than in normal hearing.

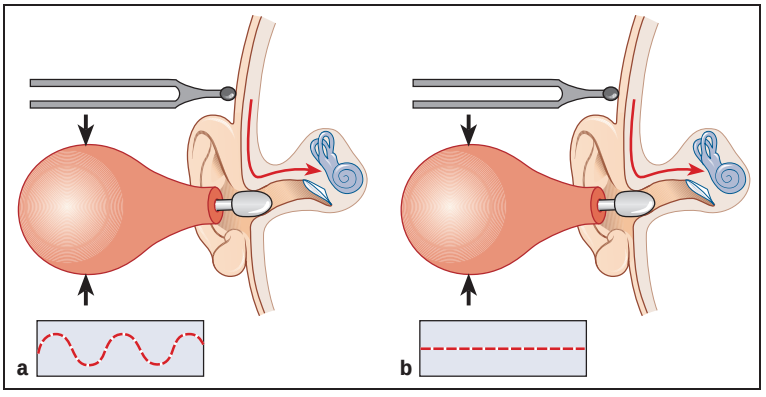


Fig. 1.42a, b The Gellé test. A fixed ossicular chain causes conductive hearing loss.
a Compression of the Politzer bag induces fluctuations of loudness in the normal ear.
b The fluctuations are absent when the ossicular chain is immobile.

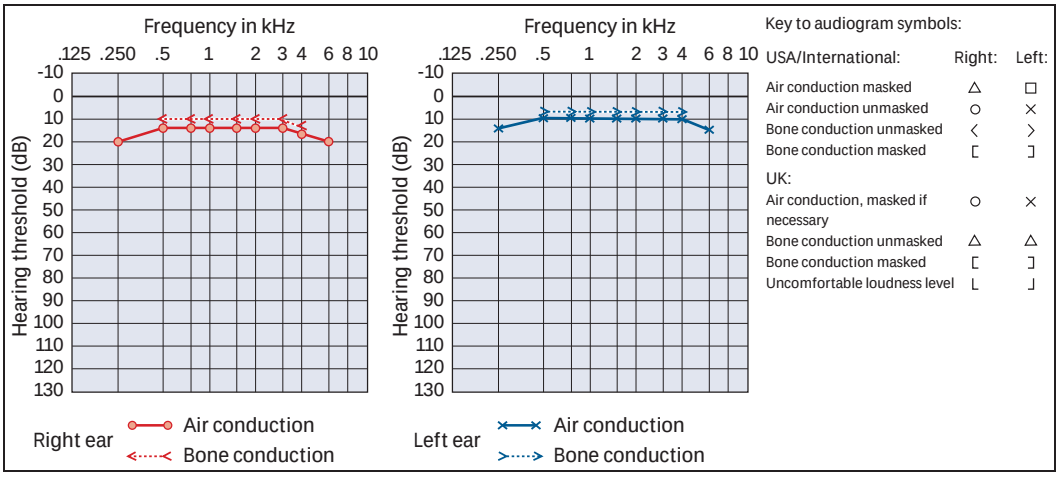


Fig. 1.43 A normal pure-tone audiogram.

Table 1.1 Evaluating the results of clinical hearing tests

	Discrepancy between hearing distance for whispered and conversational speech	Weber test	Rinne test
Normal individual	None	Midline	Positive
Conductive hearing loss	Usually small	Lateralized to the poorer-hearing ear in unilateral hearing loss	Negative or equivocal
Sensorineural hearing loss	Usually large	Lateralized to the better-hearing ear in unilateral hearing loss	Positive
Degree of hearing loss	Distance for hearing conversational speech: Slight (> 4 m) Medium (< 4 m, > 1 m) Severe (< 1–25 cm) Total deafness < 25 cm		

Table 1.2 Properties of sound

Sound	A molecular vibration of an elastic medium propagated as a waveform (in air, water, bone, and all other media)
Speed of sound	340 m/s in air, 1400 m/s in water
Sound pressure (Pa)	This is the predominant change of pressure in a sound field. It is a function of time at any particular point and is expressed in pascal units
Mass unit	The old-fashioned unit was the microbar (µb), dynes/cm^2. The SI unit for absolute sound pressure is the pascal (Pa), equivalent to newtons per m^2 (1 Pa = 1 N/m^2 = 10 µb)

Table 1.3 Hearing or dynamic range and sound pressure level

Hearing range (0 dB)	The lower limit—i. e., the hearing threshold at 1000 Hz—is 20 µPa
(120 dB)	The upper limit or pain threshold is 20 Pa
Sound pressure level (SPL)	The unit is the decibel, a logarithmic unit calculated as follows: $Lp = 20\log_{10}(p/p_0)$ dB, where p ist sound pressure being measured and p_0 ist reference pressure, defined as 20 µPa.

Table 1.4 Hearing range and decibel (dB) scale

Sound source	Intensity ratio	dB
Jet engine	1 : 10^{13}	130
Riveting hammer	1 : 10^{12}	120
Drilling machine	1 : 10^{11}	110
Printing machine	1 : 10^{10}	100
Weaving machine	1 : 10^{9}	90
Machine workshop	1 : 10^{8}	80
Street traffic	1 : 10^{7}	70
Normal speech	1 : 10^{6}	60
Soft radio music	1 : 10^{5}	50
Soft speech	1 : 10^{4}	40
Whispering	1 : 10^{3}	30
Quiet living room	1 : 10^{2}	20
Rustling of leaves	1 : 10	10
Hearing threshold	1 : 10^{0}	0

Table 1.5 Sound intensity, sound volume, and loudness

Scale of sound intensity	This is a physically defined decibel scale based on the square amplitude value of tones, rather than on a subjective assessment of the loudness of the tone
Volume of loudness level	Measured in phons, a logarithmic unit. The tone is compared subjectively with a reference sound of 1000 Hz. The sound pressure level (SPL) of the reference tone is adjusted so that the test tone and reference tone sound equally loud. The result in decibels SPL is expressed in phons. A sound with a loudness level of 50 phon produces the same sensation of loudness as a reference tone at 1000 Hz at an SPL of 50 dB
Loudness	The unit is the sone, which is a linear scale depending on subjective comparison with a measured value. The loudness of a test tone is compared with that of a reference tone of 1000 Hz and 40 dB SPL
Isophon curves	See Fig. 1.40. Consist of curves of the same loudness level measured in phons, but at different frequencies (in Hz) and SPL (dB)
Hearing range	Between the hearing threshold at 4 phons and the threshold of pain at 130 phons (see Fig. 1.40)

Table 1.6 Tone, timbre, noise

Tone	A pure sinusoidal vibration in the audible range characterized by frequency
Frequency	Vibrations per second in hertz
Timbre	A sound contains overtones in addition to the basic tone, which determine the subjective color of the sound
Noise	Sound whose pressure in the sound field is not a periodic function of time
White noise	Consists of equal components of all the audible frequencies from 18 to 20 000 Hz
Loud noise	May be distressing or cause actual damage

Table 1.7 Impedance

Acoustic impedance	Resistance to the flow of sound pressure waves through a medium, proportional to: The mass of the vibrating system Its resistance Its elasticity
Resistance	The frictional resistance in the joints, ligaments, and muscles of the sound-conducting apparatus
Reactance	An imaginary component determined by the stiffness and mass of the system
Compliance	The flexibility of the tympanic membrane

Note: The audiometric characteristic of conductive or middle ear hearing loss is that the threshold for air conduction is poorer than that for bone conduction, producing an air–bone gap.

Conductive hearing loss results from an increase in impedance (Fig. 1.45a–d). If the elastic recoil due to air in the middle ear and mastoid process increases, mobility in the middle and low tones decreases at constant mass and tension. The resonance point of the middle ear is displaced to the upper frequencies. Conductive hearing loss is characterized by greater loss of hearing for air conduction in the lower frequencies—as seen, for example, in ossification of the stapes annulus in *otosclerosis* (Fig. 1.45c; see also Fig. 1.101). The conduction system is increasingly damped by the increase in mass and tension, and the resonance point of the middle ear is thus displaced into the lower tones. The hearing loss that results that is greater for air conduction in the middle and higher tones—e. g., as occurs with glue ear exudate in the middle ear (Fig. 1.45b) and impacted wax.

Conductive hearing loss independent of frequency is caused by simultaneous elastic stiffening and dampening of the sound conduction apparatus. This may occur in advanced *otosclerosis,* in *middle ear cholesteatoma* with destruction of the ossicular chain, in *tympanosclerosis,* and in *congenital anomalies.* A flat air conduction curve is found in such cases.

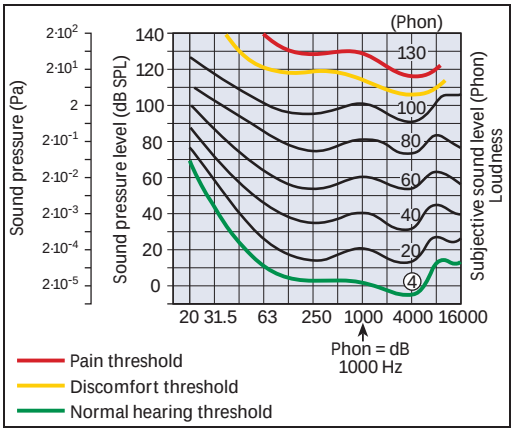


Fig. 1.44 Human auditory field. The sound pressure (in Pa), the sound pressure level (in dB), and the loudness (in phon) are shown together in a coordinate system with the spectrum of human hearing in hertz. The abscissa shows frequencies, the ordinate decibels and phon. Isophons are curves of equal loudness. The curves for decibels and phon coincide only at 1000 Hz and deviate from each other above and below this frequency.

Note: The bone conduction threshold curve is an expression of the function of the inner ear and, to a limited extent, of its central connections.

This rule applies with a few unimportant exceptions—e. g., bony closure of one or both windows.

The audiometric characteristic of all forms of sensorineural hearing loss (inner ear and retrocochlear hearing loss) is that the thresholds for air and bone conduction coincide (Fig. 1.46a–d). Supplementary *suprathreshold tests* have to be performed to differentiate inner ear hearing loss from retrocochlear hearing loss.

Demonstration of Recruitment

Patients with *inner ear hearing loss* showing *recruitment* often have difficulty in hearing relatively soft tones. In contrast, they hear loud conversational speech as well as individuals with normal hearing. They find excessive loudness upsetting due to distortion and painful sensations, as the *threshold of discomfort* is exceeded. In inner ear deafness, recruitment occurs in the frequency range of the damaged hair cells, which require a considerably higher sound pressure in comparison with the normal hair cells to produce a response. The resulting reduction of the dynamic hearing range has extremely deleterious effects as far as hearing of speech is concerned (see Fig. 1.44 and pp. 108–110).

The following tests are used:

Fowler test. *Principle:* This test is based on a subjective comparison of loudness between the right and left ears. A tone of the same frequency and loudness is presented alternately. A recruitment phenomenon is present if a difference in the hearing threshold between the two sides disappears as the loudness of the test tone increases (Fig. 1.47).

Note: Demonstration of the recruitment phenomenon is currently accepted as indicating an inner ear or hair cell lesion, whereas this phenomenon is usually absent in retrocochlear neural hearing loss due to vestibular schwannoma, for example.

Tone intensity difference threshold (Lüscher test). *Principle:* The threshold of intensity difference in decibels at the same distance above the hearing threshold is smaller in an ear affected with recruitment than in the normal ear.

Short increment sensitivity index (SISI) test. *Principle:* A test tone is produced 20 dB above the patient's threshold and is increased by 1 dB every 5 seconds with a duration of 0.2 s. Negative results are obtained in retrocochlear lesions with pathologic fatigue. The score is greater than 80 % in patients with cochlear hearing loss showing recruitment.

Demonstration of Pathologic Fatigue

Pathologic auditory fatigue is a sign of a retrocochlear hearing loss. It can be demonstrated using the technically simple *tone decay test* and the *Békésy test.* These two methods have now been largely replaced by measurement of auditory evoked potentials (AEPs; see p. 37), which allows objective testing of auditory functions and considerably more accurate diagnosis of retrocochlear hearing disorders.

▪ Speech Audiometry

Speech audiometry is an integral part of audiometric methods of investigation. The ability to hear and understand speech is more important in human communication than the ability to hear pure tones. Speech audiometry therefore has both diagnostic and therapeutic significance. To understand the re-

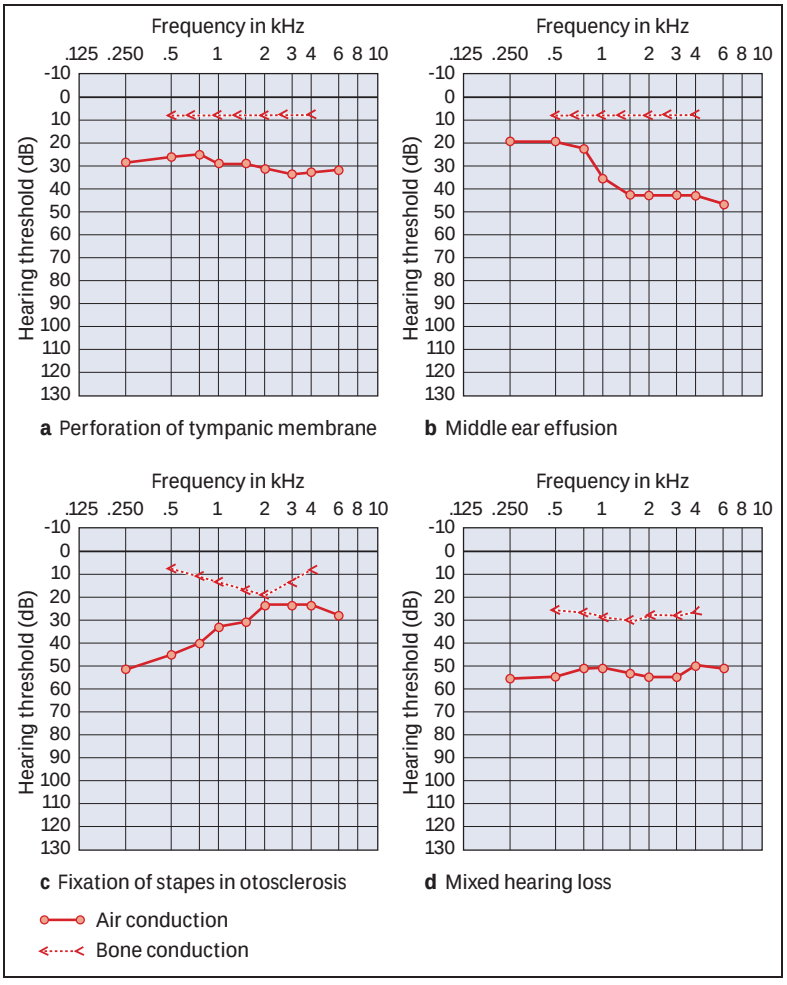


Fig. 1.45a–d Audiograms showing conductive hearing loss (right ear).

sults of speech audiometry, it is necessary to know the frequencies contained in speech. The fundamental vocal frequencies for men (125 Hz) and women (250 Hz) are shown in a tone threshold audiogram in Fig. 1.48.

The loudness of speech is perceived as an acoustic image, the frequencies of which extend from 100 to 8000 Hz. *Hearing loss for speech* is assessed using two-syllable test words, and maximum discrimination is also measured using one-syllable test words (Fig. 1.49a).

Speech audiometry is not performed in the same way as testing of the vocal speech (see p. 127)—i. e., with an increasing distance between the patient and the sound source—but rather by varying the loudness as measured in decibels, i. e., with a speech sound level above 20 µPa (see p. 30, Table 1.2).

The speech or test material is recorded on a disk and is presented to the patient either using earphones or in a free field using a loudspeaker with varying loudness levels. The percentage of numbers, words, or sentences understood correctly at each loudness level is then assessed.

The dependence of speech comprehension on the loudness level is tested using speech audiometry. In the standardized test (e. g., the *Freiburg speech test*), multisyllable numbers are first used. This can provide a rapid rough estimate of the extent of hearing loss.

An individual with normal hearing understands 50 % of numbers presented at 18.5 dB. This normal value forms the basis for assessing hearing loss for numbers. The patient's ability to comprehend monosyllabic words is also tested.

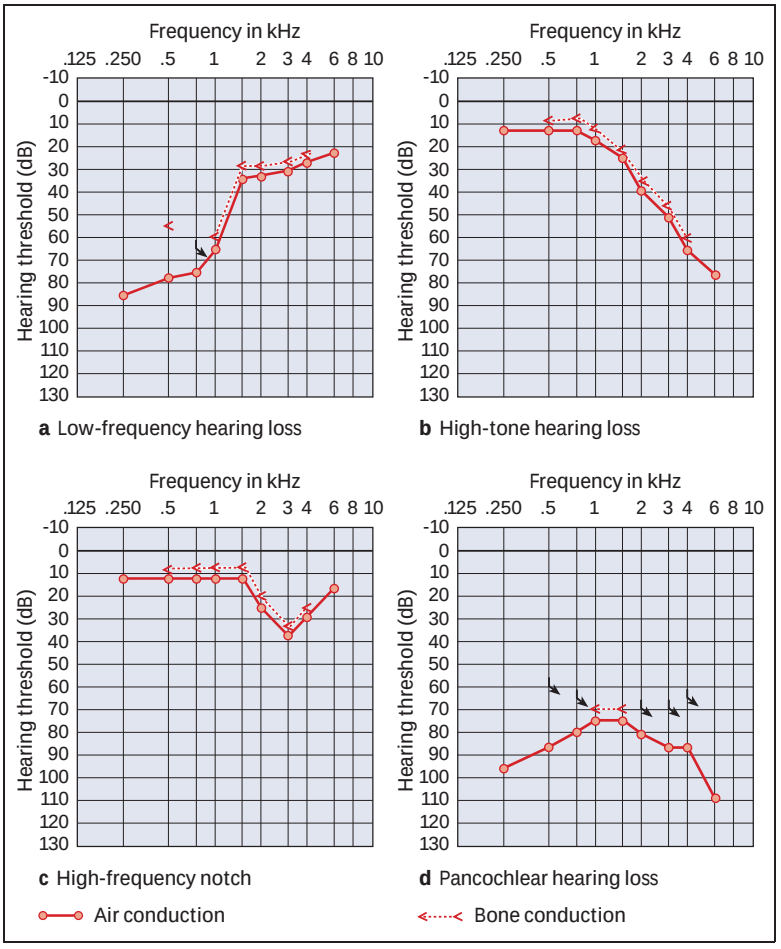


Fig. 1.46a–d Audiograms showing sensorineural hearing loss.
↘ = no response

These words are considerably more difficult to understand than multisyllabic numbers. The purpose of the monosyllable test is to assess percentage comprehension and ultimately to achieve 100 % comprehension values, if possible, by increasing the loudness level. Normal individuals hear 100 % of monosyllables at 65 dB, and in favorable conditions at 50 dB, whereas 100 % speech comprehension cannot be achieved even in normal individuals at a sound pressure level of less than 50 dB.

Speech audiometry allows quantitative measurement of hearing. The speech audiogram indicates the percentage of syllables, words, or sentences that the individual has heard correctly in each test series. The result of a speech audiogram depends not only on hearing, but also on higher cognitive functions such as memory, language comprehension, and motor speech. Other factors that influence the results include whether the patient's mother tongue is being used and the patient's vocabulary range.

Comparison of pure-tone and speech audiograms. Discrepancies between the results of pure-tone and speech audiometry are mainly found in retrocochlear hearing disorders. In such cases, hearing for speech is considerably worse than hearing for pure tones. The pathophysiologic basis for this is described on p. 17.

Diagnosis of *central hearing disorders* is based on tests of the central understanding of speech. The classic methods of testing hearing fail in such cases due to the phenomenon of *redundancy.* This is the safety margin within the auditory pathways, which can transmit and analyze billions of information units, whereas only 100 are necessary for recogniz-

ing and decoding acoustic information. A disorder of the central summation and integration capacity can only be demonstrated with difficulty—e. g., by distorting speech by filtering out high frequencies and inserting periodic interruptions of the speech signal, or with binaural application of garbled test words, reducing the information content of normal speech to a minimum (Feldmann dichotic speech test).

Note: Speech audiometry is indispensable for:

- Assessing residual hearing for speech. This makes it possible to predict the probable benefit to be expected from a hearing aid. The loss of discrimination and the threshold of discomfort can be measured.
- Assessing the need for hearing aids and surgery to improve hearing.
- Investigating central hearing loss. This allows assessment of the integrative performance of the auditory centers.
- Assessment, for insurance purposes, of a loss of hearing for speech leading to a loss of earning capacity.

Objective Hearing Tests

Behavioral, pure-tone audiometry is based on a subjective response from the patient. In contrast, objective audiometry makes it possible to carry out testing without eliciting a patient response. This method uses tests based on involuntary physiologic reactions and "objective" parameters. These objective responses support the interpretation of pure-tone audiometry and are very important for audiometric diagnosis in infants, small children, and patients with mental and cognitive impairment.

Three main methods are used in objective audiometry:

- Measurement of changes in the acoustic impedance of the tympanic membrane*: impedance audiometry.*
- Measurement of acoustically evoked bioelectric responses of the cochlea, vestibulocochlear nerve and tract, or cerebral cortex: *auditory evoked potentials (AEPs).*
- Measurement of spontaneous or acoustically evoked vibrations of the cochlea: *otoacoustic emissions (OAEs).*

Fig. 1.47 Fowler's loudness balance test in a patient with unilateral left-sided sensorineural hearing loss, with a 40-dB loss at 1 kHz.

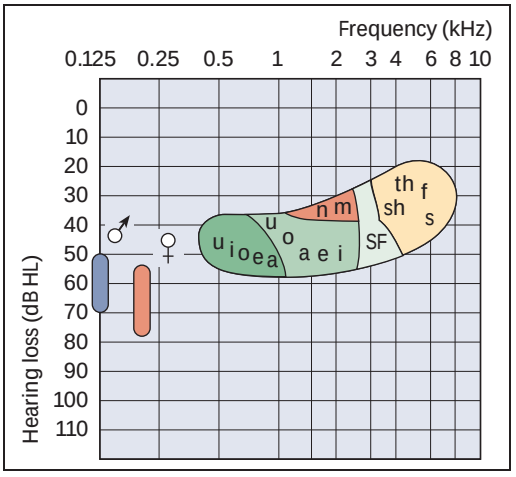


Fig. 1.48 The speech field. The fundamental voice frequency is 125 Hz in men and 250 Hz in women. Vowels are formed between 500 Hz and 4000 Hz and are spoken ≈ 10–20 dB louder than consonants in normal conversational speech. Several consonants lie in a higher frequency range (s, t) and therefore cannot be perceived by patients with high-frequency deafness; "e" as in "bed," "a" as in "bar." Dark green area = region of the first formant; medium green area = region of the second formant; light green area = region of the speaker's formant (SF); red area = resonance of the nasal tract.

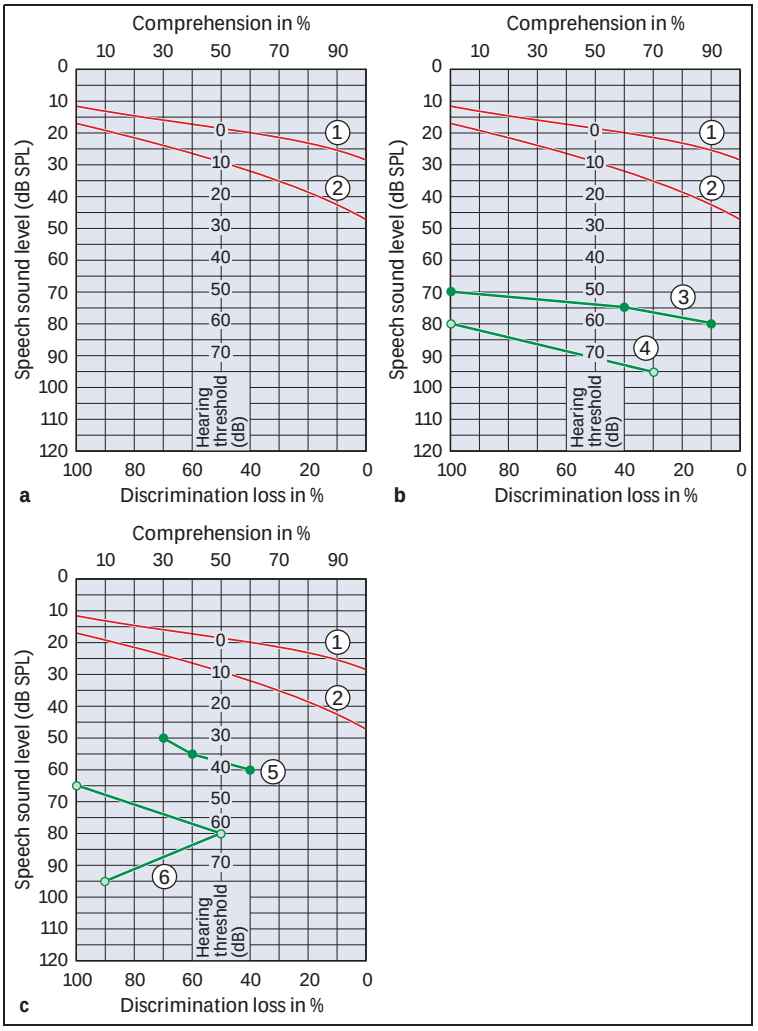


Fig. 1.49a–c Speech audiometry is carried out using uniform test content consisting of multisyllabic numbers and monosyllabic words.
a An individual with normal hearing understands 50 % of numbers heard at 18.5 dB and 100 % of those heard at 30 dB (1). For monosyllabic test words (2), intelligibility is 50 % at 30 dB and 100 % at 50 dB.
b In patients with conductive hearing loss, a parallel shift toward higher sound levels occurs in the performance–intensity function (3), but nearly 100 % comprehension can still be achieved at sufficiently high levels (4).
c Sensorineural hearing loss leads to a flattening of the performance–intensity function for monosyllabic words (5). Loss of intelligibility and a decline in speech recognition at higher sound levels are signs of abnormal speech processing, such as that caused by cochlear damage or neural disturbances (6).

Impedance Audiometry

This technique is part of the functional diagnosis of the sound conduction apparatus. It includes the following two investigation methods:

- *Tympanometry:* This involves recording the impedance (see p. 14) or indirect measurement of pressure in the middle ear, when the tympanic membrane is intact, by means of pressure in the external meatus. This is an indirect test of tubal function.
- *Measurement of the acoustic reflex:* The change in impedance caused by the acoustic stapedial reflex is measured.

Technique

The external auditory meatus is closed by an airtight plug, through which three tubes pass. One tube carries the test tone; the second is connected to the pressure regulator, which allows positive or negative pressure (± 400 mmH_2O) to be produced in the external auditory meatus. A microphone is connected to the third tube, allowing measurement of the sound pressure of the test tone reflected from the tympanic membrane as the impedance changes (Fig. 1.50).

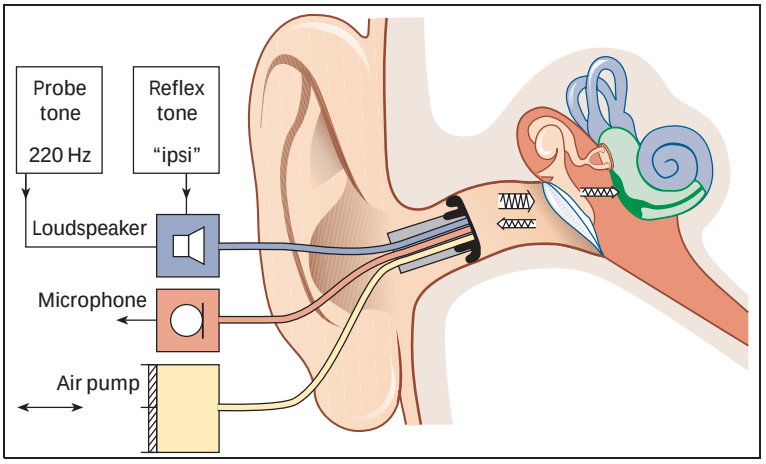


Fig. 1.50 Tympanometry. Tubes passing through an air-tight plug transmit the test tone and the reflected tone. The flexibility of the tympanic membrane, compliance, is calculated from the measured sound level. Pressure-dependent displacement of the tympanic membrane is regulated by an air pump.

Tympanometry. Normally, there is no pressure differential between the two sides of the tympanic membrane, so that the acoustic resistance of the tympanic membrane is minimal. Recording the impedance of the tympanic membrane during a change in pressure in the external auditory meatus allows the pressure difference on the two sides of the tympanic membrane to be determined by measuring its compliance. The greater the pressure differential, the greater is the impedance of the tympanic membrane. Recording the impedance at pressures from –300 mmH_2O to +300 mmH_2O produces a curve with a peak at zero for a normally mobile tympanic membrane. This represents the maximum flexibility—i.e., compliance—of the tympanic membrane, and thus minimal impedance. The apex of this curve is lower if the tympanic membrane is stiffened by scar tissue or damped by exudate in the middle ear. It becomes higher with increasing compliance due to atrophic scars of the pars tensa (Fig. 1.51a–d).

Stapedial reflex. The *principle* of this test is that a sound stimulus greater than 70 dB above the threshold induces a reflex contraction of the stapedius muscle. This causes a change of impedance at the tympanic membrane, which can be recorded graphically. The effect is absent when the tympanic membrane is immobile, when the ossicular chain is disrupted, and when the stapes is fixed in the oval window by otosclerosis. In simulated deafness, this reflex is activated by loudness approaching the norm. In this case, simulation can be assumed.

The stapedial reflex is an acousticofacial reflex. The afferent limb is the vestibulocochlear nerve and parts of the central auditory pathway up to the auditory centers. The efferent limb is formed by the connections between the auditory centers and the facial nucleus, and finally by the facial nerve. Measurement of the stapedial reflex is therefore very useful in topical diagnosis of facial paralysis.

Testing the threshold for the stapedial reflex is of considerable diagnostic importance for assessing the following hearing disorders: otosclerosis, recruitment (Metz recruitment is reduction of the difference between an elevated hearing threshold and the threshold for the stapedial reflex, with increasing hearing loss for high tones), retrocochlear deafness, and brain stem lesions.

The stapedial reflex is absent in:

- Retrocochlear sensorineural deafness as a result of auditory fatigue—i.e., in vestibular schwannoma.
- Otosclerosis and other middle ear diseases.
- Facial nerve damage proximal to the point at which the stapedius muscle is innervated.
- Brain stem lesions with damage to the central reflex arc.

Auditory Evoked Potentials (AEPs)

The patient is repeatedly exposed to an acoustic stimulus, either regularly or irregularly, and an electroencephalogram (EEG) is used to assess whether there is any change in brain activity. The AEPs are recorded from the scalp using needle or surface electrodes. As the amplitudes of the AEPs

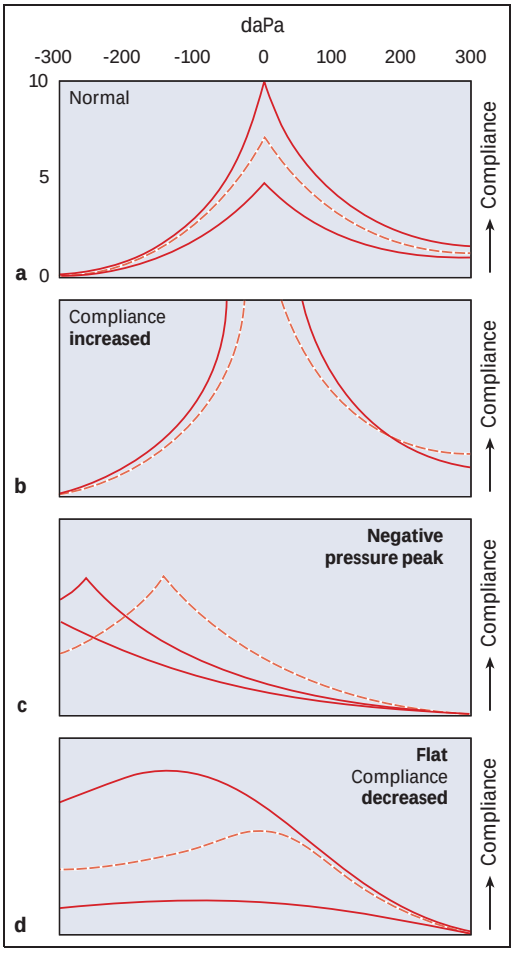


Fig. 1.51a–d Summary of the four most important results of a tympanogram. The curve shows the compliance of the tympanic membrane to changes in pressure in the external canal.
a Normal: the apex of the curve (daPa) lies close to 0 on the pressure scale when the pressures in the meatus and in the middle ear are equal.
b Increased compliance: the apex of the curve will be abnormally high if the tympanic membrane is extremely mobile. This situation may occur with atrophic scars of the pars tensa or interruption of the ossicular chain.
c Negative peak pressure: the apex of the curve is displaced below 100 daPa due to reduced pressure in the middle ear.
d Flat tympanogram with no compliance peak. This is seen when the tympanic membrane is dampened due to the middle ear effusion. This type of curve also occurs if the tympanic membrane is perforated, but the equivalent (or ear) canal volume (ECV) will then be high.

are very small relative to the total activity of the brain, averaging the potentials is necessary (see Fig. 1.49). Averaging means that the individual response, which is concealed on the EEG by the "noise" of brain activity, can be distinguished by mathematical analysis of numerous evoked individual potentials. The intermittent acoustic stimulus produces a uniform potential during a time interval that always occurs at the same time and can be amplified by repetitive summation of the EEG segment.

The properties and shape of AEPs depend partly on the time at which they occur after presentation of the acoustic stimulus, or their latency (in milliseconds). Several types of AEP can be distinguished on the basis of different sites of origin and latency.

Classification

Electrocochleography (ECochG). This measures the potentials arising in the cochlea and vestibulocochlear nerve. These potentials occur ≈ 1–3 ms after the stimulus is presented. The two most useful diagnostic parameters are *cochlear microphonics* (CM) and the *action potential* of the vestibulocochlear nerve (PI).

Auditory brain stem response (ABR) audiometry (brain stem evoked response audiometry, BERA). This measures the potentials arising in the vestibulocochlear nerve and brain stem structures, with a latency of up to ≈ 10 ms. The latency of individual potentials, particularly between potential peaks I and V, is very important for recognizing retrocochlear hearing disorders (Fig. 1.52).

Auditory middle latency potential (AMLP) audiometry. This measures potentials with a latency of 10–100 ms that originate in the thalamus and primary auditory cortex.

Cortical evoked potentials (CEPs). This measures potentials with a latency of 100–1000 ms, which express generalized higher-order cortical function.

Measurement of the auditory brain stem response (ABR) and electrocochleography (ECochG) are two of the most important diagnostic methods for accurate differentiation between cochlear and retrocochlear deafness. The latter is due to space-occupying formations in the CPA (e.g., vestibular schwannoma), a tumor of the posterior cranial fossa, or multiple sclerosis. AEP is also very useful for investigating deafness in infants and young chil-

dren. It can also be used to assess residual function of the central nervous system in patients with severe head injuries, coma, or other conditions marked by a complete loss of consciousness. It does not, however, replace pure-tone audiometry or tympanometry (including the stapedial reflex), which still form the basis for audiometric evaluations. ABR is also tested intraoperatively in order to monitor hearing.

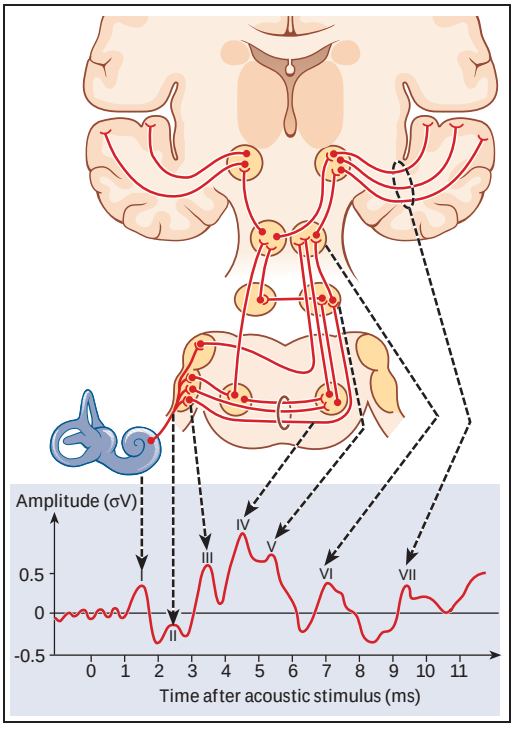


Fig. 1.52 Auditory brain stem response (ABR). The typical waveform consists of five or seven waves (I–VII), which reflect the acoustically induced activity of the anatomical structures of the auditory system (see Fig. 1.20).

Otoacoustic Emissions (OAEs)

Otoacoustic emissions are sound signals emitted from the inner ear in response to acoustic stimulation. The signals are vibrations produced by the biomechanical cochlear amplifier (see p. 16). They occur spontaneously or in response to an acoustic stimulus and are transmitted in retrograde fashion across the ossicles to the tympanic membrane. The membrane acts like a loudspeaker membrane, so that emitted vibrations can be measured as sound waves in the external ear canal. These active cochlear vibrations can be detected by a sensitive microphone.

Otoacoustic emissions are clinically important, as they reflect the functional integrity of the cochlea. OAE detection depends on normal middle ear function for good transmission to the tympanic membrane.

Classification

Spontaneous OAEs (SOAEs). Vibrations can arise spontaneously in the cochlea without any external stimulus. They are detectable as low-level, continuous tones in ≈ 50 % of individuals with normal hearing.

Transient evoked otoacoustic emissions (TEOAEs). Emissions are detected in response to an acoustic stimulus (click) in individuals with normal cochlear function. An averaging technique is used (as in ABR; see p. 38). This measurement is used as an objective audiometric testing method (Fig. 1.53a). TEOAEs occur in normal hearing and confirm cochlear integrity; they are absent in patients with middle ear disease or cochlear hearing loss with a threshold increase of ≈ 30 dB. The amplitude in infants with normal hearing is usually higher than in adults.

Distortion product otoacoustic emissions (DPOAEs). Acoustic distortions in the cochlear amplifier can be detected by stimulation with two continuous tones that have different, but adjacent, frequencies. DPOAEs are intimately linked to outer hair-cell function. This is another frequently used objective audiometric testing method (Fig. 1.53b).

The most important application of otoacoustic emissions is for screening cochlear function in newborns, infants and small children. DPOAEs can be used to detect early discrete lesions of the outer hair cells, and they provide an important noninvasive screening method for cochlear impairment that can even be used without sedation or general anesthesia. OAEs can also be used to investigate nonorganic hearing loss, to objectify audiometric findings in adults, and to assess cochlear function in risk groups (ototoxic medication).

Note: In the absence of OAEs, additional audiologic tests such as auditory evoked potentials and pure-tone audiometry should be used.

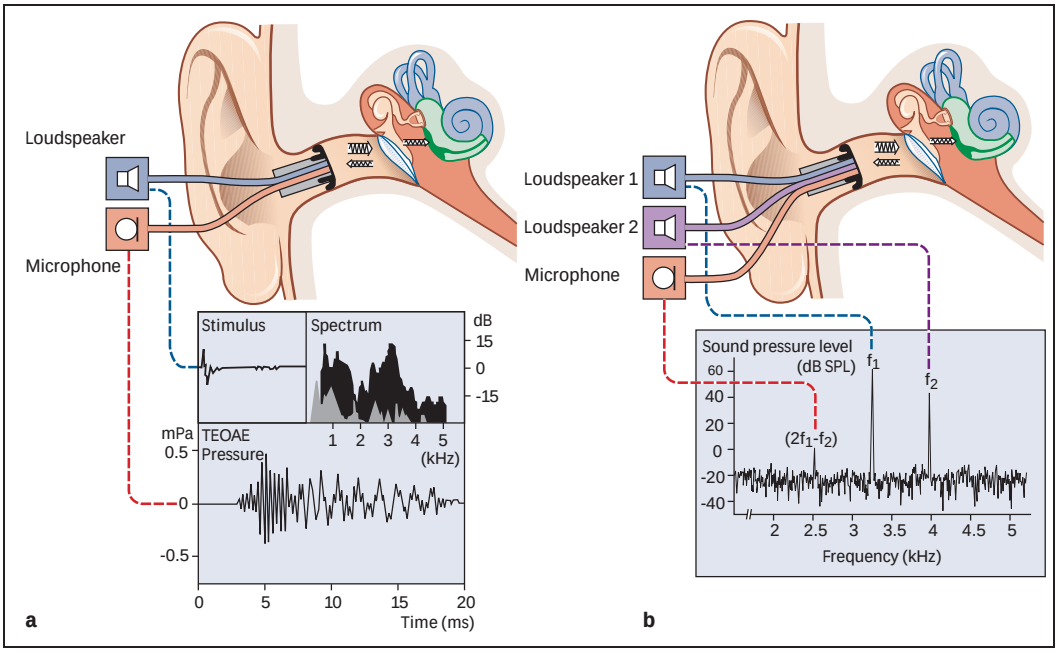


Fig. 1.53a, b a The system for measuring transient evoked otoacoustic emissions (TEOAE). A measuring probe with a microphone and loudspeaker is placed in the external ear canal. A click impulse is induced by the loudspeaker, and evoked emissions from the cochlea are recorded by the miniature microphone. b The system for measuring distortion products of otoacoustic emissions (DPOAE). The cochlea is stimulated with two tones (f_1 and f_2). The sound pressure changes as a response in the external ear canal are recorded by the microphone along with the primary tones. The curve represents the frequency spectrum of the microphone signal.

Hearing Tests in Infants and Young Children

Note: Every child who does not respond normally to sound stimuli soon after birth—and at the latest after the first 6 months—must undergo otologic examination.

Since even a completely deaf child passes through a period of crying and babbling, serious hearing loss only begins to be suspected when speech does not develop. Most children with hearing disorders are therefore presented to the general practitioner or otologist between the first and third years of life. As hearing is not an obvious condition in the newborn, it needs to be detected using screening, which is indicated in particular in the following cases:

- Necessary treatment for more than 48 hours in an intensive-care unit.
- Positive family history of hearing impairment.
- Manifest craniofacial anomalies.

Universal screening: Every newborn should be screened on the second or third day after birth during the second routine examination. Eighty percent of all hearing problems can be detected using this method. The organization required for this method of screening depends on the local health-care system.

Additional screening should be performed at routine pediatric visits or in a preschool medical examination.

Tests

Otoacoustic emissions (OAEs). If OAEs are present, the peripheral hearing is satisfactory, but this does not exclude a hearing disorder. The degree of any hearing loss cannot be determined.

Auditory brain stem response (ABR). If an ABR is not elicited, severe hearing loss is present. The hearing threshold can be determined when auditory evoked potentials are measurable.

Pediatric audiology with behavioral tests. Subjective responses in pediatric audiometric testing are important methods and can be performed at virtually any age, but should be age-appropriate. The reliability of the test results is variable.

Test Methods

Reflex audiometry. Nonspecific responses to auditory stimuli, such as sucking responses, motor responses (Moro reflex, acousticopalpebral reflex) or breathing responses, can be elicited in normal infants from birth on. The reflexes can be stimulated only by a loud noise (nearly 80 dB).

Response audiometry. By the second half of the first year of life, acoustic stimuli evoke typical response patterns. A normally hearing infant turns his or her head toward the sound source that is out of the range of vision.

Distraction test. A tester attracts the child's attention with a toy, and the examiner presents an acoustic stimulus invisible to the child and observes the reaction.

Visual reinforcement audiometry (VRA): An acoustic stimulus is combined with the activation of a moving toy. After conditioning, the child moves toward the toy when it hears the acoustic stimulus.

Play audiometry. As a variation of pure-tone audiometry, tasks and responses to tone testing are incorporated into a play setting (e.g., while playing, the child has to react when an acoustic stimulus is presented).

Pediatric speech audiometry. Children aged 3–4 years can be examined using audiometric speech tests specially designed for children (e.g., the Pediatric Speech Intelligibility Test).

Table 1.8 Checklist for suspected congenital or early acquired hearing loss

Family history	Hearing and speech disorders, psychiatric and neurologic diseases, congenital anomalies
History of pregnancy	Virus infection with rubella, measles, influenza, herpes zoster, coxsackievirus, or *Toxoplasma;* drugs such as thalidomide or aminoglycosides; diseases such as diabetes or neuropathy; or vaccination
Perinatal history	Forceps or other mechanical damage, asphyxia, prematurity, kernicterus
Postnatal history	Infectious disease, vaccination reaction, diseases of the central nervous system, trauma to the skull, intoxication, and drugs
Hearing	Reaction to noise and speech, directional hearing, the time when the hearing disorder began, and the progress of the symptoms
Speech	Age at which the first sounds, words, and sentences were uttered

Note: The sense of hearing is a vitally important factor for acquiring speech. It is therefore essential for hearing loss in a child to be recognized and treated. The earlier the treatment is instituted, the more successful it is. Treatment should be started in the second half of the first year (Table 1.8; see also Table 1.25).

Vestibular Function Tests

Investigations of the vestibular system comprise:

1. Case history and analysis of symptoms.
2. Testing of the vestibulospinal reflexes.
3. Testing for spontaneous and provoked nystagmus.
4. Experimental testing of the vestibular and optokinetic systems.

Case History

The subjective feeling of dizziness is generally regarded as being an expression of a disturbed neuronal discharge pattern in the cortical projection

Fig. 1.54 The Unterberger stepping test. The patient is asked to walk on the spot with the eyes closed.

Fig. 1.55 The patient's position for static positional tests. A spontaneous deviation reaction and spontaneous tone reaction in the arm are observed while the patient is sitting on a chair.

areas. A thorough case history is required in order to achieve a structured analysis, allowing differential-diagnostic classification of:

- Peripheral vestibular dizziness.
- Central vestibular dizziness.
- Nonvestibular dizziness.

The case history should include questions about previous illnesses, medications, and noxae. Questions regarding the type of subjectively perceived dizziness, as well as its duration and intensity, are important. Dizziness-causing factors and secondary symptoms are also important details to clarify.

■ Vestibulospinal Reflexes

In peripheral vestibular lesions, the body's center of gravity is usually displaced to the side on which the labyrinthine lesion is located. In central disturbances of balance, the pattern of unsteadiness of gait and the direction of falling are irregular. Body sways can also be registered on an electronic scale (posturography).

Romberg test. The patient is asked to stand with the feet together (touching each other) and to close the eyes. A check is made to see whether there is then any unsteadiness or a tendency to fall.

Blindfold gait and walking a straight line. Only gross abnormalities of gait are diagnostically important. The patient deviates to the same side as in the Romberg test.

Unterberger stepping test (Fig. 1.54). *Stepping on the spot with the eyes closed:* patients with peripheral disorders show rotation of the body axis to the side of the labyrinthine lesion; in central disorders, the deviation is irregular. Only deviations of more than 40° are of diagnostic significance.

Static positional tests
(see also p. 45)

Spontaneous deviation reaction, past pointing. Parallel displacement of *both* arms (with arms in the supine position) occurs in accordance with the vestibulospinal reflexes.

Spontaneous tone reaction in the arms. The arm on the side of the cerebellar lesion sinks as a result of loss of tone of the muscles (Fig. 1.55).

Finger–nose pointing test. The index finger of the outstretched arm is brought to the tip of the nose with the eyes closed. Ataxia and disorders of coordination (overshooting) indicate an ipsilateral cerebellar lesion or a disorder of positional sense and deep sensation.

■ Spontaneous and Provoked Nystagmus

Nystagmus. This is a conjugated, coordinated eye movement around a specific axis; the movement consists of rhythmically alternating slow- and fast-beating phases. The direction of the fast component of the nystagmus determines the laterality of the nystagmus.

Tests

Observation with and without Frenzel's glasses. This is used for the diagnosis of a *spontaneous nystagmus.* The patient is examined in a darkened room with +15-diopter lenses that almost completely suppress optical fixation, so that the visual fixation suppression of the vestibular nystagmus is eliminated (Fig. 1.56).

Direct gaze, with and without fixation, is used to recognize *fixation nystagmus.* Lateral gaze and gaze upward and downward are used to confirm *gaze-directional* or *gaze-paretic* nystagmus.

The direction (←), frequency (>>—), and amplitude (=) of the eye movements observed are recorded on a Frenzel's chart (Figs. 1.57).

Fig. 1.56 Frenzel's glasses with magnifying lenses allow assessment of nystagmus.

Electronystagmography (ENG). The eye is a dipole in which the cornea is electropositive and the retina electronegative. The periocular electrical field therefore changes when the eyes move. This change in the corneoretinal potential is proportional to the amplitude, frequency, and speed of the nystagmus. It can be picked up and recorded by electrodes and analyzed. The direction of the eye movements is demonstrated by a positive or negative corneoretinal potential (Fig. 1.58a–c).

Video nystagmography (VNG). The eye movements are recorded by a touchless video camera. The position of the dark pupil of the eye can be recorded by a processor that analyzes the eye's horizontal and vertical rotation.

Spontaneous Nystagmus

This term includes all eye movements that have the character of nystagmus and are not induced by external stimulation of the vestibular and visual systems (Fig. 1.59). The fast component usually beats toward the side of the functionally dominant vestibular center.

Three main forms of spontaneous nystagmus can be distinguished:

Spontaneous vestibular nystagmus. This disorder may be due either to a peripheral vestibular disorder, in which case the fast component of the nystagmus always beats toward the dominant labyrinth; or it may be caused by a central vestibular disorder. The inhibitory impulses on the vestibular center are suppressed (see p. 20). The nystagmus beats on the side of the lesion.

Recovery nystagmus may be due either to a central compensatory process after a peripheral lesion, or to the recovery of peripheral function. In both cases, it is directed toward the side of the dominant vestibular center—i. e., in this case toward the affected ear.

Gaze-evoked and gaze-paretic nystagmus. This form of nystagmus is always induced by a central lesion. Often it beats to both sides and in both the horizontal and vertical planes. It only appears after deviation of the globe by more than 30° for at least 30 s.

An exceptional form of toxic gaze-evoked nystagmus may occur after barbiturate or alcohol poisoning, due to release of the central inhibitory effect.

This form of nystagmus is due to a lesion affecting voluntary motor control of gaze, which in serious cases is accompanied by paralysis of gaze. Transitions from gaze-evoked to gaze-paretic nystagmus are fluid. The latter is characterized by a nystagmus to the side of the gaze paresis.

This is due to a congenital or acquired disorder (such as multiple sclerosis) of the gaze centers of the reticular formation of the pons (the center for horizontal gaze movement) and of the tegmentum of the midbrain (the center for vertical gaze movements). These centers are involved in central voluntary motor control of gaze (integration of voluntary gaze impulses and visual and vestibular afferents), binocular coordination via the medial longitudinal bundle (see Fig. 1.21), and the rhythm of nystagmus. Lesions in this area of the brain stem therefore lead to serious abnormalities of gaze movements and nystagmus—such as changes in the rhythm and form of beat, dissociation of movements of the right and left eyes, extinction of the fast phase of nystagmus, unilateral or bilateral enhancement of optokinetic nystagmus, gaze-evoked and gaze-paretic nystagmus, and internuclear ophthalmoplegia.

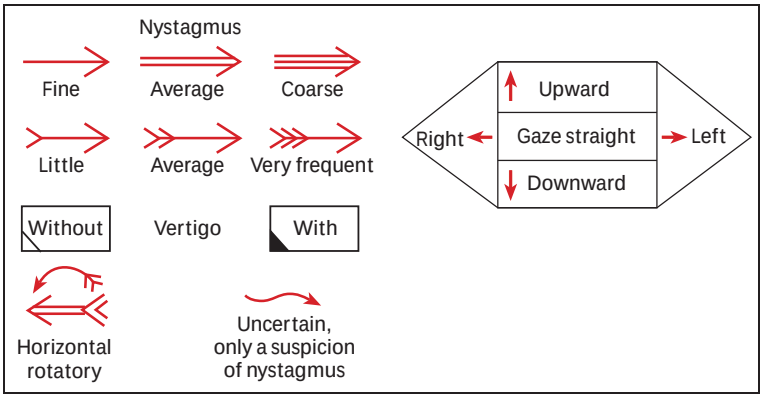


Fig. 1.57 Symbols for recording nystagmus and vertigo (left). The direction is recorded on a Frenzel's chart (right).
→ = direction,
≡ = amplitude,
⋙ = frequency.

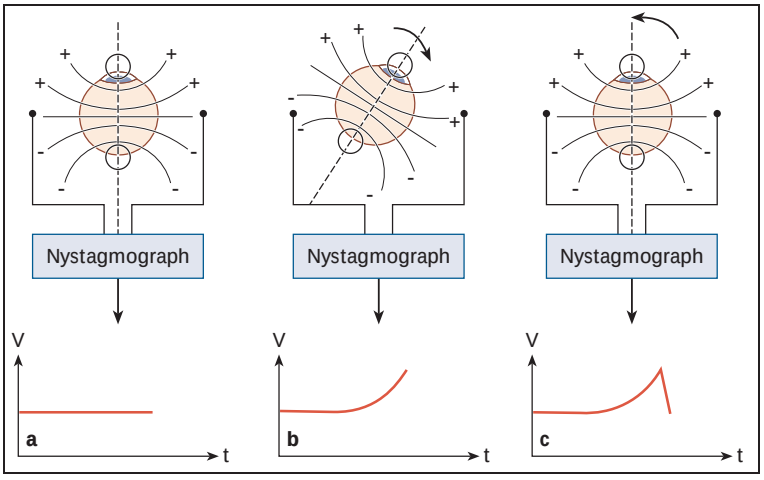


Fig. 1.58a–c The principle of nystagmography.
a Gaze straight ahead. The nasal and temporal electrodes are positive, and the isoelectric baseline is horizontal.
b The eyeball is turned slowly to the right (slow phase). The nasal electrode is positive, the temporal electrode is negative, and the baseline is displaced superiorly.
c The eyeball returns quickly (fast phase), the baseline returns to the neutral position, and both electrodes are positive.

Classification of spontaneous nystagmus

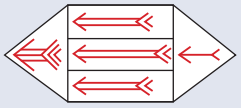
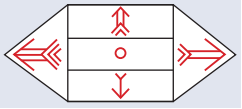
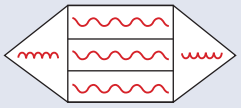
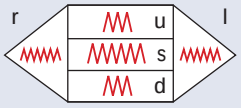
Spontaneous vestibular nystagmus		Eyes open in the dark with Frenzel's glasses
Gaze-evoked or gaze-paretic nystagmus		Even open in the light, 30° lateral gaze
Fixation nystagmus a) Pendular nystagmus		Binocular fixation
b) Latent nystagmus		Monocular fixation

Fig. 1.59 Classification of spontaneous nystagmus (u = upward, s = straight, d = downward).

Fixation nystagmus. This form of nystagmus does not have any typical fast or slow components, but rather a pendular movement. It almost always occurs with binocular fixation, but may rarely be seen with monocular fixation. It is often congenital and may even be hereditary. Synonyms for it include *congenital* or *hereditary pendular nystagmus.*

The three main forms of spontaneous nystagmus should not be confused with the following:

- *Endpoint nystagmus,* a short-lived, nonpathologic, rapidly decaying beat at the extremes of gaze—i. e., more than 50° deviation.
- *Fatigue nystagmus,* which occurs during prolonged lateral gaze due to fatigue of the lateral rectus muscle, similar to tremor in skeletal muscles. This is also nonpathologic.
- *Adjustment nystagmus,* which is due to adjustment of movements of a nystagmoid character when fixing on an object in the visual field. There is a rapid beat that fatigues quickly. This, too, is nonpathologic.

Provoked Nystagmus

Unlike spontaneous nystagmus, this is exclusively a vestibular-induced nystagmus that only appears after specific stimuli, such as changes in the position of the body or of the head.

Frenzel's glasses are used to investigate this condition. The same criteria are used for assessing provoked nystagmus as for spontaneous nystagmus; however, the duration of eye movements is also taken into account. One of the following patterns of nystagmus may be seen:

- *Transitory* nystagmus, which lasts less than 60 s.
- Continually beating *persistent* nystagmus.
- *Head-shaking nystagmus*—i. e., "release" spontaneous nystagmus of peripheral or central origin. This may be transitory or persistent.

Provocation Measures

Head-shaking. Spontaneous nystagmus can be provoked by gentle, passive, horizontal shaking of the patient's head.

Positional testing (static). The nystagmus is induced by adopting various body positions in slow motion (supine, lateral decubitus, head-hanging). The vestibular apparatus and the otolithic organs in particular are exposed to various gravitational stimuli in the different positions (Fig. 1.60a).

Positional testing (dynamic). This involves a unilateral quick movement of the patient back to a head-hanging position for 60 s. The test is repeated on the opposite side (*Dix–Hallpike maneuver*) (Fig. 1.60b).

It is a useful test for diagnosing *benign paroxysmal positional vertigo* (BPPV, see p. 100). The classic finding includes, after a short latent period of 5–10 s, a horizontal or rotatory downward-beating nystagmus toward the underlying ear, with an increase in intensity. Later, after 15–30 s, it decreases and a marked subjective feeling of vertigo arises. Occasionally, a transitory nystagmus beating in the opposite direction occurs when the patient sits up. In addition to this *paroxysmal positional nystagmus*, persistent or transitory direction-determined nystagmus, regular direction changing, and irregular positional nystagmus may be observed. The latter is always of central origin.

■ Experimental Tests of the Vestibular System

These tests and analysis of them should be performed by a specialist.

Turning test. The rotatory or turning test uses *angular acceleration* as the stimulus for investigating the sensitivity of the horizontal semicircular canals. Rotation around an axis passing through the head stimulates one or more semicircular canals on each side, depending on the head position. The left and right sides are stimulated in an opposing fashion. Rotation of the head in one direction (e. g., to the right) induces nystagmus toward the same side (rotatory nystagmus). When the rotation ceases, the nystagmus reverses to the opposite direction (postrotatory nystagmus). The test is carried out in a darkened room using a rotating chair.

Using the rotatory test, it can be determined whether the vestibular apparatus is functioning properly or whether there are signs of a functional or regulatory disorder (imbalance to one side, swinging).

Caloric labyrinthine testing. The principle involved in these tests is illustrated in Fig. 1.61a–c. The horizontal semicircular canal is brought into a vertical position in the supine patient. Cooling or warming the labyrinthine capsule by irrigation with water at 30°C or 44°C for 30–40 s induces convection currents and a slight change in the volume of the endolymph. This produces move-

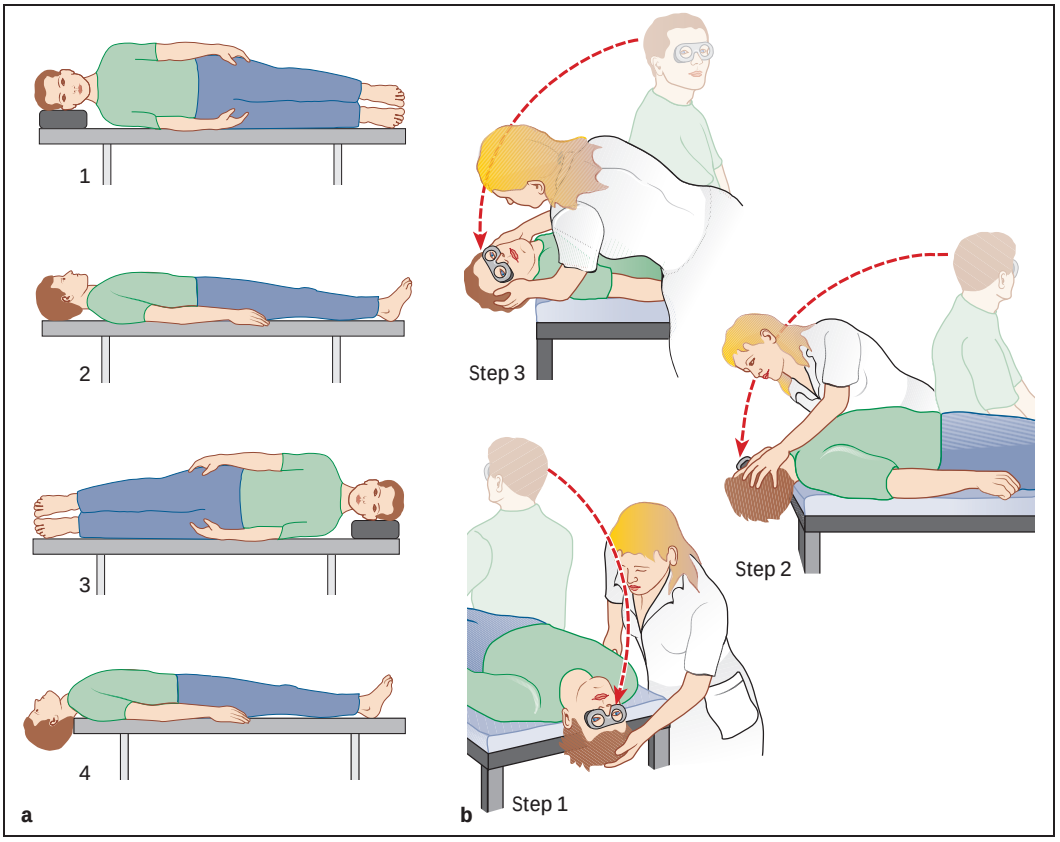


Fig. 1.60a, b a Positional testing (static). Beginning with a supine position on the examining table, the patient rolls onto the right side and then rolls back to the supine position and continues rolling onto the left side. After that, the patient adopts a head-hanging position. b Positioning test (dynamic). A dynamic provocation is carried out. Starting in the sitting position, the patient adopts a head-hanging position, and the upper body is then swiftly brought back to the sagittal plane (step 1). The head is then turned to the left (or right) and the patient adopts the head-hanging position for each side (steps 2 and 3).

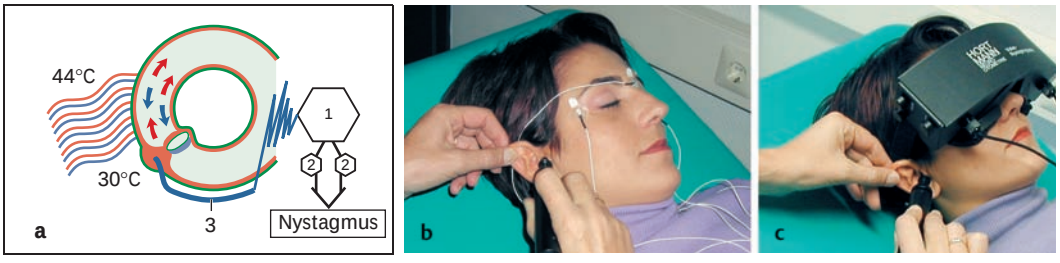


Fig. 1.61a–c Principle of caloric labyrinthine tests.
a Temperature changes at the lateral labyrinthine capsule cause a change in the density of the perilymph, leading either to an influx away from the ampulla (cold stimulus) or to an influx toward the ampulla (warm stimulus). The oscillation of the cupula produces a neural stimulus that is transmitted by the vestibular nerve (3) to the vestibular nucleus (1) and ocular muscle nucleus (2).
b Examination with electrode deflection.
c Examination with video nystagmography.

ment of the endolymph, which deflects the cupula. This process has exactly the same electrophysiologic effect as deflection of the cupula by angular acceleration (see Fig. 1.28a–c)—i. e., it induces nystagmus via the vestibuloocular reflex.

The *extent of the caloric response* (the nystagmus and the subjective feeling of dizziness) gives some indication of the function of the stimulated labyrinth. *Reduced excitability* indicates partial functional loss, and *lack of response indicates subtotal or complete loss of function.* The advantage of caloric tests is that the labyrinth on each side can be investigated separately.

Fistula test. In the presence of a fistula in the horizontal semicircular canal or elsewhere in the labyrinthine capsule (see p. 8), caused by an inflammatory osteolytic process such as cholesteatoma, a sudden increase in pressure in the external auditory meatus produces a subjective feeling of dizziness, objective nystagmus, and lateropulsion (Fig. 1.62). The same phenomenon can also occur when there are adhesions between the membranous labyrinth and the stapes footplate (fistula symptom without a labyrinthine fistula, see p. 81).

Technique: A Politzer balloon with a perforated olive is introduced into the meatus. Compression induces nystagmus toward the affected ear and aspiration to the other side.

Pseudofistula symptom: Inflation or aspiration of air in a patient with a large defect in the tympanic membrane produces cooling of the horizontal semicircular canal, which induces a caloric labyrinthine reaction and thus nystagmus. However, this always beats toward the sound ear for both compression and aspiration.

Note: The fistula symptom always has to be tested in chronic otitis media with marginal perforation of the tympanic membrane, especially when there is a suspicion of cholesteatoma of the middle ear.

Optokinetic Function and Pursuit Tracking

The optokinetic and eye-tracking tests are among the most sensitive methods available for detecting central oculomotor lesions. They are indispensable for distinguishing peripheral from central disorders of balance, as the two systems are functionally very closely linked.

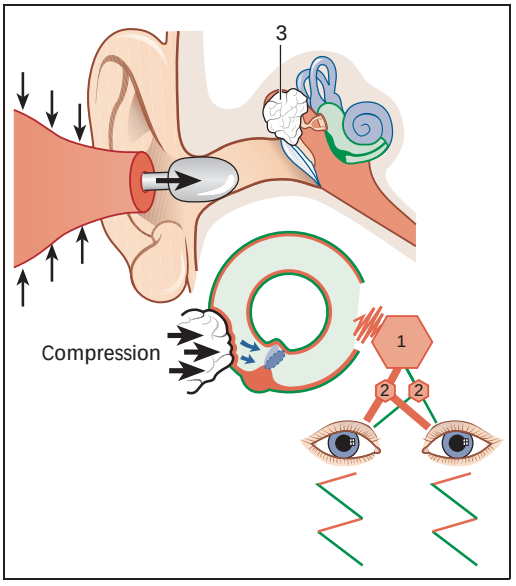


Fig. 1.62 A positive fistula test due to erosion of the right lateral semicircular canal by cholesteatoma. 1, Vestibular nucleus; 2, oculomotor nerve nuclei; 3, cholesteatoma.

Principle: Observation of an object moving within a stationary visual field (foveal stimulation) or observation of the displacement of the entire visual field (foveoretinal stimulation). Only the latter induces the optokinetic reflex—a conjugated reflex eye movement that shows a slow movement in the direction of the displacement of the moving object or visual field (gaze following movement) and a fast phase (the central correction movement) in the opposite direction. This is described as *optokinetic nystagmus.*

Brain stem lesions, especially of the pons and cerebellum, produce the changes involved in optokinetic nystagmus, such as unilateral directional preponderance, disintegration of coordinated movement of the left and right eye, and complete unilateral or bidirectional disintegration of coordinated movement. Gaze-evoked or gaze-paretic nystagmus and simultaneous abnormalities of optokinetic nystagmus are characteristic early symptoms of multiple sclerosis.

Investigation of the Facial Nerve

The first and most important investigation serves to differentiate central from peripheral paralysis:

- In *central paralysis,* the function of the branches to the forehead is preserved.

- In *peripheral paralysis,* all three branches are affected. The secretion of tears and sensitivity for taste are affected, and hyperacusis may occur due to disruption of the stapedial reflex.

The *topographic diagnosis* of peripheral lesions of the facial nerve is shown in Fig. 1.22.

Taste. The anterior two-thirds of the tongue is innervated by the chorda tympani. The test stimulus is 20 % sugar, 10 % saline, or 5 % citric acid solution (see Fig. 3.2 c).

Gustometry. The peripheral taste fibers are stimulated electrically, and the threshold is measured in milliamperes (see p. 242).

Schirmer test. The Schirmer test uses paper strips inserted into the eye for several minutes to measure the production of tears. The eyes are closed for 5 min. The paper is then removed and the amount of moisture is measured. Sometimes a topical anesthetic is administered to the eye before the filter paper, to prevent tear production due to irritation from the paper. The use of the anesthetic ensures that only basal tear secretion is measured. The reduction in the secretion of tears due to interruption of the lacrimal anastomosis in the greater superficial petrosal nerve is measured on the paralyzed side (see Fig. 1.22).

The *stapedial reflex* is measured using impedance audiometry (see p. 37).

Electrical and Magnetic Excitability Tests

The severity and prognosis of a paralysis can only be determined by *electrodiagnosis.*

> Note: Every facial paralysis, of whatever cause, has to be investigated as early as possible using electrodiagnostic methods.

Three stages of a lesion can be distinguished:

- *Neurapraxia:* There is complete absence of function, but without interruption of the axon. This stage is reversible.
- *Axonotmesis:* There is disruption of the axon with preservation of the connective-tissue framework of the nerve (endoneurium, perineurium, and epineurium). These lesions usually do not recover completely.
- *Neurotmesis:* There is disruption of the axon and of the supporting tissues. This is irreversible without surgical intervention.

Electromyography (EMG). Muscle action potentials are picked up by a needle electrode on voluntary contraction of the facial musculature. The EMG is of relatively little value in the acute phase of facial paralysis, since denervation potentials only appear 12 days after the start of the paralysis.

Electroneuronography (ENoG). The sums of the action potentials of the facial musculature induced by contraction in response to maximal percutaneous faradic stimulation are measured. The proportion of degenerated fibers can be assessed approximately by comparing the summation potential between the healthy and the paralyzed side.

Nerve excitability test (NET). The strength of current in milliamperes that is sufficient to induce a muscle twitch at a constant duration of impulse of 0.3 ms is assessed. The threshold for the two facial nerves varies to an insignificant extent (0.4 mA) in the same individual. A difference in threshold between the two sides that is greater than or equal to 3.5 mA is abnormal. An increase in the threshold indicates progressive degeneration of the nerve fibers or progressive axonotmesis.

Transcranial magnetic stimulation (TCMS). The stimulation is administered by applying a quickly changing magnetic field above the motor cortex and the facial nerve. The summation potential is diverted by the mimetic muscles. This makes it possible to measure the motor transmission speed and the location of the nerve lesion.

In the acute phase of paralysis, the results of NET and ENoG (rheobase and chronaxy) provide important information about the extent and progression of degenerative processes in the nerve and are decisive in determining the choice of treatment.

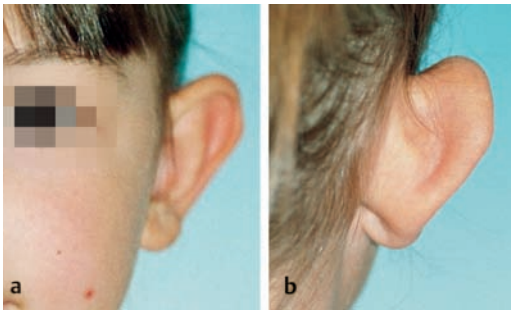


Fig. 1.63a, b Bat ears are characterized by hypertrophy of the conchal cartilage or failure of the auricle to fold.

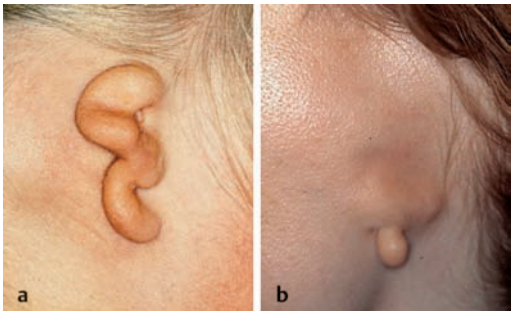


Fig. 1.64a, b Dysplasia of the auricle: type II microtia (a) and anotia, absence of the auricle (b).

Clinical Aspects of Diseases of the External Ear

Congenital Anomalies

In addition to wide variation in the position, size, and shape of the auricle, there are also numerous disfiguring anomalies. Classifying auricular deformities provides an overview of these congenital dysplasias, at increasing degrees of severity:

- *First-degree dysplasia*: minor deformities (e.g., prominent ear, macrotia, scaphoid deformity, lobular deformities, cup ear deformities types 1 and 2).
- *Second-degree dysplasia*: some characteristics of a normal auricular structure are present (e.g., first- and second-degree microtia, cup ear type 3).
- *Third-degree dysplasia*: absence of a normal auricular structure (e.g., third-degree microtia, anotia).

Lop ear (bat ear). This is one of the most common disfiguring anomalies (Fig. 1.63a, b). It is often caused by hypertrophy or excess curvature of the conchal cartilage, or by failure of the auricle to fold due to underdevelopment or absence of the antihelix.

Treatment: The deep conchal cavity is corrected, and an antihelixplasty is performed. The angle between the auricle and the head is reduced to the ideal value of 30° and the scaphoconchal angle to 90°. The operation should be performed during preschool years.

Microtia, anotia. *Microtia* (abnormal smallness of the auricle of the ear, Fig. 1.64a) and *anotia* (absence of the auricle or the ear, Fig. 1.64b) are often associated with auditory canal stenosis or atresia and with middle ear deformities. Deformities of the external ear and the middle ear can also occur in connection with other facial deformities (e.g., Franceschetti syndrome (Fig. 1.65), mandibulofacial dysostosis, Treacher–Collins syndrome).

Congenital aural fistulas and auricular appendages. These are usually located in front of the auricle. They are due to incomplete closure of the first branchial cleft or incomplete fusion of the auricular hillocks. Three groups can be distinguished on the basis of the embryologic association and site:

1. Preauricular fistulas, between the angle of the mouth and the tragus.
2. Fistulas that begin in front of the ascending helix and lead toward the meatus, or open externally inferior to the angle of the jaw as a hyomandibular fistula (Fig. 1.66).
3. A small fistula or pitted depression affecting any part of the auricle.

Treatment: Treatment is excision, with careful consideration of the danger of damaging the parotid gland or facial nerve (see pp. 112 and 435).

Reconstructive Operations on the Auricle

Reconstructive procedures on the auricle vary in difficulty. Correcting an anomaly of the auricle's shape (remodeling the auricular cartilage) can be relatively simple in the hands of a skilled surgeon

Fig. 1.65 Franceschetti Syndrome: deformity of the auricle and middle ear, with facial deformities.

Fig. 1.66 Preauricular fistula.

Fig. 1.67 Perichondritis of the auricle.

and can lead to very satisfactory results. However, reconstruction of the entire auricle is one of the most difficult tasks in plastic surgery in this area.

Thorough familiarity with skin and cartilage transplantation techniques is a prerequisite for the success of this reconstruction procedure. Costal cartilage is an ideal source for graft, and specialist instruments are required for modeling the graft. Staged surgery is necessary. Revision surgery may be required. Providing skin to create the new auricle is not easy and requires the construction of rotation and sliding flaps from the neck and from the region of the hairline. Prostheses should be considered as an alternative in difficult cases.

Reconstruction of the external auditory meatus and the sound-conducting apparatus is discussed in the section on anomalies of the middle ear (see p. 94).

Inflammations of the External Ear

Nonspecific Inflammation

Classification. Bacterial or fungal inflammation of the skin of the external auditory canal, with or without inflammation of the tympanic membrane (myringitis).

Clinical Features. *Acute exudative inflammatory phase:*

- Swelling of the skin of the auditory canal with lubricious, often fetid secretion.
- The tympanic membrane is often not visible, due to shifting of the auditory canal.
- There is an accumulation of detritus in the auditory canal.
- The cartilaginous part of the meatus is painful.
- Painful limitation of motion of the adjacent temporomandibular joint.
- The regional retroauricular lymph nodes may be so enlarged and tender to pressure in serious cases that the condition bears a strong resemblance to *pseudo-mastoiditis*.

Chronic inflammatory phase:

- The meatus is wide, the epithelial lining is atrophic, and dry scales of epidermis accumulate.
- There is intense itching, which causes scratching by the patient. This promotes the development of *superinfection* with acute dermatitis, with or without perichondritis (Fig. 1.67).

Pathogenesis

- Maceration of the meatal skin by exogenous and endogenous factors, such as fluid, mechanical and chemical damage, allergy, and diabetes.
- Reduction of the elasticity of the meatal skin and atrophy of the ceruminous and sebaceous glands.
- Loss of the protective film of secretion, with drying of the meatal skin, disturbance of its chemical balance, and increased susceptibility to infection by bacteria and fungi.
- Disturbance of the pH balance of the meatal skin, leading to the growth of anaerobic bacteria.

Factors that encourage the elimination of pathogens in the meatal skin include:

- Low pH values.
- Fatty acids in the secretions of the sebaceous glands.
- Normal lysozyme content of the secretions of the ceruminous glands.
- A normal self-cleaning mechanism, with external migration of the meatal epithelium.

Disturbance of protective factors—as may occur in allergy, with displacement of the pH to alkaline values, reduction of the protective film, and changes in the composition of the secretions due to mechanical stimulation or recurrent inflammation—can cause chronic or recurrent otitis externa.

Diagnosis and differential diagnosis

- The inflammation is localized to the auricle, external auditory meatus, and regional lymph nodes.
- Inflammation is often associated with etiological skin conditions such as eczema or psoriasis.
- Extension to the outer and middle layers of the tympanic membrane (myringitis) is only an exceptional occurrence.
- The middle ear and mastoid are not affected by the disease.
- Conductive deafness due to obstruction of the external auditory meatus is unusual and mild in nature.
- It is important to exclude acute otitis media, mastoiditis, and the acute phase of a chronic middle ear inflammation with cholesteatoma.
- Pain when pressure is applied to the tragus strongly suggests otitis externa.

Investigations. These include otoscopy, a specimen of pus for bacteriology, possibly irrigation of the ear, hearing and tuning-fork tests, audiography, and imaging. If there is any clinical suspicion of mastoiditis or other infective complications, a CT scan should be done.

Treatment. The external auditory meatus is cleaned manually under vision, ideally with a microscope, or by irrigation with water at 37°C. The external auditory meatus is dried, and eardrops consisting of locally active broad-spectrum antibiotics and a corticosteroid are instilled several times a day during the moist phase. In severe cases, systemic antibiotics are given. Once the acute inflammatory phase subsides, local applications of ointments based on a combination of an antibiotic and a steroid are used. However, certain antibiotics, notably neomycin, can themselves cause an allergic skin reaction. In these cases, the local use of 70–80 % pure alcohol is indicated, and in the acute inflammatory phase, a fine gauze wick should be introduced into the ear. This is moistened repeatedly with alcohol, which opens the external auditory meatus and reduces the swelling of the meatal skin by absorbing moisture.

Fig. 1.68 Erysipelas of the skin.

Specific Forms of Inflammation of the External Ear

Bacterial and Viral Diffuse Otitis Externa

A characteristic form of *erysipelas* occurs in streptococcal infection. In *swimmer's otitis,* due to maceration of the skin by halogen-containing swimming-pool water and deep penetration of virulent organisms, there is usually a deep-seated cellulitic inflammation (Fig. 1.68), with *perichondritis.* In addition to swimmer's otitis externa, confined to the external ear, a tubal form of swimmer's otitis media may also occur.

Clinical Features. These include fever, generalized illness, regional lymphadenitis, and pain when the auricle is pulled or pressure is applied to the tragus. A cellulitic form can extend to surrounding tissues and organs, such as the parotid gland, mastoid, and skull base, and in exceptional cases can cause osteomyelitis of the temporal bone and septicemia *(necrotizing otitis externa;* see p. 53). In severe cases of otitis externa, especially in infants and young children, there is complete obstruction of the ex-

Fig. 1.69 Edematous swelling of the lobule of the auricle is a pathognomonic sign in patients with Lyme disease.

Fig. 1.70 Herpes zoster oticus.

ternal auditory meatus, with an accompanying retroauricular lymphadenitis. The ear is displaced laterally, and the patient may appear to have mastoiditis.

Treatment. Systemic antibiotics; local reduction of the swelling of the skin of the external auditory meatus with 70–95% alcohol, chloramine in a 1 : 1000 irrigation, or local application of topical antibiotics with or without a steroid.

Furuncle of the Ear (Otitis Externa Circumscripta)

The patient is in good general condition, but has local pain in the ear. A characteristic finding is a circumscribed, exquisitely painful swelling in the cartilaginous part of the external auditory meatus (hair follicles), modest regional lymphadenitis, and pain when pressure is applied to the tragus or the auricle is pulled.

Treatment. Gauze wicks soaked with alcohol (70–95%) are used until the furuncle points and bursts spontaneously. Incision and systemic antibiotics are only exceptionally indicated in patients with severe pain, a protracted course, or marked swelling.

> **!** Note: The glucose content of the urine and blood should be checked in patients with recurrent furunculosis of the external meatus.

Lyme Disease (Lyme Borreliosis)

Lyme disease is a multisystemic bacterial infection caused by species of bacteria belonging to the genus *Borrelia*. It is transmitted to humans by the bite of an infected hard tick.

The early infection often starts with flu-like symptoms such as headache, stiff neck, fever, muscle aches, and fatigue. An enlarging rash, known as erythema migrans (EM), develops days to weeks later at the site of the bite (Fig. 1.69). Neurological problems such as severe headaches, meningitis, or cranial nerve involvement can occur at a later stage. Changes in smell or taste, vocal cord and facial nerve paralysis, vertigo with nystagmus, and hearing and swallowing disorders are typical otolaryngological symptoms. Progression of the infection is characterized by central nervous symptoms such as cognitive changes (memory problems, difficulty in finding words, confusion, decreased concentration, problems with numbers) and behavioral changes (depression, personality changes).

Herpes Zoster Oticus

This disease is characterized by multiple herpetic vesicles arranged in groups on the auricle, the external auditory meatus, and occasionally the tympanic membrane (Fig. 1.70). In severe cases, disorders of hearing and balance and facial paralysis may occur (see Table 1.16, p. 99, and p. 111).

Bullous Myringitis

This disease usually occurs in association with an influenzal infection. It is occasionally combined with otitis media, in which case the patient has conductive deafness. Initially there is a moist, bluish-livid bullous inflammation, which can extend to the tympanic membrane. After a few days, the hemorrhagic vesicles dry out and heal without complications. The patient usually reports extremely severe pain.

Note: If the middle ear is involved, systemic antibiotics must be administered immediately due to the danger of superinfection.

In most cases, treatment is limited to simple cleansing of the external meatus and otoscopic control. Antibiotics are only given for protection against secondary infection.

Necrotizing Otitis Externa

Severe necrotizing inflammation can develop from commonplace otitis externa, especially in patients with diabetes, whether latent or manifest. The infection is generally caused by optionally anaerobic Gram-negative organisms, usually *Pseudomonas aeruginosa*. The infection spreads through the tissue clefts of the cartilaginous meatus and extends into the depths of the retromandibular fossa and along the base of the skull as far as the jugular foramen, and it leads to insidious osteomyelitis of the temporal bone.

Diagnosis. The hallmark of this condition is pain in and around the ear. In addition to examining the auditory canal and carrying out a hearing test, it is necessary to examine the neighboring cranial nerves (examination with Frenzel glasses for spontaneous nystagmus; examination of the facial nerve, trigeminal nerve, and abducent nerve), ultrasound examination of the parotid and neck, microbiological culture and sensitivity tests, CT/MRI of the base of the skull, blood sugar profile, C-reactive protein (CRP), erythrocyte sedimentation rate (ESR), hemoglobin, and full blood count.

Treatment. Treatment of diabetes and intensive anti-inflammatory therapy with broad-spectrum antibiotics (e.g., ciprofloxacin) is fundamental. Local treatment of the external acoustic meatus promotes the drainage of infective secretions. In severe cases, generous drainage of the retromandibular space, infratemporal fossa, and pterygopalatine fossa together with debridement of necrotic tissue from the external meatus may become necessary. Ligation or resection of the internal jugular vein may also be indicated if it is involved.

Prognosis. Even with massive antibiotics and extensive surgery, the prognosis is poor, due to septicemia and sinus thrombosis.

Chronic Inflammation

Tuberculosis and *syphilis* (stage 2) cause local, circumscribed lesions of the external ear and auditory meatus. The therapy is focused on local dermatological treatment and specific high-dose antibiotic treatment.

Otomycosis and Eczema

Otomycosis

Clinical Features. The infection is caused by a fungus and is limited to the external auditory meatus. There may be a recent history of antibiotic drops being used to treat bacterial infection. Typically, a fine, easily removable coating that is loose and fluffy, varying in color from whitish-yellow to greenish-black, is visible. The patient reports itching, but rarely describes pain.

Diagnosis. Culture and sensitivity tests demonstrate fungal mycelium.

Treatment. The mainstay of treatment is manual cleaning, but irrigation should be avoided if possible to prevent the formation of a "moist chamber," which promotes the growth of the fungus.

Antimycotic agents, such as 1% clotrimazole or clioquinol, can be applied locally unless contraindicated by perforation of the tympanic membrane. Antibiotics should not be used. Daily painting of the meatus with thiomersal or 2% acetic acid spray helps. In severe cases, systemic antimycotics such as fluconazole can be used.

Course. The course is chronic and recurrent.

Eczema of the Ear

Clinical Features. The disease follows an episodic course, with intermittent acute exacerbations. The entry of fluid such as sweat or water during washing, or the presence of a moist exudate, promote colonization by pathogenic bacteria or fungi in the relatively enclosed external meatus.

Fig. 1.71 Contact dermatitis caused by jewelry containing nickel.

In the acute stage, there is deep-red inflammatory swelling with moist vesicles and pustules. Later, crusts accumulate. Rhagades form around the meatal introitus, and fetid debris collects. The usual picture is one of nonspecific acute otitis externa, but occasionally the appearance may progress to chronic myringitis with superficial granulations.

In the chronic stage, the skin is atrophic, dry, scaly, and may be partly lichenified. The patient reports chronic irritation. Occasionally, stenosis may occur.

Diagnosis and differential diagnosis. *Contact eczema* may be due to cosmetic solutions, hair sprays, glasses frames made of metal or plastic, cement or flour dust, or medications—e. g., antibiotics. Skin tests have to be performed in order to identify the antigen (Fig. 1.71).

Microbial eczema is mainly due to infection with staphylococci or to oral mycosis. A swab should be taken for culture and sensitivity tests.

Seborrheic eczema is the most common form. It is often combined with acne.

Endogenous eczema is a localized manifestation of a generalized eczema.

Treatment. Elimination of the allergen and local treatment, as detailed above.

■ Trauma

Sharp or Blunt Trauma to the External Ear

Any injury to the auricle and cartilaginous part of the external auditory meatus can damage the perichondrium, causing cartilaginous necrosis. A distinction is made between auricle lacerations and avulsions either of the whole auricle or parts of it.

Treatment. For lacerations, primary closure of the wound with monofilament suture material (6–0 or 7–0) is recommended, and the duration of antibiotic treatment depends on the degree of contamination. In comprehensive lacerations with cartilaginous involvement and extensive skin damage, it is recommended—following wound debridement if necessary—to close the wound, starting on the posterior aspect of the auricle, with monofilament suture material.

The cartilage should be repaired with 5–0 absorbable suture material, and the skin of the anterior surface with monofilament 6–0 suture material. Replantation of the avulsed part of the auricle in the acute situation is also the treatment of choice for partial avulsion injuries; ischemia periods of up to 8 hours are tolerable and still allow the graft to take. If the graft does not take, plastic reconstruction is necessary in the interval.

Prognosis. Bacterial infection can cause perichondritis, with partial or complete destruction of the cartilaginous framework, leading to cauliflower ear (see Fig. 1.73) or atresia of the meatus.

Hematoma of the Auricle

This arises from closed blunt injury, with dissection of the skin and perichondrial layer from the cartilage and the formation of a subperichondrial hematoma (Fig. 1.72).

Treatment. The aim of treatment is to remove the seroma or hematoma and to achieve long-term adaptation of the perichondrium to the cartilage in order prevent leakage into the seroma cavity.

In general, drainage from the anterior surface can be carried out, with the incision being made in the fossa of the helix (scapha). Following curettage and readaptation of the skin with monofilament suture material, light compression for 3–5 days with a dental roll, secured with through-and-through sutures, is recommended. For drainage from the posterior surface of the auricle, an incision has to be made in the cartilage to reach a seroma/hematoma on the anterior surface, and a cartilage window may be necessary to achieve adherence of the perichondrium layers. Antibiotic treatment is recommended in order to prevent perichondritis.

> ! Note: Repeated aspiration may cause a seroma or superinfection, leading to perichondritis.

If a hematoma is not treated, connective-tissue organization, secondary calcification, and deformity of the auricle occur, leading to cauliflower ear (Fig. 1.73).

Frostbite
Grade 1: Cyanosis of the skin due to vascular spasm.
Grade 2: Ischemia with formation of vesicles.
Grade 3: Deep necrosis of tissue.

Treatment. Sterile dressings, antibiotics, intravenous vasodilators, and possibly stellate ganglion block are used, depending on the severity of the injury. The area has to be kept dry.

Burns require the same treatment as burns to the skin; particular attention needs be given to the close relationship between the skin and the cartilage.

Late complications include necrosis of the auricle and atresia or stenosis of the external auditory meatus.

Fig. 1.72 Auricular hematoma (othematoma).

Fig. 1.73 Untreated hematoma or infection can lead to a deformity of the auricle—cauliflower ear.

Wax and Foreign Bodies

Wax

Note: Collections of wax and cell debris are unusual if the self-cleaning mechanism of the external auditory meatus is undisturbed. The present widespread habit of cleaning the external ear daily with ready-made cotton swabs is inappropriate and can sooner or later lead to the development of chronic otitis externa.

Wax is a yellowish-brown mass consisting of secretions of the sebaceous and ceruminous glands, desquamated epithelium, hair, and particles of dirt.

Plug of wax. The normal symptom is deafness when the external auditory meatus is completely closed, but occasional complaints include a roaring noise in the head and a feeling of dizziness.

Differential diagnosis. A plug of epidermis, foreign body, dried blood, purulent exudate, and cholesteatoma of the meatus or of the middle ear.

A *plug of epidermis* is a compact white mass consisting of desquamated epithelial crusts, which usually adhere firmly to the skin of the meatus.

Treatment. The ear is irrigated with water at 37°C. Manual syringes are now rarely used, and electrically controlled water jets with controlled pressure are used instead. It should be ensured that a perforation of the tympanic membrane is not hidden behind the plug of wax. This should be excluded by taking a careful history. If the patient does have such a perforation, the wax should be removed manually by a specialist. Hard wax should be first softened with soft soap drops, 5 % sodium bicarbonate, olive oil drops, or 3 % hydrogen peroxide for up to 1 week before syringing. The moist external auditory meatus should be mopped out with cotton applicators. Local steroid or antibiotic creams or eardrops should be prescribed for patients with inflammation of the meatus. For medicolegal reasons, the patient's hearing should be tested after this procedure.

Foreign Bodies

Foreign bodies are diagnosed using careful otoscopy. The findings vary depending on the object and the length of time it has been in the ear. In children, a careful history should be taken to establish the nature of the foreign body. Depending on their age, children may be able to indicate that they have a foreign body in the ear, or they may present with symptoms of ear pain or discharge. Insects can cause injury to the canal or tympanic membrane as a result of scratching or stinging.

Physical pain or bleeding may occur with objects that abrade the ear canal or rupture the tympanic membrane, or as a result of the patient's attempts to

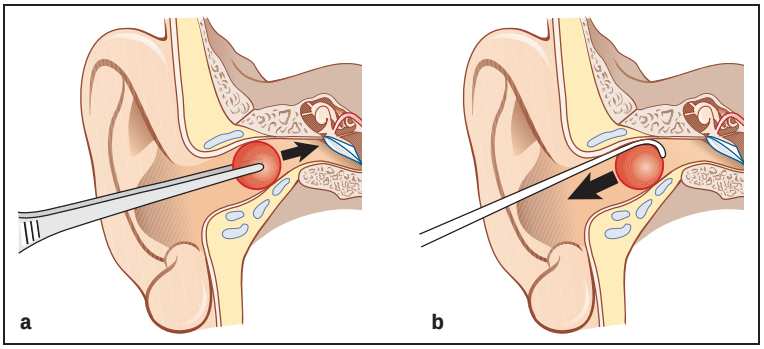


Fig. 1.74a, b Attempts to remove a foreign body with simple forceps (a) may displace the foreign body more deeply, and it may perforate the tympanic membrane, causing dislocation of the ossicles and injury to the facial canal. The foreign body can be removed easily without danger to the patient using a hook, under otoscopic or microscopic guidance (b).

remove the object. Hearing loss may be observed. If the presentation is delayed, erythema and swelling of the canal and a foul-smelling discharge may be present.

Note: Foreign bodies that cannot be removed by syringing should be removed manually, with general anesthesia being administered in small children.
If a perforation of the tympanic membrane is known of or suspected, the ear should not be syringed.
Blind attempts to extract foreign bodies without otoscopic control, or attempts to extract them under vision with unsuitable instruments and inadequate anesthesia, should be avoided. Instrumental removal of foreign bodies from the meatus should therefore only be carried out by a specialist, except for the simplest cases.
Figure 1.74a,b shows correct and incorrect methods of extracting a foreign body from the meatus.

Tumors

Benign Tumors

These include retroauricular atheroma, cicatricial keloid, hemangioma and lymphangioma, dermoid tumors, fibroma, papilloma, keratoma, lipoma, and nevi.

Treatment. The treatment consists of tumor resection.

Meatal tumors include *hyperostosis,* caused by periosteal stimulation and resulting in appositional bone growth with progressive narrowing of the lumen of the meatus; and *exostosis,* a true bony tumor arising from the ossification centers in the anulus tympanicus. This tumor should only be removed if it is causing stenosis. *Chondrodermatitis nodularis circumscripta helicis* causes painful nodes on the ear that should be distinguished from a premalignant lesion by the marked pain that occurs when pressure is applied.

Precancerous and Malignant Tumors

The classification, symptoms, diagnosis, and differential diagnosis are shown in Table 1.9.

Precancerous Lesions

Premalignant lesions often progress to true carcinoma, and they should therefore always be treated with radical surgery, in the same way as true tumors. Biopsy excision is not advisable, and any tumor of the external ear suspected of being malignant should be excised with a wide margin. The mainstay of treatment is surgery; radiotherapy or cryosurgery are second-best options.

Precancerous lesions such as senile keratosis (Fig. 1.75a) or Bowen disease are excised with a healthy margin. Periodic follow-up is necessary.

Malignant Tumors

Basal cell carcinoma. Surgical excision of basal cell carcinoma in an otherwise healthy patient is decisive for the prognosis (Fig. 1.75b). A two-step surgical procedure is the method of choice. The first step involves temporary wound coverage of the defect after resection of the tumor. After histological confirmation that the wound margins are tumor-free, the defect can be covered in a second operation, which may require reconstruction of the auricle. For very small tumors, a one-step procedure is possible, with primary wound closure after intraoperative histological control of the wound margins using frozen-section histology.

Table 1.9 Precancerous and malignant tumors of the external ear

	Form	Color	Skin surface	Cartilage invasion	Regional lymph-node metastases
Cutaneous horn	A sharply limited warty growth of the epidermis	Inconspicuous	Slightly nodular, intact	None	None
Senile keratosis	Smooth mass with indistinct borders	Yellowish-brown	Rough, intact, occasionally covered by crusts	None	None
Bowen disease	Smooth round lump	Intensive brownish-red	Smooth but intact	None	None
Basal cell carcinoma	Sharply demarcated smooth, slowly growing mass	Hyperemic, sometimes much more pigmented than the surrounding skin (special form: pigmented basal cell carcinoma)	Often superficially ulcerated, crusted, raised edges with an atrophic center (or central ulcer)	Perichondrium sometimes infiltrated, tumor relatively immobile at its base	Rarely
Squamous cell carcinoma	Exophytic tumor of relatively rapid growth with indistinct margins	Often hyperemic	Ulcerated, raised edges, superficially nodular, firm	Always The tumor is not movable; occasionally perichondritis	20 %
Malignant melanoma	A round mass, sometimes verrucous, rapidly growing	Dark-brown to black, occasionally weakly pigmented, the amelanotic melanoma	Smooth to slightly nodular, occasionally ulcerated or bleeds easily to touch	Perichondrium often infiltrated and the tumor is relatively immobile at its base	Frequent Also early distant metastases, especially to the lung

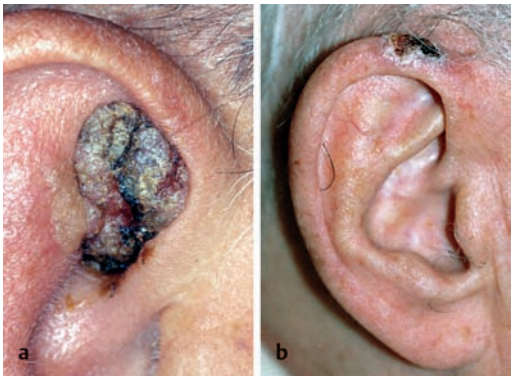


Fig. 1.75a, b a Senile keratosis of the auricle.
b Basal cell carcinoma.

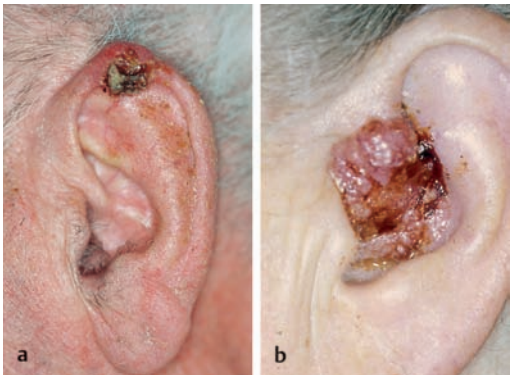


Fig. 1.76a, b Squamous cell carcinoma.
a A small erosive carcinoma invading the helix.
b Carcinoma growing out of the external meatus.

Keratinizing squamous cell carcinomas. These are infiltrating, often ulcerated carcinomas (Fig. 1.76a, b). In 20 % of patients, the lesions have early metastases to the regional lymph nodes. The treatment of choice is *radical surgery,* with no regard for the cosmetic result. Radiotherapy is only successful for tumors less than 1 cm in diameter that are not ulcerated, have not infiltrated the perichondrium, and have not metastasized to the regional lymph nodes. In all other cases, *partial or complete excision of the auricle* is the method of choice. Neck dissection is performed when there are regional lymph-node metastases.

Carcinoma of the external auditory meatus. This represents ≈ 5 % of all aural carcinomas. In comparison with tumors of the auricle, the prognosis is unfavorable due to late diagnosis and because the tumor penetrates early into the parotid space or the middle ear. The treatment of choice is extensive resection of the tumor with radical neck dissection and, if necessary, parotidectomy.

Pigmented nevus and suspected malignant melanoma. These should be treated with primary excision of the tumor, without previous biopsy. Depending on the histological results, it may be necessary to carry out a secondary surgical excision, dictated by the extent and depth of the tumor (see p. 405), with regional neck dissection or radical neck dissection and, if necessary, parotidectomy. Postoperative radiotherapy, chemotherapy, or immunotherapy may also be necessary. Surgery should only be considered when distant metastases have been ruled out.

Clinical Aspects of Diseases of the Middle and Internal Ear

Disorders of Ventilation and Drainage of the Middle Ear Spaces

Pathophysiology. Most chronic middle ear diseases are based largely on two functional disturbances: *impaired middle ear ventilation* and *inflammation*. Impairment of ventilation due to eustachian tube dysfunction is based on a mucosal inflammation in the nasopharynx, leading to inflammation of the middle ear mucosa.

Pathogenesis. The eustachian tube does not open regularly on swallowing, due to one of the following factors:

- A dysfunctional tensor veli palatini muscle.
- Swelling of the tubal mucosa caused by chronic inflammation of neighboring structures, such as the sinuses or tonsils or by allergy.
- Obstruction of the ostium of the tube by hypertrophic adenoids, in a child or adult.
- Infiltration of the tube by a malignant tumor of the nasopharynx.

The middle ear is thus no longer aerated, and the remaining air is resorbed, producing a decrease in pressure that acts as an irritant to the middle ear mucosa.

In *short-term tubal occlusion* or persistent, reduced pressure in the middle ear, the following changes may occur:

- Edema of the mucosa.
- Middle ear fluid, due to transudation of the constituent parts of the serum.
- Stiffening of the ossicular chain, with retraction of the tympanic membrane.

> Note: Ventilation and drainage disorders of the middle ear are usually caused by a dysfunctional opening mechanism of the tube, but mechanical obstruction of the tube by a lesion in the nasopharynx may also occur.

In *long-standing tubal occlusion* and reduced pressure in the middle ear, the following changes may occur:

- Metaplasia of the middle ear mucosa from the flat epithelial cells of the mucoperiosteum to form columnar, ciliated, mucus-producing goblet cells.
- Increase in secretory activity of the goblet cells and mixing of the mucus with the transudate already present in the middle ear, causing a *seromucinous exudate.*
- Formation of cholesterin-containing mucosal cysts (cholesterol granuloma).

The seromucinous exudate and mucosal changes substantially reduce the aeration of the middle ear, thus causing a vicious circle.

The metaplasia of the middle ear mucosa also affects the submucosa, causing firstly a proliferation of the connective tissue and secondly maturation of a local immunologically active cellular defense mechanism. The active secreting goblet cells produce a mucus blanket, which serves to transport newly formed immunoglobulins to the mucosal surface. The previously inactive mucoperiosteum of the middle ear is thus converted into a secreting hyperplastic respiratory mucosa, characterized by its newly acquired property of responding to every new stimulus (mechanical, chemical, bacterial, enzymatic, allergic, or autoimmune) with a completely mature defense mechanism. This consists of the following:

- Mucociliary elements, which form the transport medium for the superficially active mucus blanket.
- Enzymatic elements, which consist in particular of bactericidal lysozymes and protease inhibitors.
- Locally produced immunoglobulins.

The result of this increased activity in the local and mucosal immune system is that each bacterial stimulus causes hyperplasia or metaplasia of the superficial epithelium and induces mucosal edema with a cellular infiltrate. The resulting increase in the volume of the middle ear mucosa thus starts a vicious circle of deteriorating ventilation and drainage.

This hyperactivity of the middle ear mucosa continues after cessation of the external stimulus and ultimately leads to *tympanosclerosis.* Enzymes and the pathologic concentration of metabolic products and mediators of inflammation (lipoids, mucopolysaccharides) cause progressive metaplasia of the middle ear mucosa, resulting in fibrosis and sclerosis of the middle ear and formation of cholesterol granulomas. These changes lead to an irreversible tympanosclerotic condition.

Basically, two main forms of disorders of ventilation and drainage can be distinguished: the first occurs acutely and is reversible, while the second follows a chronic course that is only partially reversible.

Acute Tubal Occlusion (Serotympanum)

Clinical Features. A feeling of pressure in the ear occurs during a head cold (rhinopharyngitis), often accompanied by stabbing pain, deafness, and a crackling noise when swallowing.

Diagnosis. Otoscopy shows a retracted tympanic membrane (Fig. 1.77a, b) and hyperemia of the handle of the malleus and of the vessels of the tympanic membrane. If a transudate occurs, there is an amber discoloration of the pars tensa and occasionally a fluid level and air bubbles in the middle ear. The patient also has mild to moderate conductive hearing loss.

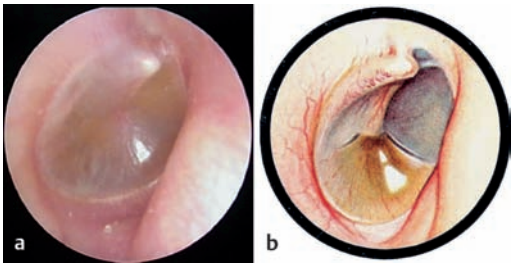


Fig. 1.77a, b Serous otitis media. The tympanic membrane is slightly retracted and transudate is visible in the tympanic cavity. The malleus handle is perceptibly shortened, the light reflex is widened.

Differential diagnosis. Acute otitis media has to be ruled out.

Treatment. This is initially directed at the underlying disease. Rhinopharyngitis is treated with decongestant nose drops, vasoconstrictors, and antihistamines to reduce hyperemia and edema in the tube. Antibiotics are given if there are dangerous signs of progression to otitis media or of acute nasal pharyngitis. Hypertrophic adenoids are removed later if necessary, and sinusitis is treated.

> Note: Oral analgesics, rather than analgesic ear drops, should be used to treat pain in the ear. Ear drops obliterate the otoscopic appearance of the tympanic membrane by macerating the superficial epithelium and can make it difficult to recognize otitis media.

Course and prognosis. The symptoms usually resolve rapidly, but occasionally the disease progresses to chronic seromucinous otitis media.

> Note: The Valsalva maneuver or air insufflation should not be performed when there is acute inflammation in the nasopharynx, due to the danger of transmitting infectious microorganisms to the middle ear and causing tubal otitis media.

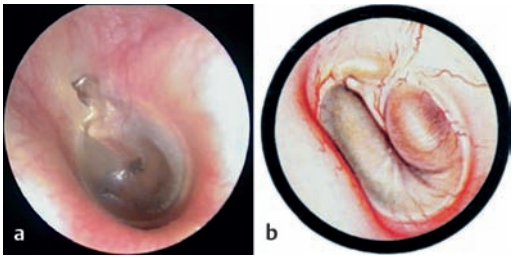


Fig. 1.78a, b Chronic seromucinous otitis media. The pars tensa of the tympanic membrane is protracted, while the pars flaccida is retracted. A bulb filled with yellow-brown liquid expands the tympanic membrane.

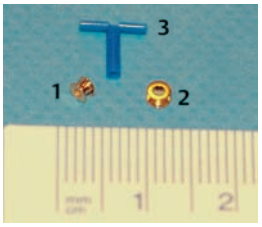


Fig. 1.79 Different types of ear ventilation tube. 1, 2, Silver ear tubes coated with gold, sizes I and II; 3, Silastic tube for long-term placement.

Fig. 1.80 Middle ear drainage is achieved by inserting a metal tube ("grommet").

Chronic Seromucinous Otitis Media

Clinical Features. There is a feeling of pressure and fullness in the ear, often following an infection of the upper airway, and there is a considerable decrease in hearing on one or both sides. Noises are heard in the ear when yawning, swallowing, and sneezing. Pain is absent.

Pathogenesis. Tubal obstruction and the resulting reduction in tympanic pressure predominate. Obstructive processes in the nasopharynx and disorders of tubal kinetics, particularly incompetence of the muscles opening the tube in cleft palate, and viral infections are the most common underlying mechanisms.

Diagnosis. Otoscopy shows a markedly retracted tympanic membrane with localized protrusion, an exudate in the middle ear, and dark discoloration behind the tympanic membrane (the so-called "blue drum"), with a blackish fluid level or air bubbles. There is conductive deafness for the entire frequency range up to 50 dB. A typical flat curve is found. A search for the primary cause shows enlargement of the adenoids, sinusitis, allergy, or tumor (Fig. 1.78a, b).

Differential diagnosis. This includes *hemotympanum* (the dark-brown exudate behind the tympanic membrane on occasion lends this a bluish tinge) and *chronic otitis media,* evidenced by perforation, cholesteatoma flakes, and purulent exudate. Adhesions may occur after recurrent otitis, shown by a markedly retracted tympanic membrane with thick scars, tympanosclerotic deposits, and abnormal tubal function. In adult patients with unilateral symptoms, biopsy from the nasopharynx should be considered to exclude tumor; some tumors are submucosal and not clearly apparent.

Treatment. If necessary, surgical restoration of tubal patency by adenoidectomy or revision adenoidectomy under direct vision, and elimination of any sinus infections.

Selective insertion of a ventilation tube after paracentesis and drainage of the middle ear (see Fig. 1.84), when clinically indicated by symptoms, is the treatment of choice.

An incision is made in the anteroinferior quadrant of the tympanic membrane, under general anesthesia in children and under local anesthesia in some compliant adults. The middle ear effusion is aspirated, and long-term drainage is provided for at least 6 months using a ventilation tube (or "grommet"; Figs. 1.79, 1.80). Alternatively, watchful waiting can make it possible to avoid surgery, as the disease resolves in many cases. Adjuvant treatment consists of steroids with antibiotic cover and mucolytic drugs. Other alternatives—such as local aerosol treatment (intranasal or tubotympanic application), α-chymotrypsin (tubotympanic, transtympanic, or systemic administration), hyaluronidase, and corticosteroids (intratympanic application)—are not generally used, but have been investigated in research studies. Antiallergic treatment includes antihistamines and intranasal steroids if there is positive evidence of allergy. Acute otitis media is often present even if the immunological findings are normal. Mastoiditis has to be ruled out, otherwise mastoidectomy should be considered.

Course and prognosis. Long-term healing is only achieved in a proportion of patients. In others, there is a progressive, chronic course leading to adhesive processes as a result of connective-tissue organization of the seromucinous exudate and development of cholesterol granuloma and tympanosclerosis. Hyaline degeneration of the mucoperiosteum may also occur, with the formation of sclerotic submucosal plaques as a result of local metabolic disturbances (Fig. 1.81a, b).

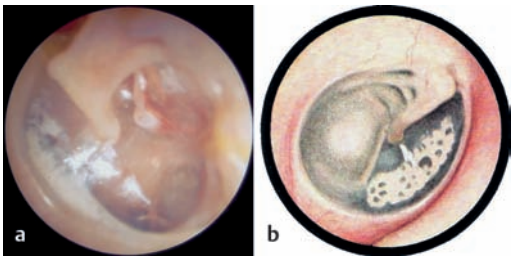


Fig. 1.81a, b Tympanosclerosis.
a Hyaline degeneration of the mucoperiosteum occurs with the formation of sclerotic submucosal plaques.
b The posterior half of the tympanic membrane shows atrophic scarring, and plaque has formed on the anterior half.

Syndrome of the Patulous Eustachian Tube

Clinical Features. These include *autophony,* which is a rumbling reverberation of the patient's own voice, and a noise in the ears synchronous with breathing, due to movements of the tympanic membrane and resonance in the nasopharynx.

Pathogenesis. The symptoms represent a masking effect of the lower and middle tones, evoked by resonance and respiratory noise. The primary cause is insufficiency of the closing mechanism of the tube (see p. 3). The secondary cause is disappearance of the fat bolster around the opening of the tube, a gaping ostium caused by hormonal disturbances, and possibly also by use of the contraceptive pill.

Diagnosis. The diagnosis is made by impedance audiometry and tubal function tests.

Treatment. If possible, the basic cause is dealt with. The patient also needs to have the cause of the symptoms explained.

■ Nonspecific Inflammation of the Middle Ear and Mastoid

> Note: Inflammatory diseases of the middle ear are important because of their frequency and the life-threatening complications associated with them due to the close relationship between the middle ear and the cranial cavity.

Acute Otitis Media

Clinical Features. In the first phase of *exudative inflammation,* lasting for 1–2 days, there is an increase in temperature to 39–40°C, and in severe cases, rigors, and occasionally meningism in children. The patient has a severe pulsating pain, which is worse at night than during the day. There is a muffled noise in the ear synchronous with the pulse, deafness, and sensitivity of the mastoid process to pressure. In older patients, there is often no fever.

The second phase, involving *resistance and demarcation,* lasts 3–8 days. The pus and middle ear exudate usually discharge spontaneously, after which the pain and fever subside. This phase can be considerably shortened by administering topical therapy (meatal cleansing, application of an astringent solution). Early antibiotic administration does not alter the clinical course of the disease significantly, and it does not prevent spontaneous perforation of the tympanic membrane.

In the third, *healing* phase, lasting 2–4 weeks, the aural discharge dries up and hearing returns to normal.

> Note: Acute middle ear inflammation may have a serious course, even if the tympanic membrane does not perforate.

Pathogenesis. *Routes of infection:* The tubal route is the most common. Hematogenous infection is unusual and occurs in measles, scarlet fever, typhus, and septicemia. Exogenous infection requires rupture of the tympanic membrane or previous perforation, allowing bathwater or dirt to penetrate during irrigation of the ear. Incorrect methods of re-

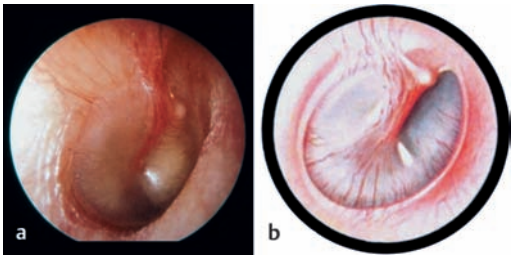


Fig. 1.82a, b Acute otitis media.
a The tympanic membrane is erythematous and bulging, and the handle of the malleus shows hyperemia.
b The tympanic membrane also has radiating hyperemia.

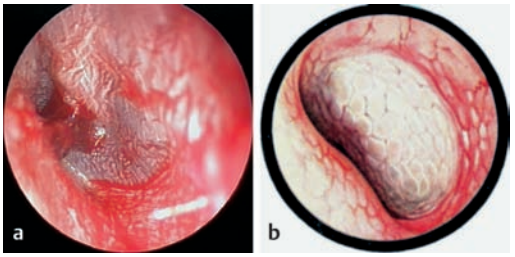


Fig. 1.83a, b Acute otitis media (desquamative phase). The tympanic membrane is severely bulging and a livid red, partially blue-gray appearance with scales, as a result of severe desquamation.

moving a foreign body from the external meatus are also another cause.

> Note: In healthy individuals, the middle ear is sterile if the tympanic membrane is intact.

Type of organism: the infection is monomicrobial. In decreasing order of frequency, the infecting organisms are: streptococci in adults, pneumococci in children, *Haemophilus influenzae, Moraxella catarrhalis,* and various staphylococci. A viral infection may prepare the way for secondary bacterial infection (herpes simplex and zoster, flu viruses). The inflammation usually affects not only the mucosa of the middle ear, but also that of the entire respiratory system.

> Note: Every attack of acute otitis media is accompanied by mastoiditis.

Diagnosis. In the first phase, otoscopy shows hyperemia, then moist infiltration and opacity of the surface of the tympanic membrane. The contours of the handle of the malleus and its short process disappear (Fig. 1.82a, b). The patient has conductive deafness. At the height of the exudative phase, the tympanic membrane bulges, particularly its posterosuperior quadrant. Pulsation is also seen. The inflammation may extend to the external meatus, obliterating the boundary between the meatus and the tympanic membrane (Fig. 1.83a, b). The accompanying mastoiditis makes the mastoid process tender to pressure. In influenzal otitis, hemorrhagic bullae form on the external auditory meatus and the tympanic membrane.

In the second phase of acute otitis media, immediately before spontaneous rupture, a pinhole-sized fistula forms, usually in the posterosuperior quadrant. This discharges a pulsating, thin, fluid, odorless pus. Radiography with the Schüller view or preferably CT scanning shows clouding of the cell system without osteolysis—i. e., the bony septa appear sharp. Imaging is indicated only if there are severe clinical symptoms (facial palsy, dizziness, vertigo, severe sensorineural hearing loss).

In the third phase of acute otitis media, the inflammation and thickening of the tympanic membrane resolve, the pulsations disappear, and the discharge becomes mucoid and finally ceases. The perforation closes spontaneously, leaving a fine scar. Hearing returns to normal. CT scans show gradual clearing of the cell system.

Differential diagnosis. *Otitis externa* needs to be considered. In otitis externa, there is pain when pressure is applied to the tragus, and the exudate does not pulsate, is usually fetid, and is never mucoid. There is little or no deafness, and the cell system appears normal on radiographs.

Treatment

1. Systemic antibiotics at high dosages are given if condition worsens after 48 h. Hospital admission and intravenous application of amoxicillin and other broad-spectrum penicillins are indicated. Culture and sensitivity tests are carried

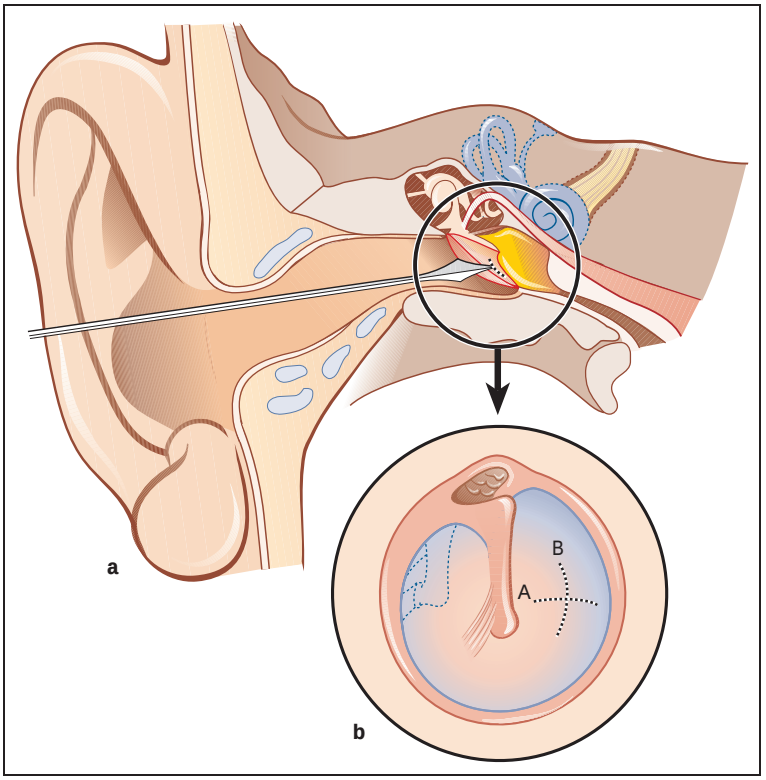


Fig. 1.84a, b The principle of paracentesis.
a The position of the myringotomy knife relative to the external meatus and tympanic membrane.
b Incision into the anterior lower quadrant of the pars tensa. A, Correct incision; B, alternative incision.

out and appropriate antibiotics are administered if the tympanic membrane perforates.
2. Nasal drops are given to decongest the nasopharyngeal mucosa around the opening of the tube.
3. Analgesics (liquid paracetamol is recommended for children) and mucolytic drugs can be given.

Paracentesis (Fig. 1.84a, b), performed using a myringotomy with placement of a tympanostomy tube, is indicated in the following circumstances:
- Marked bulging of the tympanic membrane.
- Persistent high fever and severe pain.
- Unsatisfactory spontaneous perforation, with incomplete differentiation of the tympanic membrane.

If *symptoms of early mastoiditis* occur, with discrete facial palsy, acute meningitis, or labyrinthitis, and if the appearance of the tympanic membrane is inconclusive, surgery is indicated.

Note: Drops containing cortisone and antibiotic solution should not be used locally for aural discharge. They are ineffective, and also carry a risk of resistance to antibiotics developing. However, regular meatal cleansing should be performed for acute otitis media after the tympanic membrane perforates. The meatus can be irrigated with water at body temperature. The external meatus is then dried. Closing the meatus with cotton wool or gauze strips provides an ideal moist environment for inoculation with Gram-negative bacteria or fungi, and the meatus therefore has to be kept open.

Course and prognosis. In the first, acute phase, there is a danger of early otogenic complications, depending on the virulence and resistance of the organism, until the patient's own resistance develops and the bacterial infection is controlled with antibiotics.

During the second phase, complications occur very rarely. However, latent otitis media and the resulting occult mastoiditis may develop during

this period due to insufficient antibiotic dosage, increased resistance of the organism, or inadequate resistance on the part of the patient. The general findings at otoscopy then do not correlate with the severity of the pathologic changes taking place in the middle ear and mastoid process. The paucity of symptoms leads to a false assumption that the otitis media has healed rapidly and completely. After an apparently symptom-free interval, late otogenic complications can occur in the third phase (see pp. 65 and 76). This course resembles that of the previously described, dreaded pneumococcal-type III mastoiditis.

In the third phase, most cases of acute otitis media and its concurrent mastoiditis heal completely. However, if latent otitis media and mastoiditis become established in the second phase, late otogenic complications may develop in the third period—i.e., 2–3 weeks after the start of the otitis. The symptoms include:

- Reappearance of fever.
- Recurrence of aural pain and discharge.
- Headaches.
- Worsening of the patient's general condition.
- Elevated erythrocyte sedimentation rate (ESR).

Specific Types of Inflammation of the Middle Ear and Mastoid

Acute Otitis Media in Infants and Children

Severe forms are unusual nowadays, thanks to effective diagnosis and treatment with antibiotics. However, they can occur in patients with immune deficiency or after inappropriate therapy. They are characterized by:

- Severe general symptoms, with high fever, meningeal and cerebral irritation, vomiting, loss of appetite, and disturbance of sleep.
- Immediate improvement of the child's condition after spontaneous perforation or paracentesis.
- A protracted course with numerous recurrences or exacerbations, with combined otitis media and bronchopneumonia, digestive and feeding upsets, and pyelonephritis.

> Note: The younger the child, the more severe the generalized symptoms are and the more discrete the local signs are. The gastrointestinal symptoms are sometimes the most pressing.

Infants and young children have a predisposition to tubal middle ear infection because the tube is short, straight, and wide; because of the uniform character of the mucosa of the middle ear and the upper respiratory tract; and due to a higher frequency of infections of the respiratory tract, hyperplasia of the lymphoid tissue of the Waldeyer ring, poor aeration of the middle ear cavity, which is still partially filled with myxomatous tissue or hyperplastic mucosa, and a difference in the way in which the general and mucosal immune system react, caused in part by the genotype and in part by the phenotype.

Local symptoms. These include pressure in the ear, tugging on the affected ear, and painful reactions to pressure and traction. The tympanic membrane is grayish-red in color and bulges only slightly. Spontaneous perforation is not common; its site of predilection is the anteroinferior quadrant. Discharge is uncommon, as pus drains through the short, wide, and straight eustachian tube. If discharge occurs, it is stringy and pulsating. Mucosal polyps may form in the middle ear; regional retroauricular lymphadenitis causes a swelling behind the ear. If the petrosquamosal fissure is open, the pus may penetrate directly from the middle ear beneath the periosteum, causing marked swelling behind the ear.

Treatment. Treatment consists of oral antibiotics. If the infection is severe, intravenous antibiotics are given. Decongestant nose drops and analgesics can also be administered. The ear is irrigated with physiologic saline solution at body temperature.

Paracentesis (see Fig. 1.84a, b) should be carried out with the patient under general anesthesia early if the tympanic membrane does not perforate spontaneously.

If indicated on clinical grounds, *cortical mastoidectomy* should be performed early, even if radiographs are normal. *Cortical mastoidectomy* is carried out under general anesthesia in infants and young children in whom the mastoid process is incompletely pneumatized or not pneumatized at all. The infected part of the mastoid process is cleared via a retroauricular access route, with wide opening and drainage of the mastoid antrum (Fig. 1.85).

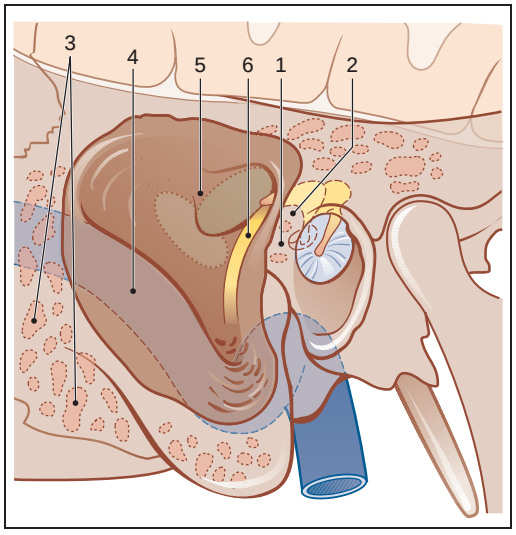


Fig. 1.85 The principle of cortical mastoidectomy. The posterior meatal wall (1) and attic wall (2) remain intact, while the cell system of the mastoid process (3) is cleared via a retroauricular approach. The anatomy of the external auditory meatus is not altered by this operation. 4, Sigmoid sinus; 5, mastoid cavity; 6, facial nerve (mastoid segment).

Course. The course is usually protracted, with intermittent exacerbations. There is often quick improvement and healing after surgical treatment of the affected ear. The prognosis is good with the correct treatment, but there is otherwise a danger of periantral osteomyelitis developing (infantile occult antritis), with vomiting and generalized toxic symptoms. In this case, immediate antrotomy is indicated.

Specific forms. *Influenzal otitis*: This is a hemorrhagic, bullous, acute middle ear inflammation. Primary infection with influenza A virus, combined with secondary bacterial infection *(Streptococcus pneumoniae)*, can take a fulminating course with complications (facial paralysis, labyrinthine irritation, meningitis).

Measles otitis: Hematogenous viral otitis media with subsequent tubal secondary infection, often leading to purulent mastoiditis.

Scarlatinal otitis: Acute necrotizing inflammation, with subtotal perforation of the tympanic membrane, necrosis of the ossicular chain, and osteomyelitis of the temporal bone.

Mastoiditis

The most frequent complication of middle ear inflammation is *mastoiditis*, an extension of the infection from the middle ear cavity to the pneumatic system of the temporal bone. In contrast to the mucosal inflammation, which always accompanies otitis media (see p. 61), the infection extends to, and causes dissolution of, bone. An unusually well-pneumatized bone infection can extend to the petrous pyramid (petrositis) and more rarely to the diploë of the temporal bone, causing osteomyelitis.

Clinical Features. Mastoiditis becomes manifest when there is a change in progress during resolution of acute otitis media.

General: Worsening of the general condition, rise in temperature, leukocytosis, and markedly increased ESR.

Local: Increasing pain in the ear, synchronous with the pulse and radiating to the temporal bone and the occiput; reappearance or increase in the aural discharge, which is creamy, odorless, and purulent. The patient also has hearing loss.

Pathogenesis. Acute otitis media with concomitant mastoiditis usually resolves without complications. The development of complications depends on:

- The anatomic relationships between the respiratory system and middle ear space. Because of the narrow connection between the antrum and the mastoid cells, there is poor aeration from the eustachian tube.
- The virulence and resistance of the organism.
- The local immune resistance of the mucosa.
- The patient's general immune resistance.
- The patient's general condition. Generalized diseases such as diabetes, immunodeficiency, allergy, and disorders of the liver and kidneys are important.

Diagnosis

- Aural discharge.
- Tenderness to pressure over the mastoid.
- Retroauricular swelling, with a protruding ear.

This classic triad of symptoms is now seldom seen, as otitis media is treated with antibiotics. This is especially true of the critical period that used to occur in the third week, during the preantibiotic era. The symptoms of mastoiditis are now more discrete and its course more insidious than they

Fig. 1.86 Subperiosteal abscess, due to acute otitis media with mastoiditis.

used to be, so that this complication is easily overlooked. For this reason, the following otoscopic findings, which are also present during antibiotic treatment, need to be treated with caution:

- A pale but still thickened tympanic membrane.
- Circumscribed inflammation and thickening of the tympanic membrane in the posterosuperior quadrant.
- Thickened opaque tympanic membrane.
- Formation of a nipple on the tympanic membrane, with a fine pinpoint fistulous opening.
- Prolapse of the posterior meatal wall, which occurs relatively often in small children (see p. 2).

Local findings over the mastoid process and in the surrounding area:

- *Subperiosteal abscess*: Soggy swelling of the skin is caused by edema due to spread of the infection. Reddening and a taut, elastic, fluctuating swelling of the skin over the mastoid process (Fig. 1.86).
- *Zygomatic bone inflammation*: There is swelling of the zygomatic process, with extension to the cheek and eyelids in an extensively pneumatized bone. This is relatively common in children.
- *Bezold mastoiditis*: Visible and palpable tender swelling of the lateral triangle of the neck, with torticollis. This is due to an abscess tracking from the mastoid apex in the fascial spaces of the digastric, sternocleidomastoid, splenius, and longissimus capitis muscles.
- *Radiographic signs* in the Schüller view and/or CT of the temporal bone show a decrease in radiolucency due to a reduction in the air content in the respiratory system, haziness and opacity of the mastoid cells, haziness of the fine bony structures as a result of decalcification and liquefaction of the bony septa between the cells, bone destruction with foci of liquefaction, and erosion of neighboring structures.

Differential diagnosis (pseudomastoiditis) (see p. 51). Furuncle of the external ear canal, parotitis, or cervical lymphadenitis.

Treatment

Note: Mastoiditis in which the inflammation is no longer confined to the mucosa but has extended to the bone should be treated surgically.

It is incorrect to assume that inflammation of the pneumatic system of the temporal bone can be cured by antibiotics once it has invaded the bony structures. The poor vascularization of the mucosa and bone makes it impossible to maintain a satisfactory concentration of antibiotic in the tissues. The poorly aerated cells, filled with hyperplastic mucosa and granulations, are an ideal culture medium for bacteria, particularly anaerobes.

Indications for mastoidectomy: The operation is indicated if bone infection is thought to be present at any stage of otitis media:

- Symptoms of otogenic intracranial complications.
- Signs of a subperiosteal collection.
- A focus of liquefaction of mastoid cells on CT scans of the temporal bone.
- Facial paralysis.

In the healing phase—i. e., in the critical third week of otitis media—the operation is also indicated when there is:

- Recurrence of aural discharge, pain, and subfebrile temperature.
- Worsening of the general condition and an increase in CRP.
- Aural discharge persisting for 4–5 weeks and resisting treatment, in the presence of good pneumatization of the temporal bone, with reduced air content on radiographs, and without serious generalized symptoms.

Principle of mastoidectomy: The diseased tissue in the cell system of the mastoid process is excised via a retroauricular skin incision under general anesthesia. A wide connection is created between the mastoid antrum and the mastoid cavity, and from the latter to the middle ear cavity (see Fig. 1.85).

Course and prognosis. Two forms of true mastoiditis with bone destruction can develop from the mastoiditis accompanying otitis media, even during antibiotic treatment:

- *Acute mastoiditis:* This is marked by purulent liquefaction of the bony septa of the pneumatic system, with external rupture to form a subperiosteal or retroauricular abscess. The course is rapid and the symptoms marked. Paralysis of the facial nerve is possible.
- *Chronic mastoiditis:* This is partly a productive inflammation, with obliteration of the spaces of the pneumatic system by inflammatory granulation tissue; and partly a continuous inflammatory breakdown of bone. The course is therefore insidious, and the symptoms are initially mild.

The prognosis is good with correct treatment, but otherwise there is a danger of late otogenic complications (see p. 76).

Chronic Otitis Media

Chronic Mucosal Inflammation (Chronic Mesotympanic Otitis)

Clinical Features. *There is a chronic discharge of mucoid, purulent, odorless exudate.* The otitis is associated with periods of complete freedom from symptoms, alternating with acute exacerbations. The exudate may be creamy and purulent in the acute phase and then becomes mucoid and stringy as the infection resolves. However, it is always odorless.

Hearing: The patient has conductive hearing loss. Pain is absent, and the general condition is good.

Pathogenesis. The disease is not one that has a single cause, but rather it is the end result of several different primary disease processes. The inflammation remains confined to the mucosa, but in certain patients it can lead in time to rarefying osteitis—i. e., chronic inflammatory destruction of the ossicles, such as the long process of the incus. In contrast to cholesteatoma, this destructive bone process is unusual and less likely to extend and progress. Vascular obliteration can occur due to heavy scar tissue deposits in the vascular subepithelial connective-tissue layer, leading secondarily to nutritional disturbances of the neighboring bony tissue (aseptic bone necrosis).

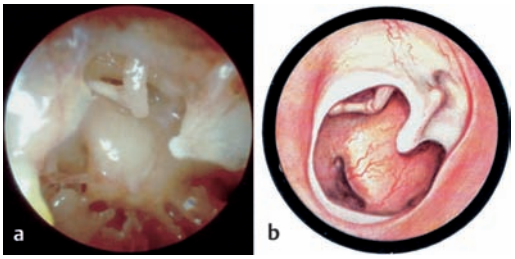


Fig. 1.87a, b Chronic otitis media (mesotympanic). There is a large central perforation in the tympanic membrane, with only a thin edge left. The promontory is visible through the perforation. The long process of the incus, the stapes tendon, and the round window niche are visible.

Pathogenetic factors:

- Constitutionally reduced mucosal (immunological) competence (see p. 7).
- Type, pathogenicity, virulence, and resistance of the bacterial organisms.
- Anatomic conditions in the middle ear, such as pneumatization, and connections between the attic, antrum, middle ear cavity, and eustachian tube.
- Disordered function of the eustachian tube—e. g., in patients with cleft palate.
- Generalized diseases such as allergy, immune defects, cachexia, and diabetes.

Diagnosis. The *history* shows a chronic recurrent aural discharge with reduced hearing. The *otoscopic findings* include a central defect of the tympanic membrane (Fig. 1.87a, b), scarring of the pars tensa, and occasionally aural polyps due to mucosal hyperplasia in acute exacerbations.

CT scanning (as well plain Schüller radiographs) show either reduced pneumatization or opacity of the cell system, if it is well pneumatized; occasionally, signs of bony destruction and formation of new bone (sclerosis) are seen. These are regarded as signs of chronic mastoiditis.

The *audiogram* shows conductive hearing loss.

Differential diagnosis. *Cholesteatoma* is associated with a marginal defect in the tympanic membrane and a fetid discharge.

Aural tuberculosis shows several central perforations of the tympanic membrane, with marked hearing loss.

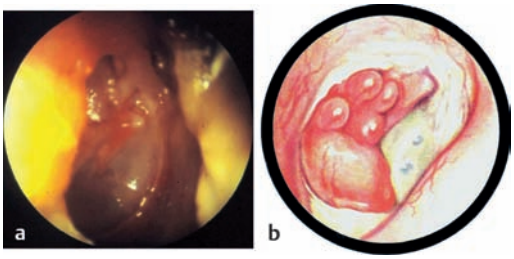


Fig. 1.88a, b Chronic otitis media (epitympanic). The marginal defect in the tympanic membrane in the posterosuperior pars tensa and in the pars flaccida is causing osteitis in the bony lateral wall of the epitympanum. Granulations or polyps are typical signs.

Middle ear carcinoma causes a marginal defect with exuberant tissue extending into the meatus, and bone destruction in the attic and the meatal wall.

Treatment. *Conservative measures for drying the middle ear:* The external meatus is cleaned periodically. It can be irrigated with physiologic saline at body temperature. In the acute phase, pus is taken for culture and sensitivity tests, and appropriate systemic and local antibiotics are given, with care being taken not to use ototoxic drugs.

Aural polyps are removed with a wire snare or microforceps. Chronic infection of the nasopharynx and paranasal sinuses must be looked for.

Surgery: Mastoidectomy can be carried out to eliminate foci of infection in the temporal bone and middle ear cavity. Tympanoplasty can be performed to reconstruct the sound-conducting apparatus—i. e., the tympanic membrane and ossicular chain.

Course and prognosis. The course is episodic, with exacerbations caused by exogenous infection—e. g., from bathwater—and tubal infection. Complications are very rare. Progression to cholesteatoma is exceptional, and hearing loss is usually progressive. The prognosis is good in relation to survival, but poor with regard to function. An early tympanoplasty should therefore be carried out after intensive preparation (see Figs. 1.95, 1.96).

Age in itself is not a contraindication to surgical treatment. The key objective of the operation is to close the perforation, to avoid recurrent infection of the middle ear mucosa via the external ear canal. Improvement of hearing can only be achieved in a small proportion of the children, and this should be taken into account and discussed during the process of obtaining consent for the operation.

Every patient presenting with active mucosal inflammation with frequent infections that do not settle with conservative medical management should be treated as early as possible using *tympanoplasty*, since the extent of the destruction of the sound-conducting apparatus increases with every inflammatory episode. If a hearing aid is necessary, tympanoplasty is also indicated, since chronic otorrhea makes it impossible to wear an earpiece.

> Note: Chronic mucosal inflammation is a form of chronic otitis media in which the inflammation is mainly confined to the mucosa. It does not usually cause progressive bone destruction and is therefore free of complications, but it has a protracted course.

Chronic Bone Suppuration (Chronic Epitympanic Otitis Media)

As a result of a marginal defect of the tympanic membrane in the posterior superior pars tensa or in the pars flaccida, inflammation spreads to parts of the bony lateral wall of the epitympanum. Granulations or polyps are typical signs of granulating ostitis (Fig. 1.88a, b).

Acquired Cholesteatoma of the Middle Ear

A cholesteatoma is a skin growth that occurs in the middle ear. It is usually due to repeated infection, which causes ingrowth of the skin of the eardrum. Progressive inflammation develops on the basis of a marginal tympanic membrane perforation, with osteitis on the lateral epitympanic wall. Cholesteatomas often take the form of a cyst or pouch that sheds layers of old skin, which builds up inside the middle ear.

Clinical Features

- Fetid otorrhea that is sometimes minimal or completely absent; when present, it is always purulent, and never mucoid.
- Progressive hearing loss, possibly dizziness.
- Otalgia and fever in acute exacerbations.
- Dull headaches or a feeling of pressure in the head.

Pathogenesis

Note: An acquired middle ear cholesteatoma is not a tumor, but rather a chronic inflammation which, unlike chronic mucosal inflammation, causes progressive destruction of the bony cells and structures.

Promoting factors:

- Disordered ventilation and drainage of the middle ear (chronic reduction of pressure) with hypopneumatization.
- Displaced squamous epithelium as a result of increased capacity for growth of the meatal skin in the upper part of the anulus tympanicus (the papillary ingrowths form the later *matrix*, either as a result of invagination of the pars flaccida or by formation of a retention pocket in the pars tensa).
- An increased proliferative tendency in the stratum germinativum (see Fig. 1.7), caused by the stimulus of inflammation.
- Incompletely resolved embryonic hyperplastic mesenchymal remnants in the submucosa of the middle ear, which later form the *perimatrix*.

Histopathogenesis: A cholesteatoma may form a compact sac of desquamated lamellae, arranged like the layers of an onion and connected with a fairly thick pedicle to its site of origin in the tympanic membrane (the pars flaccida or tensa). Alternatively, it may consist of a widely fanned-out cholesteatoma matrix lining the antrum and mastoid cavity and sending off shoots into the furthest bony niches of the bony process. The latter type of cholesteatoma therefore has a reticular or dendritic, branched structure. The latter occurs more commonly in a tensa cholesteatoma than in a flaccida cholesteatoma. Bone destruction is caused firstly by enzymes (e. g., collagenase) formed in the perimatrix, and secondly by osteoclastic destruction of bony tissue—i. e., chronic osteomyelitis.

Note: A prerequisite for the development of a cholesteatoma is direct contact between the keratinizing squamous epithelium in the external meatus and mucoperiosteum of the middle ear that has been damaged by inflammation.

This can happen as a result of:

- A *marginal perforation* with a destruction of the protective barrier of the anulus fibrosus.
- *Papillary ingrowths*—e. g., in the region of the pars flaccida or of the destroyed anulus fibrosus.
- Formation of a *retraction pocket* in the pars tensa.
- *Traumatic displacement* of keratinizing squamous epithelium after longitudinal pyramidal fractures or ruptures of the tympanic membrane, with the development of posttraumatic otitis media.

The same pathogenetic factors (see p. 67) are involved in the genesis of cholesteatoma as to that of chronic mucosal inflammation.

Diagnosis. An inflammatory middle ear cholesteatoma can be classified from several different points of view (primary, secondary, or topographic anatomy). The method using the site of origin appears to be the most sensible from the diagnostic point of view and clearer than the other classifications. Cholesteatoma can thus be classified as follows:

- *Tensa cholesteatoma* (synonym: secondary middle-ear cholesteatoma).
- *Flaccida cholesteatoma* (synonyms: primary or genuine attic or epitympanic cholesteatoma).
- *Occult cholesteatoma* (synonyms: cholesteatoma behind an intact tympanic membrane, or congenital cholesteatoma).

The *tensa cholesteatoma* (Fig. 1.89a, b) develops from a retraction pocket caused by chronic inflammation, usually in the posterosuperior quadrant of the pars tensa. It is characterized by a posterosuperior marginal perforation. Inflammatory granulations, fetid exudate with flakes of cholesteatoma, and circumscribed destruction of the surrounding posterosuperior meatal wall are often found in the region of the edge of the perforation. Conductive hearing loss is also present, due to destruction of the ossicular chain. The site of predilection is the incudostapedial joint.

The *flaccida cholesteatoma* (Fig. 1.89c, d; Fig. 1.90a–c; Fig. 1.91a, b) arises from papillary ingrowth of the keratinizing squamous epithelium in the region of the Shrapnell membrane, in the presence of simultaneous chronic epitympanitis. The stimulus of inflammation induces increased proliferation of the squamous epithelium. Circumscribed perforation of the pars flaccida, often covered by a crust and usually also accompanied by destruction of the lateral attic wall, is therefore

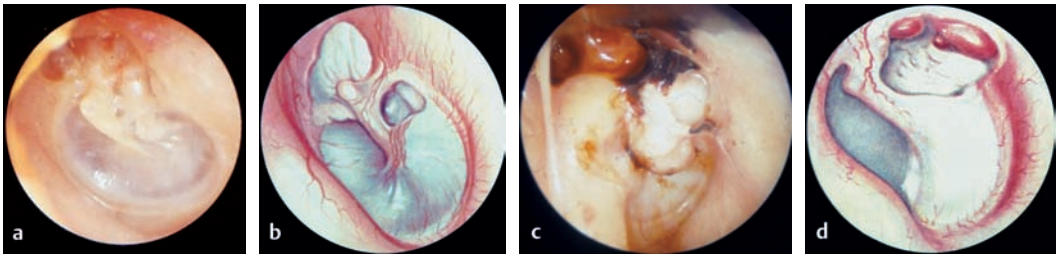


Fig. 1.89a–d Chronic otitis media with cholesteatoma.
a, b A marginal tympanic membrane defect in the anterior upper quadrants.
c, d A large marginal perforation in upper posterior quadrant. The chronic inflammation has destroyed the posterosuperior auditory wall.

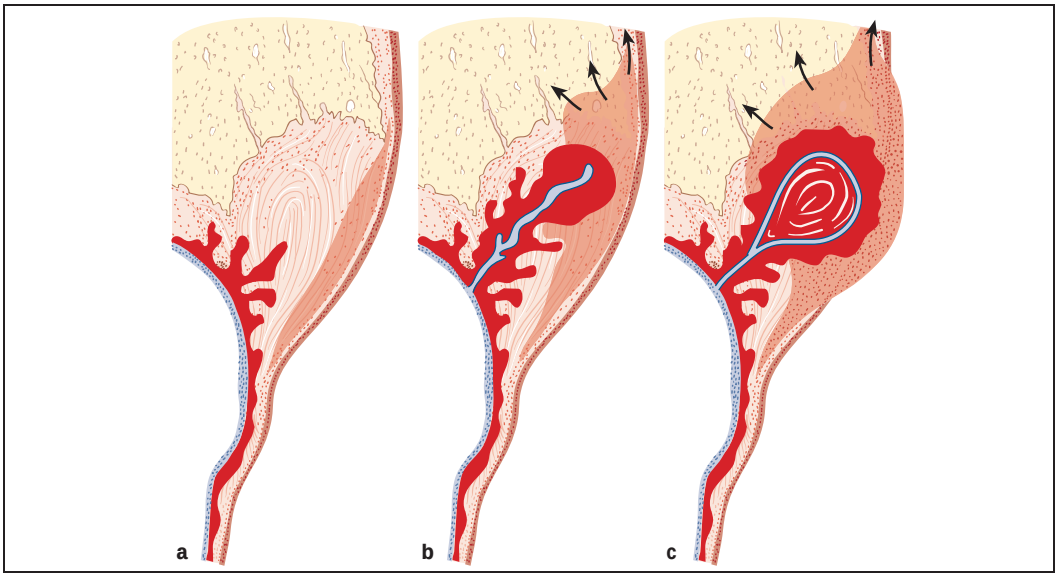


Fig. 1.90a–c Histological pathogenesis of an attic cholesteatoma.
a Papillary growth of the stratum corneum (the future matrix) of the pars flaccida down toward the attic. The connective tissue is loose, and the submucosa (the future perimatrix) is hyperplastic and chronically inflamed.
b Invagination of the pars flaccida into the attic as a result of hypoventilation and reduced pressure in the epitympanum. The future matrix is in contact with the perimatrix and there is early bone destruction.
c Activation of the basal cell layer of the matrix by chronic inflammation. Proliferation of keratinizing squamous epithelium and destruction and undermining of the mucosa in the middle ear, with replacement by the "foreign" squamous epithelium from the invaginated pars flaccida and formation of a cholesteatoma sac.

characteristic of this type of cholesteatoma. Medial to this lie inflammatory granulation tissue and fetid exudate with flakes of cholesteatoma. The patient has conductive hearing loss and also, in the advanced stages, involvement of the inner ear.

Occult cholesteatoma develops gradually and remains asymptomatic for a long time behind an intact tympanic membrane with no demonstrable perforation. In most cases, this involves a flaccida cholesteatoma that develops without a perforation by papillary ingrowth into the epitympanum, extending from there to the neighboring middle-ear space (Fig. 1.92). The pneumatization and the mucosal folds determine the pathways of spread of the cholesteatoma. In rare cases, it may be a congenital cholesteatoma.

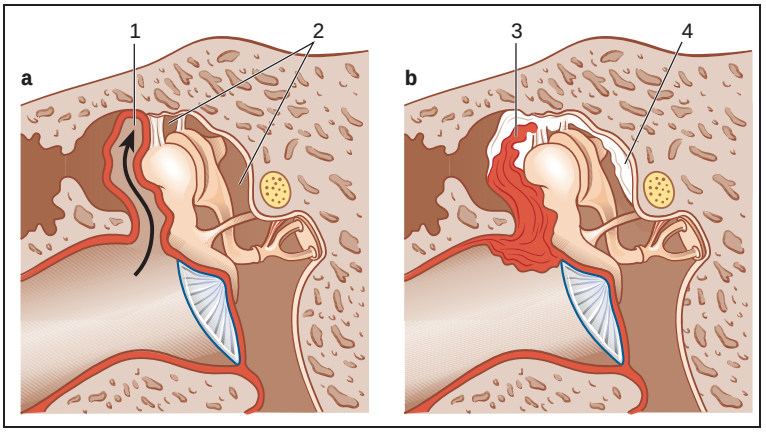


Fig. 1.91a, b Development of an attic cholesteatoma.
a Retraction and invagination of the pars flaccida into the epitympanum occurs as a result of persistently reduced pressure in the middle ear. The keratinized squamous epithelium is thus displaced into the middle ear. 1, Invaginated pars flaccida; 2, attic.
b Chronic inflammation causes hyperplasia of the mucosa of the attic and thus prevents adequate aeration of the region. 3, Cholesteatoma sac; 4, hyperplastic mucosa and "mesenchymal cushion" of the attic.

Common diagnostic characteristics. *Radiology: Axial and coronal high-resolution CT* of the temporal bone is the most useful and versatile procedure for demonstrating bone destruction in the petrous pyramid, soft-tissue abnormalities in the middle ear, and extension of the cholesteatoma into the cranial cavity. Schüller and Stenver views can partly show the grade of pneumatization, but have limitations for demonstrating destructive processes.

Audiometry: The audiogram shows conductive hearing loss, possibly combined with sensorineural deafness. Extension into the labyrinth causes progressive sensorineural deafness.

Vestibular tests: If the labyrinthine capsule is intact, spontaneous and provoked vestibular nystagmus do not occur. Erosion of the lateral semicircular canal causes a positive fistula test sign (see p. 47).

Facial nerve function: Erosion of the bony canal of the facial nerve and its mastoid or tympanic segment, or extension into the internal meatus, first cause neurapraxia and later progressive axonotmesis.

Differential diagnosis

- *Inactive chronic mucosal inflammation* with adhesions between the promontory and an atrophic pars tensa.
- *Carcinoma of the middle ear or external meatus.*
- *Tuberculosis of the middle ear.*

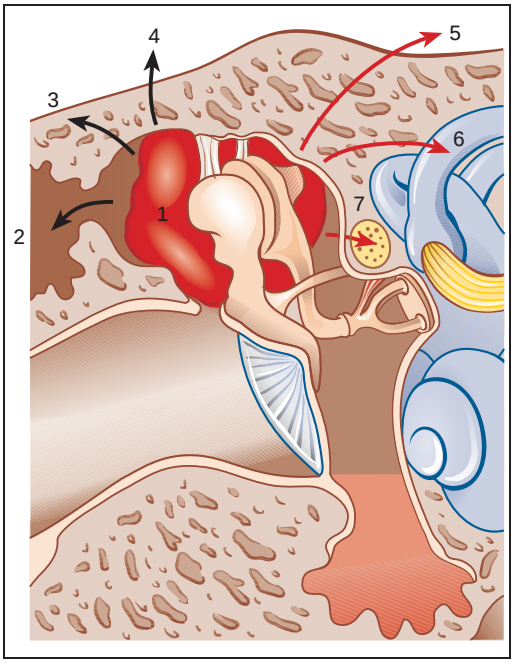


Fig. 1.92 Pathways of cholesteatoma extension: from the epitympanum into the attic (1); posteriorly, toward the mastoid and the sigmoid sinus (2) and the posterior (3) and middle cranial fossa (4); or medially, toward the internal meatus (5); and anteriorly toward the labyrinth (6) or the facial canal (7). 1, attic (see also Fig. 1.4.).

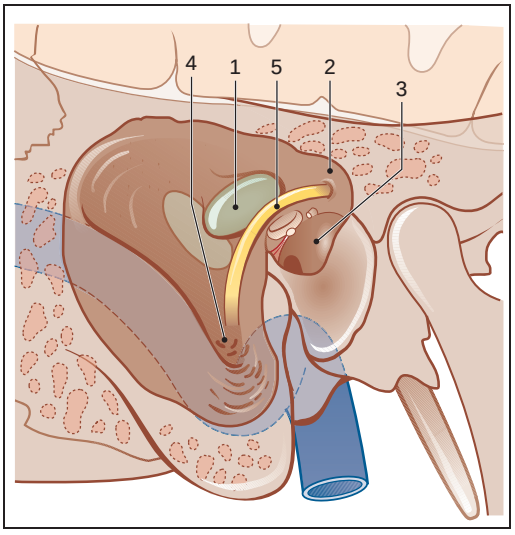


Fig. 1.93 Principle of radical mastoidectomy. There is eburnation of the mastoid, and the antrum (1), attic (2), middle ear cavity (3), and small mastoid cavity (4) are opened widely. Dissection of the bony facial canal as far as the second knee (tympanic segment) (5). These structures are united in a common cavity opening into the external meatus. If a tympanoplasty is to be performed, then a modified radical operation is carried out: the middle ear cavity is closed off from the mastoid cavity, and the sound-conducting apparatus is reconstructed.

Treatment. *Conservative treatment* with antibiotics, irrigation of the ear (also with a drainage tube), and antibiotic and corticosteroid-containing eardrops are ineffective, since antibiotics (whether systemic or local) are not able to control the local inflammation or the displacement of squamous epithelium into the middle ear space.

Surgical treatment: Surgical treatment of chronic otitis media with cholesteatoma involves a curative procedure, with radical removal of the inflammation, and a reconstructive procedure, with reconstructive surgery of the middle ear.

Note: There are two goals in surgery for cholesteatoma:

1. Radically eliminating inflammation by removing the matrix, perimatrix, and invaded bone
2. Maintaining and reconstructing the sound-conducting apparatus

Mastoidectomy: In the curative part, the attic, middle ear cavity, antrum, and infected pneumatic spaces of the mastoid process are exposed using a retroauricular or endaural approach. The mastoid and antrum cells are exposed and resected. Two mastoidectomy principles are used here, depending on the extension of the cholesteatoma:

1. The *closed* or *intact canal wall* technique. The bony posterior meatal wall and lateral attic wall are preserved (see Fig. 1.85).
2. The *open* or *canal wall–down* technique: After removal of the posterior bony meatal wall, the mastoid, attic, epitympanum and middle ear cavity are brought into continuity with the external meatus (Fig. 1.93).

Subtotal petrosectomy: After a mastoid cavity has been created using the canal wall–down technique, additional portions of the temporal bone are resected. The surgical cavity is obliterated with abdominal fat, and the eustachian tube and external ear canal are permanently closed.

Radical removal of the cholesteatoma matrix and perimatrix is carried out under microscopic control. The radical cavity heals within a matter of weeks, developing an epidermal layer (Fig. 1.94a,b). The disadvantage of this operation is that the sound-conducting chain is not reconstructed, so that the functional result is generally poor. In addition, there is an open connection between the middle ear, the mastoid cavity, and the external auditory meatus, so that chronic otorrhea is common due to tubal or exogenous infection.

Tympanoplasty: The purpose of this part is to reconstruct sound pressure protection of the round window and sound pressure transformation between the tympanic membrane and the oval window, by:

- Closing the perforation of the tympanic membrane with fascia or perichondrium.
- Reconstructing the direct connection between the tympanic membrane and stapes footplate if the ossicular chain is defective (ossiculoplasty).
- Separating the middle ear cavity from the external meatus by reconstructing the posterior meatal and lateral attic walls with a bony or cartilaginous graft or preserving the intact bony meatal wall.

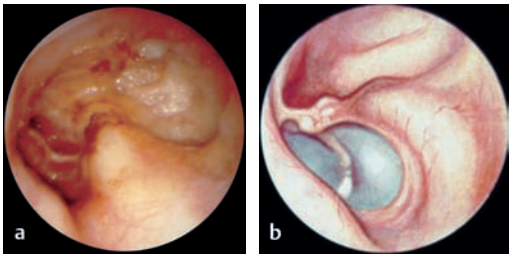


Fig. 1.94a, b View into an open cavity following a radical mastoidectomy. The cavity is completely epithelialized.

Radical elimination of the infection and reconstruction of the sound-conducting apparatus are usually performed in a one-stage procedure, but a radical mastoidectomy is still occasionally performed in preference to a tympanoplasty for the following indications:

1. Cholesteatoma with intracranial complications.
2. Cholesteatoma with facial nerve paralysis.
3. Extensive reticular–dendritic cholesteatoma in a well-pneumatized temporal bone.

The main goal of radical mastoidectomy is to control the disease. This therapeutic approach can be expanded by performing a tympanoplasty, which is done as a secondary procedure after complete healing of the infection; the aim is to achieve ossicular reconstruction. This expanded approach is not practised in all countries, however, as the theories underlying the treatment vary.

Principles of tympanoplasty: In 1952, Wullstein introduced a scheme of five basic types of operation for reconstructing the middle ear (Fig. 1.95a–e). Only type I and type III are of practical importance nowadays, as fenestrations of the semicircular canal are no longer performed and the importance of the lever mechanism of the ossicles was overestimated. Closure of the perforation of the tympanic membrane using fascia or perichondrium is the type I operation and is called *myringoplasty.* Type III involves reconstruction of the ossicular chain (ossiculoplasty), the aim being to restore the sound-pressure transformation from the tympanic membrane to the footplate in the oval window. The condition of the stapes is particularly important for this procedure. Three primary situations are encountered (Figs. 1.95a–e, 1.96a–f):

1. *Stapes intact*: The reconstructive procedure establishes a bridge between the tympanic membrane and/or the malleus handle and the head of the stapes. The connection is provided by an alloplastic (synthetic) prosthesis, known as a *partial ossicular replacement prosthesis* (PORP) (Figs. 1.96 d, 1.97a).
2. *Stapes crura absent, footplate mobile*: An alloplastic prosthesis, known as a *total ossicular replacement prosthesis* (TORP), is placed between the footplate and the tympanic membrane (Figs. 1.96 f, 1.97b).
3. *Footplate absent or fixed*: An opening is made in the oval window and subsequently closed with autologous tissue (type V tympanoplasty; Fig. 1.95e).

Ossiculoplasty can be carried out using different materials, mainly consisting of alloplastic implants (gold, titanium, plastic, ceramic). Remnants of true ossicles are used whenever possible.

Course and prognosis. Untreated cholesteatoma is the most dangerous form of chronic middle ear inflammation (see Fig. 1.92). At any time, healthy patients can develop the following life-threatening intracranial complications:

- Labyrinthitis and meningitis.
- Sinus thrombosis and septicemia.
- Epidural or subdural abscess, with meningitis.
- Temporal lobe or cerebellar abscess.

Note: Conservative treatment almost never achieves healing of a cholesteatoma. Surgery is therefore always indicated, due to the danger of intracranial complications.

Congenital Cholesteatoma of the Temporal Bone

This is a rare form of cholesteatoma, with no obvious connection with the external meatus or tympanic membrane. It is usually an occult lesion behind an intact tympanic membrane and only exceptionally develops from an embryonic ectodermal remnant within the temporal bone. It is also known as true or primary cholesteatoma.

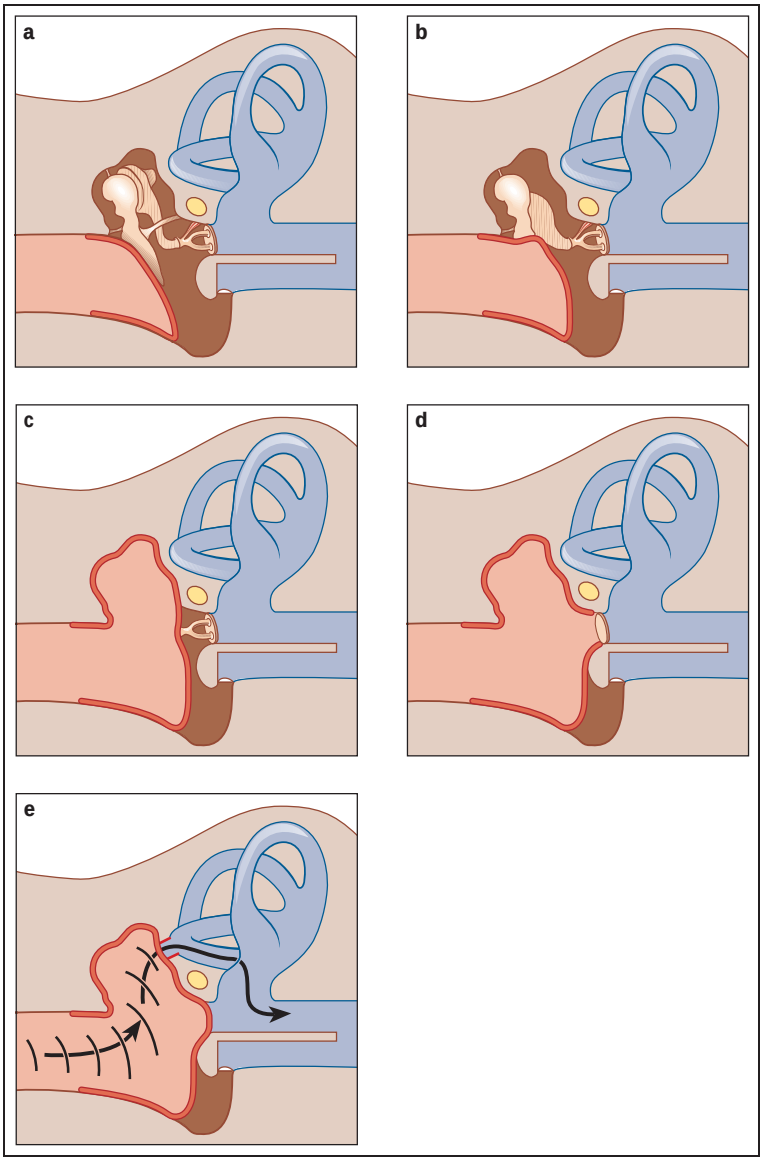


Fig. 1.95a–e The five classical types of tympanoplasty as defined by Wullstein.
a Type I: simple myringoplasty. The perforation of the tympanic membrane is closed with fascia or perichondrium.
b Type II: reconstruction of the defective ossicular chain by bridging the defect with autologous bone or cartilage grafts.
c Type III: direct transmission of sound waves from the tympanic membrane to the stapes by the columella effect. A shallow tympanum is created.
d Type IV: the ossicular chain is absent; sound is transmitted directly to the oval window, and sound protection is provided for the round window. A small tympanum is formed.
e Type V. In this case, the oval window is completely closed by bony fixation of the footplate. A window is made in the horizontal semicircular canal so that sound is transmitted directly to this fenestration, as in the similar operation for otosclerosis. Types IV and V have both now been abandoned in favor of interposition of an artificial columella (type IV) or—instead of type V—by removing the footplate and interposing a bony, cartilaginous, or synthetic prosthesis, as in stapedectomy for otosclerosis (see Fig. 1.102).

Course of Specific Forms of Cholesteatoma

Cholesteatoma is rare in infants and young children, but increases in frequency after the age of 6 years.

Cholesteatoma is relatively rare in old age. It is then usually due to reactivation of an old inflammatory process, which proceeds insidiously and may become manifest initially through the development of complications such as:

- Dizziness.
- Rapidly progressive hearing loss.
- Facial paralysis.
- Meningitis.

Petrositis due to progressive bone destruction, formation of sequestra, and invasion of the labyrinth, facial canal, and cranial cavity, with an epidural or subdural abscess, may develop insidiously. One pathogenetic factor is diabetes, due to the reduction in general resistance and local tissue metabolic disorders. The postoperative healing process is often prolonged, and the prognosis on the whole is poor.

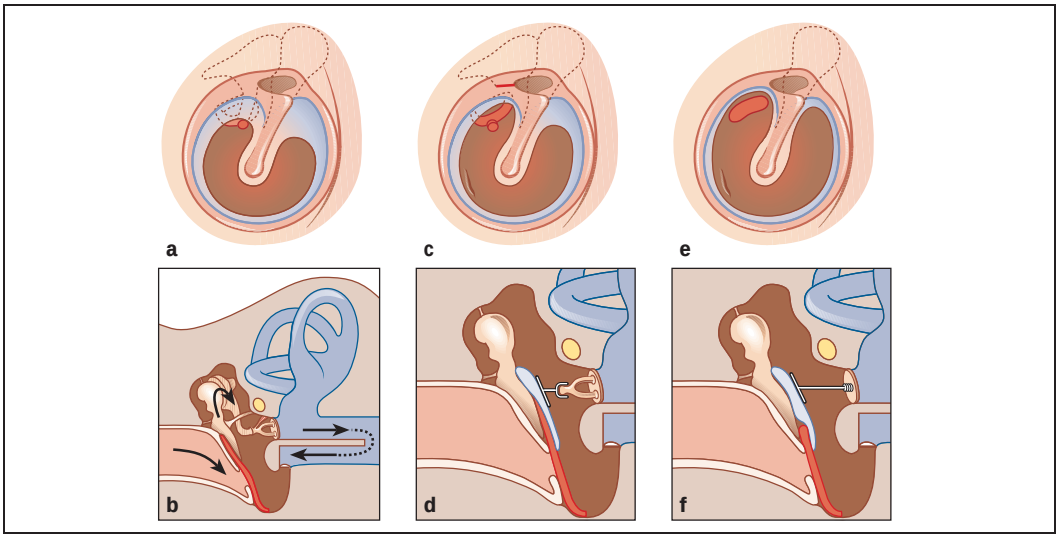


Fig. 1.96a–f Principles of tympanoplasty.
a Simple perforation of the tympanic membrane; the ossicular chain is intact.
b Closure of the defect in the pars tensa with underlay graft (fascia or perichondrium, *red*) and reconstruction of the sound pressure protection and transformation mechanism.
c Tympanic membrane perforation with a defect of the long process of the incus.
d Closure of the perforation. Reconstruction of the sound-conducting apparatus by interposing an alloplastic prosthesis (partial ossicular replacement prosthesis, PORP) between the malleus and the stapes—the columella effect. There is a slice of cartilage (*blue*) between the prosthesis and tympanic membrane.
e Defect of the incus and stapes with retention of the long process of the malleus and the footplate of the stapes, in a subtotal perforation of the tympanic membrane.
f Bridging of the defect of the ossicular chain by interposing an alloplastic prosthesis (total ossicular replacement prosthesis, TORP).

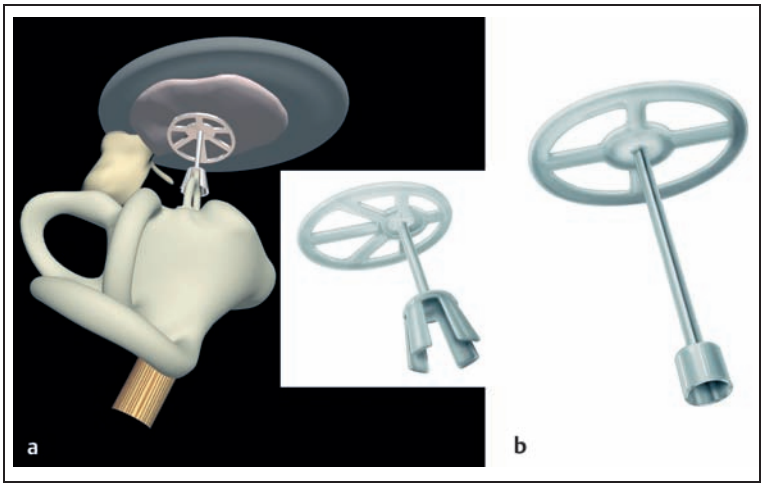


Fig. 1.97a, b a Positioning of an alloplastic PORP prosthesis, which is placed on the head of the stapes.
b TORP prosthesis. The oval plate is connected to the tympanic membrane; the small end is placed on the stapes footplate.

Meatal cholesteatoma can arise either from the pathologic proliferative tendency of the meatal epidermal lining in the bony part, or it may be the result of a rupture of a middle ear cholesteatoma from the antrum into the part of the meatus immediately lateral to the tympanic membrane.

Posttraumatic cholesteatoma arises as a result of a longitudinal pyramidal fracture extending into the external meatus. This allows meatal epidermis or a part of the tympanic membrane to be displaced into the middle ear space. The displaced keratinizing squamous epithelium initially forms an innocent epidermal cyst; a bone-destroying cholesteatoma with a matrix and perimatrix arises later, after the cyst has become infected.

Fig. 1.98 Adhesive otitis. The tympanic membrane is retracted, scarred, and thickened, but intact.

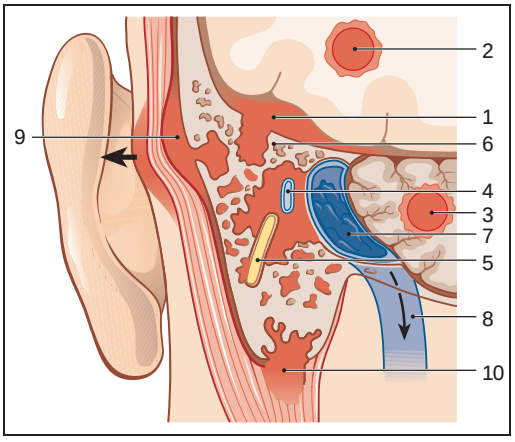


Fig. 1.99 Pathogenesis and pathways of spread of otogenic complications. 1, Extension into the middle cranial fossa, causing a subdural abscess; 2, meningitis or temporal lobe abscess; 3, extension into the posterior cranial fossa, causing meningitis or cerebellar abscess. Intratemporal complications: 4, labyrinthitis; 5, otogenic facial paralysis; 6, petrositis; 7, sinus thrombosis with thrombophlebitis; and 8, sepsis. Extratemporal complications: 9, external rupture of mastoiditis to form a subperiosteal abscess; 10, rupture of the mastoid apex to form an infratemporal Bezold abscess.

Adhesive Otitis (Middle Ear Fibrosis)

Clinical Features. The tympanic membrane is retracted, scarred, and thickened, but intact. The patient has severe conductive hearing loss (Fig. 1.98).

Pathogenesis. The condition is caused by recurrent otitis with cicatricial fibrosis of the poorly aerated middle ear, formation of cholesterol granuloma, and fixation of the ossicular chain.

Otogenic Infective Complications

Otogenic complications (Fig. 1.99) are otologic emergencies that should be investigated and treated by a specialist without delay. Evaluation using audiometric and neurovestibular examinations, as well as CT scanning, should be carried out as soon as possible. Indications for surgery have high priority.

Labyrinthitis

Clinical Features. Dizziness, nausea, vomiting, whistling noises in the ears, and deafness develop within a short period. The patient has no fever and no pain.

Pathogenesis. In *acute otitis media,* toxins diffuse through the labyrinthine windows and the infection extends along the vessels (see p. 77).

In *chronic otitis media* with cholesteatoma, a fistula forms in the labyrinth and the infection extends directly into the perilymphatic space. After a transverse fracture of the temporal bone or operative trauma, there is direct damage to the membranous labyrinth, with secondary infection (see Fig. 1.62).

Diagnosis. The patient has the cochlear and vestibular symptoms of rapidly progressive inner ear failure.

Differential diagnosis. Ménière disease, sudden deafness, and acute vestibulopathy.

Treatment. Intravenous antibiotics are administered in high doses by continuous infusion. The middle ear should be drained, and a mastoidectomy may have to be performed. Mastoidectomy using the canal wall–down technique or subtotal petrosectomy is performed for cholesteatoma. An ad-

ditional labyrinthectomy has to be performed in cases of purulent labyrinthitis with meningitis. After trauma with temporal bone fractures, surgery is only undertaken if there is simultaneous cerebrospinal fluid (CSF) otorrhea, facial nerve paralysis, or meningitis.

Course and prognosis. Because of the varying pathologic anatomy, several different clinical forms of labyrinthitis are possible: serous in posttraumatic or viral cases; purulent due to bacterial invasion of the perilymphatic space; circumscribed due to a labyrinthine fistula in cholesteatoma; and generalized with involvement of the entire labyrinth due to extension of infection or generalized infection. In the latter form, the course is fulminant, with irreversible total loss of function. The infection is capable of extending to the meninges.

Epidural Empyema

Clinical Features. Dull pulsating pain in the head, otorrhea, and subfebrile temperature occur. There is no completely characteristic pattern of symptoms.

Pathogenesis. Acute or chronic infection extends from the mastoid process into the epidural space, due to destruction of the inner table by infection. The infection may also spread via preformed pathways along the perforating vessels in the bone, which remains intact.

Diagnosis. Characteristically, the condition has very few symptoms. It is usually an incidental finding at mastoidectomy, but it can be demonstrated using neuroradiographic techniques, including angiography, CT, and MRI.

Treatment. Immediate mastoidectomy with wide exposure of the dura, drainage, and antibiotics are indicated.

Course and prognosis. The prognosis is good if the disease is recognized early. Otherwise, there is a danger of pachymeningitis developing, with extension to the leptomeninges.

Subdural empyemas are rare and only occur during the course of diffuse meningitis.

Otogenic Meningitis

Clinical Features. These include headaches, stiffness of the neck, scaphoid abdomen, increasing loss of consciousness, photophobia, restlessness, tonic–clonic convulsions, and facial paralysis. Otorrhea, otalgia, and even deafness may be absent or occult. Typically, there is a bounding pulse, irregular breathing, and a fever of 39–40°C. The patient may have an oculomotor or abducens paralysis and abnormal optic fundi.

Pathogenesis. The cause is the spread of an acute or chronic bacterial infection, usually due to pneumococci, into the subarachnoid space:

- By *direct continuity,* due to inflammatory destruction of the bony walls.
- *Along preformed pathways,* via the perforating vessels and nerves in the bone—e. g., via the caroticotympanic nerves.
- By a thrombophlebitis extending along the diploic veins.
- *Via the labyrinth* and as a result of spread into the internal auditory meatus (rare).

Diagnosis. In addition to the typical clinical symptoms of acute meningitis, including CSF abnormalities, there are signs of acute, subacute, or chronic middle ear inflammation of varying degrees of severity. If the hearing is more or less normal and the otoscopic findings are equivocal, the signs of inflammation of the pneumatic system on radiographs of the temporal bone (in CT scan, Schüller or Stenver views) can be decisive in making the diagnosis and establishing the indication for surgery.

The CSF shows pleocytosis, a marked increase of protein, and a reduced sugar and chloride content. The pressure is greater than 200 mmHg. In the acute phase, bacteria—usually pneumococci—can be found in the fluid.

Differential diagnosis. This includes viral or epidemic meningococcal meningitis and tuberculous meningitis.

Treatment. High dosages of antibiotics are administered intravenously on the basis of an antibiogram—e. g., penicillin 40–60 million U/24 h for a pneumococcal infection. Repeated lumbar punctures are carried out to assess the CSF.

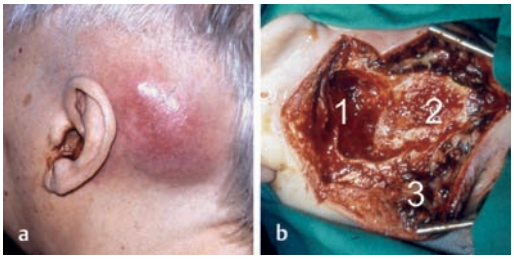


Fig. 1.100a, b Otogenic sinus thrombosis.
a Painful swelling of the mastoid.
b The intraoperative situation after drainage of the thrombophlebitis. 1, Opened mastoid cavity; 2, ligated sigmoid sinus with infected thrombus; 3, severely inflamed tissue on the mastoid.

The *blood–brain barrier* for antibiotics varies greatly, depending on the severity of the meningitis. Some of the antibiotic is inactivated by binding to the CSF protein. Third-generation cephalosporins such as ceftazidime (an antipseudomonal cephalosporin and therefore useful for otogenic meningitis due to chronic otitis media), cefotaxime, and ceftriaxone penetrate the blood–brain barrier very well. Intrathecal antibiotics are only used in exceptional cases. Immediate drainage of the middle ear cavity by mastoidectomy is required, with wide exposure of the dura.

Course and prognosis. The disease ends fatally if it is untreated or not treated correctly. The prospects of recovery are ≈ 90 %, provided that the otitis media is recognized as causing the meningitis at an early stage and provided that energetic treatment is started.

Note: Any unexplained attack of meningitis must be suspected of having a nasal or otologic origin.

In doubtful cases with obscure otoscopic and radiographic findings, the risk of an exploratory operation is less than that of an expectant policy if the middle ear space is infected. Intensive antibiotic treatment does not lead to resolution unless the primary focus of infection has been eliminated by surgery.

Otogenic Sinus Thrombosis

Clinical Features. A perisinus abscess, periphlebitis, and incipient sinus thrombosis lead to the same diagnostic difficulties as an epidural empyema. The release of emboli of infective thrombi alone causes the characteristic signs of *septicemia.* These include:

- Chills.
- A spiking temperature chart, with several peaks on the same day.
- Increased pulse rate.
- Headaches.
- Vomiting.
- Somnolence.
- Neck stiffness (accompanying meningitis).
- Dyspnea due to septic lung metastases or pneumonia.
- Jaundice due to septic metastases in the liver or to nonspecific reactive hepatitis.

Pathogenesis. Infection due to mastoiditis or cholesteatoma destroys bone in continuity, so that it can rupture into the perisinus space. A perisinus abscess forms, with periphlebitis of the sigmoid sinus, followed by sinus phlebitis. The thrombus is initially mural, but later it occludes the lumen and extends superiorly to involve the transverse and sagittal sinuses and the mastoid emissary vein, and inferiorly toward the internal jugular vein.

The thrombus undergoes thrombolysis due to bacterial infiltration, and septic metastases are caused by blood-borne infected emboli.

Diagnosis. The following symptoms during acute otitis media or chronic otitis media with cholesteatoma suggest sinus thrombosis (Fig. 1.100a, b):

- High fever above 40°C.
- Spiking temperature with chills.
- Swelling and sensitivity to pressure over the mastoid emissary foramen at the posterior border of the mastoid process (Griesinger sign).
- Induration and tenderness of the internal jugular vein and of the anterior border of the sternocleidomastoid muscle.
- Petechiae in septic coagulopathy.
- Splenomegaly.

Laboratory investigations: Blood culture is strongly positive. Urinalysis shows hematuria due to septic interstitial focal nephritis, albuminuria, and cylindruria.

Radiology: CT scans show bone destruction in the area of the sinus. Angiography shows narrowing or occlusion of the sigmoid sinus in the venous phase of carotid angiography.

Differential diagnosis. This includes miliary tuberculosis, typhus, malaria, brucellosis, viral pneumonia, and cystopyelitis.

Treatment. Immediate surgical excision of the primary inflammatory focus in the mastoid and sigmoid sinus by mastoidectomy for cholesteatoma is performed (Fig. 1.93 b; see also p. 72). The sigmoid sinus has to be widely exposed; an incision is performed or the sinus wall is incised, and thrombectomy is performed. The internal jugular vein is ligated and divided, with a margin of healthy tissue. Parenteral antibiotics are given in high dosages for a long period, as well as anticoagulants, if possible determined by the results of culture and sensitivity tests.

Course and prognosis. The patient should be monitored with serial CT scans to check the changes in the surrounding tissue. The disease is fatal if it is not treated correctly, or if the basic cause and its secondary consequences are not recognized promptly. Eighty percent of patients are cured when adequate treatment is started early.

Note: Every unexplained case of septicemia requires rigorous investigation of the ear, including radiographic diagnosis, since the otitis media that is primarily responsible may go unrecognized due to a lack of other typical clinical signs.

Otitic Hydrocephalus

This disease is caused by increased intracranial pressure due to obstruction of CSF drainage caused by a sterile otogenic sinus thrombosis.

Clinical Features

- Failing vision.
- Vomiting.
- Double vision.
- Jacksonian epilepsy.
- Pareses and disorders of sensation.

Pathogenesis. A relatively asymptomatic chronic mastoiditis follows an acute otitis media, leading to sterile erosion of perisinus bone and sigmoid sinus phlebitis with formation of a mural thrombus, extending to the confluence of the sinuses and the superior sagittal sinus. This causes occlusion of the pacchionian granulations, which interferes with resorption of CSF, leading to increased CSF pressure.

Diagnosis

- Abducens paralysis without Gradenigo syndrome (see p. 80).
- Increased CSF pressure without pleocytosis.
- Free CSF circulation without CSF obstruction.
- Congested optic fundi with failing vision.
- Opacity and osteolytic perisinus lesions in temporal bone CT scans.
- A history of acute otitis media 3–5 weeks previously.

Differential diagnosis

- Petrositis (see p. 80) with arachnoiditis in the cerebellopontine angle.
- Carotid aneurysm at the petrous apex.

Treatment. Treatment includes mastoidectomy, exposure of the sinus, thrombectomy, and neurosurgical decompression to allow drainage of the CSF.

Course and prognosis. These are good if the disease is recognized and treated early. Otherwise, the disease progresses to blindness and the development of Jacksonian epilepsy.

Otogenic Brain Abscess

This is one of the most serious late complications of chronic inflammatory middle ear cholesteatoma.

Clinical Features. See Table 1.10.

Pathogenesis. The disease can spread by *direct continuity* via one of the following pathways:

1. Through the tegmen tympani, to form a temporal lobe abscess.
2. Through the sigmoid sinus to the posterior cranial fossa, to form a cerebellar abscess.
3. From the labyrinth to the endolymphatic sac, to form a cerebellar abscess (see Fig. 1.99).

Another pathway is via vessels (the diploic veins, advancing septic thromboangiitis of the cerebral veins), or via the internal auditory meatus in cases of labyrinthitis.

Clinical Features of temporal lobe abscess

- Speech disturbances, revealed by a history of aphasia and difficulty in understanding words. (This disorder of speech is exclusively sensory and is never motor.)
- Central hearing disorders, which are mostly discrete.
- Acoustic hallucinations.
- Disorders of smell, which are usually discrete.
- Visual disturbances such as quadrant hemianopsia and gaze paresis.

Table 1.10 Clinical Features of otogenic brain abscess

1. Initial stage	Meningism, nausea, headache, psychological changes, fever
2. Latent stage	Epileptiform attacks, neurologic signs
3. Manifest stage	Vomiting and bradycardia, psychological changes, focal signs of aphasia, alexia, agraphia, hemiplegia, epileptic attacks, and ataxia in cerebellar abscess. Symptoms due to spread to neighboring organs include cranial nerve paralyses, visual field defects, disorders of the oculomotor system and of posture
4. Terminal stage	Stupor, coma, conjugated deviation to the side of the lesion, bradycardia, and Cheyne–Stokes respiration

- Neuropathies of cranial nerves III–VII.
- Crossed lesions of the pyramidal tracts.

Differential diagnosis of a temporal lobe abscess. Intracerebral tumor.

Clinical Features of a cerebellar abscess
- Disorders of the oculomotor and postural system.
- Nystagmus: coarse spontaneous nystagmus to the side of the lesion, vestibular provocation nystagmus with irregular positional nystagmus, gaze-directional nystagmus due to secondary damage to the pons, gaze-paretic nystagmus to the side of the lesion.
- Ataxia, intention tremor, dysmetria, adiadochokinesia, hypotonia, and symptoms due to spread to neighboring organs such as paralysis of cranial nerves III, V, VI, VII, IX, and X.

Differential diagnosis of a cerebellar abscess. Labyrinthitis, acute vestibulopathy, multiple sclerosis, cerebellar tumor, and cerebellopontine angle syndrome need to be considered.

Investigations. In addition to otologic examination, these include:
- Neurotologic investigations: AEP, ENG, and electrodiagnostic methods for the facial nerve.
- Neuro-ophthalmologic investigation of the optic fundi, visual field, and ocular motor nerves.
- Neurologic and neuroradiologic investigations, including CT, MRI, EEG, brain scan, and possibly angiography, echoencephalography, and investigations of the CSF.

Treatment. The primary focus is removed by an otologist, using mastoidectomy. Intracranial drainage can be performed during the same operation. Primary removal of the brain abscess can be achieved by craniotomy in a neurosurgical procedure. Isolated aspiration of the abscess has now been abandoned, except in a few exceptional circumstances. It is more effective to eliminate the focus of infection radically by surgery along the pathway of infection. In addition to surgery, appropriate, intense antibiotic therapy is administered.

Course and prognosis. Despite intensive surgical treatment, the mortality is still 5–8 %. The defect often heals, but there may be neurologic deficits after neurosurgical excision of the abscess.

Petrositis (Petrous Apex Syndrome)

Pathogenesis. Good pneumatization of the entire petrous pyramid is a prerequisite for the development of petrositis. As a result of extension of inflammation from the middle ear to the perilabyrinthine cells, purulent liquefaction in the cells of the petrous apex results, often accompanied by osteomyelitis. The classic Gradenigo triad is due to the close relationship of the pyramidal apex to the trigeminal nerve and abducens nerve. The symptoms of this triad include otorrhea, ipsilateral irritation of the trigeminal nerve, and abducens paralysis. The facial, vagus, and glossopharyngeal nerves are also often paralyzed. In addition, the patient may have signs of labyrinthitis due to extension of the inflammatory process. The symptoms of primary otitis media may be hidden due to antibiotic treatment. Predisposing factors include advanced age and diabetes.

Diagnosis. The main symptoms are:
- Trigeminal neuralgia.
- Abducens paralysis with double vision.
- Dizziness and deafness.
- Deep, throbbing headache.
- Abnormal CT scans of the temporal bone.

Differential diagnosis. Brain abscess and sinus phlebitis.

Treatment. A combination of administering high-dose and prolonged parenteral antibiotics and surgery. Mastoidectomy is performed, with translabyrinthine clearance and drainage of the apical cells, ensuring that the facial nerve is protected.

Prognosis. If the disease is recognized early and treated effectively, the prognosis is relatively good. However, the prognosis is poor in patients of advanced age and those with diabetes (development of circumscribed meningitis at the base of the brain, with subsequent multifocal microinfarction of the pons).

Specific Diseases of the Middle Ear and Mastoid Process

Wegener Granulomatosis

The disease is characterized by a combination of necrotizing granuloma formation and vasculitis. It can become manifest in adult patients as chronic bilateral serous otitis media. It can cause facial nerve paralysis and conductive hearing loss, as well as sensorineural hearing loss secondary to vasculitis of the stria vascularis. The inflammation damages renal and pulmonary structures. In consultation with a rheumatologist, treatment with cyclophosphamide and steroids is recommended. Inserting tympanic ventilation tubes may help alleviate the hearing loss.

Tuberculosis

Tuberculosis of the middle ear may be due either to a miliary process or to tubal extension of a localized infection from the nasopharynx. Multiple and persistent perforations of the tympanic membrane, severe loss of hearing, facial paralysis, and otorrhea are suspicious signs, but the diagnosis can be difficult to confirm. Confirmation is based on a positive tuberculin skin test and demonstration of acid-fast bacilli in the otorrhea or on tissue staining. The first-line treatment is medical, with antituberculous antibiotics. Surgical therapy may be necessary in order to debride bony sequestra.

Syphilis

Syphilis of the middle ear is extremely rare in comparison with syphilis of the internal ear and vestibulocochlear nerve. It becomes manifest in the secondary and tertiary stages and also as metasyphilitic or congenital disease.

Clinical Features. These include dizziness, tinnitus, rapidly progressive nonfluctuant hearing loss, and headaches due to chronic syphilitic meningitis.

Pathogenesis. The disease is due to a specific labyrinthitis and neuritis of the vestibulocochlear nerve, the latter arising during the course of a meningovascular syphilitic meningitis. In the tertiary stage and also in congenital and metasyphilitic disease, the nervous apparatus degenerates and atrophies due to meningovascular or parenchymatous syphilis, with progressive demyelinization of the vestibulocochlear nerve.

Diagnosis. The diagnosis is established by serum specimens that test positive on the *Treponema pallidum* hemagglutination (TPHA) or fluorescent treponemal antibody absorbed (FTA-abs) tests.

Differential diagnosis

- Ménière disease with a fluctuating course.
- A cerebellopontine angle tumor, which has typical neuroradiologic findings.
- Vertebrobasilar insufficiency, which occurs in the elderly, with discrete neurologic findings.

Treatment. Long-term penicillin is given in high doses.

Course and prognosis. Early diagnosis and correct treatment can stop progression, but the outlook for function being restored is poor.

Noninflammatory Diseases of the Labyrinthine Capsule

Otosclerosis

Otosclerosis is a localized disease of the bony labyrinthine capsule, the cause of which has not yet been identified. Spongy bone hardens around the base of the stapes. This condition fixes the stapes to the opening of the inner ear, so that the stapes no longer vibrates properly. Otosclerosis can also affect the malleus, incus, and the bone that surrounds the inner ear, resulting in disruption of sound transmission to the inner ear. Untreated otosclerosis eventually results in total deafness, usually in both ears. The exact diagnosis is made by histology, but only 10 % of patients with histologic evidence of the disease have clinical otosclerosis: 8–10 % of whites, 1 % of Japanese, and fewer than 1 % of blacks have histologic diseases.

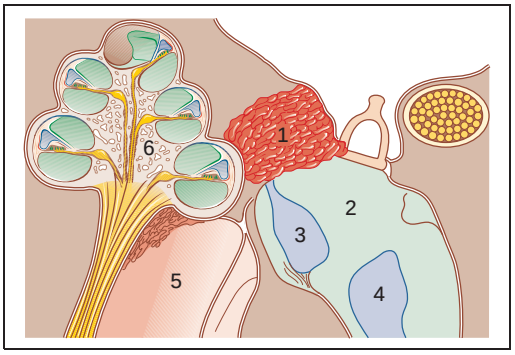


Fig. 1.101 Pathology of otosclerosis. 1, Focus of otosclerosis; 2, vestibule; 3, utricle; 4, saccule; 5, internal acoustic canal, 6, cochlea.

Clinical Features. Depending on the site of the otosclerotic focus, the symptoms include:

- Conductive hearing loss of the middle ear type, in ≈ 80 % of patients.
- Mixed conductive and sensorineural hearing loss, in ≈ 15 % of patients.
- Pure sensorineural hearing loss, in ≈ 5 % of patients.

The disease becomes manifest *subjectively* through:

- Slowly progressive hearing loss, which usually affects one ear initially, but later affects both ears in most patients.
- Constant, progressive tinnitus.

The disease never causes otalgia, otorrhea, dizziness, or disorders of balance.

Pathogenesis. This disease appears to have multifactorial causes, with the following being the most important:

Heredity: In 50–60 % of patients, there is a familial disposition with dominant inheritance, possibly due to a hereditary enzyme defect. As clinical otosclerosis only occurs in 10 % of patients with histologic disease, the pattern of inheritance appears to be recessive. The chance of inheriting the disease from a parent with clinically manifest disease is ≈ 20 %, and ≈ 10 % from a parent with histologic disease.

Disorders of hormone and bone metabolism: Pregnancy coincides with a period of progression of clinically manifest otosclerosis in half of female patients with the disease. Abnormal lysosome formation of and increased enzyme activity of the histiocytes and osteocytes in the labyrinthine capsule, as well as enzymatic collagenolysis and bony remodeling have been demonstrated. The newly re-formed bone fixes the stapes footplate in the oval window (Fig. 1.101).

Local infection with measles virus: As elevated levels of measles virus–specific immunoglobulins, acting as antigens, have been found in the perilymph in otosclerotic patients, it is thought that infection with measles virus may be an etiological factor. Measles-like structures have also been found in otosclerotic foci using transmission electron microscopy.

Autoimmunity: Some immunological findings have suggested that autoimmune reactions play a role in the pathogenesis of otosclerosis.

Note: Otosclerosis is due to an extremely localized disorder of mineral or bone metabolism, with an abnormal increase of enzyme activity in the mesenchymal cells of the labyrinthine capsule, mainly determined by genetic factors but also by hormonal disturbances.

Diagnosis.

- Positive family history.
- Otoscopy occasionally shows hyperemia of the promontory as it shines through the tympanic membrane (Schwartze sign).
- Functional symptoms: Pure-tone audiography usually shows pure middle-ear hearing loss, occasionally mixed hearing loss, and rarely pure sensorineural deafness with positive recruitment. There is often a characteristic notch in the bone conduction curve at 2000 Hz (the Carhart notch). Impedance audiometry usually shows a normal curve at normal pressures. However, the stapedial reflex is often suppressed due to otosclerotic fixation of the footplate.
- Radiography usually shows very good pneumatization of the temporal bone.

Differential diagnosis.

- Congenital anomalies of the middle ear.
- Posttraumatic dislocation or fracture of the ossicles.
- Postinflammatory fixation of the ossicles (malleus head).
- Adhesive processes.
- Tympanosclerosis.

Treatment. *Surgery* is indicated when there is sufficient inner ear function and the contralateral ear is not deaf. Three major components need to be taken into account when recommending surgery for otosclerosis: overall hearing loss, the extent of the air–bone gap, and the degree of handicap experienced by the patient. Although these three components are interrelated, it is not possible to predict the extent of one on the basis of the others. They have to be assessed independently.

In some countries, a trial of a hearing aid is recommended initially before surgical options are used, as hearing aids are usually very effective early in the course of the disease.

Two surgical techniques are used in otosclerosis:

Stapedectomy: The principle of this operation is to open the middle ear and expose the stapes footplate in the oval window niche. The fixed stapes is removed. The oval window is then closed with connective tissue, and the stapes is replaced with an alloplastic or wire prosthesis, or with an autologous cartilage graft. Complete removal of the footplate is no longer recommended.

Stapedotomy: After exposure of the oval niche, the footplate is perforated and a tiny, piston-like prosthesis is inserted and fixed to the long process of the incus (Fig. 1.102).

In cases of far-advanced otosclerosis with high-grade sensorineural hearing loss and no benefit from hearing aids, bilateral cochlear implantation can optimize performance.

The alternative treatment in sensorineural hearing loss is fitting a hearing aid. Other less successful forms of treatment include fluoride administration, which theoretically becomes incorporated into bone and inhibits progression of the otosclerosis. This treatment is not able to reverse conductive hearing loss, but may slow the progression of both the conductive and sensorineural components of the disease process.

Course and prognosis. Bilateral manifestations of the disease occur in 70 % of cases. Both ears can be operated on with stapedotomy, with an interval of at least 6 months between the procedures. If one ear has severe sensorineural hearing loss, surgery on the better-hearing ear is not indicated.

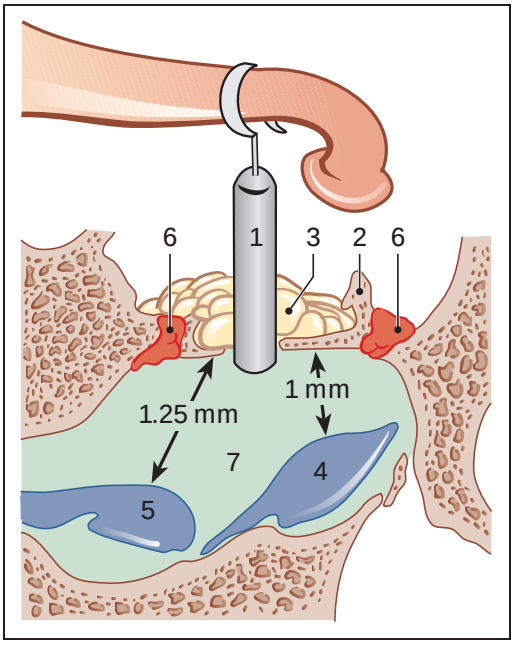


Fig. 1.102 The principle of stapedectomy, with reconstruction of the sound-pressure transformation mechanism. A suitable piston (1) is placed in the opening in the footplate (2) and its wire loop is fixed to the crura of the incus. Loose connective and fat tissue (3) is placed around the piston and the footplate to close off the perforation. The small distances between the stapedial footplate and the saccule (4) and utricle (5) are surgically important. 6, Otosclerotic focus on the footplate; (7) vestibule.

Note: The earlier in life otosclerosis becomes manifest, the more rapid and unfavorable is its course.

In the vast majority of cases, the prognosis with surgical treatment is excellent. The rate of hearing deterioration over time after stapedotomy has not been found to exceed that due to presbyacusis. Patients with otosclerosis very seldom develop total deafness, and they continue to be able to hear with a hearing aid. The older the patient, the less the tendency for further hearing loss to occur due to the process of otosclerosis.

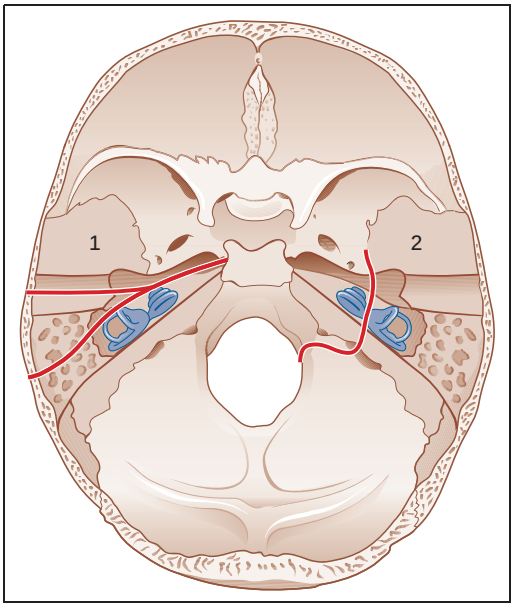


Fig. 1.103 Temporal bone fractures. 1, Longitudinal fractures; 2, transverse fracture.

Otologic Manifestations of Generalized Skeletal Disorders

The temporal bone, and the labyrinthine capsule in particular, can be affected by other generalized skeletal disorders:

- Osteogenesis imperfecta.
- Paget disease.
- Localized fibrous dysplasia.
- Osteogenesis imperfecta tarda with blue sclera (Lobstein type).

Trauma of the Middle and Inner Ear

Thorough familiarity with injuries to the ear and their consequences is essential for every practicing doctor. These lesions are usually caused by accidents, and the patients are therefore first seen by doctors working in accident and emergency departments or by general practitioners.

Frequency. Although injuries to the ear only make up 2–3 % of all injuries, 45 % of fractures of the base of the skull extend to the temporal bone, affecting the middle and inner ear.

Note: The ear and nasal sinuses should be examined as early as possible after every head injury. It is incumbent on the doctor who first sees the patient to investigate the following:

- Fresh bleeding or CSF leakage from the ear or nose.
- Evidence of blood or brain tissue in the external auditory meatus or the nose.
- Facial paralysis.
- Hemotympanum, rupture of the tympanic membrane, or a break in the outline of the anulus tympanicus and the external meatal wall.
- Hearing loss.
- Dizziness, disorders of balance, and nystagmus.
- Bleeding from the nasopharynx.

Temporal Bone Fracture

Pathogenesis. *Direct fractures* are caused by the effect of external force concentrated on a small surface—e. g., by gunshot wounds. The result is a penetrating, perforating fracture with brain damage.

Indirect fractures are due to diffuse external forces. The course of the fracture can run either:

- Along the pyramidal axis (i. e., *a longitudinal fracture*), extending into the middle ear.
- Across the pyramidal axis (i. e., *transverse fracture*), extending into the bony labyrinth and internal auditory meatus.

In both cases (Fig. 1.103), the dura may be torn, producing an open connection between the pneumatic system of the temporal bone and the subarachnoid space of the cranial fossae. The patient is then in danger of latent infection ascending via the eustachian tube to the meninges.

Clinical Features of longitudinal pyramidal fractures (mainly affecting the middle ear)

- Hemotympanum or CSF in the middle ear cleft (Fig. 1.104).
- Tearing of the tympanic membrane (Fig. 1.105).
- Bleeding from the external auditory meatus.
- A break in the contour of the tympanic ring.
- Step formation in the external auditory meatus, which should be differentiated from a posteriorly displaced fracture of the mandibular condyle.
- Middle ear deafness.
- Facial paralysis in ≈ 20 % of patients—usually neurapraxia or partial axonotmesis.
- Occasionally, CSF otorrhea.

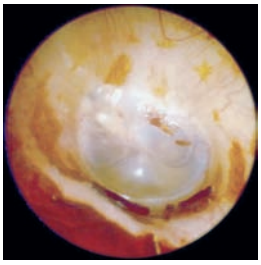


Fig. 1.104 Cerebrospinal fluid (CSF) in the tympanic cavity after a temporal bone fracture.

Fig. 1.105 Traumatic perforation of the tympanic membrane.

> **!** Caution: Syringing or manipulations must not be performed within the external auditory meatus.

Diagnosis. This is based on otoscopic findings and high-resolution CT scans.

Clinical Features of transverse pyramidal fractures (mainly affecting the inner ear)

- Intact external auditory meatus.
- Intact tympanic membrane, possibly with hemotympanum or CSF in the middle ear cleft (see Fig. 1.104).
- Hearing loss.
- Vertigo.
- Spontaneous nystagmus beating to the healthy ear.
- Facial paralysis in ≈50% of patients, usually showing axonotmesis or neurotmesis.
- Cerebrospinal fluid leak via the eustachian tube to the nasopharynx.

Diagnosis. This is based on otoscopic and functional findings and high-resolution CT scans. Additional investigations include EMG and neuronography, and the Schirmer test and gustometry for facial nerve paralysis (see p. 47).

Treatment of longitudinal and transverse pyramidal fractures. The treatment is dictated by the ever-present risk of otogenic meningitis. Prophylactic antibiotics are therefore administered in the form of high-dose, long-term parenteral broad-spectrum agents.

The temporal bone has to be explored if early or late complications occur (Table 1.11).

Emergency surgery definitely needs to be performed for the indications detailed above, as soon as the general condition of the patient permits. As the patient has usually suffered multiple injuries, the various disciplines have to be prioritized as follows:

1. Traumatology.
2. Neurosurgery.
3. Otology.
4. Maxillofacial surgery.
5. Ophthalmology.

Course and prognosis. The following complications are possible, particularly as a result of unsatisfactory treatment or missed diagnoses:

Table 1.11 Pyramidal fracture

Indications for early otologic intervention
• Early meningitis, treated by mastoidectomy
• Bleeding from the sigmoid sinus, treated by opening of the mastoid and packing or ligature of the sinus
• Persistent CSF otorrhea, treated by repair of the dura and possible obliteration of the mastoid cavity.
• Facial paralysis with signs of progressive axonotmesis, treated by decompression if there is more than 90% denervation shown by neuronography
• Depressed fracture of the external auditory meatus, treated by reconstruction of the meatus because of the danger of secondary atresia
• Gunshot wounds of the temporal bone, treated by debridement of the fragmented area
Indications for late otologic intervention
• Antibiotic-resistant traumatic otitis media
• Chronic mastoiditis, treated by mastoidectomy
• Late facial nerve paralysis with symptoms of denervation, treated by facial nerve decompression
• Posttraumatic hearing loss, treated by tympanoplasty and ossiculoplasty
• Posttraumatic cholesteatoma, treated by mastoidectomy and tympanoplasty

CSF, cerebrospinal fluid.

Early complications:
- Acute otitis media with mastoiditis.
- Extension of the infection to the subarachnoid space, causing early meningitis or an infected labyrinthitis extending to the meninges.

Late complications:
- Chronic otitis media with mastoiditis.
- Late otogenic meningitis.
- Epidural abscess.
- Otogenic brain abscess.
- Posttraumatic cholesteatoma.

Labyrinthine Concussion

Posttraumatic disorders of inner ear function (deafness and dizziness) in the presence of normal otoscopic and radiographic findings are summed up under the heading of labyrinthine concussion.

Clinical Features. These include tinnitus, unilateral or bilateral sensorineural hearing loss with positive recruitment and high-tone loss or a notch at 4000 Hz; dizziness particularly after changing position or after rapid movements of the head; and disorders of balance.

Pathogenesis. The condition is usually due to organic mechanical damage to the membranous labyrinth, similar to acute acoustic trauma (see p. 88). Microfractures of the labyrinthine capsule accompanied by bleeding into the perilymphatic and endolymphatic space, and mechanical disturbances of the microcirculation causing degeneration of the cochleovestibular sensory cells, may also occur.

Diagnosis
- Normal otoscopic findings.
- Normal radiographs (Schüller and Stenver views; CT scan).
- A pure-tone audiogram showing a sensorineural hearing loss with a notch at 4000 Hz or high-tone loss with recruitment.
- Vestibular provocation nystagmus in the presence of vertigo, and more rarely spontaneous nystagmus; possibly reduced sensitivity to caloric stimulation.

Differential diagnosis
- Acute acoustic trauma in which vestibular symptoms are absent.
- Posttraumatic psychogenic hearing loss in which the findings are inconsistent and vestibular symptoms absent.

Treatment
- Intravenous low-molecular-weight dextran infusion (if there is no general contraindication—e.g., hypertension, heart rhythm disturbance, or allergy).
- Corticosteroids intravenously (if there is no contraindication—e.g., diabetes, hypertension).
- Antivertigo drugs are administered for dizziness.

Course and prognosis. In young patients and patients with normal circulation, recovery is rapid. Cochlear symptoms often only resolve incompletely. Irreversible vestibular defects are compensated centrally (see p. 20). Cochleovestibular symptoms, especially dizziness, often progress in elderly patients.

Direct Injuries to the Tympanic Membrane and Middle and Internal Ear

Clinical Features. *Injury to the tympanic membrane:* This is accompanied by momentary pain, slight bleeding from the ear, and slight hearing loss.

Middle ear injuries: There is profuse bleeding, pain and hearing loss, a pulsating sound in the ear, and occasionally facial paralysis.

Inner ear injuries: There is immediate tinnitus, hearing loss, dizziness, nausea, and vomiting.

Pathogenesis. Damage to the tympanic membrane and to the middle and internal ear can be caused by slapping injury or by the introduction of pointed objects such as matchsticks, toothpicks, knitting needles, hairpins, and twigs into the ear; and by careless removal of foreign bodies, by occupational injuries (hot cinders, welding sparks), acid burns, or gunshot wounds.

Diagnosis. Otoscopic findings include a tympanic membrane perforation with jagged, frayed, blood-streaked, and occasionally rolled-in edges (see Fig. 1.105). Blood is found in the external auditory meatus, mixed with perilymph, in injuries to the inner ear. *Conductive deafness* is found in middle ear injuries, *sensorineural deafness,* or mixed deafness if the inner ear is involved, and in severe lesions there is *complete deafness* and spontaneous nystagmus.

Immediate total peripheral *facial nerve paralysis* is found in fractures involving the bony tympanic segment of the facial nerve canal (see Fig. 1.22). CT can reveal a retained bullet or a bullet track.

Treatment. In simple tympanic membrane rupture, the fragments of the membrane are repositioned and splinted aseptically using a thin Silastic or plaster sheet under the operating microscope. Systemic prophylactic antibiotics are administered.

Caution: Nonsterile instruments should not be used in the ear, and the ear should not be syringed.

In *combined injuries* of the tympanic membrane, middle ear, and inner ear, immediate exploration of the middle ear cavity and labyrinthine capsule is carried out with the operating microscope (tympanoplasty). Any perilymph leakage—e.g., from the round window—is stopped by closing the labyrinthine fistula. Prophylactic antibiotics are administered. Simultaneous facial nerve paralysis is treated by surgical decompression of the facial nerve if a disruption of the bony facial nerve canal is detectable. Otherwise, steroids and expectant management are recommended (see p. 112 and Fig. 1.115).

Course and prognosis. Simple injuries to the tympanic membrane and middle ear usually heal smoothly and without functional deficit if they are treated correctly by an otologist. Involvement of the labyrinth is usually followed by irreversible cochleovestibular failure. The prognosis for the facial paralysis is very good if continuity of the nerve has been preserved, but after neurotmesis there is irreversible paralysis if surgery is not undertaken.

Barotrauma

Clinical Features. Acute pain, pulsating tinnitus, hearing loss, occasionally vertigo and disturbance of balance.

Pathogenesis. Sudden changes in air pressure, producing an absolute or relative reduction of pressure in the middle ear, cause bleeding into the middle ear mucosa and into the tympanic membrane, and on occasion even rupture of the tympanic membrane and of the round window membrane. This may occur after rapid decompression or recompression from a low-pressure or high-pressure chamber, a rapid dive from a great height in a non-pressurized aircraft, or after surfacing too quickly from deep-sea diving.

The condition is caused by a sudden closure of the tube, which is compressed by the rapid rise in atmospheric pressure or by the associated increase in tissue pressure. After ≈ 2 h of closure of the tube, the Valsalva maneuver and politzerization are ineffective, as mucosal edema and serous–hemorrhagic exudate in the middle ear cavity have occurred due to the reduced pressure in the middle ear. The disorder is called *aerotitis* or *barotitis*.

Diagnosis. This is based on the history. The otoscopic findings show retraction of the tympanic membrane, occasionally subepithelial hemorrhage in the pars tensa, transudate behind the tympanic membrane, or a hemotympanum. Audiological tests show conductive hearing loss.

Treatment. This includes decongestant nose drops, possibly paracentesis, analgesics, and oral anti-inflammatory agents. Prophylaxis is important, and the patient should avoid flying or diving during inflammation of the nasopharynx, nose, and paranasal sinuses. Anatomic deformities in the nose and nasopharynx obstructing nasal respiration and favoring the development of inflammatory diseases of the eustachian tube should be dealt with. These diseases include septal deformity, hypertrophy of the turbinates, and adenoidal hypertrophy. Immediate tympanotomy should be performed for barotrauma with severe sensorineural hearing loss, to allow assessment of the round window so that a possible perilymph fistula due to rupture of the round window membrane can be closed.

Caisson Disease and Diving Accidents

These accidents occur in people who work underwater at depths where the pressure is several times that of atmospheric pressure. They can also occur in amateur sports divers who surface too quickly from too great a depth.

Clinical Features

- Dizziness, vomiting, headache.
- Severe tinnitus and rapidly progressive hearing loss.
- The above symptoms have a latent period of minutes to hours.
- Disorders of vision, ataxia, and clouding of consciousness in severe cases.

Pathogenesis. When working at several times atmospheric pressure, either in a caisson or when diving to depths greater than 10 m, a considerable quantity of air, including relatively insoluble nitrogen, is taken into solution. Gaseous nitrogen is released into the blood if the patient decompresses too quickly from the caisson or surfaces too rapidly from depths greater than 10 m. This can lead to the formation of small gas emboli within the cerebral end arteries. These cause deficits in the area of supply of the cerebral vessels, including that of the inner ear. This explains the symptoms described above.

Diagnosis. Previous history of an accident. Sensorineural hearing loss, spontaneous and provocation vestibular nystagmus, and in serious cases ataxia and neurologic deficits.

Treatment. Hyperbaric oxygen is given.

Course and prognosis. This depends on the degree of severity of the gas emboli and on the time taken to institute treatment. In serious cases, the patient may become blind or deaf, suffer from balance disorders, may be paralyzed, or may even die.

Acute Acoustic Trauma

> Note: There is an important basic difference between explosion and gunfire injuries. The physical-acoustic properties of an explosion are qualitatively identical to those of gunfire, but are completely different quantitatively. In an explosion, there is a high-pressure wave, but the shock wave lasts more than 1.5 ms, whereas with gunfire the peak of the pressure wave lasts less than 1.5 ms.

Clinical Features. *Blast trauma:* There is marked, persistent earache, occasionally with bleeding from the affected ear, deafness, and tinnitus.

Gunshot trauma: There is a short stabbing pain in the ear, marked continuous tinnitus, and deafness.

Pathogenesis. In both explosion and gunshot trauma, the causes are partly direct and mechanical, due to bleeding, and partly indirect, with a metabolic effect on the microcirculation causing partially reversible damage to the sensory cells of the organ of Corti. The severity and site of the lesion in the cochlea directly depend on the sound pressure level of the acoustic energy and its maximum frequency. In explosion trauma, ruptures of the tympanic membrane and other middle ear lesions often occur.

Diagnosis. Only *explosion injury* causes abnormal otoscopic findings; in addition, the audiogram shows sensorineural or mixed hearing loss. In *gunshot trauma,* there is a notch at 4000 Hz, or a high-tone loss, and positive recruitment.

Treatment. Intravenous infusion of low-molecular-weight dextran and a vasodilator such as pentoxifylline within 24 h of the trauma if possible. Tympanoplasty has to be performed in cases of visible middle ear injuries such as tympanic membrane perforation and defects in the ossicular chain.

Course and prognosis. Traumatic *middle ear lesions* usually heal without complications after surgical repair. The prognosis is good.

Inner ear lesions are partially reversible, but in some patients there is continuing degeneration of sensory cells and secondary increased degeneration of the peripheral neurons.

Chronic Noise Trauma

In contrast to acute acoustic trauma, this condition is the result of damage to the inner ear caused by weaker, but more prolonged, noise. Chronic noise-induced hearing loss is now a considerable hazard in the younger population, as they are exposed to loud music and other noises associated with leisure activities. The severity of the lesion therefore depends not only on the sound pressure peaks of the noise (noise level), but also on the exposure time and on the individual patient's sensitivity to the effect of noise. Exposure to levels below 85 dB (A) for 8 h/day is considered safe. Emotional factors also play an important role and produce autonomic symptoms that have deleterious effects on the entire body.

Clinical Features. *Subjectively,* there is a sense of pressure in the ears and in the head, a feeling of hearing loss, generalized tiredness and lack of concentration, and often tinnitus. The subjective symptoms are often reversible, since the patient becomes used to the noise. Few patients are aware of the developing deafness in the early phases.

Objectively, a pure-tone audiogram of chronic noise trauma initially shows a notch at 4000 Hz, typically in both ears. Later, the threshold for the lower frequencies rises, and finally the deafness spreads to the speech frequencies. Further loss of hearing follows as a result of the physiologic aging process. In the early phases of its development, chronic noise deafness shows a certain tendency toward recovery when the patient is no longer exposed, but with increasing exposure this tendency declines.

Pathogenesis. Depending on the intensity and duration of exposure, the ear may react to sounds in one of the following ways:

1. A physiologic *adaptation of threshold* may develop.
2. After more prolonged exposure, the ear may react with *fatigue* or with the appearance of a temporary threshold shift (TTS), which can be related directly to acoustic damage that is proportional to the exposure time and has a linear relationship with the sound intensity. The "physiologic" TTS usually recovers within minutes and at the most 2 h after the end of exposure to the noise.
3. A *permanent threshold shift* (PTS) may develop, which is an expression of pathologic fatigue and irreversible damage to the hearing organ. This is caused by metabolic decompensation of the sensory cells due to a disturbance of the balance between supply and demand in energy metabolism. This is caused by increased oxygen consumption or by decreased supply during permanent intensive acoustic exposure. The outer hair cells degenerate first, and the inner cells last. Not all individuals are affected equally. Some highly susceptible people lose their hearing ability faster than others. Intracellular conditions presumably have an influence on this—e. g., the adenosine triphosphate (ATP) level. In a person who is constantly exposed to noise, it is not possible to determine whether or not any hearing loss is due to the noise.

Diagnosis. The hearing loss is of long standing, and a social history reveals occupational exposure or lifelong social habits that are responsible. A pure-tone speech audiogram is important.

Differential diagnosis

- Endogenous heredodegenerative sensorineural deafness in which there is a positive family history.
- Infective and toxic damage to the inner ear and vestibulocochlear nerve, particularly by ototoxic antibiotics.
- Progressive sensorineural deafness in severe generalized diseases such as diabetes, chronic nephritis, and hypertension.

Note: The limits of noise causing damage to the ear are as follows:

- Equivalent continuous sound pressure (Leq) in the range of 85–90 ± 2.5 dB and higher must be regarded as damaging to the ear.
- Single sound impulses exceeding a peak of 135 dB also damage the ear.

Treatment. No active methods of treatment for dealing with the cause are available. If hearing in social situations becomes inadequate, a hearing aid should be prescribed. Prophylaxis, including the provision of hearing protectors, is very important.

Protection of hearing:

- Elimination or reduction of noise through technical improvements to machinery.
- Protection of personnel from noise using ear protectors.
- Limitation of the time of exposure to noise and frequent rest periods.
- Medical prophylaxis against damage to hearing, prescribed by occupational medical health staff.

Note: A cotton wool plug does not protect against noise trauma to the ear.

Course and prognosis. The disease progresses to advanced hearing loss and tinnitus, which may decompensate.

Tumors of the Middle and Internal Ear, Vestibulocochlear Nerve, and Facial Nerve

Nonchromaffin Paraganglioma (Glomus Tumor)

This is the most frequent true tumor of the middle ear. It develops from neuroectoderm. The structure

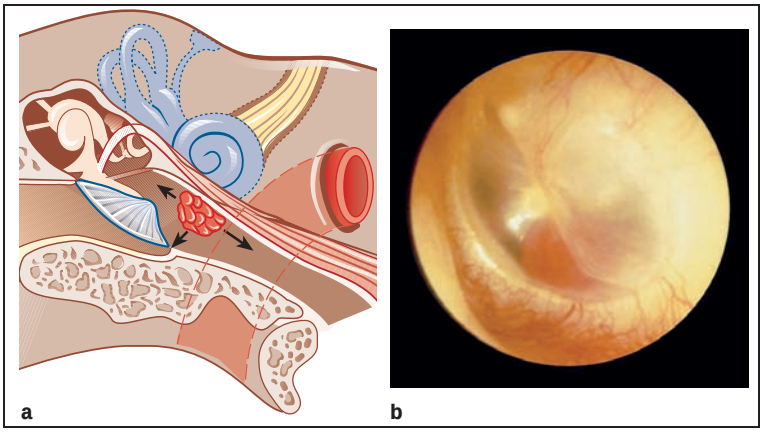


Fig. 1.106a, b Glomus tympanicum tumor.
a The arrows mark the possible direction of tumor growth.
b A small red tumor becomes visible.

of the tumor is similar to that of the chemoreceptor tissue of the carotid body. It is characterized by extensive vascularity. Because of its structural and functional relationship to the carotid body, this tumor is also considered in the section on chemodectomas (see p. 397).

Clinical Features. These are very variable, depending on the site of origin and extent of the tumor, and include:

- Unilateral tinnitus, synchronous with the pulse.
- Unilateral hearing loss and feeling of pressure in the ear.
- Disorders of balance.
- Lesions of the lower cranial nerves causing facial paralysis, paralysis of the soft palate, hoarseness, disorders of swallowing, and paralysis of the tongue in the late stage.

Pathogenesis. The tumor develops from nests of epithelial cells surrounded by a high vascular stroma. The sites of predilection are the bulb of the internal jugular vein, the tympanic plexus, and the lesser superficial petrosal nerve. They can be grouped by site and extent as follows:

1. Glomus tympanicum tumors limited to the middle ear cavity (Fig. 1.106a).
2. Glomus jugulare tumors limited to the middle ear cavity and the bulb of the jugular vein, without destruction of bone (Fig. 1.106b).
3. Glomus jugulare tumors destroying bone and invading the whole of the mastoid as far as the petrous apex.
4. Glomus jugulare tumors with intracranial extension.

Diagnosis. Otoscopy shows the tumor shining through the tympanic membrane. The tumor is often red. If the tumor breaks through into the external meatus, a polyp that bleeds easily can be seen (Fig. 1.107a, b).

The hearing loss is conductive in the early stage, but later becomes sensorineural, and finally the patient may be completely deaf if the tumor erodes into the labyrinth.

The hypoglossal, glossopharyngeal, or vagus nerves may be paralyzed in tumors arising primarily from the bulb of the jugular vein (the jugular foramen syndrome).

Intracranial extension is accompanied by pontine and cerebellar syndromes, in addition to facial paralysis, trigeminal hypoesthesia, deafness, and vestibular symptoms.

Supplementary investigations include CT scans with contrast enhancement, to allow three-dimensional comparison of the two temporal bones and the jugular foramen and jugular bulb area. The tumor extension is best evaluated using MRI, magnetic resonance angiography, and subtraction angiography (which allows embolization of the main afferent vessels in selected patients during the same session).

Treatment. Tumors in groups 1 and 2 (see above) can be removed easily and radically with otologic surgery. Tumors in group 3 require a combined cervicotemporal approach, and those in group 4 a two-phase neurotologic procedure designed to remove the tumor completely while preserving the facial nerve.

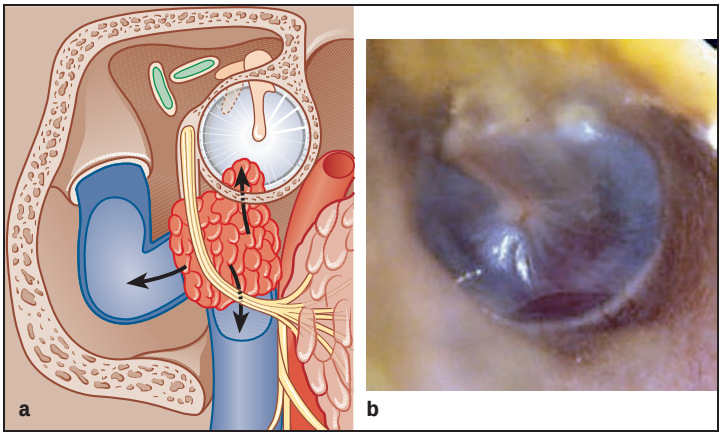


Fig. 1.107a, b Glomus jugulare tumor.
a The arrows mark the possible direction of tumor growth into the epitympanum, the hypotympanum, and through the tympanic membrane.
b There is a blue-colored tumor shining through the tympanic membrane.

Preoperative angiographic embolization of the larger and tumor-feeding vessels reduces intraoperative blood loss. In group 4 tumors, tumor control using radiotherapy has been proposed as an alternative form of treatment.

Course and prognosis. The tumor grows slowly. In advanced stages with extensive intracranial invasion, life-threatening situations occur due to compression of the brain stem or thrombosis of the carotid artery.

Middle Ear Carcinoma

In most patients, this involves a keratinizing squamous cell carcinoma, arising at the junction between the external auditory meatus and the tympanic membrane, which has penetrated into the middle ear.

Adenocarcinomas and adenoid cystic carcinomas arising primarily from the middle ear mucosa are very rare, as are sarcomas.

Clinical Features

- Neuralgic pain around the ear.
- Blood-stained otorrhea, which is very fetid.
- Progressive hearing loss.
- Occasionally, dizziness, disorders of balance, facial nerve paralysis, and intense headache if the tumor is infiltrating into the dura.

Diagnosis

- Otoscopy shows a readily bleeding aural polyp (Fig. 1.108).
- Destruction of the tympanic membrane by hemorrhagic granulations.
- Destruction of the posterior meatal wall.

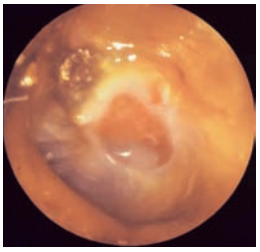


Fig. 1.108 Middle ear carcinoma.

- Peripheral facial paralysis, enlargement of the regional lymph nodes, conductive hearing loss, combined conductive and sensorineural hearing loss or even deafness, spontaneous vestibular nystagmus to the healthy side, depending on the extent of the tumor.
- Tumor expansion into the parotid gland and regional lymph-node metastases.
- CT scans show extensive destruction of the temporal bone arising from the middle ear cavity and the external auditory meatus.

Differential diagnosis

- Chronic otitis media with cholesteatoma.
- Necrotizing otitis externa.

Treatment

- Radical expansive operation with subtotal petrosectomy, parotidectomy, neck dissection, and reconstruction of the facial nerve.
- Postoperative radiotherapy.

Course and prognosis. Despite combined surgery and radiotherapy, the prognosis is poor due to early expansion and the lesion's tendency to metastasize.

Table 1.12 Subjective symptoms of vestibular schwannoma

Focal symptoms
• Tinnitus (incidence 70%)
• Unilateral progressive deafness (45%)
• Sudden hearing loss (40%)
• Fluctuating deafness (10%)
• Dizziness (30%)
Associated symptoms
• Unilateral facial nerve spasm or paralysis
• Double vision
• Ataxia
• Clumsiness on moving the arms
• Unilateral disturbances of sensation of the face
Symptoms of increased cranial pressure
• Occipital headache
• Projectile vomiting
• Decrease in vision and papilledema
• Personality changes

Vestibular Schwannoma (Acoustic Neuroma)

Clinical Features. See Table 1.12.

Pathogenesis. This is a histologically benign tumor arising from the Schwann cells of the neurilemma. It usually arises in the transitional zone between the neuroglia and the neurilemma of the pars superior of the vestibular nerve. The tumor is a generally slow-growing one and may also grow inside the labyrinth or arise from the cochlear nerve, but only in rare cases.

Depending on their site of origin, these tumors may be divided into:

- *Lateral vestibular schwannomas,* which lie in the internal auditory meatus and exclusively cause localized symptoms; and *mediolateral vestibular schwannomas* in the region of the porus, lying partly in the internal meatus and partly in the cerebellopontine angle and causing both localized symptoms and symptoms due to impairment of neighboring organs.
- *Medial vestibular schwannomas,* which arise in the cerebellopontine angle and cause slight symptoms in cranial nerve VIII, but marked symptoms due to involvement of the adjacent cranial nerves, brain stem, and cerebellum, and finally the symptoms of increased intracranial pressure.

Three stages can be distinguished, depending on the size of the tumor (Fig. 1.109a-c):

1. Small intrameatal tumors with a diameter of 1–8 mm, which cause focal symptoms.
2. Medium-sized tumors with a diameter of up to 2.5 cm, with intrameatal and intracranial extension, which cause focal symptoms and slight symptoms caused by involvement of neighboring neural structures.
3. Large tumors more than 2.5 cm in diameter, causing focal symptoms, symptoms due to involvement of neighboring neural structures, and symptoms of increased intracranial pressure, depending on their size.

Diagnosis. See Table 1.13. Diagnostic investigations include:

- Pure-tone and speech audiometry, stapedial reflex.
- Auditory evoked potentials.
- Vestibular tests with nystagmography.
- MRI with Gd-DTPA enhancement.

Differential diagnosis

- Ménière disease (see p. 97).
- Sudden sensorineural hearing loss (see p. 101).
- Primary congenital cholesteatoma of the cerebellopontine angle.
- Vascular compression syndrome.
- Secondary acquired occult middle ear cholesteatoma with perilabyrinthine extension and rupture into the internal auditory meatus (see p. 68).
- Meningioma and facial nerve neuroma.
- Congenital syphilis causing vascular cochleovestibular symptoms (see p. 81).

Treatment. Intrameatal tumors (stage 1) can be removed using an extradural transtemporal approach through the middle cranial fossa (Fig. 1.110a). In asymptomatic patients with good hearing, a "wait-and-scan" approach with regular follow-ups can be considered. Stereotactic radiosurgery (with the gamma knife) is a possible treatment. The indication depends on the quality of hearing and the patient's age and wishes.

Medium-sized tumors (stage 2) are removed via a translabyrinthine (Fig. 1.110b) or retrosigmoid route (Fig. 1.110c). The mortality is zero, but 85% of patients become totally deaf. Facial nerve function, however, can be preserved in 85% of patients. Large tumors (stage 3) are treated using a retrosigmoid or suboccipital neurosurgical approach.

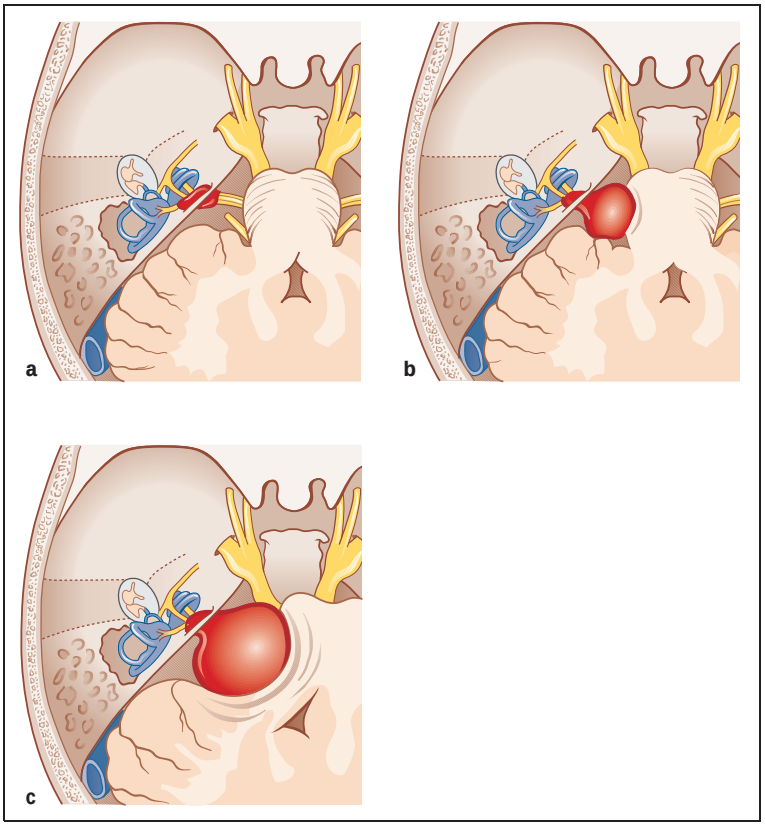


Fig. 1.109a–c The three stages of a vestibular schwannoma.
a Intrameatal tumor.
b Intrameatal and extrameatal tumor.
c A mainly extrameatal medial tumor compressing the brain stem and the cerebellum.

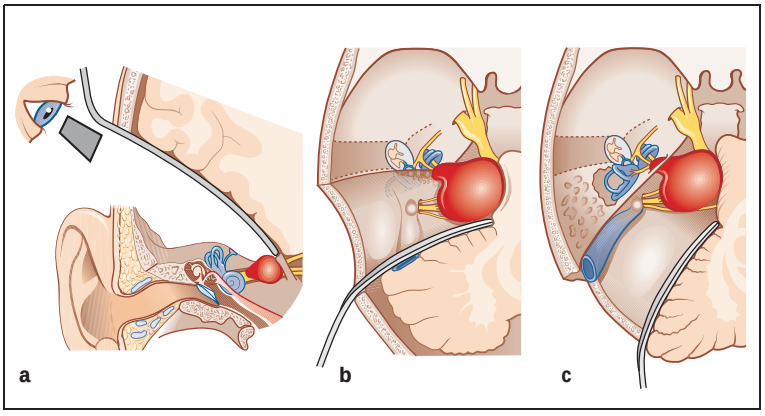


Fig 1.110a–c Otologic and neurosurgical routes of access to the internal meatus.
a The transtemporal approach through the middle cranial fossa.
b, c The translabyrinthine and retrosigmoid approaches.

Table 1.13 Objective symptoms of vestibular schwannoma

Focal symptoms
• Retrocochlear sensorineural hearing loss, with negative recruitment, pathologic hearing fatigue: abnormal SISI test, abnormal decay on Békésy audiogram, and discrepancy between pure-tone and speech audiogram. Stapedial reflex absent. Pathologic ABR and electrocochleography • Vestibular symptoms include spontaneous nystagmus to the healthy ear and loss of caloric labyrinthine responses
Associated symptoms
• Peripheral, usually slight, facial paralysis, neurodiagnostic axonotmesis and positive Hitselberger sign • Abducens paralysis • Loss of the corneal reflex • Hypoesthesia of the trigeminal nerve • Occasionally, palatal paralysis
Symptoms due to brain stem compression
Disorders of oculomotor nerves. In stage III tumors, pontine compression causes the following: • Gaze-evoked nystagmus to the side of the tumor • Direction-changing irregular positional nystagmus • Abnormal or absent optokinetic nystagmus
Cerebellar symptoms
In stage III, often include dysdiadochokinesia and ataxia
Symptoms of increased intracranial pressure
Papilledema and projectile vomiting

ABR, auditory brain stem response; SISI, short increment sensitivity index.

Note: The possibility of a vestibular schwannoma must always be suspected in all patients with unilateral progressive sensorineural hearing loss or unilateral recurrent loss of hearing, and a thorough neuro-otologic examination and neuroradiologic examination (including MRI and evoked response audiometry) must be performed.

Note: In patients with von Recklinghausen neurofibromatosis (NF2), one or more neuromas of the auditory or facial nerve may be suspected.

Facial Nerve Neuroma

Clinical Features. The tumor causes slowly progressive facial paralysis or hemifacial spasm. Depending on its site, the tumor may cause symptoms similar to those of acoustic neuroma (vestibular schwannoma).

Bony Tumors

These rare tumors include:

- Osteoma.
- Giant cell tumor.
- Solitary plasmacytoma.
- Histiocytosis (reticuloendotheliosis; Letterer–Siwe disease, Hand–Schüller–Christian disease, eosinophilic granuloma).

Congenital Anomalies of the Middle and Internal Ear

Simultaneous congenital anomalies of the middle and internal ear are rare, but those affecting the middle and external ear together are common, occurring in one in 10 000 normal births.

Combined Anomalies of the External and Middle Ear

Clinical Features

- Dysplasia.
- Microtia.
- Meatal atresia.
- Facial deformities.
- Deafness.

Pathogenesis. The following factors may be responsible for these anomalies:

- Genetic factors, chromosomal abnormalities, or mutation of genes.
- Exogenous factors, including hypoxia, radiation, ultrasound, and thalidomide.
- Combined exogenous and genetic factors, causing polygenic multifactorial anomalies.

The developmental process (described on p. 1) is restricted or disturbed by these factors.

Diagnosis

- Inspection.
- High-resolution CT scan.
- Auditory evoked potentials.
- Vestibular tests.

Treatment. Surgery is performed or a hearing aid is prescribed.

Purpose of treatment:

1. Construction of an esthetically satisfactory auricle using rib cartilage, followed by creation of an external meatus in a second procedure.
2. Improvement of hearing, either by reconstructing the sound conducting apparatus with tympanoplasty (see Fig. 1.96 d, f) or by prescribing a hearing aid.

Timing of the operation:

A patient with bilateral meatal atresia with a conductive hearing loss of 50 dB should undergo surgery on one ear in the third to fourth year of life. However, it is preferable to reconstruct the external auditory meatus and to carry out a tympanoplasty after pneumatization is complete and when the auricle has reached its final size—i. e., after the eighth year of life. Otoplasty is performed after the fifth year of life.

A bone conduction hearing aid is prescribed for patients with bilateral atresia as early as possible—i. e., in the second half of the first year of life—to allow speech to develop normally.

Congenital Anomalies of the Internal Ear

These are rare and are summarized in Table 1.14.

Table 1.14 Congenital anomalies of the internal ear

Mondini syndrome	Isolated dysplasia of the cochlea
Congenital CSF otorrhea	Due to a fistula of the oval window with a patent cochlear aqueduct (gusher syndrome)
Thalidomide embryopathy	A labyrinthine dysplasia with aplasia of the petrous pyramid
Rubella embryopathy	Labyrinthine anomalies with dysplasia of the middle ear

CSF, cerebrospinal fluid.

Clinical Aspects of Cochleovestibular Disorders

Toxic Damage to the Hearing and Balance Apparatus

Exogenous ototoxins include drugs such as aminoglycosides, cytostatics and diuretics, as well as industrial products, tobacco, and alcohol.

Endogenous ototoxins include bacterial toxins and the toxic metabolites that arise in metabolic disorders such as diabetes and renal disease.

Clinical Features

- Tinnitus is usually the first symptom.
- Progressive bilateral sensorineural hearing loss, initially for high tones, but later from high to middle and lower frequencies.
- Positional vertigo with nausea.
- Balance disorders with persistent vertigo and unsteadiness of gait.
- Oscillopsia—i. e., a weakness of fixation

Ototoxic Drugs

Aminoglycoside Antibiotics

Pathogenesis. Aminoglycoside antibiotics are retained for a longer period and in higher concentrations in inner ear fluids than in other body tissues and fluids. Ototoxic aminoglycosides destroy both cochlear and vestibular sensory cells; streptomycin is mainly vestibulotoxic, whereas dihydrostreptomycin is ototoxic. Neomycin and kanamycin are strongly ototoxic, and gentamicin is both ototoxic and vestibulotoxic.

> Note: The development of inner ear damage depends on the dose and the half-life of the ototoxic antibiotic and on the patient's renal function.

Treatment

- Immediate cessation of aminoglycoside administration.
- Intravenous infusion of low-molecular-weight dextran solutions.
- Intravenous infusion of a vasodilator and systemic corticosteroids.

Course. The disease usually progresses for up to 6 months after the antibiotics are stopped; the prognosis therefore has to be guarded, as the inner ear damage is largely irreversible.

Cytostatics
Cisplatin and cyclophosphamide produce a direct toxic effect on the hair cells of the cochlea, while cisplatin also has neurotoxic effects. Other platinum-based cytostatic drugs, such as carboplatin, appear to be less ototoxic, but hearing loss may still occur.

Note: Any patient being treated with aminoglycosides or cytostatic drugs must have the following investigations: renal function tests and an audiogram once or twice a week during the course of treatment.

Diuretics
Diuretics, including furosemide and ethacrynic acid, can also damage the inner ear. The extent of the lesion varies, and it affects mainly the outer hair cells, due to disturbance of the regulation of ion concentration caused by a lesion in the stria vascularis. These lesions are reversible in ≈ 90 % of patients.

Other Ototoxic Antibiotics
Metabolic damage to the cochlear hair cells can be caused by quinine and salicylate. The initially reversible effect can become irreversible in overdosed, long-term treatment.

Ototoxic Occupational Toxins

- Arsenic compounds.
- Mercury salts.
- Lead salts.
- Organic phosphate compounds.
- Sulfur and tetrachlorocarbon compounds.
- Carbon monoxide.
- Benzol, nitrobenzol, and aniline.

Inflammatory Lesions of the Hearing and Balance Apparatus

Herpes Zoster Oticus (Ramsay Hunt Syndrome)
The most common site of zoster infection in the head and neck, after herpes zoster ophthalmicus, is that affecting the ear. Herpes zoster oticus can occur at any age, but it mainly does so between 40 and 60 years of age.

Clinical Features
- Reduced general condition and/or subfebrile temperature.
- Erythema and vesicles on the auricle and the external meatus (see Fig. 1.70).
- Regional lymphadenitis (discrete).
- Severe neuralgic pain.
- Peripheral facial paralysis in 60–90 % of patients.
- Severe sensorineural (retrocochlear) hearing loss in 40 % of patients.
- Vertigo and balance disorders in 40 % of patients, with release nystagmus to the healthy side.

Pathogenesis. This disorder is due to a viral infection. The portal of entry is unknown. The virus may spread via the bloodstream to the CSF and meninges, causing encephalomyelomeningitis with neuritis of the spiral or vestibular ganglion.

Diagnosis
- Inspection of the auricle and otoscopy.
- Audiogram and impedance test.
- Vestibular tests.
- Tests of facial nerve function.
- Schirmer test.
- Testing of glossopharyngeal and vagus nerve function.
- Viral serology.
- Lumbar puncture with CSF examination (for diagnosis of serous meningitis).

Differential diagnosis
- Myringitis bullosa.
- Idiopathic facial paralysis.

Treatment
- Antiviral treatment with aciclovir or famciclovir (antiviral agents that inhibit DNA synthesis of type I and II herpes simplex virus, as well as varicella-zoster virus).
- Gammaglobulin.
- Antibiotics (to avoid superinfection).
- Local dermal treatment with lotions or ointment.

Course and prognosis. Complete recovery is possible within 4 weeks, but recovery is often only partial in cases of facial nerve paralysis. The cochleovestibular loss is usually irreversible.

Other Viral Infections

Influenza, measles, adenovirus, chickenpox, coxsackievirus, and mumps viruses often cause a vestibular neuritis, with corresponding symptoms.

The virus of *epidemic parotitis* (mumps) has a particular affinity for the cochlea and usually causes unilateral serous labyrinthitis, destruction of the hair cells, or degeneration of the organ of Corti. Neurolabyrinthitis with destruction of the spiral ganglion may occur. The vestibular part of the labyrinth is almost never attacked by the mumps virus.

Note: Infection with the mumps virus is the most frequent cause of unilateral complete deafness in young children.

Course. The course is usually mild or abortive with respect to the primary disease.

Prognosis. This is poor, with irreversible damage.

Serous Labyrinthitis

This condition is a toxic or virally determined abacterial serous inflammation of the perilymphatic and endolymphatic spaces, with partial or total destruction of cochlear and vestibular sensory cells. The loss of cochlear and vestibular function is usually irreversible.

Immunologic Diseases of the Inner Ear

Recognized autoimmune syndromes can lead to inner ear disease. Rapid bilateral and often asymmetrical cochlear hearing loss, occasionally with a fluctuating course, is typical. There may be accompanying vestibular symptoms.

- Cogan syndrome.
- Wegener granulomatosis.
- Recurrent polychondritis.
- Primary autoimmune disease.

Trauma

(See pp. 84–89).

Vestibular Disorders

Ménière Disease

Clinical Features. The classic triad originally described by Ménière in 1861 consists of attacks of:

- Tinnitus—continuous low frequency with varying intensity (Table 1.15).
- Fluctuating hearing loss and dysacusis.
- Attacks of rotatory vertigo.

The typical attack begins acutely and consists of the three symptoms listed above, followed by nausea, vomiting, and other autonomic symptoms. Patients feel sensations of aural fullness. The tinnitus and hearing loss may change before and during the attack. Characteristically, the tinnitus improves, while hearing becomes poorer. Mostly, hearing improves again following the attack. A marked improvement in hearing during the attack itself is called *Lermoyez phenomenon*.

The symptoms are usually unilateral. In the disease-free intervals, hearing often returns to normal and the tinnitus disappears, at least in the early phases of the disease. In the later stages, there is

Table 1.15 Classification of tinnitus

Subjective tinnitus	
Conductive tinnitus	Obstruction of the ear canal
	Middle ear disease
Sensorineural tinnitus	Cochlear damage
	Cochlear nerve disturbance
Central tinnitus	Damage to the central auditory pathway
Objective tinnitus	
Vascular tinnitus	Vascular malformations
	Arteriovenous fistulas
	Paragangliomas
Myogenic tinnitus	Velopharyngeal myoclonus
	Middle ear myoclonus

fluctuating hearing loss for low tones. In the end stage, hearing loss is unilateral, severe, and pancochlear. The patient also reports persistent tinnitus. Initially, the cochlear symptoms predominate.

There is no predisposing factor, apart from psychological stress.

Pathogenesis. The disease is caused by a disturbance of the *quantitative* relation between the volume of the perilymph and endolymph. In addition, a disorder of the *qualitative* electrolyte composition of the two fluids causes abnormal osmotic pressure regulation within the membranous labyrinth. This causes endolymphatic hydrops.

The primary cause of this increase of pressure within the endolymphatic space is a disorder of resorption of the potassium-rich endolymph. This leads to an increase in osmotic pressure. When this exceeds a certain level, the Reissner membrane lying between the endolymphatic and perilymphatic spaces ruptures, allowing the endolymph and perilymph to mix. Sites of predilection for the rupture are the helicotrema, the basal turn of the cochlea and the utricle, and part of the saccule lying opposite the ampulla of the semicircular canals. This explains the genesis of the cochleovestibular symptoms in the classic Ménière attack.

Rupture of the hydropic labyrinth and the resulting mixing of the potassium-rich endolymph with perilymph, which is normally low in potassium, leads to a considerable increase in potassium content in the intercellular spaces of the perilymphatic network, in which the afferent neurons of the acoustic and vestibular nerves run. These are paralyzed by depolarization due to the increase in potassium, thus causing the symptoms of cochleovestibular failure. The process may last from a few minutes to several hours and is reversible in the early phases of the disease—which explains the clinical recovery, especially the fluctuating hearing loss.

Psychological factors such as stress can act as a trigger mechanism for an attack, but are not primarily responsible for the development of the disease.

Diagnosis. See Table 1.16.

Treatment. *During an attack:*
- Bed rest.
- Intravenous infusion of fluid and electrolytes for prolonged vomiting.
- Intravenous antivertigo and antiemetic drugs.
- Low-molecular-weight dextran and systemic vasodilator infusions to improve the labyrinthine circulation and to increase the flow of perilymph and endolymph.
- Psychotropic drugs should not be given during an attack because of the danger of central disinhibition of the vestibular system, causing worsening of the symptoms.

In the symptom-free interval:
- Antivertigo antihistamine (betahistine).
- Thiazide diuretics.
- Psychological support, if necessary psychotropic drugs.

When there is a protracted course, with repeated attacks that disable the patient:
- Destruction of the vestibular end organ while retaining social hearing: intratympanic application of gentamicin to the round window, which selectively destroys the sensory cells of the vestibular end organ.
- Neurectomy of the vestibular nerve via a transtemporal approach to the internal meatus.
- Surgical exposure and drainage of the endolymphatic sac (saccotomy; the operation is based on the hypothesis of hydrops decompression).

In patients with poor hearing and marked tinnitus:
- *Labyrinthectomy*: Surgical ablation of the severely damaged inner ear using transtympanic or translabyrinthine exposure of the internal meatus, with resection of the intrameatal portion of the vestibulocochlear nerve. This operation produces complete deafness and is only indicated in selected cases when other alternatives are not adequate.

Course. One of the characteristics of Ménière disease is that its course is unpredictable. At one end of the spectrum, there are *abortive forms,* which heal after a few attacks without permanent deafness. At the other end, the disease may progress episodically over several years with symptom-free intervals of varying duration. However, over the course of time, the fluctuating hearing loss becomes irreversible. In addition, there is also an *acute form,* in which several attacks lead rapidly to almost complete deafness and to severe disability due to the disturbance of balance. In these cases, elimination of the diseased vestibular part of the inner ear by transtympanic application of gentamicin, or by one of the surgical routes described above, is indicated.

Table 1.16 Synopsis of the most important disorders of balance and hearing

Diagnosis	Subjective symptoms	Audiogram	Vestibular symptoms	Associated neurologic signs	Other clinical signs
Acute unilateral vestibular paralysis	Acute rotatory dizziness and prolonged dizziness, possibly positional. Nausea, vomiting, no loss of consciousness, normal hearing and no tinnitus	Normal	Spontaneous nystagmus to the sound side, possibly positional nystagmus, no response to caloric stimulation, ataxia. Galvanic tests are usually abnormal, with an increased threshold	None	Often diabetes, acute infection by viruses or toxoplasmosis, or hypertension or hypotension, immune disorder
Ménière disease	Episodic vertigo, unilateral tinnitus, unilateral hearing loss, and fluctuating hearing loss	Unilateral fluctuating sensorineural hearing loss with positive recruitment, and often a flat or low tone curve	Spontaneous nystagmus to the healthy side after the attack, reduced caloric response	None	None
Vestibular schwannoma	Progressive unilateral hearing loss, progressive disorder of balance, occipital headache, occasionally also recurrent hearing loss with partial remission, tinnitus	Unilateral retrocochlear sensorineural hearing loss, at first for high tones. Stapedial reflex lost. Absent recruitment and marked loss of discrimination for speech and with a discrepancy between the pure-tone and speech audiograms, and pathological ABR	Spontaneous nystagmus to the healthy side, possibly gaze-evoked nystagmus to the diseased side, positional nystagmus, abnormal caloric responses, possibly abnormal optokinetic responses in the presence of compression of the brain stem and cerebellum	Associated symptoms, absent corneal reflex, abnormal glabellar tap, facial weakness, and positive Hitselberger sign, papilledema, and abducens paralysis in large tumors	None

ABR, auditory brain stem response.

Bilateral disease is relatively uncommon, occurring in only 10 % of patients.

Acute Vestibular Loss (Vestibular Neuronitis, Vestibular Neuritis)

Clinical Features. See Table 1.17.

Pathogenesis. The site of the pathophysiological lesion (labyrinth or vestibular nerve) is not known. The cause may be a disturbance of the microcirculation due to an infection (e. g., neurotropic virus or other agents such as *Rickettsia,* or protozoa such as *Toxoplasma gondii*), an autoimmune disease, or a metabolic disorder.

This disorder has some similarities with sudden sensorineural hearing loss in its course, unilateral involvement, and acute onset.

Diagnosis. See Table 1.16.

Differential diagnosis. See Tables 1.16, 1.17.

Treatment. *Early acute phase:*

- Bed rest.
- Symptomatic treatment with antivertigo drugs and sedatives.
- Intravenous rheological drugs.
- Antibiotics, when there is objective evidence of bacterial infection.
- Corticosteroids for suspected autoimmune disease.

Table 1.17 Clinical Features of acute vestibular paralysis (vestibular neuronitis)

A previously healthy patient suffers from:
• Rotatory dizziness
• Nausea and vomiting
• Dizziness persisting for days or weeks
• Ataxia
Typically, the following are absent:
• Tinnitus
• Hearing loss
• Loss of consciousness
• Double vision or visual field defects

Soon after the acute phase:
- Active physiotherapy, including exercises to train balance.
- No psychotropic drugs!

Course and prognosis. The symptoms subside within a few days, with spontaneous recovery of equilibrium (vestibular compensation). Younger patients tend to recover more quickly than older patients. Despite a persistent caloric hypoexcitability of the affected labyrinth, vestibular function recovers. Latent disequilibrium with a tendency to fall may persist in older patients.

Benign Paroxysmal Positional Vertigo (BPPV) (Cupulolithiasis)

Clinical Features. The characteristic symptoms are sudden frequent attacks of severe rotatory vertigo provoked by certain movements. Symptoms sometimes occur at night during movement while asleep.

Pathogenesis. Benign paroxysmal positional vertigo is a peripheral lesion of the end organ. The theory is that vertigo is incited by particles floating in the endolymph of a semicircular canal (canal lithiasis). The particles are usually otoconia separated from the macula. During certain movements, the increased mass of the otoconia causes a nonphysiologic deflection of the cupula and subsequently the typical symptoms.

Diagnosis. The Dix–Hallpike maneuver is used to test for BPPV (see p. 46).

Treatment. Initial treatment consists of therapeutic maneuvers designed to displace and reposition the canaliths (canalith repositioning procedure or Epley maneuver).

Neural and Central Vestibular Disorders

Clinical Features. The following symptoms indicate that a balance disorder has a central cause:
- Sudden attacks of dizziness of short duration (1–2 s).
- Fluctuating dizziness with loss of consciousness.
- Dizziness with "drop attacks," which are brief attacks of muscle-tone loss in which the patient sinks to the ground but does not become unconscious.
- Dizziness with double vision and other disturbances of vision such as hemianopsia, scotoma, etc.
- Dizziness with dysarthria and change of personality.

Pathogenesis. Central vestibular disorders are usually the result of *multifocal lesions* of the brain stem. They are therefore usually accompanied by symptoms of disorders of the visual oculomotor and somatosensory system (Tables 1.18, 1.19).

The investigation and treatment of this group of vestibular disorders fall within the specialty of neurology. The most frequent cause is vascular insufficiency in the brain stem region, which can lead to ischemic lesions or hemorrhages.

Bilateral Vestibular Loss

This balance disorder is caused by severe, bilateral hypofunction or complete loss of the peripheral vestibular apparatus. Systematic causes are possible, as well as bilateral diseases of the vestibular apparatus.

Systemic causes
- Ototoxicity (drugs, industrial toxins).
- Endogenous lesions (renal disturbances).

Local disturbances
- Labyrinthitis (bacterial, viral, autoimmune).
- Acquired labyrinthine disorders (anomalies, acute loss).
- Vestibular nerve disease (polyneuropathy, NF2).

Clinical Features
- No nystagmus, but oscillopsias.
- Balance disturbance.
- Bilateral sensorineural hearing loss.

Different Causes of Peripheral Vestibular Disorders

Cervical syndrome includes cervicobrachial neuralgia and brief attacks of dizziness dictated by the position and changes in position of the head, occasionally associated with tinnitus and pain in the nape of the neck, which may radiate to the occiput and the forehead area. Objective findings include neck-torsion nystagmus. The cause is a lesion in the joints of the cervical spine and in the muscles on the back of the neck (see p. 375). An acute onset of deafness with vestibular failure in this condition may also be due to embolism of the labyrinthine artery. In the majority of cases, the symptoms develop independently. The prognosis for functional recovery is unfavorable.

Posttraumatic cervical syndrome is mostly due to whiplash injury and causes almost the same symptoms after a symptom-free interval of several weeks, but the objective vestibular signs such as positional nystagmus are more pronounced. In addition, sensorineural hearing loss can occur and may persist for weeks and months. Other causes of posttraumatic vestibular disorders are blunt head trauma with labyrinthine contusion and concussion, temporal bone trauma with fractures, and postsurgical conditions (ear and temporal bone surgery).

Motion sickness is a sensory conflict, or mismatch, between the visual and vestibular systems that produces symptoms of malaise, fatigue, yawning, hypersalivation, nausea, and vomiting. Prevention relies on anticipating and visually perceiving the motion with as much of the visual field as possible. Training can induce the development of central patterns to reduce susceptibility. Medical treatment with antivertigo drugs (scopolamine, dimenhydrinate) is possible.

Perilymphatic fistula: A connection—either congenital or induced by trauma (e.g., barotrauma, acoustic trauma)—between the perilymphatic space and middle ear, usually through the round or oval window, can lead to vestibular or acoustic impairment. The fistula can be inspected and repaired by covering the window niches with fat tissue under local anesthesia, and this should be considered immediately.

Tullio phenomenon is a vestibular symptom that is induced by acoustic stimulation, usually with the sound at high levels on high and low frequencies. Possible causes are a hypermobile stapes footplate, adhesions between the footplate and saccule, or perilymphatic fistula.

Table 1.18 Pathogenesis of central vestibular disorders

Inflammation
• Meningitis • Meningoencephalitis • Cerebellar abscess
Trauma
• Cerebral concussion and contusion
Space-occupying processes
• Infratentorial tumors • Cerebellopontine angle tumors • Glomus tumors • Arachnoid cysts
Vascular processes
• Vertebrobasilar insu□ciency • Basilar artery migraine • Arteriovenous anomalies
Intoxication
• Barbiturates • Alcohol
Degenerative diseases of the central nervous system
• Multiple sclerosis • Syringobulbia • Cerebellar degeneration

Hearing Disorders

Sudden Sensorineural Hearing Loss

Clinical Features. Initially, there is a feeling of pressure in the ear, followed by tinnitus, which is usually marked, followed by severe hearing loss beginning within minutes and occasionally leading immediately to complete deafness.

Table 1.19 Synopsis of central balance disorders

	Neurotologic signs	Neuroophthalmologic signs	Neurologic sign
Encephalitis	Spontaneous nystagmus, often disassociated; positional nystagmus (irregular); gaze-evoked nystagmus; pathologic optokinetic reflexes	Papilledema	Meningism, somnolence, restlessness, cerebellar symptoms, focal neurologic symptoms, abnormal spinal fluid chemistry
Cerebral concussion	Positional (provocation) nystagmus, pathologic vestibulospinal reflexes	–	Headaches, memory defects, poor mental concentration
Infratentorial tumor with brain stem compression	Spontaneous nystagmus, positional nystagmus (irregular) gaze-evoked and gaze-paretic nystagmus, pathologic optokinetic nystagmus, pathologic vestibulospinal reflexes with ataxia, pathologic caloric responses	Eye muscle paresis, papilledema, horizontal and vertical gaze paresis	Focal neurologic symptoms of cranial nerves V, VII, IX, and X, dysphagia, pyramidal signs, extremity paresis, dissociated sensation, headache, vomiting
Vertebrobasilar insufficiency with involvement of pons and medulla	Vestibular spontaneous nystagmus, vestibular provoked nystagmus, pathologic vestibulospinal reflexes, pathologic optokinetic reflexes	Horner syndrome	Cerebellar ataxia, motor hemiparesis, disturbed sensation, involvement of cranial nerves V, VII, IX, and X
Multiple sclerosis	Vestibular spontaneous nystagmus, irregular positional nystagmus (irregular), gaze-evoked and gaze-paretic nystagmus, abnormal caloric responses, abnormal optokinetic reflexes, pathologic vestibulospinal reflexes, ataxia	Dissociated (disconjugate) nystagmus horizontal and vertical gaze paralysis, bilateral internuclear ophthalmoplegia, retrobulbar neuritis	Multiple attacks of multilocular demyelinating disorders of the central nervous system with spastic paralysis, urinary incontinence, intention tremor, dysdiadochokinesis, and paresthesia and dysesthesia
Barbiturate intoxication	Vestibular spontaneous nystagmus, irregular positional nystagmus (irregular), gaze-evoked nystagmus, abnormal vestibular ocular and optokinetic reflexes	Oculomotor disorders	Cerebellar ataxia, dysarthria, somnolence

Pathogenesis. *Idiopathic sudden hearing loss:*
- Viral infection.
- Disturbance of microcirculation of the inner ear.
- Autoimmune reaction.

The types of disorder are shown in Table 1.20.

Diagnosis. The symptoms are usually unilateral. Audiography shows sensorineural hearing loss restricted to the higher and middle frequencies, with recruitment or complete deafness. Causes of symptomatic sudden hearing loss should be excluded by a test diagnostic battery including: impedance audiometry, otoacoustic emissions, and vestibular tests. Additional tests focus on arterial hypertension, hyperlipidemia, infections (e.g., neurotropic viruses, syphilis, borreliosis, toxoplasmosis). After a period of recovery, cerebellopontine angle tumors are ruled out by ABR testing and MRI (but caution is needed with the latter, as there is a danger of acoustic stress during the test procedure).

Treatment. *Idiopathic sudden hearing loss:*

The aim is to improve the microcirculation and oxygenation of the cochlea as soon as possible through:
- Intravenous low-molecular-weight dextran infusions and/or a systemic vasodilator.
- Corticosteroids.

Symptomatic sudden hearing loss:

Therapy is initiated immediately to treat the above-mentioned causes.

Course and prognosis. If treatment is instituted within 24 h, there is often partial or complete recovery within several days. Spontaneous remission also often occurs. The prognosis is poor for patients with diabetes, arterial hypertension, or established irreversible vascular disease.

Chronic Progressive Idiopathic Sensorineural Hearing Loss

This disease is defined as having an onset before the age of 50, usually starting between 30 and 50 years of age. The cause is unknown; intrinsic or extrinsic pathophysiological factors are not evident. A recessive genetic defect may be involved.

Clinical Features. Bilateral involvement, with a variable course of hearing loss, is characteristic. There is a sudden onset and gradual progression of hearing loss, frequently accompanied by tinnitus. Severe hearing loss or complete deafness can develop over a period of years or decades.

Diagnosis
- Progressive bilateral sensorineural hearing loss.
- Exclusion of etiologic factors (metabolic, genetic, autoimmune, infectious, toxic, etc.).
- Exclusion of retrocochlear, psychogenic, and central hearing loss.

Treatment and prognosis
- Hearing rehabilitation (hearing aid, lip-reading training).
- Cochlear implant.

Presbyacusis

This type of symmetrical sensorineural hearing loss is an age-related disease affecting persons over 50 years of age. Approximately one-third of persons over the age of 65 have significant hearing loss, averaging 35 dB or more on pure-tone audiometry.

Clinical Features. These include bilateral hearing loss, initially for high tones and later for middle frequencies. A “social deafness” is gradually established, i.e., the patient can no longer take part in conversation with several people at the same time. Hearing for speech is affected by loud noise, and loud sounds cause discomfort due to positive recruitment. Pure-tone hearing is often better than the hearing for speech, and hearing for syllables often better than that for sentences *(schizacusis).* Other symptoms include noises in the ears and psychological disturbances, which increasingly isolate the elderly deaf patient from his environment, causing depression, mistrust, and delusions of persecution.

Pathogenesis. *Physiologic presbyacusis.* This is a degenerative process in the inner ear and central nervous system (CNS), without exogenous damage. The degenerative process is caused by a disorder of DNA synthesis, deposition of pigment (lipofuscin), extracellular deposition of cholesterin and lipids, conversion and breakdown of collagen substances, and loss of intercellular fluid.

Pathologic presbyacusis. Supplementary inner ear and CNS lesions are caused by exogenous factors, such as environmental noise, and by lifestyle factors such as diet, smoking, alcohol, physical and psychological stress, hypertension, and maturity-onset diabetes. In summary, this is a multifactorial disease of the peripheral and central auditory systems that starts earlier than physiologic presbyacusis.

Four types of presbyacusis can be distinguished on the basis of morphologic degenerative lesions:

1. *Sensory presbyacusis* due to hair cell degeneration. The audiogram shows high tone loss.
2. *Neural presbyacusis:* A large proportion of the population of cochlear neurons is lost, so that the predominant symptom is loss of discrimination for speech.
3. *Strial presbyacusis,* due to degeneration of the stria vascularis. This causes abnormalities in endolymph production and secretion, with repercussions on the energy metabolism of the hair cells. Audiography shows a flat curve of pancochlear hearing loss with retained discrimination for speech.
4. *Conductive cochlear presbyacusis:* An age-related degenerative process in the cochlear duct causes lesions in the structure of the basilar membrane. This affects stimulus transport in the cochlea (see p. 14) and is demonstrated on audiography by bilateral symmetrical sensorineural deafness causing a characteristic sloping curve, with a linear increase in hearing loss above 1000 Hz.

Diagnosis. The patient has bilateral (usually symmetrical) sensorineural deafness. The audiometric curve depends on the type.

Differential diagnosis. Marked unilateral sensorineural deafness has to be distinguished from vestibular schwannoma or a cerebellopontine angle tumor. Marked unilateral or bilateral tinnitus, synchronous with the pulse, has to be differentiated from an intracranial aneurysm of the posterior cranial fossa or a glomus tumor. Other causes, ototoxic or metabolic, should be taken into consideration.

> Note: Elderly patients with hearing loss require early rehabilitation of communication to the same extent as young patients with hearing loss, provided that their mental powers are not severely limited.

Treatment

- Auditory rehabilitation (hearing aid, auditory training, lip-reading course).
- Psychosocial rehabilitation (family, social programs for integration of hearing-disabled people).

Course and prognosis. The degree and time course of the hearing loss is highly variable. Its progression cannot be predicted.

Clinical Aspects of Central Hearing Disorders

See Table 1.20.

Acoustic Agnosia

Synonyms for this condition include sensory aphasia, psychogenic or word deafness, sensory deaf-mutism, and central deafness.

Clinical Features. The main symptoms include acoustic inattention due to an absence of the acoustic ability to differentiate, disturbances of perception, slurred articulation, animated gestures, and miming. Directional hearing is lost, and the patient suffers paramusia, amusia, and loss of musicality.

Pathogenesis. The causes may be cerebral or skull trauma, encephalitis, or prenatal, perinatal, or postnatal damage to the central nervous system.

Diagnosis. A clinical diagnosis of acoustic agnosia can only be made using neuropsychological tests. Investigations also include electroencephalography, CT and MRI, and evoked response audiometry.

Treatment. Long-term treatment includes acoustic differentiation exercises, articulation training, and prescription of a hearing aid for objectively confirmed sensorineural deafness. A hearing aid is of no use for true sensory agnosia. Rhythmic exercises, music therapy, and exploitation of visual observations can improve the patient's comprehension (e. g., through lip-reading).

Table 1.20 Synopsis of hearing disorders and their treatment

Anatomic substrate	Function	Type of disorder	Effect on hearing	Treatment	Prognosis for auditory rehabilitation
Conductive hearing loss (middle ear)					
Middle ear	Conduction	Discontinuity due to infection of the ossicular chain, ventilation disorders of the middle ear, or stiffening of the ossicular chain and the tympanic membrane with increasing acoustic resistance—i. e., increased impedance	Quantitative hearing loss due to mechanical loss of sound energy	Surgery or a hearing aid	Good
Sensorineural hearing loss (inner ear)					
Inner ear	Mechanical frequency analysis, stimulus transformation of a mechanical into a bioelectrical stimulus, and possibly coding	Destruction of the sensory cells in the inner ear due to traumatic, vascular, metabolic, toxic, or inflammatory damage	Quantitative hearing loss combined with a qualitative worsening of speech intelligibility, due to loss of frequency analysis and stimulus transformation coding; distortion effect due to recruitment	Hearing aid, hearing training, and lip reading. Trial of medical treatment	Relatively good. Depends on the degree of hearing loss
Peripheral neuron	Coding, transmission of nerve impulses; lateral inhibition and interneural inhibition ensure acoustic selectivity	Degeneration of the peripheral neuron due to inflammatory, vascular, traumatic, or metabolic injury	Quantitative and qualitative deterioration of hearing for speech due to abnormal coding, loss of neurons, unsatisfactory selectivity, and inability to discriminate	Hearing aid, hearing training, and lip reading	Doubtful
Auditory perception (central)					
Central auditory pathways and centers	Integration—i. e., assembly of individual nervous impulses in a functional modulated activity; storage of auditory memory; decoding of acoustic information	Degeneration of the central pathways and the ganglion cells in the primary and secondary hearing centers due to inflammatory, vascular, traumatic or metabolic lesions	Mainly complete loss of the information content due to unsatisfactory integration and decoding of the acoustic signal. Partial loss of the auditory memory. The final phase is complete central deafness	Hearing aid useless. In selected cases, hearing training or lip reading	Poor

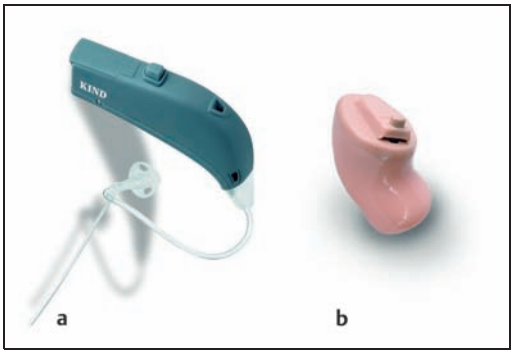


Fig 1.111a, b Different types of hearing aid.
a A behind-the-ear device.
b An in-the-ear device with an earmold fitted.

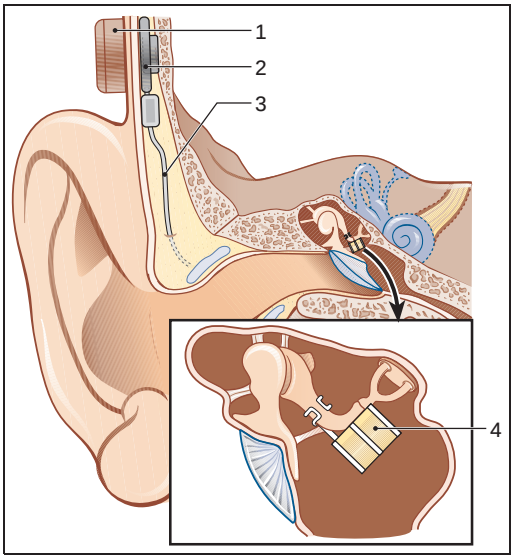


Fig. 1.112 The principle of an implantable hearing aid. An externally worn microphone and speech processor (1) transmits sound through the skin to an implanted receiver (2). A cable (3) connected to a tiny transducer (4) directly vibrates the ossicles by mimicking the natural motion of the ossicular chain, sending an enhanced signal to the cochlea.

Rehabilitation of Hearing Disorders with Hearing Aids

Note: The assessment of hearing disorders in relation to the potential for social rehabilitation should be based not only on the degree of severity and site of the hearing loss, but also on the patient's age and physical, mental, and speech development. The aim of auditory rehabilitation is to restore or improve auditory communication, which is crucial for social functioning and the patient's well-being.

A variety of rehabilitative options are available, depending on the nature and degree of the hearing loss and the degree of handicap it involves.

Surgery. This is an option for patients with conductive hearing loss due to functional disturbances of the middle ear (see p. 73, Fig. 1.96 a–f).

Hearing aids. These selectively amplify auditory signals. The acoustic signal is received by a microphone, amplified and processed, and then delivered to the ear through a loudspeaker. Hearing aids can be worn behind the auricle or inside the ear canal (Fig. 1.111a, b). The fitting of a hearing aid is an important procedure involving several steps: establishing the indication in an audiological examination; selecting and trying out hearing aids; final selection of the hearing aid and provision of follow-up care.

Bone-anchored hearing apparatus (BAHA). Devices with an acoustic amplifier and a transducer are connected to the cranial bone via a bone-anchored system. Mechanical vibrations are transduced directly to the cranial bone to improve sound transmission. BAHA systems are mainly used in patients with untreatable diseases of the external meatus or with severe hearing loss.

Active middle ear implants. An amplifier implanted under the skin transforms sound waves into electrical impulses, which are converted into mechanical vibrations in a transducer. The vibrations are transmitted directly to the ossicles or cranial bone (Fig. 1.112).

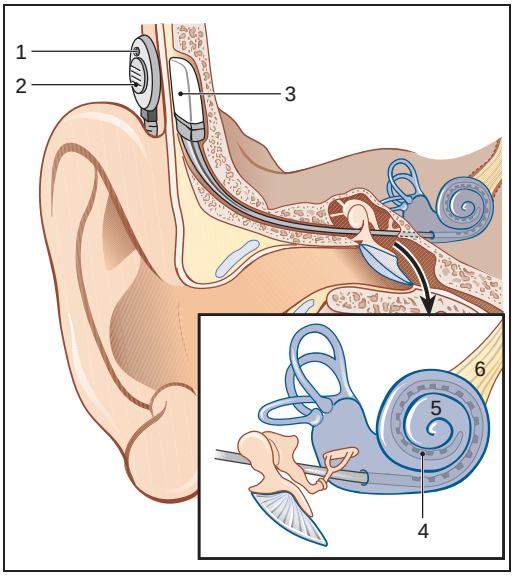


Fig. 1.113 The principle of a cochlear implant. The acoustic signal is received by a microphone (1) worn behind the ear and is processed by an external speech processor (2). An electronic receiver (3) is implanted into the temporal bone under the skin. It is connected to an electrode array (4) inserted into the cochlea (5). The electrodes directly stimulate the vestibulocochlear nerve (6).

Cochlear implant (CI). A subcutaneously implanted device can transform sound waves into electrical impulses that directly stimulate the vestibulocochlear nerve with intracochlear electrodes. Patients with almost complete deafness due to an absence of cochlear function are candidates for implantation (Fig. 1.113). In children, best results are obtained when the CI procedure is done before the age of 1 year because of the early brain plasticity in the growing child. Before implantation, the responsiveness of the vestibulocochlear nerve has to be tested using the promontory test with a transtympanic electrode. Subjective hearing impressions can be recorded in adults, whereas electrical brain stem responses are used for recordings in children. The surgical procedure is standardized and safe. Modern trends in CI procedures include bilateral implants and implants in children with a low auditory threshold. Children with sensorineural hearing loss more than 60 dB are also provided with CI because they can benefit from better acoustic information arriving at the auditory cortex. The success of the implantation depends on further rehabilitation being provided by speech and language therapists and teachers for hearing-impaired people. Speech recognition is also supported by teaching the patients to lip-read. Over 90 % of patients derive definite benefit from a CI and more than 50 % can achieve open speech. Patients who have only had hearing loss for a short period benefit most from implantation.

Vibrotactile aids. Acoustic signals are picked up by a microphone and converted into vibrations that are transmitted to the wrist or fingers.

Assistive devices. Devices include optical or vibrating wake-up alarms, light flashers, telephone amplifiers, text telephones, television headphones, and digital communication devices such as fax machines, text messaging, and the Internet.

Training. Hearing-impaired patients can be trained in selective listening, using listening tactics or auditory training. Using hearing aids and other assistive devices, as well as learning how to lip-read and how to improve their own speech are also important training exercises for patients.

Pediatric Hearing Disorders (Pediatric Audiology)

Significant hearing impairment is present at birth in 0.8–2.3 per 1000 newborns in Europe. During the first year of life, permanent hearing loss occurs in one in 1000–2000 children. Causes of pediatric hearing disorders, which can be classified into genetic and acquired forms, are listed in the Tables 1.21, 1.22, 1.23, 1.24.

The presence of hearing loss profoundly affects the subsequent development of speech and has specific consequences for rehabilitation and personality development in these children. If there is the slightest sign of hearing impairment in an infant or young child, an audiologic evaluation should be performed. Parents and special services for infants and children should be informed about the time course of speech development in children (Table 1.25).

Table 1.21 Genetic forms of deafness and hearing loss

Sporadic recessive deafness	
Progressive morphologic lesion present at birth, usually with total degeneration of the cochlea and the peripheral cochlear neuron	
Types: • Michel • Mondini • Scheibe	Hypoplasia or aplasia of the labyrinth with almost complete loss of function of the cochlea and vestibule
Dominant hereditary degenerative hearing loss	
Progressive disorder with an irregular course. First becomes manifests at or after puberty. Often combined with other inherited symptoms to form a circumscribed syndrome such as those detailed below	
Waardenburg syndrome	Congenital anomalies of the facial skeleton, dystopia canthorum; blepharophimosis; disorders of pigmentation of the eyes, hair and skin (albinism); degenerative atrophy of the cochlea and the ganglion cells
Usher syndrome	Hereditary progressive sensorineural hearing loss with retinitis pigmentosa. Degeneration of the cochlea and the spiral ganglion
Refsum syndrome	The same symptoms are found as in Usher syndrome, with the addition of polyneuropathy and ataxia. Often, this condition first becomes manifests between the ages of 10 and 20
Alport syndrome	Progressive inner ear hearing loss, bilateral, but often asymmetrical, beginning in the second decade of life, associated with a nonspecific chronic glomerulonephritis and interstitial nephritis. The frequency is one per 200,000 births. The renal disorder is inherited, and the inner ear lesion may be a secondary nephrogenic effect
Pendred syndrome	Perceptive hearing loss with labyrinthine dysplasia associated with thyroid gland disorders
Chromosomal anomalies	
Anomalies of the external and middle ear associated with numerous other organ anomalies and developmental disorders of the inner ear	
Trisomy 13	Labyrinthine hypoplasia with aplasia of the spiral organ of Corti and the stria vascularis
Trisomy 18	Similar changes, with atresia of the vestibulocochlear nerve
Cri-du-chat syndrome	Similar malformations associated with laryngeal anomalies

Audimutism

This term is inaccurate, as it does not adequately characterize delayed development of speech. The child is mute despite being able to hear. The term "audimutism" in the strictest sense should be confined to children who, after the third year of life, can only make themselves understood by gestures and are unable to speak any words, but are not mentally defective and demonstrate normal hearing. There is a connection between audimutism and brain damage in early childhood. The term "deaf-mutism" has therefore been replaced by "absent or delayed speech development."

Classification and Effects of Hearing Disorders Depending on the Degree of Severity

Normal hearing. The hearing threshold of the pure-tone audiogram does not exceed 20 dB for any frequency.

Slight hearing loss. On the pure-tone audiogram, there is hearing loss of > 20 dB or at most 40 dB over the entire frequency range. Hearing loss of this extent does not usually cause delay in the development of speech in a child of normal intelligence.

Conductive hearing loss can usually be treated medically. Sensorineural hearing loss has to be

treated individually with a hearing aid, supplemented by language training by a hearing therapist. Individual schooling and hearing training are not indicated.

Moderate hearing loss. In this case, the average hearing loss in the main speech frequencies of 250–4000 Hz is between 40 and 60 dB. This causes considerable difficulty in understanding speech, so that speech development is delayed. Prescription of a hearing aid is absolutely indicated, and children require early hearing and speech education. Initially, the children should attend a special school for the deaf, but they can usually attend a normal school later, provided they have a hearing aid and normal intellectual development.

Severe hearing loss. The average hearing loss in the main speech frequencies is greater than 60 dB, and tones above 1000 Hz are most severely affected. Provided that the child has normal intellectual development, a hearing aid is certainly indicated, since good speech communication can be developed with even the least residual hearing using electroacoustic methods and early speech and hearing training. These children should attend special schools for the deaf. They should have several years' training in lip-reading to encourage visual compensation for the loss of auditory function.

Bilateral complete deafness. This form is fortunately very rare. Even patients with almost total deafness often have residual hearing, which can be demonstrated using electroacoustic methods and exploited using high-performance hearing aids. The child should be educated in a separate school for the deaf. In these schools, sign language is currently being replaced by the use of real speech: practice in articulated speech is provided using the sense of vibration and training in lip-reading, partly directly and partly with a video recorder. In selected cases, both children and adults can be considered for a *cochlear implant,* which involves the implantation of a multichannel cochlear prosthesis.

Multiple handicap. A special group of deaf children also have intellectual retardation, behavioral disturbances, extensive organic brain damage, or delay in motor development causing failure to speak. A demanding therapeutic program needs to be established for this group with a team including an otologist, phoniatrist, neurologist, child psychiatrist, psychologist, and speech pathologist.

Table 1.22 Prenatally acquired (exogenous) deafness and hearing loss

Intrauterine infection	
Rubella embryopathy	Developmental disorders of the middle and inner ear, severe bilateral sensorineural hearing loss
Congenital syphilis	Progressive degeneration of the inner ear and the peripheral neuron associated with interstitial keratitis and dental defects (Hutchinson triad)
Toxoplasmosis	Inflammatory damage to the inner ear
Viral infections	Mumps, herpes zoster, poliomyelitis, influenza, cytomegalovirus
Toxic damage due to:	Quinine, aminoglycosides, and thalidomide which causes multiple anomalies
Further exogenous damage due to:	Diabetes mellitus in the mother, fetal hypoxia, irradiation

Table 1.23 Perinatally acquired (exogenous) deafness and hearing loss

Perinatal hypoxia	Injury to the cochlea and its centers in the brain stem
Premature birth	Hemorrhage into the cochlea
Kernicterus (perinatal hyperbilirubinemia)	Rh-incompatibility, massive deposits of bilirubin in the cochlear centers and occasionally in the cochlea itself with corresponding cochleoneural deafness

Voice and speech disorders due to deafness. Deafness mainly affects the ability to communicate in children, but it can also affect adults. Physiologic auditory verbal communication requires both a normal voice and speech apparatus and also normal feedback through the ear.

Altered speech in those born without hearing or in those who become deaf or partially deaf in later life, is known as otogenic dyslalia.

Table 1.24 Postnatally acquired (exogenous) deafness and hearing loss

Infective diseases	
Meningitis and meningoencephalitis	Labyrinthitis and cochleovestibular neuritis with damage to the sensory cells and peripheral neurons; central lesions
Mumps	Cochlear and neural lesions (see pp. 96 and 421)
Measles	Degeneration of the cochlea and its peripheral neurons due to infective damage, serous labyrinthitis
Otitis media	Recurrent otitis, infective toxic damage to inner ear associated with sensorineural deafness

Table 1.25 Timetable of speech development

Up to the 7th week	Crying
6 weeks–6 months	First babbling period, beginning of auditory feedback
6–9 months	Second babbling period, hearing predominant
8–9 months	Echolalia, imitation and first understanding of speech
9–12 months	Beginning of deliberate speech
13–15 months	Early precise use of significant words, symbolic function of speech
12–18 months	One-word sentences, developmental stammering
18–24 months	Two-word sentences; unformed sentences of several words, first age of questioning
End of 2nd year of life	Nongrammatical sentences, establishment of consciousness of symbols
From 3 years onward	Formed sentences of several words, adoption of first grammatical principles
From 4th year of life	Second questioning age, first establishment of logical and emotional relationships, maturation of the thinking–speaking process

The Completely Deaf Child

In a child with no hearing or only residual islands of hearing, there is no auditory feedback, and the child develops a typical, monotonous form of speech, which distinguishes the totally deaf child from a child with normal hearing or partial hearing loss. The best rehabilitation results are achieved through:

- Early diagnosis and treatment.
- Early prescription of a hearing aid.
- Prolonged auditory training and early voice and speech training in special preschools and schools for the deaf to develop the missing sound components of speech and speech content.
- Cochlear implant in selected cases.

Cochlear Implantation

Indications

- Bilateral cochlear deafness.
- Residual hearing that is not usable for speech understanding or speech development despite the use of hearing aids.
- Intact acoustic nerve and central hearing tract.
- Postlingual deafness. Acquired deafness after definitive speech development.
- Prelingual deafness. Acquired deafness before speech development, or congenital deafness.

Procedure

Preoperative examination and evaluation. After the cause of deafness has been determined and the functional integrity of the vestibulocochlear nerve and central hearing tract has been tested, the child's psychosocial environment also has to be assessed. This involves assessing the child's general development and language development level, as well as the general communication situation.

Surgical procedure. A subcutaneous device is implanted after complete mastoidectomy. An electrode is inserted into the scala tympani via an opening in the basal turn of the cochlea (cochleostomy).

Postoperative fitting of the speech processor. After complete wound healing, the implanted speech processor is adapted to optimize the hearing result. Lifelong technical checking and adjustment are necessary.

Hearing and speech training. Postlingually deaf patients are able to compare auditory impressions

conveyed by the implant with their stored auditory impressions. They learn how to understand these new impressions within a few months. By contrast, prelingually deaf children experience hearing as a new sense modality and learn and develop language in the same way that a normally hearing child does. This process usually takes several years, and it requires special rehabilitation centers for the children.

Clinical Aspects of Disorders of the Facial Nerve

The symptoms and differential diagnosis of central and a peripheral lesions of the facial nerve are discussed above (see pp. 13 and 47). Only the effects of peripheral lesions of the facial nerve and their causes, characteristics, and treatment are therefore discussed here.

Inflammatory or Otogenic Facial Paralysis

A form of peripheral facial paralysis that is at least partly reversible may be caused by neurotropic viruses such as herpes zoster (see pp. 52 and 96), coxsackievirus, or poliomyelitis virus, and by neuroallergic polyradiculitis, Guillain–Barré disease, or sarcoidosis. The latter is also known as Heerfordt–Mylius syndrome, with uveitis and parotitis in addition to facial paralysis (see p. 417). However, the most important and most common form for the otologist is *otogenic facial paralysis*, which occurs as:

- A complication of acute otitis media or mastoiditis, which is treated with antibiotics and immediate mastoidectomy; the lesion is usually a neurapraxia. The prognosis is always good, provided the patient is not affected by diabetes or immunodeficiency.
- A complication of middle ear cholesteatoma invading the bony facial canal. Extension of the inflammation to the nerve sheath and endoneurium causes axonal degeneration and demyelinization of the mastoid, tympanic, and labyrinthine segments. The condition is treated with mastoidectomy and decompression of the nerve. The prognosis is occasionally poor, despite intensive antibiotics and decompression of the facial nerve, as the nerve is usually only partially reinnervated and does not fully heal.

Idiopathic Facial Paralysis (Bell's Palsy)

As the term "idiopathic" indicates, the cause of this is unknown. It may be a disturbance of microcirculation leading to serous inflammation, with edema developing. The bony canal is rigid, and the resulting compression of the nerve leads to ischemia and venous congestion, so that a vicious circle starts. The annual incidence of Bell's palsy is ≈ 20–30 per 100,000 population. The condition occurs most often in the 15–45-year-old age group and it is much less common in children under the age of 10.

More recent evidence now suggests that the condition may be caused by herpesvirus—specifically, herpes simplex virus type 1.

Diagnosis. Topographic diagnosis of the facial nerve is carried out (see p. 47) and causes involving inflammation, trauma, or tumor are excluded.

In ≈ 80% of patients, electrodiagnosis shows neurapraxia, with a good prognosis. In 20% of patients, there is partial axonotmesis. In ≈ 5–10% of patients with idiopathic facial paralysis, the axon degeneration progresses between days 4 and 10 despite treatment with corticosteroids. If neuronography shows that more than 90% of neurons have degenerated, decompression is indicated.

Treatment

- Intravenous infusion of low-molecular-weight dextran.
- Corticosteroids. Schedule for prednisone in the treatment of idiopathic facial paralysis: 60 mg for 4 days, reducing by 5 mg daily to 5 mg on the 15th day, followed by intermittent dosage of 5 mg for 10 days.
- Antiviral drugs (aciclovir). The evidence for using aciclovir is inconclusive, but the agent is generally recommended, as undetected slow virus infections are suspected.
- It has been recommended that surgical decompression needs to be performed within 2 weeks of the onset of total paralysis in order for it to be effective. This is a rather controversial area, and there is a lack of consensus regarding the value of facial nerve decompression in this setting.

During the first 2 weeks of the paralysis, electromyography does not provide prognostically valid information. During this period, therefore, electrodiagnostic and neuronographic investigations

should be carried out every 2 days to identify patients who require decompression treatment.

Melkersson–Rosenthal syndrome is an unusual form of idiopathic facial paralysis. Symptoms include recurrent facial paralysis, furrowed tongue and cheilitis, and facial edema. The cause is unknown, treatment with steroids is unsatisfactory, and the prognosis is uncertain.

Traumatic Facial Paralysis

Causes. The second most common cause of facial paralysis is trauma, which includes blunt, penetrating, and iatrogenic forms. Most commonly, traumatic injuries to the facial nerve are caused by temporal bone fractures.

Frequency. Less than 5% of temporal bone fractures involve the otic capsule; of these, half are associated with facial nerve injury. Facial paralysis occurs in 10–20% of all longitudinal pyramidal fractures and in 50% of all transverse fractures. Seventy-five percent of all paralyses develop completely within 24 h of the trauma and 90% of late-onset paralyses resolve spontaneously.

Prognosis. The two most important prognostic factors for recovery from facial nerve paralysis are the severity of the condition and the time of onset. Nearly 40% of patients with immediate-onset complete paralysis have a poor recovery. The severity of a pyramidal fracture is determined by the type of the lesion, such as depression of the canal, damage to the nerve by a splinter of bone, shearing of the nerve without fracture of the bony canal, or a nerve sheath hematoma leading to edema. Electrodiagnosis is the only reliable method of determining which patients will develop rapid degeneration of the axons and will therefore not recover fully without decompression.

Treatment. The functional goals in facial nerve surgery are to protect the eye, restore the nasal airway, and reestablish oral competence. Cosmetic goals include improved balance and symmetry of the face at rest and coordinated movement of the facial musculature. In the acute setting, the best functional results are obtained with direct facial nerve anastomosis and interpositional nerve grafts. If transection of the nerve occurs in cases of traumatic injury, end-to-end anastomosis is recommended. If part of a facial nerve is sacrificed and reananstomsis leads to unwanted tension, interpositional grafting can be as good as direct reanastomosis. Conservative treatment is limited to steroids and is indicated when any suspicion of direct nerve injury has been excluded. The expected rate of peripheral axonal nerve regeneration is ≈ 1 mm per day, but it varies widely. Adequate facial tone can recover within 6 months, and motion can progressively improve over a further 1–3 months.

Reconstructive Surgery after Facial Paralysis

Facial paralyses are due to various lesions of the facial nerve. Their severity depends on the extent to which the continuity of the nerve has been preserved and which segment of the nerve is affected (Fig. 1.114).

In cases of crushing and constricting injury, depression of the bony canal, and direct injury to the nerve by bone splinters, exposure of the nerve along its intratemporal course in the region of the bone damage is sufficient for decompressing the nerve. Occasionally, slitting of the nerve sheath is also required in order to release a hematoma. Lesions of the facial nerve in the facial region due to skull fractures, knife and stab wounds, and surgical trauma are relatively unusual. A lesion of *all* nerve branches is caused solely by damage to the main trunk of the nerve (see Figs. 1.21, 1.114); peripheral injuries usually only affect individual branches.

Principles of treatment. A search should be made for the site of injury to the nerve, if possible within 48 h. The nerve should be resutured using microsurgery, if necessary with a nerve graft from the great auricular or sural nerve. An old injury should be managed by attempting to reconstruct the nerve in the region of the lesion, and if this is not possible, by anastomosing the branches of the facial nerve to the opposite side in the form of a “crossover,” using sural nerve grafts.

Loss of continuity may be due either to a clean division of the nerve or to an extensive defect. requiring one of the following procedures:

1. *End-to-end suture* is indicated after division of the nerve if the proximal and distal stumps can be approximated without tension. The epineurium is resected from the nerve stumps. Suturing is performed by passing the needle through the connective-tissue sheath surrounding the fascicular bundles (Fig. 1.115).

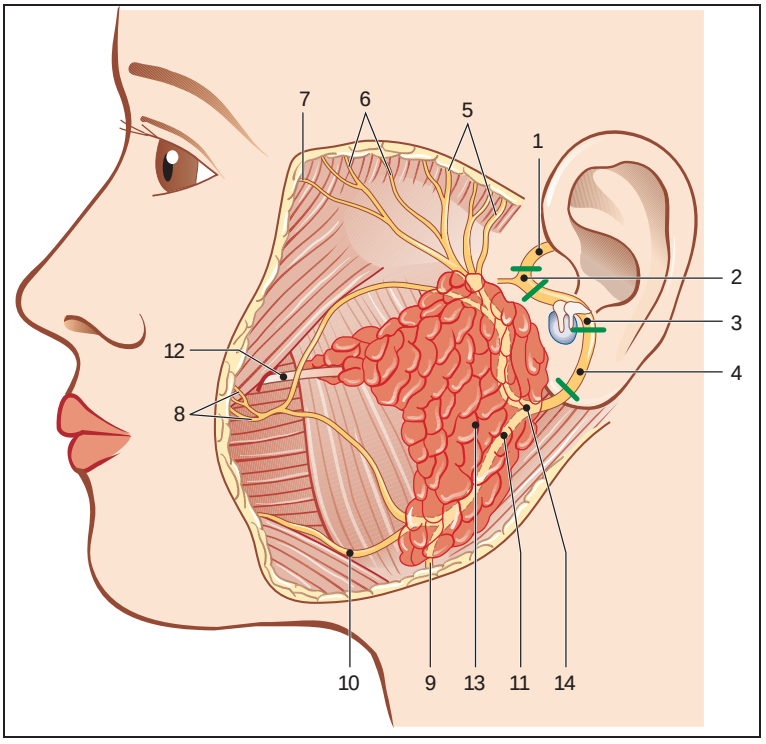


Fig. 1.114 Anatomy of the course of the facial nerve relative to exposure of the nerve. *Intratemporal section:* 1, meatal segment; 2, labyrinthine segment; 3, tympanic segment; 4, mastoid segment. *Extratemporal section:* 5, temporal rami; 6, zygomatic rami; 7, temporofacial portion; 8, buccal rami; 9, cervical rami; 10, marginal mandibular ramus; 11, cervical part; 14, extratemporal part. *Other structures:* 12, parotid duct; 13, parotid gland.

2. *Rerouting* consists of exposing the damaged nerve over a wide area and shortening its route by lifting the nerve out of the bony canal. This allows tension-free repair with end-to-end suture, even for extensive defects (Fig. 1.116).
3. *Autogenous nerve grafts* (cable nerve grafting), with free grafting from the great auricular nerve, sural nerve, or medial and lateral antebrachial nerves, are used for large defects if the freshened nerve stumps cannot be apposed without tension.

In most cases of irreversible facial nerve damage, satisfactory recovery of function can be achieved by one of these surgical techniques. When there is extensive damage of the nerve and its immediate surroundings, and especially when there are defects in the cerebellopontine angle, reinnervation of the muscles of facial expression can only be achieved using nerve replacement techniques.

The peripheral stump can be anastomosed to a cranial nerve such as the ipsilateral hypoglossal or accessory nerve. Individual branches of the still-functioning healthy side can be anastomosed with free autologous nerve grafts to the facial nerve using a cross-face (or faciofacial) anastomosis.

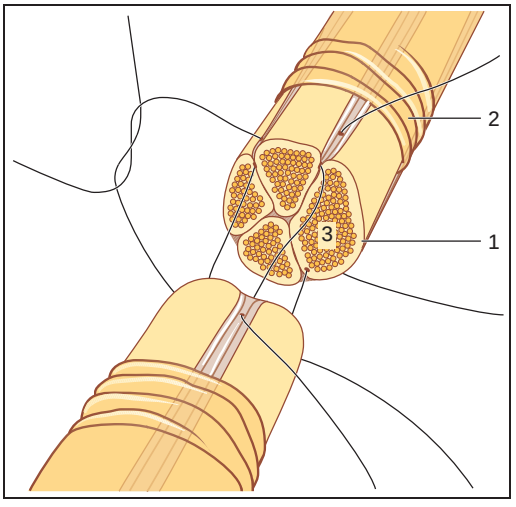


Fig. 1.115 Perineural–fascicular nerve suture. Perineural reconstruction starts with the central fascicle, followed by suture of the epineurium. 1, Perineurium; 2, epineurium; 3, fascicle.

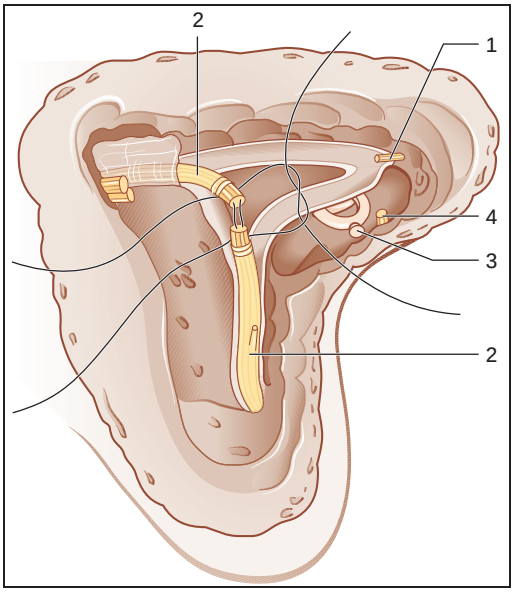


Fig. 1.116 Facial nerve decompression and reconstruction. Decompression and reconstruction in the internal auditory canal can be performed using the translabyrinthine approach. The greater petrosal nerve (1) is cut and the facial nerve (2) is rerouted and anastomosed in the mastoid. 3, Stapes; 4, malleus handle.

The best results are achieved with *hypoglossal–facial anastomosis*, which has to include resection of the epineurium.

In cases of facial nerve defects in the area of the cerebellopontine angle after tumor resection, end-to-end anastomosis with fibrin adhesive or nerve transplantation is performed (Dott's operation).

Facial function should not be expected to return earlier than 6 months after nerve repair or grafting, and 12–18 months may be needed before maximal recovery is seen. In long-standing paralyses, irreversible changes develop over an interval of more than 18 months. These involve fibrosis of the nerve, degeneration of motor end plates, and disuse atrophy of the muscle fibers. Irreversible changes of this type may preclude successful nerve repair or grafting. Reconstruction procedures using regional muscle transfer, such as temporalis muscle or masseter muscle, or free tissue transfer with reinnervation, can be used. Static sling techniques, additional ancillary lid procedures, and brow lifts can also be performed.

Synopsis of Ear Symptoms

See Tables 1.26, 1.27.

Table 1.26 Local symptoms and signs of ear disease

Pressure sensation	*Unilateral or bilateral, constant or intermittent:* cerumen, tubal problem, Ménière disease, glomus tumor
Pain (otalgia)	*Dull, penetrating sensation:* early otitis media or parotitis *Pulsating:* caused by acute otitis media or furuncle in the external canal *Stabbing or intermittent:* otalgia secondary to radiation therapy; bad teeth; temporomandibular joint syndrome; parotitis; tumor of the base of the tongue, tonsil, hypopharynx, or larynx
Aural discharge (otorrhea)	*Smell:* fetid in otitis externa and cholesteatoma *Color:* yellow with cerumen and pus; hemorrhage in influenzal otitis *Consistency:* watery in cerebrospinal otorrhea, mucoid in chronic secretory otitis, greasy in external otitis and cholesteatomas *Pulsating:* otitis media with perforation *Nonpulsating:* external otitis

Table 1.27 Functional ear symptoms

Tinnitus	*Type:* whistling, buzzing, ringing, hissing; crackling are often due to tubal problems *Occurrence:* constant, intermittent, may occur in attacks, as in Ménière disease *Character:* synchronous with pulse in hypertonia, cranial aneurysm, or glomus tumor
Hearing loss	High-tone loss caused by acoustic trauma or presbyacusis Low-tone loss caused by Ménière disease, otitis media, otosclerosis Decreased speech intelligibility, due to a loss of discrimination (inner ear or cranial nerve VIII lesion) Fluctuating, episodic, as in Ménière disease and sudden hearing loss Improvement in noisy environment, reported to be typical of conductive hearing loss Unilateral in Ménière disease, cranial nerve VIII tumors Bilateral in age-induced and noise-induced losses
Vertigo	Turning vertigo in Ménière disease and acute vestibular paralysis (vestibular neuronitis) Elevator and swing vertigo due to inner ear involvement Blackout spells due to extravestibular or orthostatic cause Episodic in Ménière disease Continuous vertigo occurs in vestibular paralysis Spontaneous and episodic in Ménière disease Provoked by head movement and position change of the body, caused by the inner ear, the cervical syndrome, vertebrobasilar insufficiency Unsteadiness in gait and standing is caused by central vestibular disturbance and multiple sclerosis Otologic disorders with vertigo: • Nausea, vomiting—occurs in acute vestibular paralysis and Ménière disease • Double vision—indicates a brain stem lesion • Loss of consciousness—is due to a brain stem or a space-occupying intracranial lesion

2 Nose, Nasal Sinuses, and Face

Applied Anatomy and Physiology

Basic Anatomy

External Nose

The supporting structure of the nose consists of bone, cartilage, and connective tissue. Figure 2.1 shows the most important elements. The *bony* superior part of the nasal pyramid is often broken in typical fractures of the nasal bones, but it may also be fractured in injuries to the central part of the face. The *cartilaginous* inferior portion is less at risk, at least in mild blunt trauma, because of its elastic structure, but it is endangered in stab wounds, lacerations, and gunshot injuries. The shape, position, and properties of the bone and cartilage of the nose have a considerable influence on the shape and esthetic harmony of the face (see p. 212) and on the function of the nasal cavity.

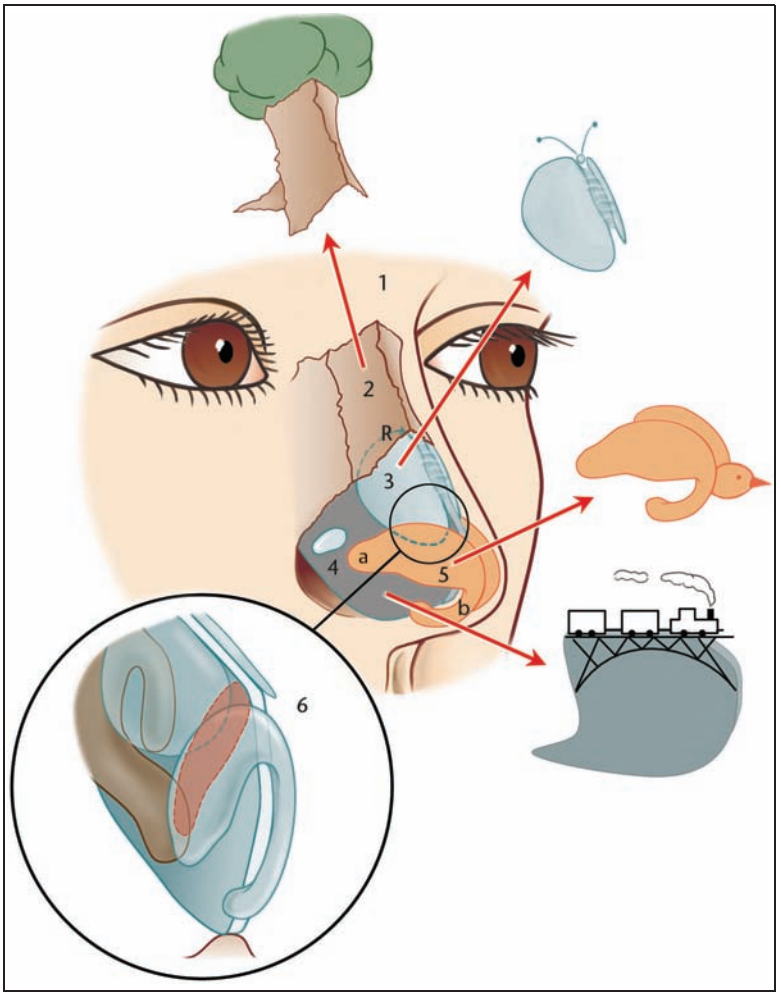


Fig. 2.1 The nasal skeleton. 1, Glabella; 2, nasal bone; 3, lateral nasal cartilage; 4, cartilaginous nasal septum; 5, alar cartilage with lateral (a) and medial crus (b); 6, nasal valve between the cranial alar and caudal lateral cartilages. R, Rhinion. The bony portion of the nose is completely rigid. The flexible cartilaginous portion begins at the rhinion. The distal caudal lateral cartilages can move like the wings of a butterfly, whereas the alar cartilages move more like the wings of a bird. The nasal septum is the center of stability.

The following *blood vessels* in the external nose are of practical importance:

- Facial artery and its branches.
- Dorsal nasal artery, arising from the ophthalmic artery.

Profuse hemorrhage can arise from these vessels when the central part of the face is injured.

The angular vein is also clinically important. Thrombophlebitis arising from a furuncle of the upper lip or the nose can spread via the ophthalmic vein to the cavernous sinus, causing a cavernous sinus thrombosis (see p. 189 and Fig. 2.2).

The external nose derives its *sensory nerve supply* from the first and second branches of the trigeminal nerve (see Fig. 2.15a, b). The muscles derive their *motor nerve supply* from the facial nerve.

Nasal Cavity

The interior of the nose is divided by the nasal septum into two cavities, which are usually unequal in size. Each side may be divided into the *nasal vestibule* and the *nasal cavity* proper (Fig. 2.3a, b). The nasal vestibule is covered by epidermis containing hairs (vibrissae) and sebaceous glands. The latter are the site of origin of a nasal furuncle, which can thus only develop in the nasal cavity in the vestibule.

The medial wall of the nasal vestibule encloses the supporting structure of the anterior part of the cartilaginous septum and the membranous septum—i. e., the *columella*. The roof of the vestibule is formed by the bird wing–shaped lower lateral or alar cartilage, the medial crus of which extends into the columella and the lateral crus of which supports the external wall of the vestibule (Figs. 2.1, 2.3a, b). The alar cartilage determines the shape of the nasal tip and the nasal apertures. Correcting this area is often an important part of rhinoplasty.

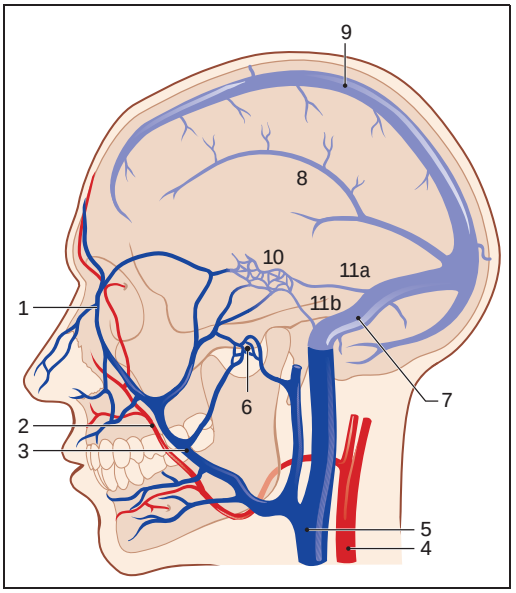


Fig. 2.2 Important vascular relationships in the face. 1, Site for ligation of the angular vein; 2, facial artery; 3, facial vein; 4, common carotid artery; 5, internal jugular vein; 6, pterygoid plexus; 7, sigmoid sinus; 8, inferior sagittal sinus; 9, superior sagittal sinus; 10, cavernous sinus; 11a, superior petrosal sinus; 11b, inferior petrosal sinus.

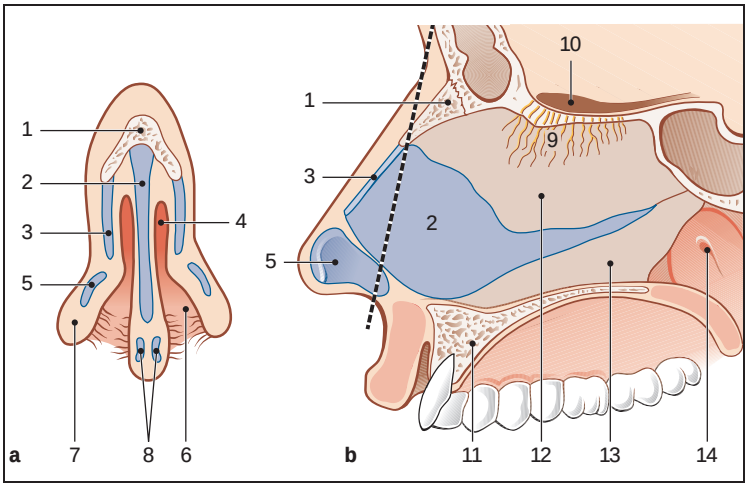


Fig. 2.3a, b a Section through the anterior nose, showing the vestibule and the limen nasi. The dashed line in b shows the sectional plane of a. The limen nasi is located at the junction of the pink and red areas.
b Medial nasal wall. 1, Bony nasal bridge; 2, nasal septum; 3, upper lateral nasal cartilage; 4, nasal cavity; 5, alar cartilage; 6, nasal vestibule; 7, nasal ala; 8, nasal columella with medial crus of the cartilaginous ala; 9, filaments of the olfactory nerve; 10, olfactory bulb; 11, palatine bone; 12, perpendicular plate of the ethmoid; 13, vomer; 14, pharyngeal eustachian tube ostium.

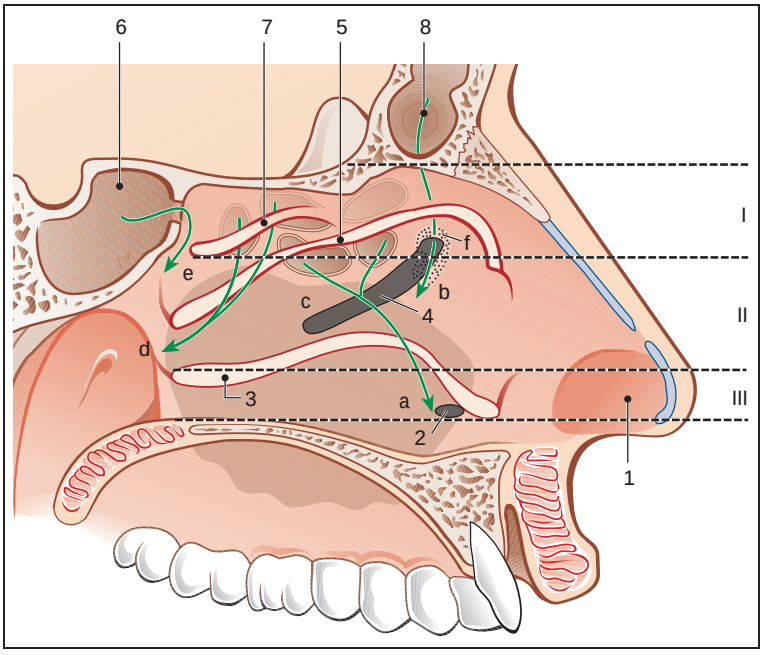


Fig. 2.4 Lateral nasal wall. I, Superior meatus; II, middle meatus; III, inferior meatus. 1, Nasal vestibule; 2, opening of the nasolacrimal duct; 3, origin of the inferior turbinate; 4, semilunar hiatus; 5, insertion of the middle turbinate; 6, sphenoid sinus; 7, insertion of the superior turbinate; 8, frontal sinus. a, Drainage of the antral cavity; b, drainage of the frontal sinus; c, drainage of the anterior ethmoid cells; d, drainage of the posterior ethmoid cells; e, drainage of the sphenoid sinus; f, area of infundibulum (dotted area).

The *internal nasal valve* or limen nasi is a very important structure from the physiologic point of view. It lies at the junction of the vestibule and the nasal cavity. It is formed by a prominence of the anterior edge of the upper lateral or triangular cartilage on the lateral wall of the nose (Fig. 2.3a). The internal nasal valve is normally the narrowest point in the entire cross-section of the nasal cavity, making it an important factor in nasal respiration (see p. 135).

The *nasal cavity* extends from the internal nasal valve to the choana. The structure of the floor and roof of the nose, the medial wall, and the *nasal septum* is shown in Fig. 2.3a, b.

The outline of the lateral wall of the nasal cavity is more complex than that of the medial wall. It contains several structures that are important in the functioning of the nose and nasal cavity (Fig. 2.4):

- Three nasal turbinates (superior, middle, and inferior)
- Drainage of the paranasal sinuses (frontal sinus and maxillary sinus through the hiatus semilunaris in the middle meatus, located between the inferior and middle turbinate; the sphenoid sinus has an ostium of its own in the sphenoethmoidal recess)
- Opening of the nasolacrimal duct into the inferior meatus

The superior, middle, and inferior meatus are located inferior to the three turbinates (Fig. 2.4); the paranasal sinuses and the nasolacrimal duct open into them. These openings are of diagnostic and therapeutic importance.

- The inferior meatus, located between the floor of the nose and the insertion of the lower turbinate, lacks a sinus ostium, but contains the opening of the nasolacrimal duct ≈ 3 cm posterior to the external nasal opening and 3 mm posterior to the head of the inferior turbinate (Fig. 2.4).
- The middle meatus, between the inferior and middle turbinate, is of clinical importance because the frontal recess, the anterior ethmoid cells, and the maxillary antrum open into it (Fig. 2.4).
- The superior meatus, between the middle and superior turbinate, contains the opening for the posterior ethmoid cells. The sphenoid ostium is located on the anterior wall of the sphenoid sinus, at the level of the superior meatus (Fig. 2.4).

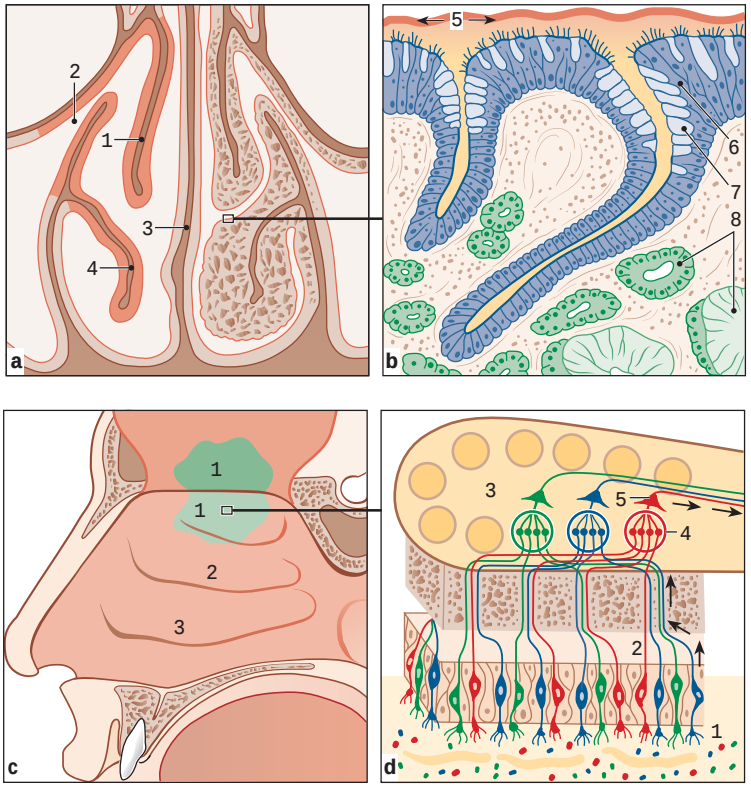


Fig. 2.5a–d a Frontal section through the nasal cavity. The nasal mucosa is constricted on the left side and normal on the right.
b Respiratory mucosa. 1, Middle turbinate; 2, antrum with ostium; 3, nasal septum; 4, inferior turbinate; 5, layer of mucus; 6, respiratory epithelium with cilia; 7, goblet cells; 8, mucosal glands.
c Sagittal section through the nose with the septum reflected superiorly. 1, Olfactory region; 2, middle turbinate; 3, inferior turbinate.
d 1, Scent molecules dock onto receptors; 2, bipolar nerve cells; 3, olfactory bulb with 4, glomerulus (microcenter) and 5, mitral cells.

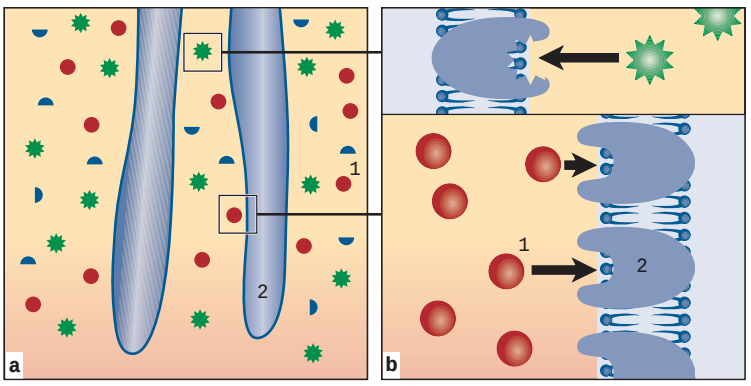


Fig. 2.6a, b a 1, Scent molecules, 2, scent receptors. Each olfactory cell bears only a single type of receptor. There are 350 types of receptor in humans. A receptor can recognize up to 100 structurally similar molecules in a scent category.
b Scent molecules only fit one specific receptor. They trigger a biochemical reaction and a subsequent electrical signal, which is transmitted to the olfactory center in the brain.

The nasal cavity is lined by two types of epithelium: respiratory and olfactory (Figs. 2.5a–d, 2.6a, b). *Respiratory epithelium* coats the entire airway and its projections and extensions (e. g., the paranasal sinuses and the middle ear), from the nasal introitus to the bronchi. It shows morphologic variation in different parts of the respiratory tract. Figure 2.5c, d shows the structure of the respiratory epithelium of the nasal cavity. The epithelium is columnar-ciliated with goblet cells and a layer of mixed glands, a fairly well demarcated lymphoid cell zone and well developed venous cavernous spaces in the turbinates and around the ostia (Fig. 2.5a).

The *olfactory mucosa,* innervated by fibers of the olfactory nerve, covers the area of the olfactory cleft, the cribri-

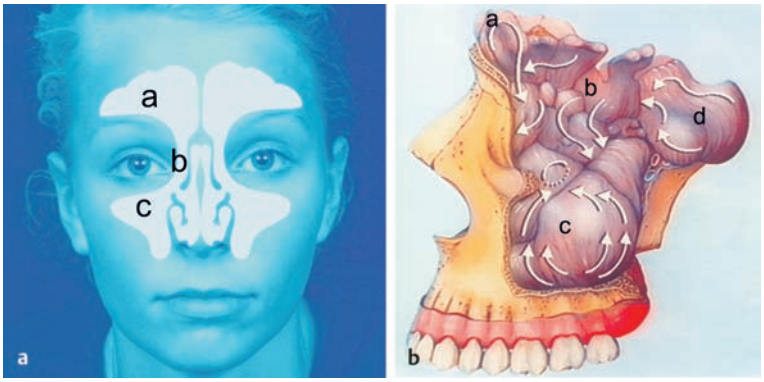


Fig. 2.7a, b The paranasal sinuses.
a Arrangement of the paranasal sinuses in the facial skeleton.
b The mucosal surface of the paranasal sinuses is substantially larger than that of the nose. The arrows indicate the mucociliary transportation of secretions out of the sinuses to the nose.
a, Frontal sinus; b, ethmoid cell system; c, maxillary sinus; d, sphenoid sinus.

Fig. 2.8 Anatomy of the ethmoid bone. 1, Labyrinth; 2, perpendicular lamina; 3, ethmoidal bulla; 4, crista galli.

form plate, part of the superior turbinate, and the part of the septum lying opposite it. The structure is shown in Fig. 2.5 d, and its topographic extent in Fig. 2.5c. Bowman glands occur specifically in this area. They produce a lipo-lipid secretion that covers the olfactory region and aids in olfactory perception due to the enzymes it contains. It is entirely dissimilar to the secretion of the glands of the respiratory epithelium.

■ Paranasal Sinuses

The paranasal sinuses are pneumatized cavities in the bone adjacent to the nose (Fig. 2.7a, b).

Ostiomeatal Complex

The ostiomeatal complex is a functional entity in the anterior ethmoid complex that represents the final common pathway for drainage and ventilation of the frontal, maxillary, and anterior ethmoid cells (see Fig. 2.9). Any or all cells, clefts, and ostia, as well as their dependent sinuses, can become diseased, contributing to the symptoms and pathophysiology of sinusitis.

Ethmoidal Labyrinth

The central structure in this system of pneumatized cavities is the ethmoid bone. The *anterior ethmoid* is the morphologic connection and secretion channel between the nose and the frontal and maxillary sinuses. The tight spaces and clefts normally ensure ventilation and drainage of the sinus mucosa. The anterior ethmoid is central to the pathogenesis of acute, recurrent, and chronic inflammations of the frontal and maxillary sinuses, since ≈ 90 % of all diseases of the frontal and maxillary sinuses begin here.

The central part of the ethmoid bone is T-shaped. In the median plane, the *crista galli* projects into the anterior fossa (Fig. 2.8). The *falx cerebri* inserts here. The *perpendicular plate* (lamina perpendicularis) abuts anteriorly onto the septal *cartilage* and dorsally onto the *vomer*. The paired *ethmoidal labyrinth* is situated between the nose and orbit. It consists of 10 to 15 pneumatized cells lined with *respiratory epithelium*. The volume varies individually; roughly, it is the size of a matchbox standing on its short side.

The *lamina cribrosa* passes into the *ethmoidal notch* (incisura ethmoidalis) of the frontal bone. The *olfactory nerves* extend from the olfactory rim to the olfactory bulb.

The ethmoid air cells are differentiated into anterior and posterior groups. They are derived from an embryonic paired anlage that coalesces in the first year of life to form a single ethmoid cell. There are communications between all of the ethmoid cells on one side. There are often also common ostia for the anterior ethmoid complex in the

middle meatus and for the posterior ethmoid complex in the superior meatus (see Fig. 2.4). Unlike all the other nasal sinuses, the ethmoid labyrinth is fully formed at birth.

Superiorly, it is related to the anterior part of the base of the skull and is an avenue of spread for rhinogenous intracranial infections. The cranial closure of the ethmoidal labyrinth is formed by frontal bone.

Laterally, the lamina papyracea separates the ethmoid cells from the orbital cavity and is a pathway of spread for orbital complications. It is very thin in children.

Posteriorly, the ethmoid labyrinth is related to the sphenoid sinus. The posterior closure of the ethmoid labyrinth is formed by the sphenoid bone. The optic nerve often runs very close to the posterior ethmoid cells, or even within them. This may explain some cases of retrobulbar neuritis.

Medially, the ethmoid labyrinth is related to the middle and superior turbinates.

Maxillary Sinus

The *maxillary sinus* is the largest sinus, with an average volume of 15 mL. The paired sinuses often develop asymmetrically, and the resulting differences in the thickness of the bony wall may give rise to incorrect radiologic diagnoses. The sinus usually consists of a single chamber, but it may have recesses and may even contain separate loculi. This can give rise to difficulties in diagnosis and treatment.

The ostium of the maxillary sinus occupies the superior part of the medial wall of the sinus; it opens not directly into the nose, but sagittally into a three-dimensional space, the *ethmoidal infundibulum.* The infundibulum opens into the nose in the *hiatus semilunaris* in the middle meatus (Fig. 2.9).

The *superior* or *orbital wall* of the maxillary sinus also forms the floor of the orbit. The infraorbital nerve passes through it.

The *medial wall* is also the lateral wall of the nasal cavity. The *anterior wall* contains the infraorbital foramen.

The *posterior wall* separates the sinus from the pterygopalatine fossa. The maxillary artery, the pterygopalatine ganglion, and branches of the trigeminal nerve and autonomic nervous system lie within the pterygomaxillary fossa (see Fig. 2.12).

The *floor* of the maxillary sinus is related to the dental roots in the alveolus, particularly those of the second premolar and the first molar. Odontogenic sinusitis can originate from this site.

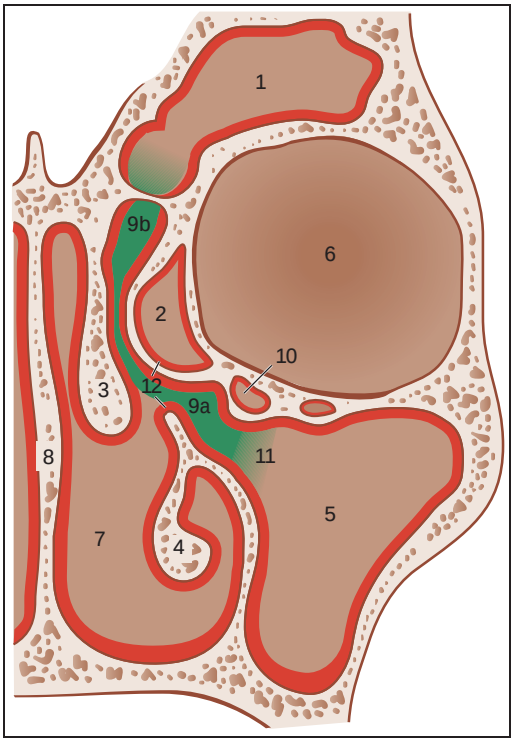


Fig. 2.9 Ostiomeatal unit (green). 1, Frontal sinus; 2, ethmoid sinus; 3, middle turbinate; 4, inferior turbinate; 5, maxillary sinus; 6, orbit; 7, nasal cavity; 8, nasal septum; 9a, ethmoidal infundibulum; 9b, frontal recess; 10, orbital ethmoidal cell; 11, natural ostium; 12, semilunar hiatus.

Before the second dentition erupts—i.e., until about the seventh year of life—the maxillary sinuses are usually very small, since the maxilla contains the tooth buds of the second dentition. The maxillary sinus does not develop its final form and size until after the second dentition appears.

Frontal Sinus

The *frontal sinus* varies in shape and extent more than the maxillary sinus. The average frontal sinus has a capacity of 4–7 mL. There is often a considerable difference in size between the right and left cavities in the same person. The frontal sinuses may be completely absent on one or both sides in 3–5% of individuals, but they may also be very extensive and contain loculi. The latter favor the development of inflammatory complications. In the drainage system of the frontal sinus, the frontal infundibulum

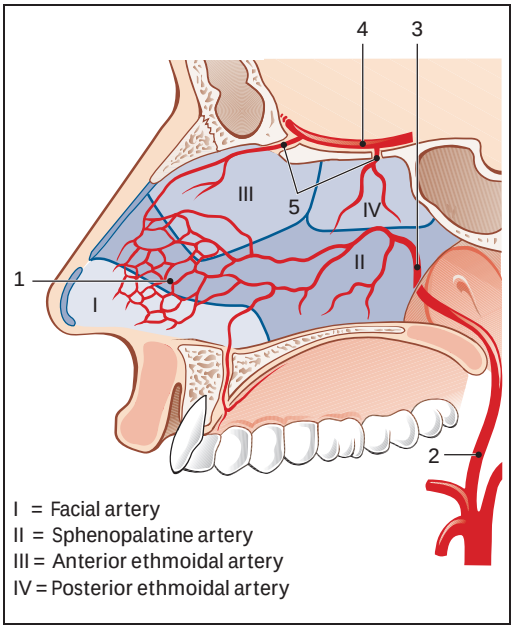


Fig. 2.10 Vasculature of the nasal cavity. 1, Kiesselbach area; 2, internal maxillary artery; 3, sphenopalatine artery; 4, ophthalmic artery; 5, anterior and posterior ethmoid arteries. I–IV: areas supplied by arteries to the nose.

merges into the frontal recess (ethmoidal bone) at the hiatus semilunaris in the middle meatus.

The frontal sinuses form after birth and are not completely developed until the second decade of life. A bony septum separates the two sinuses. The floor of the frontal sinus forms part of the roof of the orbit and is a pathway for the spread of inflammatory orbital complications. The canal of the supraorbital nerve traverses the floor of the frontal sinus.

The posterior wall of the frontal sinus forms part of the bony anterior cranial fossa, making it a typical pathway for the spread of rhinogenous intracranial complications due, either to frontonasal injury or as a complication of sinusitis (see p. 188).

Sphenoid Sinus

The sphenoid sinus does not begin to develop until the age of 5 years.

The *sphenoid* is the most posterior of the sinuses. It occupies the skull base at the junction of the anterior and middle cranial fossae in the body of the sphenoid bone. There are marked individual variations in shape and size, and the capacity of the sinus is 0.5–3.0 mL. The sphenoid sinus may be entirely absent in 3–5% of individuals. The ostium of the sphenoid sinus lies on the anterior wall of the body of the sphenoid bone in the sphenoethmoidal recess, behind and somewhat above the superior turbinate (see Fig. 2.4).

The *superior wall* of the sphenoid sinus is related to the anterior and middle cranial fossae and is a pathway of spread for rhinogenous intracranial complications. The optic chiasm and optic foramen are closely related. The sella turcica and pituitary gland lie on the roof of the sphenoid sinus, which can therefore be used for surgical access to the pituitary.

On the *lateral wall* of the sphenoid sinus lie the cavernous sinus, the internal carotid artery, and the second, third, fourth, fifth, and sixth cranial nerves. The optic canal may lie freely in the lateral wall of the sphenoid sinus, as may the internal carotid artery and the third and sixth cranial nerves.

The *floor* of the sphenoid sinus is related to the roof of the nasopharynx and the choana (see Fig. 2.4).

Mucosa of the Paranasal Sinuses

The *mucosa* of the paranasal sinuses (see Fig. 2.13) consists of *respiratory epithelium. Goblet cells* and *seromucous glands* produce a secretion forming a two-layered film on the mucosa surface.

The lining of the paranasal sinuses is simpler than that of the nasal cavity. Cavernous erectile tissue may be found in the mucosa around the ostia, and this can affect the patency of the ostia (see p. 119). This variability in the opening of the ostia is supplemented by simultaneous variations in the volume of the neighboring turbinates (see Fig. 2.5a). Functionally, they form the *ostiomeatal unit.*

Blood Supply

The *blood supply* to the nasal cavity and nasal sinuses is provided by both the internal and external carotid arteries and their accompanying veins. Figure 2.10 shows the blood supply of the *medial* nasal wall, and Fig. 2.93 shows the supply of the *lateral* nasal wall.

The *external* carotid artery supplies the nose internally via the internal maxillary artery and externally via the facial artery. The sphenopalatine artery is an important branch of the internal maxillary artery.

The *internal* carotid artery gives rise to the ophthalmic artery, and from there to the anterior and posterior ethmoidal arteries.

There is a particularly rich and relatively superficial plexus of small vessels (the *Kiesselbach plexus*) located on the anterior part of the nasal septum (Fig. 2.9). It is supplied ultimately by both the internal and external carotid arteries.

Venous drainage is provided by the ophthalmic and facial veins and the pterygoid and pharyngeal plexuses. It is therefore located both partially intracranial to the cavernous, coronary, and transverse sinuses, and partially extracranial (see Fig. 2.2).

The *cavernous spaces* within the mucosa of the nasal turbinates are very important clinically, as are those on the nasal septum and around the ostia of the nasal sinuses. The filling of these spaces with venous blood is highly variable and is under autonomic control. By regulating the thickness of the mucosa, the cavernous spaces of the turbinates can change the cross-sectional areas of the nasal cavity and of the ostia of the nasal sinuses, thus controlling respiration, ventilation, and drainage (see Fig. 2.5a).

Lymphatic Drainage

The lymphatic drainage consists of two parts: an *anterior system*, which collects the lymph from the nasal pyramid and drains to the submandibular superficial cervical lymph nodes; and a *posterior system,* which drains the posterior part of the nasal cavity and the nasopharynx to the retropharyngeal lymph nodes and jugular nodes.

Nerve Supply

The nasal cavity and nasal sinuses have a sensory and autonomic (secretory and vasomotor) nerve supply. In addition, they include the special sensory function of the olfactory nerve.

The *sensory nerve supply* is provided by the first and second branches of the trigeminal nerve. The complex autonomic innervation for secretion and vasomotor supply is shown in Fig. 2.11.

Autonomic innervation: The *sympathetic fibers* for vasoconstriction arise from the first to the fifth thoracic segments of the spinal cord, and synapse in the superior cervical ganglion. The postganglionic fibers follow the blood vessels to the mucosa of the nose and nasal sinuses. Some fibers run through the pterygopalatine ganglion without synapsing.

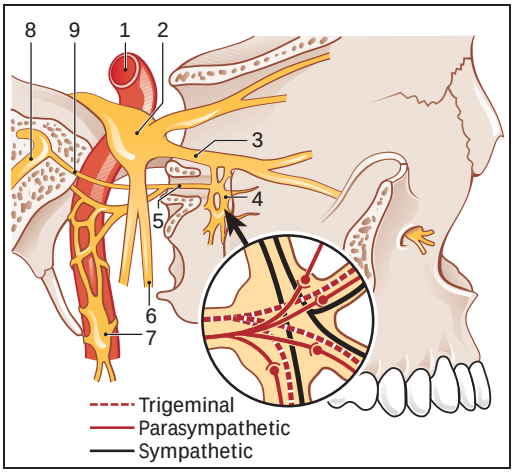


Fig. 2.11 Nerve supply of the nasal mucosa. 1, Internal carotid artery with sympathetic plexus; 2, Gasserian ganglion; 3, maxillary nerve; 4, pterygopalatine ganglion; 5, nerve of the pterygoid canal; 6, mandibular nerve; 7, superior cervical ganglion; 8, facial nerve with intermediate nerve; 9, greater petrosal nerve. The inset shows the course of fibers in the pterygopalatine ganglion: the black lines indicate sympathetic fibers, the solid red lines indicate parasympathetic fibers, and the dotted red line indicates the trigeminal nerve.

The pathway for the *parasympathetic fibers* for vasodilation is from the lacrimomuconasal nucleus along the intermediate nerve to the geniculate ganglion and then along the facial nerve, the greater superficial petrosal nerve, and the nerve of the pterygoid canal to the pterygopalatine ganglion.

The preganglionic parasympathetic fibers synapse in the pterygopalatine ganglion. From there, they supply the mucosa of the nose and nasal sinuses with secretory and vasodilator fibers.

The *pterygopalatine* or *sphenopalatine ganglion* plays a key role in the functioning of the nose and nasal sinuses. It is the main site of its autonomic innervation and has three roots:

1. Parasympathetic fibers, which supplement secretory and vasodilator functions.
2. Sympathetic fibers for vasoconstriction and inhibition of secretion.
3. Sensory fibers from the trigeminal nerve, which arise in the trigeminal ganglion and run in the maxillary nerve.

The nose and maxillary sinus are closely related both anatomically and functionally to the *maxilla.* This bone forms the upper half of the masticatory

system and also forms the main part of the middle third of the facial skeleton. The upper jaw is important in diseases of the nose, due to its immediate relationship to the nose and nasal sinuses.

Basic Physiology and Pathophysiology

The nose is both a *sensory organ* and a *respiratory organ.* In addition, the nose performs an important function for the entire body by providing both physical and immunologic protection from the environment. Finally, it is also involved in the formation of speech sounds. The nose is also a major esthetic unit in the center of the face. It balances and unites the other esthetic units, paired and unpaired, such as brows, cheeks, forehead, and lips.

The Nose as an Olfactory Organ

The human sense of smell is poorly developed in comparison with that of most mammals and insects. Nevertheless, it is still very sensitive and almost indispensable for the individual. For example, taste is only partly a function of the taste buds, which can only recognize the qualities of sweet, sour, salty, and bitter. All other sensory impressions caused by food, such as aroma, bouquet, are mediated by olfaction. This *gustatory olfaction* is due to the fact that the olfactory substances of food or drink pass through the olfactory cleft during expiration while eating or drinking. The sense of smell can stimulate appetite but can also depress it. It also provides a warning against spoiled or poisonous foods and toxic substances—e. g., gas. The sense of smell is particularly important in the field of psychology; marked affects may be induced or inhibited by smells. It should also be remembered that a good sense of smell is essential for people in certain occupations—e. g., cooks and confectioners; wine, coffee, and tea merchants; perfumers; tobacco blenders; and chemists. Finally, physicians need a "clinical nose" for making diagnoses.

The *olfactory area* of the nose is relatively small, at ≈ 2–5 cm^2. The cell bodies of the olfactory cells are located in the olfactory epithelium covering the superior nasal passage, the cranial nasal septum, and the middle turbinate (see Fig. 2.5c). The olfactory cells are primary sensory cells that project directly into the olfactory bulb. Their axons are bundled into the olfactory filaments, which pass through the cribriform plate to the olfactory bulb. Between a hundred and a thousand axons together reach each mitral cell. Glutamate is the neurotransmitter between the sensory cells and mitral cells and the messenger of the mitral cells. Other neurotransmitters are γ-aminobutyric acid and dopamine. Important ganglia for further processing within the central nervous system are located in the hippocampus and amygdala, which are responsible for processing emotions and memory content. Finally, conscious olfactory perception occurs at the level of the cortex (superior temporal gyrus, orbitofrontal cortex, insula region).

The olfactory epithelium can regenerate within 100 days, which is unique for sensory organs.

During olfaction, scents first dock onto the olfactory binding protein in the olfactory region. In this way, they reach the scent receptors located on the bipolar olfactory receptor neurons. The receptors are located within the membrane, where they bind to a specific olfactory G protein (second messenger; cyclic adenosine monophosphate).

Richard Axel and Linda Buck were the first to examine the sense of smell at the molecular level. They discovered that every olfactory cell is specialized for only a single class of scent and is equipped with only one type of receptor. Human beings have 350 types of receptor. One receptor can recognize up to a hundred molecules with structural similarities. Scent molecules have to fit into a receptor like a key into a lock. Docking of a molecule activates a chemical cascade. The scent is transformed into a electrical signal, which is transmitted to a scent center in the brain. The two scientists were awarded the Nobel Prize in 2005 for this discovery.

Only volatile substances can be smelled by humans. These substances have to be soluble in water and lipids. Only a few molecules can be sufficient to stimulate the sense of smell. On average, 10^{-15} molecules per milliliter of air are enough to exceed the stimulation threshold.

It is said that there are ≈ 30 000 different olfactory substances in the atmosphere; of these, humans can perceive ≈ 10 000 and are able to identify ≈ 200.

The sense of smell, like other senses, demonstrates the phenomenon of *adaptation.* The sensitivity of the olfactory organ is also dependent on hunger. A very hungry individual can perceive several olfactory stimuli better than one who has just eaten, and this is a useful physiologic regulating mechanism.

Olfactory Disorders (Table 2.1)

Sinonasal olfactory disorders. These are the most common disorders encountered in otolaryngology. Inflammatory or noninflammatory changes impede

the transportation of scents to the olfactory cleft or damage the olfactory epithelium directly.

- Infectious causes: chronic recurrent rhinosinusitis
- Noninfectious causes: allergies, polyposis, hyperplastic rhinosinusitis, postirritative and toxic, postinfectious, rhinitis sicca
- Noninflammatory causes: anatomic (malignant and benign tumors, stenoses, choanal atresia, adhesions, septum deviations) or nasal congestion, nerve reflexes, idiopathic rhinitis (formerly called nasal hyperreactivity), side effects of drugs

Note: Esthesioneuroblastoma is a highly malignant lesion, the most serious tumor that can affect the olfactory epithelium. It needs to be considered in every patient with an olfactory disorder.

Nonsinonasal olfactory disorders. Viral infections can lead to primary damage to the olfactory cells. Typically, patients experience a subjective loss of the sense of smell immediately after an infectious disease. Neurologic, congenital, toxic, and psychiatric conditions may involve olfactory disorders, as well as head injuries. Dysosmias may be an initial symptom of Alzheimer disease, preceding loss of cognition and abnormal behavior. In Parkinson disease, olfactory disorders often occur before motoric disorders. Noxious substances such as carbon dioxide, formaldehyde, or tobacco smoke can directly damage the olfactory cells. Congenital dysosmias (e.g., Kallmann syndrome) are rare.

A large body of clinical evidence suggests that olfactory disorders are associated with certain neurological and psychiatric diseases (Table 2.2), and as such may be associated with pronounced emotional changes. One of the most common findings is temporal lobe epilepsy, which is associated with an olfactory aura. Olfactory hallucinations are usually unpleasant and lead to corresponding emotional effects, frequently with feelings of "déjà vu," which in turn elicit motor and sensory phenomena.

An olfactory disorder is regarded as idiopathic if all other causes have been ruled out.

Table 2.1 Disorders of olfaction—dysosmias

Quantitative disorders	Hyperosmia—oversensitivity
	Normosmia—normal sensitivity
	Hyposmia—reduced sensitivity
	Anosmia—complete inability to perceive scents
Qualitative disorders	Parosmia—altered perception of scents in the presence of a stimulus
	Phantosmia—perception of scent in the absence of a stimulus
	Anosmia—inability to identify perceived scents

Table 2.2 Dysosmia associated with neurological or psychiatric disorders

Abnormal recollection of smells (déjà vu, jamais vu)
Illusions/hallucinations
Schizophrenia
Alcoholism
Senile dementia
Depression

The Vomeronasal Organ

Adolf Butenandt (1903–1995), a Nobel prizewinner from Munich, coined the term *pheromones* for molecules that are produced by a species and evoke certain reactions in animals of the same species. The vomeronasal organ (Jacobson organ) is essential for social and mating behavior in all mammals. With the exception of some of the higher primates, mammals mate only when the female is fertile. Information on the timing of ovulation is conveyed to males by means of pheromones. Numerous observations and studies indicate that pheromones also convey signals in humans.

The vomeronasal organ consists of tiny, paired, blindly terminating channels located in the anterior nasal septum. Its morphology suggests that the organ is a functioning sensory epithelium. Further studies are needed to identify its central connections with the hypothalamus and elucidate its functional significance.

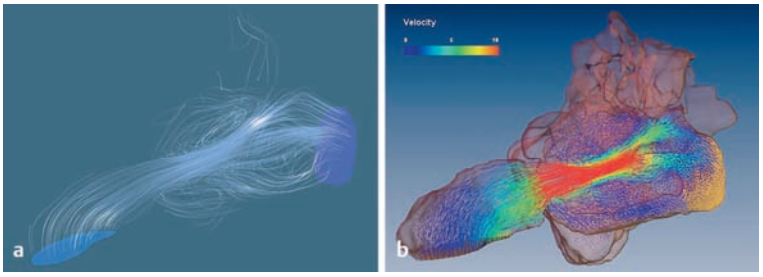


Fig. 2.12a, b Flow patterns in the nose. Images courtesy of T. Hildebrandt, Birkenwerder, Germany.

The Nose as a Respiratory Organ

In humans, the only physiologic respiratory pathway is via the nose. Mouth breathing is abnormal and is activated only as an emergency supplement to nasal respiration. The physiology of the air stream through the normal nose during inspiration and expiration can be summarized as follows. Average ventilation through a normal nose during normal breathing is 6 L/min, and 50–70 L/min during maximum ventilation. The internal nasal valve or limen nasi is the narrowest point in the normal nose. It acts as a nozzle, and the speed of the airstream is very high at this point (Fig. 2.12a, b)

The nasal cavity between the valve and the head of the turbinates acts as a diffuser—i. e., it slows the air current and increases turbulence. The central part of the nasal cavity, with its turbinates and meatus, is the most important part for nasal respiration. The column of air consists of a laminar and a turbulent stream. The ratio of laminar to turbulent flow considerably influences the functioning and condition of the nasal mucosa.

The airstream passes in the reverse direction through the nasal cavities during expiration. The expiratory airstream shows much less turbulence in the central part of the nose, reducing the exchange of heat and products of metabolism between the airstream and the nasal wall in comparison with inspiration and enabling the nasal mucosa to recover during the expiratory phase. Inspiration through the nose followed by expiration through the mouth causes the nasal mucosa to dry out rapidly.

Nasal resistance—i. e., the difference in pressure between the nasal introitus and the nasopharynx—is normally between 8 mm and 20 mmH_2O. If resistance exceeds 20 mmH_2O, the internal nasal valves expand during breathing. Supplemental mouth breathing begins at levels exceeding 40 mmH_2O.

Complete exclusion of the nose from breathing leads in the long term to deep-seated mucosal changes. Mechanical obstruction within the nose—e. g., due to septal deviation, hypertrophy of the turbinates, stenoses from scarring, etc.—can lead to mouth breathing and the resulting deleterious effects (see p. 184), as well as causing mucosal diseases of the nose and the paranasal sinuses.

Computational fluid dynamics (CFD)—simulation of nasal airflow. CFD is the most recent digital method for fluid examination. It can be used to analyze airflow phenomena in the nose (Fig. 2.12a, b). It provides data on the important value of integral pressure loss, as well as extensive information about flow, involving velocity vectors, pressure, and turbulences with high resolution, providing detailed information about the fluid flow.

CFD requires five working steps:

1. Creating a geometric model (computed tomography).
2. Generating a computational grid.
3. Preprocessing (physical modeling).
4. Executing calculations (mathematical modeling, equation solving).
5. Postprocessing (graphic and quantitative analysis of results).

In the future, CFD may become an important tool for testing and designing prototypes for the shape of the nasal airway before surgical correction.

Nasal patency. *Nasal patency* can be influenced by many different factors, including the temperature and humidity of the surrounding air; the position of the body; physical activity; changes in body temperature; the effect of cold on different parts of the body, such as the feet; hyperventilation; and psychological stimuli. The state of pulmonary function and of the heart and circulation, endocrinologic conditions such as pregnancy, hyperfunctioning or hypofunctioning of the thyroid gland, and some topical, oral, or parenteral drugs may have a considerable influence on the patency of the nose (see

p. 135). Methods of measuring nasal patency are described on p. 135.

During normal nasal respiration, the inspired air is warmed, moistened, and purified as it passes through the nose.

The *warming* of air inspired through the nose is very effective, and the constancy of the temperature in the lower airways is remarkably stable. The nasal mucosa humidifies and warms the air. The temperature in the nasopharynx during normal (exclusively nasal) respiration is constant at 31–34°C, independently of the external temperature. The heat output of the nose increases as the external temperature falls, so that the lower airways and lungs can function at the correct physiologic temperature.

The *optimal relative humidity* of ambient air for subjective well-being and function of the nasal mucosa lies between 50 % and 60 %. The water vapor saturation of inspired air in the nasopharynx is 80–85 %, and in the lower airway it is fairly constant at between 95 % and 100 %, independent of the relative humidity of the surrounding air. The amount of water vapor secreted by the entire respiratory tract per 1000 L of air can reach 30 g, most of it supplied by the nose. On the other hand, the mucosal blanket makes the nasal mucosa watertight, preventing the release of too much water into the air and consequent drying of the mucosa.

The *cleansing function* of the nose includes firstly, clearing the inspired air of foreign bodies, bacteria, dust, etc.; and secondly, cleansing the nose itself (see below). About 85 % of particles larger than 4.5 µm are filtered out by the nose, but only ≈ 5 % of particles less than 1 µm in size are removed.

Foreign bodies entering the nose come into contact with the moist mucosal surface and the mucosal blanket, which continually sweeps foreign bodies away. The details are described below.

Note: The nose warms, moistens, and cleanses atmospheric air as the most important preconditions for normal breathing.

The Nasal Mucosa as a Protective Organ

In addition to warming, humidifying, and cleansing the inspired air, the nose also has a protective function, consisting of a highly differentiated, efficient, and polyvalent resistance potential against environmental influences on the body. A basic element of this defensive system is the *mucociliary apparatus* (Fig. 2.13a, b)—i. e., the functional combination of the secretory film and the cilia of the respiratory epithelium, by which the colloidal secretory film is transported continuously from the nasal introitus toward the choana. A foreign body is carried from the head of the inferior turbinate to the choana in ≈ 10–20 min. The efficiency of this cleansing system depends on several factors, such as pH, temperature, the condition of the colloids, humidity, the width of the nose, or the presence of toxic gases. Disturbances in the composition or physical characteristics of the mucosal blanket or of ciliary activity can markedly influence the physiology of the nasal cavity. The nasal mucosa protects the entire body by making contact with and providing resistance against animate and inanimate foreign material in the environment.

Local specific immune defense of the nasal mucosa is ensured by humoral and cellular mechanisms (antibodies and immunologically competent cells).

Humoral defense mechanisms. Paraglandular plasma cells produce antibodies—immunoglobulin A (IgA), IgM, and IgG. The glands of the lamina propria absorb the immunoglobulins (e. g., IgA) and then release them as secretory antibodies (sIgA) at the epithelial surface.

Cellular defense mechanisms. The main forces of cellular defense are neutrophilic, basophilic, and eosinophilic granulocytes, macrophages, mast cells, and T and B lymphocytes. B lymphocytes are able to differentiate into plasma cells, which can secrete immunoglobulins. Neutrophils are increased in chronic rhinosinusitis, eosinophils predominate in chronic rhinosinusitis with polyposis, and degranulating mast cells and basophils are important in type I allergic reactions. Intercellular adhesion molecule-1 (ICAM-1) is expressed by the epithelium of the nasal mucosa; it serves as a receptor for viruses (rhinoviruses). Cells of the vascular endothelium are stimulated by inflammation mediators such as tumor necrosis factor-α (TNF-α) and interleukin-1 (IL-1) to express adhesion molecules that ensure diapedesis by immunologically competent cells.

The Nose as a Reflex Organ

Specific nasal reflex mechanisms can arise:

- Within the nose and affecting the nose itself.
- In other parts of the body or organs and affecting the nose.
- In the nose and affecting other parts of the body.

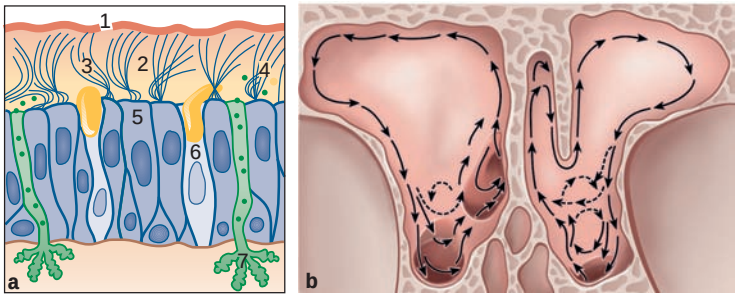


Fig. 2.13a, b a The mucociliary apparatus is the most important defense mechanism in the respiratory epithelium. Goblet cells and seromucous glands produce a two-layered film consisting of a low-viscosity sol phase and a higher-viscosity gel phase. The cilia beat in the sol phase to transport the gel along distinct pathways across the mucosa of the nose and paranasal sinuses toward the choana. The mucociliary apparatus simultaneously facilitates secretion and absorption, and it is the region where initial nonspecific and specific immunological responses to antigens and allergens occur. 1, Gel layer; 2, sol layer; 3, cilia (beating); 4, cilia (recovering); 5, ciliated epithelial cell; 6, goblet cell; 7, seromucous gland.
b Mucociliary transportation of secretions in the frontal sinus (adapted from Messerklinger).

The *nasal cycle* is a reflex system that is obviously confined to the nose, but its purpose is unknown. One cycle lasts between 2 and 6 hours. If both halves of the nasal cavity are of normal patency, the lumen widens and narrows alternately, lowering or increasing respiratory resistance in each half of the nose. Ideally, the resistance of the entire nose remains constant. This reflex phenomenon is controlled by the action of the autonomic nervous system on the cavernous spaces of the vascular system of the nasal mucosa.

Nasopetal reflexes arise in situations such as cooling of the extremities, which changes the respiratory resistance. They may also arise from the lungs and bronchi and from other autonomic control points.

Important *nasofugal* communications exist between the nose and the lung, the heart and circulation, the metabolic organs, and the genitals.

In addition, there are sneezing, lacrimal, and cough reflexes, and in certain emergency situations, reflexive respiratory arrest.

■ Influence of the Nose on Speech

The nose influences the sound of speech. During the formation of the resonants “m,” “n,” and “ng,” for example, the air streams through the open nose, whereas during the formation of vowels the nose and nasopharynx are more or less closed off by the soft palate from the resonating cavity of the mouth.

■ Function of the Nasal Sinuses

The function of the paranasal sinuses is one of the oldest and most fascinating controversies in medicine. Many theories have been proposed—e. g., that they serve to reduce the weight of the facial skull, provide resonation spaces for the voice, absorb facial trauma forces, moisturize and warm inspired air, provide olfactory function and thermic insulation, are functionless hollows, or provide a source of secretions for the main nasal cavity.

In an experimental study, one important question was answered in 1990. The mucosa of the paranasal sinuses plays a role in secretion production and control of the nasal mucosa. In other words, when the human organism needs higher humidity to moisten the incoming air, such as during sports activity, the sinuses increase secretion and convection through the ostium to the lateral wall mucosa.

The presence of the *ostia* causes particular pathophysiologic problems affecting ventilation and drainage. Ostial obstruction interrupts the self-cleansing mechanism of the affected sinus, causing the secretions to stagnate and change in composition. The retained secretions are an ideal medium for saprophytic bacteria, which are often present in normal sinuses. Ostial obstruction can lead to a vicious circle, as illustrated in Fig. 2.14.

The main cause of closure of the ostium is rhinogenous swelling due to a reaction or infection in

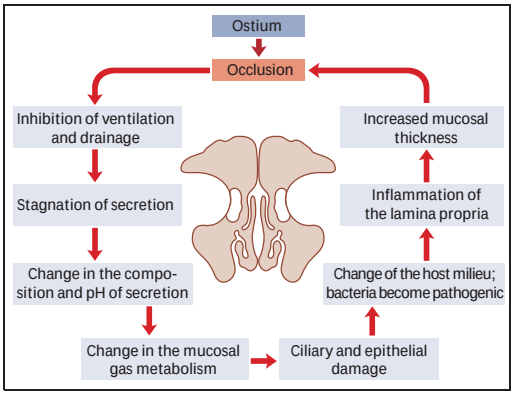


Fig. 2.14 Pathophysiology of the sinus ostium.

the nasal mucosa. Ninety percent of all sinusitis cases begin in the anterior ethmoid *(ethmoidal infundibulum, facial recess, and middle meatus).* The closure mechanism can be induced by viral or bacterial infection of the nose, barotrauma, hyperreactivity, environmental factors such as relative dryness of the nose, toxic gases, or airborne toxic agents. Other causes include localized congenital or acquired anomalies such as deviation of the septum, scars, lesions of the turbinates, infections of the nose or nasal cavities, allergic diseases of the nose or nasal sinuses (particularly in children), vasomotor dysfunction due to neurogenic or hormonal factors, metabolic diseases such as vitamin deficiencies, diabetes, electrolyte disorders, mechanical obstruction due to crusts, ethmoidal polyps, foreign bodies, prolonged use of a nasogastric tube or prolonged nasal tracheal intubation, and benign and malignant tumors.

The vicious circle of ostial occlusion can only be broken in the long term by treating the causative factors with appropriate medical or surgical measures (Fig. 2.14)

The nasal sinuses are only minimally involved in respiratory phases in the nasal cavity. Only slight changes in pressure are recorded in the sinuses during respiration. When the ostium is occluded, a relatively small drop in pressure in the sinuses (from –20 to –50 mmH_2O) occurs, which is enough to elicit the symptoms of what has been inappropriately termed *vacuum sinusitis.* The symptoms include moderately severe headaches, which disappear when ventilation of the sinus is restored.

Methods of Examining the Nose, Paranasal Sinuses, and Face

External Inspection and Palpation

During external examinations, attention is given to the following points:

- The properties of the overlying skin—e. g., hardness, firmness, discolorations, inflammatory swellings, and tenderness to pressure.
- Externally visible changes in shape of the cartilaginous or bony structure due to congenital or acquired deformities—e. g., saddle nose, hump nose, broad nose, or scoliotic nose; the early or late results of trauma; painful swelling due to inflammation; nonpainful swelling due to tumor.
- Palpable masses in neighboring structures—e. g., the forehead, cheeks, upper lip, or eyelids; proptosis, displacement of the bulb or limitation of its movement.
- The nasal alae during respiration, inspection for indrawing or flaring of the ala.
- The nasal vestibule, the anterior border of the nasal septum, the roof of the vestibule, and the internal part of the nasal cavity, inspected by lifting the tip of the nose.
- Crepitation and mobility of the nasal bony framework.
- The sites of exit of the various nerves (Fig. 2.15a, b).
- Sensitivity to pressure on the forehead, cranial vault, or cheek.

Anterior Rhinoscopy

Anterior rhinoscopy using a nasal speculum, a strong light source, and a head mirror or headlamp is only carried out after inspection without instruments (Fig. 2.16 a, b). The method of using the nasal speculum is shown in Fig. 2.17 a, b. Usually, the left hand holds the speculum when inspecting both nasal cavities. Anterior rhinoscopy alone is now considered insufficient, but it is the first step in examining the nose.

Technique. The speculum is introduced into the nasal vestibule with its blades together. The point of the speculum is directed slightly laterally in the nasal vestibule.

The speculum blades are then spread within the nasal vestibule and fixed to the nasal ala with the index finger. The instrument is held slightly open when it is being removed, to prevent pain due to avulsion of vibrissae. The right hand is used to adjust the position of the face and head. As shown in Fig. 2.17a, the patient's head is initially in the vertical position, so that the examiner's gaze is parallel to the floor of the nose and along the inferior turbinate and the inferior meatus (position I). If the nasal cavity is wide, the choana and the posterior wall of the nasopharynx can be seen in this position. To allow inspection of the upper part of the nasal cavity, the patient's head is tilted slightly backward. The middle meatus, which is very important clinically, and the middle turbinate are thus brought into view (position II) (Fig. 2.17b). If the head is tilted far backward, the olfactory cleft may also be visible.

Small children and infants, it is advisable to use an otoscope rather than a nasal In speculum for anterior rhinoscopy.

When the head position has been adjusted satisfactorily, the hand holding the speculum can be used simultaneously to fix the head, leaving the right hand free for manipulating instruments, performing aspirations, etc., within the nasal cavity.

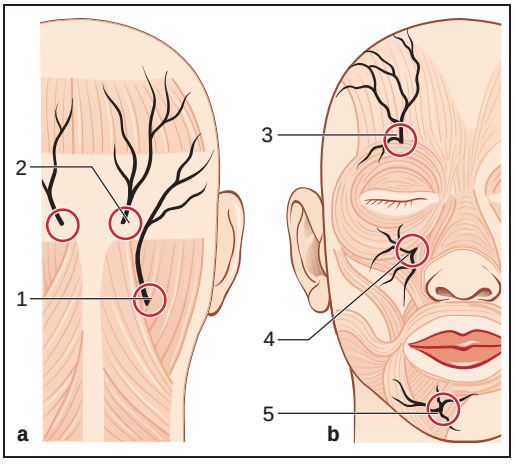


Fig. 2.15a, b Clinically important nerve exit sites.
a At the occiput: 1, lesser occipital nerve; 2, greater occipital nerve.
b On the face: 3, supraorbital nerve; 4, infraorbital nerve; 5, mental nerve.

> Note: The nasal mucosa is often so turgescent that the view of the nasal cavity is minimal. In such cases, a decongestant spray is introduced into the nose and allowed to act for 10 min, after which a good view is usually possible.

The following are noted during anterior rhinoscopy:

- Nasal secretions, their color, quantity, and properties; mucus, pus, and crust formation.
- Location of abnormal secretion.
- Swelling of the turbinates, narrowing or widening of the nasal meatus.

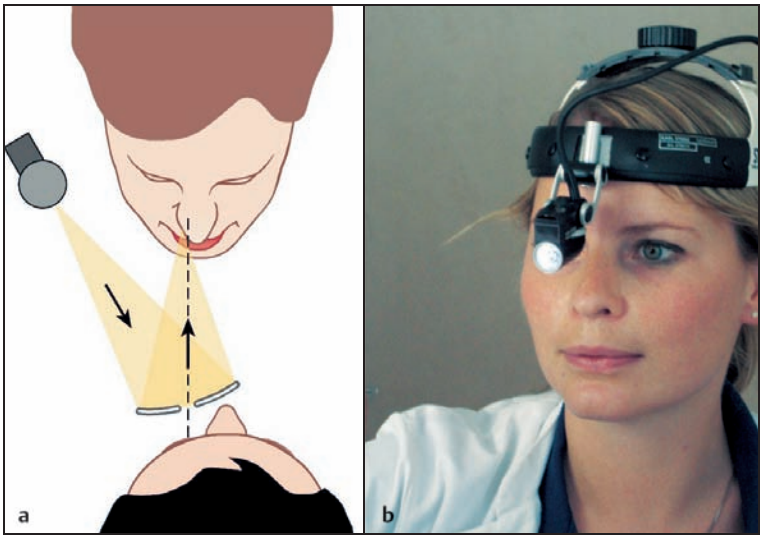


Fig. 2.16a, b a Traditional rhinoscopy, using a head mirror.
b Head lamps with cold-light or light-emitting diode (LED) illumination have now largely replaced mirrors.

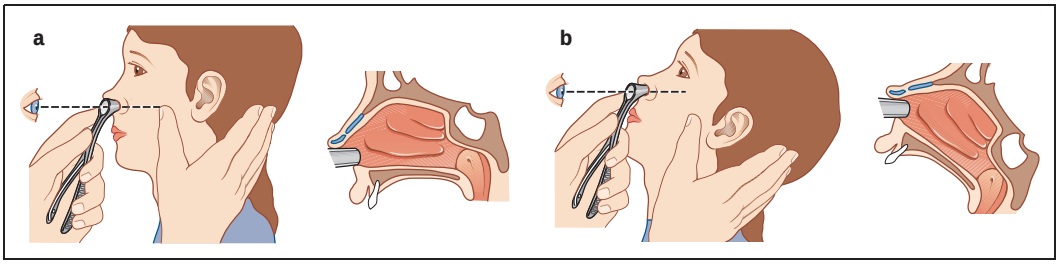


Fig. 2.17a, b Anterior rhinoscopy. a Position I. b Position II.

- Properties of the mucosal surface (including its color)—e. g., whether it is moist, dry, smooth, cornified, or uneven.
- Position of the nasal septum and septal deformities.
- Sites of major blood vessels—e. g., Kiesselbach plexus.
- Abnormal pigmentation or color of the mucosa.
- Presence of abnormal tissue.
- Ulcerations or perforations.
- Foreign bodies.

The clinically important region of the middle meatus may be anatomically narrow and difficult to examine. It can be visualized by using a long Killian nasal speculum, provided that local anesthesia of the nasal mucosa has been induced (e. g., by local application of lidocaine (Xylocaine); or 1 % pantocaine with 1 : 1000 epinephrine, one drop to 1 mL of anesthetic solution; or with a 5 % lidocaine and 0.5 % phenylephrine spray). Gustav Killian (1860–1921) developed this instrument for the medial rhinoscopy technique, having recognized as early as 100 years ago that the lateral nasal wall is important in pathogenesis. The technique of anterior rhinoscopy using nasal endoscopy was added more recently.

■ Posterior Rhinoscopy

Figure 2.18a, b shows the method of examining the nasopharynx with a mirror, with a composite view of the inspected region. Posterior rhinoscopy is used to examine the posterior part of the nasal cavity: the choana, the posterior ends of the turbinates, the posterior margin of the septum, and the nasopharynx, including its roof and the ostia of the eustachian tube.

Nasal endoscopy, including the nasopharynx, should nowadays be an integral part or an adjunctive measure in any examination assessing an otorhinolaryngology patient. It has superseded posterior rhinoscopy and can be regarded as the gold standard for obtaining nasal findings, regardless of the further management.

Technique. Posterior rhinoscopy requires a considerable amount of practice on the part of the examiner, as well as cooperation by the patient. A tongue depressor is placed on the center of the base of the tongue with one hand, and the base of the tongue is pressed slowly downward, increasing the distance between the surface of the tongue and the soft palate and posterior pharyngeal wall. The glass side of a small mirror is warmed and then tested on the hand to make sure it is not too hot. The other hand is used to introduce the mirror into the space between the soft palate and the posterior pharyngeal wall. The mirror must not touch the mucosa, otherwise it will elicit the gag reflex. If the soft palate remains tense, the patient is asked to breathe gently through the nose, to sniff, or to say “ha,” causing the palate to relax and providing an unobstructed view into the nasopharynx. A view of the various parts of the nasopharynx is obtained by moving and tilting the mirror (Fig. 2.18b). The vertical posterior edge of the septum is used for orientation to locate the normal structures. If a satisfactory view cannot be obtained due to the gag reflex, a successful examination can often be achieved by applying a local anesthetic—e. g., a 1 % lidocaine spray—to the oropharynx, particularly to the soft palate and posterior wall of the pharynx.

If it is not possible to examine the nasopharynx with this method, either endoscopy (see p. 133) or palatal retraction (or both combined) can be carried out, although endoscopy has now made palatal retraction largely obsolete.

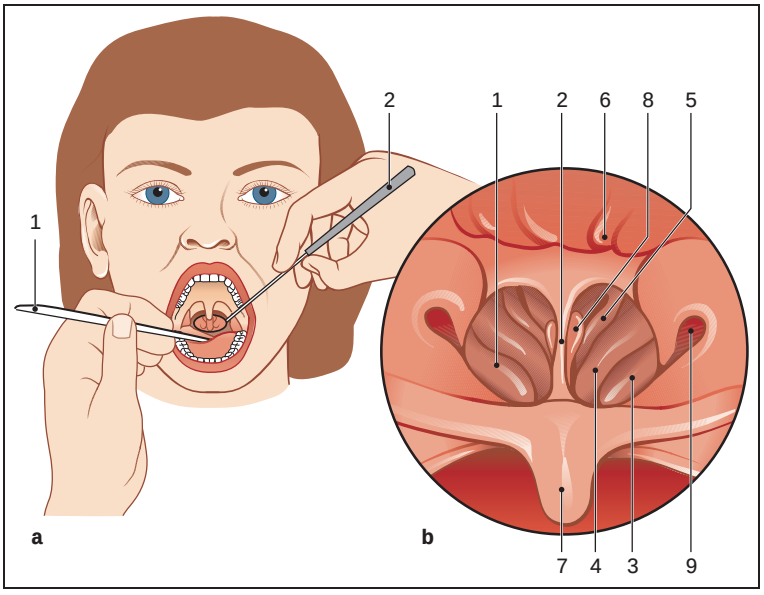


Fig. 2.18a, b Posterior rhinoscopy.
a The method of holding the tongue depressor (1) and mirror (2).
b Composite picture of the nasopharynx composed of individual views: 1, choana; 2, posterior edge of the septum; 3, inferior turbinate; 4, middle turbinate; 5, superior turbinate; 6, adenoid; 7, uvula; 8, septum padding; 9, tubal ostium.

The following should be noted during posterior rhinoscopy (Fig. 2.19):

- The opening and width of the choanae.
- The shape of the posterior end of the inferior and middle turbinates.
- Scars or deformities in the nasopharynx—e. g., due to trauma.
- The shape of the posterior part of the nasal septum.
- Nasal polyps.
- The shape of both tubal ostia and the Rosenmüller fossa (pharyngeal recess).
- Obstruction of the nasopharynx by enlarged adenoids in children.
- Tumors of the nasopharynx.
- Abnormal secretions in the choanae.
- The properties of the mucosa of the posterior part of the nose and the nasopharynx—e. g., for moisture, dryness, thickening, and color.

Computed tomography (CT) is used to evaluate the spread of paranasal sinus lesions to adjacent structures, particularly the skull base, cranial cavity, retromaxillary space, and orbits. Also used in trauma patients, CT is indispensable for differentiated evaluation of bony structures. It is supplemented by magnetic resonance imaging (MRI) for soft-tissue investigations.

Nasal Endoscopy

The principle used in the rod lens system was submitted to the Patent Office in the United Kingdom by its inventor, the English physicist Harold Horace Hopkins (1918–1994), on July 16th, 1959. The rod lens system uses precision-polished glass rods with optically processed ends instead of conventional lenses. The system has substantial advantages over conventional lens systems: better resolution and contrast, a wider angle of view, and extremely clear and bright images that resolve even the smallest details throughout the entire field of view.

The German inventor Karl Storz (1911–1996) introduced the system into otorhinolaryngology and developed cold light illumination. This marked the beginning of a new era in otorhinolaryngology—the era of endoscopy.

The Austrian physician Walter Messerklinger (1920–2001) studied mucociliary transport in the paranasal sinus mucosa over a period of several years. He used the new scopes to explore the lateral nasal wall and obtained significant pathogenetic findings:

- The vast majority of cases of recurrent and chronic sinusitis are rhinogenous in origin.
- Inconspicuous signs of mucosal disease or anatomical configurations predisposing to sinusitis are detectable with endoscopy.

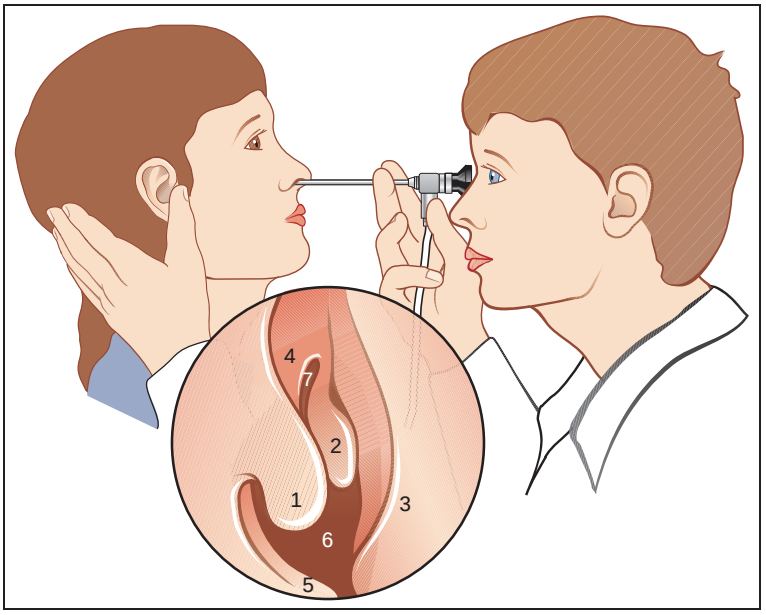


Fig. 2.19 The examination setting for nasal endoscopy. The inset shows typical landmarks when viewing the interior of the right side of the nose through a 0° lens. 1, Inferior turbinate; 2, middle turbinate; 3, nasal septum, 4, agger nasi; 5, floor of the nose; 6, inferior nasal meatus, 7, middle nasal meatus.

- The endoscope is thus an important optical device for elucidating the spreading pathway of spread of rhinogenous sinusitis.
- Endoscopic devices have therefore made new surgical approaches possible. Endoscopic surgery can be directed at the central pathogenetic mechanism of recurrent or chronic sinusitis (see functional endoscopic sinus surgery, p. 171).

Nasal endoscopy is the most important method for evaluating intranasal findings (Fig. 2.20a–d).

Clinical indications for nasal endoscopy. These include acute, recurrent and chronic sinusitis; head and facial pain; chronic nasal catarrh; epistaxis; epiphora; chronic pharyngitis and laryngitis; epipharyngeal disease; chronic otitis media; hyposmia and anosmia; suspected or known cerebrospinal fluid (CSF) rhinorrhea; foreign-body removal; sample biopsies; and sleep disorders.

Normal mucosal findings during nasal endoscopy. The mucosa has a pale pink color and a moist consistency.

> Note: The nasal mucosa is continually responding to numerous antigens or allergens via humoral and cellular reactions. Mucosal color, swelling, and secretory status are therefore subject to continuous change.

The *diagnostic evaluation criteria in nasal endoscopy* are shown in Table 2.3.

Technique. The examination is carried out with the patient in a sitting, semireclining, or fully reclined position, without premedication. If the mucosa is severely swollen, vulnerable, or sensitive, it can be decongested and anesthetized with a 5 % lidocaine and 0.5 % phenylephrine spray, or with a single puff of tetracaine plus epinephrine. It is useful to soak a few cotton swabs in a solution of this type for 5 min before the examination. Swabs should be inserted under endoscopic guidance to prevent mucosal injury.

The endoscopic examination should follow a systematic approach in which different regions are explored (Figs. 2.19, 2.21). The 0° wide-angle scope (4 mm) is the standard instrument for all endoscopic nasal examinations. Small-diameter endoscopes should be used only if access is limited due to substantial deviation of the anterior septum, or in small children (2.7 mm, 1 mm).

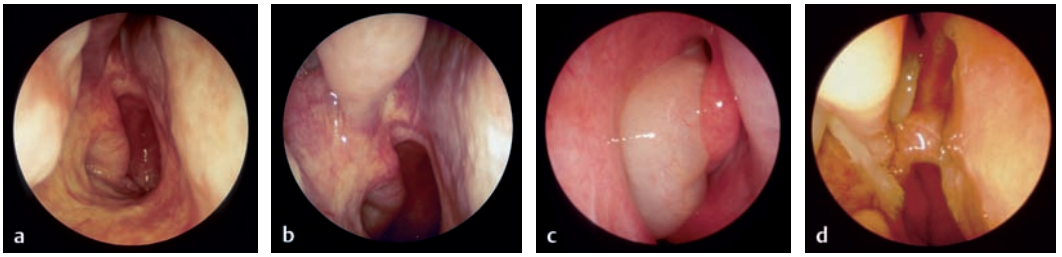


Fig. 2.20a–d Findings in nasal endoscopy.
a View of the posterior nasal cavity on the right side, 2 cm from the choana. The eustachian tube orifice is at the center of the image, and the posterior end of the lower turbinate is on the left side of the image.
b The insertion of the middle turbinate, lateral to the choana and anterior–cranial to the tubal ostium.
c A solitary polyp in the nasal meatus (right side). (a–c: 0° lens, 4 mm.)
d View of the sphenoethmoidal recess, showing a polyp in the ostium of the sphenoid cavity. A stream of mucoid pus is visible from the posterior ethmoid to the eustachian tube orifice (30° lens, 4 mm).

Table 2.3 Nasal endoscopy: diagnostic evaluation criteria

Mucosal surface	Moist, dry (e. g., rhinitis sicca), crusted (e. g., postoperative after FESS, diseases of the mucociliary apparatus), telangiectasia (e. g., Rendu–Osler–Weber disease), perforation (with multiple etiologies), petechiae (vascular disease)
Color of mucosa	Pink (normal appearance of the mucosa), livid (e. g., allergic rhinitis, nasal hypersensitivity), pale (e. g., chronic hyperplastic rhinosinusitis)
Track of pus	Present / absent (e. g., in acute inflammations, in semilunar hiatus or sphenoethmoidal recess)
Secretions (rheologic changes)	Viscous (chronic rhinosinusitis with polyps; the Sampter triad—asthma, NSAID sensitivity, and nasal polyps; fluid (anterior or posterior rhinorrhea)
Swelling	Hyperplasia of the inferior (anterior/posterior) turbinate (e. g., nasal hypersensitivity)
Anatomical variations that may predispose to recurrent sinusitis	Septal deviations, vomer crest deviation, spurs, prominent premaxilla, concha bullosa, paradoxically curved middle turbinate, medialized uncinate process
Early indications of chronic inflammation of the ethmoid infundibulum	Mucosal hyperemia of in region of infundibulum, prolapse of medial wall of infundibulum, localized edema or polypoid mucosa

FESS, functional endoscopic sinus surgery; NSAID, nonsteroidal anti-inflammatory drug.

Step 1: 0° scope: Initially, the nasal vault and nasal isthmus are inspected. Positioning the endoscope in front of the nasal cavity or the isthmus region—without deforming the nostrils by using a nasal speculum—makes it possible to evaluate the functional condition as well as low-pressure phenomena of the alar cartilages during normal and forced nasal breathing, in physiological conditions.

The endoscope is advanced into the nasal cavity to visualize the nasal floor. It is then carefully positioned between the septum and body of the lower turbinate, to advance toward the choanae. The lower turbinate and pharyngeal tubal ostium have the same orientation. The nasopharynx, soft palate motility, and function of the pharyngeal tubal ostium should be inspected. In children, the size and status of the adenoids need to be evaluated.

Step 2: 0° scope: The endoscope is now moved backward to visualize the middle nasal meatus, the "window to the ethmoid." This is particularly important, as sinusitis may originate here. The middle turbinate may be pneumatized by a system of ethmoidal cells and may be quite large (in the case of concha bullosa); it may also be the site of origin for recurrent sinusitis.

Step 3: 0° scope: Gentle medialization of the middle turbinate using a Freer elevator provides a view of the typical

relief structure of the lateral nasal wall. The uncinate process is situated ventral to the ethmoidal bulla, the size of which varies depending on its degree of pneumatization. The inferior semilunar hiatus is situated between the posterior margin of the uncinate process and the anterior surface of the ethmoidal bulla. The ethmoidal infundibulum extends in a sagittal direction and opens into the inferior semilunar hiatus. Advancing cranially from the semilunar hiatus, one approaches the frontal recess.

Step 4: 30° or 45° scope: The sphenoethmoidal recess and sphenoid sinus ostium can be inspected with a 30° or 45° scope. The endoscope is advanced along the nasal floor in a cranial direction until the choana is reached. A 45° or 70° scope can be inserted underneath the entire middle nasal meatus; it is used to inspect this area endoscopically in a cranial direction. The olfactory cleft can also be inspected—e. g., to distinguish sensorineural from respiratory hyposmia or anosmia. Angled endoscopes are suitable for inspection of the nasal ostium, which contains the nasolacrimal duct. The ostium is oval in shape and is located just a few millimeters dorsal to the anterior insertion of the lower turbinate.

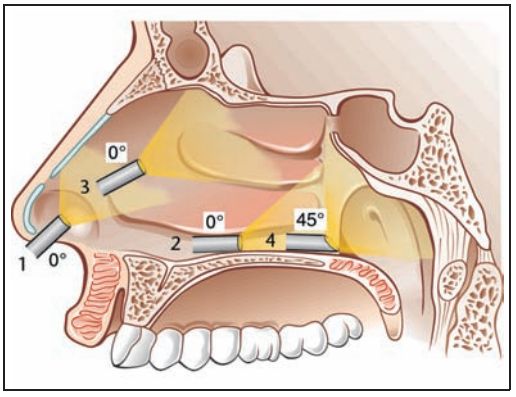
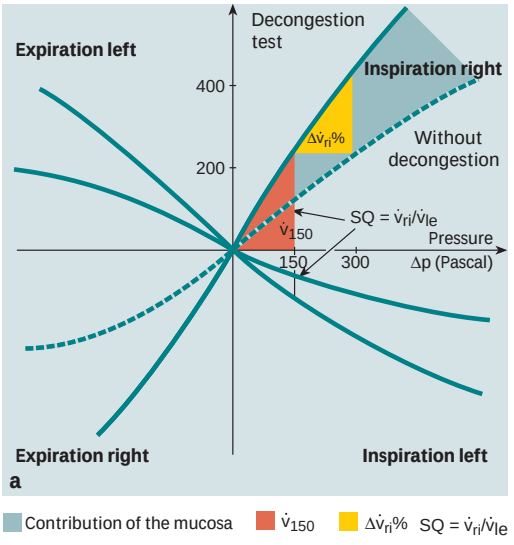


Fig. 2.21 Systematic nasal endoscopy using scopes with different angles of view. 1, Nasal vestibule; 2, nasal floor and inferior meatus; 3, middle meatus; 4, sphenoethmoidal recess.

Assessment of Nasal Patency

Rhinomanometry. During spontaneous respiration, the nasal airflow ($\dot{V}$, cm^3/s) is measured using a flowmeter (pneumotachograph, mass flowmeter) attached to a mask. Simultaneously, the differential pressure between choana and the mask (ΔP, Pa) is determined. Various resistance values can be calculated from the directly measured results for pressure and flow. In addition, comparison of the ascending and descending inspiratory and expiratory breathing phases in standard graphs makes it possible to assess the influence of the elasticity of the nasal structures. Further information can be obtained by performing rhinomanometry before and after decongestion with nose drops (temporary or permanent nasal resistance), or by provocation of the nasal mucosa with allergens (nasal provocation test) (Fig. 2.22a, b).

Measurements are usually performed during spontaneous respiration. The volume of air passing through the nose during active nasal respiration is recorded at the same time as the pressure differential across the nose. The results can be recorded either using a pair of curves or as an *xy* function (Fig. 2.22a), and they provide information about unilateral or bilateral nasal patency (Fig. 2.22b) (see p. 126).



Obstruction	Partial nose	Entire nose
No	> 500	> 800
Light	300–500	500–800
Medium	180–300	300–500
Severe	60–180	100–300
Near-complete	< 60	< 100

b

Fig. 2.22a, b a Principle of computer-supported rhinomanometry.
b Reference values in adult patients (flow at 150 Pa, in cm^3).

Acoustic rhinometry. This method uses acoustic reflection for separate measurement on each side of the cross-sectional area of the nose, from the vestibule to the nasopharynx. In contrast to rhinomanometry, it assesses the cross-sectional area of various parts of the nose, rather than flow parameters in the nose. Another difference is that it also serves to determine static parameters without the need for patient cooperation.

A rough *qualitative* test of nasal patency can be achieved by holding a polished metal plate in front of the nose during inspiration and expiration. The surface area of the resulting fogging gives an approximation of the patency of the two sides of the nose.

In *infants,* nasal patency is tested by holding a wisp of cotton wool or a feather in front of the nose.

Olfactometry

First, a precise case history is taken that includes triggering events, associated symptoms, relevant illnesses, operations, medications, and noxious influences. This is followed by nasal endoscopy, with exploration of the nasopharynx and the olfactory cleft.

Assessment of olfactory function is based on a standardized, validated test. The following olfaction tests are available and widely used:

- *Sniffin' Sticks:* Sixteen scents on felt-tip pens are presented to the patient, who is asked to identify them. This test is widely used in Europe. It combines threshold, identification, and discrimination of scents; the identification test is suitable for screening.
- *University of Pennsylvania Smell Identification Test (UPSIT):* Forty scents in microcapsules are deposited on paper, where they can be released mechanically by rubbing. The individual scents have to be identified from a list, with four alternatives each. The cross-cultural smell identification test (CCSIT) is a simplified version of this.
- *Connecticut Chemosensory Clinical Research Center Test (CCCRC):* This is a combination of a threshold test for butanol and an identification test for ten scents. The scents are contained in polypropylene flasks that can be pressed open. A disadvantage with this test is low validation.
- *Evoked response olfactometry (ERO):* Objective assessment of olfactory disorders is possible by analyzing the potentials evoked by olfactory stimuli. Derivation of olfactory evoked potentials is the only validated method of confirming a loss of the sense of smell objectively. ERO involves application of chemosensory stimuli via a tube inserted into the middle nasal meatus, at intervals averaging 20–40 s. Each stimulation lasts 250 ms. Phenylethyl alcohol or hydrogen sulfide can be used as scents.

The ability to identify different substances may be of importance in neurologic differential diagnosis (Table 2.4).

Two different parameters can be tested:

1. The *threshold* at which the substance is *perceived.*
2. The *threshold* at which it is *recognized.*

The former threshold is lower than the latter. Apart from ERO, all of the methods mentioned above depend on cooperation from the patient, and the results are therefore largely subjective. Objective results can only be obtained with ERO.

Tests for simulation include ERO and the cinnamon test. The taste of cinnamon is mediated by the olfactory nerve. Cinnamon cannot be recognized in the absence of the ability to smell.

Anosmia is complete loss of the ability to smell; *hyposmia* is a reduced ability to smell; and *parosmia* refers to a state in which the subjective impression does not correspond to the substance offered. In *cacosmia,* all smells appear to be offensive. Cacosmia is often an indication of a central nervous system lesion (see also Table 2.1).

> **!** Note: Ideally, the sense of smell should always be tested thoroughly before any surgery in the nose or sinuses, for medicolegal reasons.

Diagnostic Imaging of the Nose and Sinuses

Diagnosis of diseases of the nasal sinuses is hardly possible without the use of radiology.

Conventional radiology. *Radiographs of the nose* in the lateral projection are necessary to demonstrate a fracture of the nasal bone. They can also be used if an intranasal foreign body is suspected.

Plain sinus radiography (Fig. 2.23a, b)*:* The diagnostic value of plain sinus radiographs is controversial today. Proponents of plain sinus radiography say the films provide rapid, noninvasive evaluation of the lower third of the nasal cavity and the maxillary, frontal, sphenoid, and posterior ethmoid sinuses. Plain films are neither useful nor cost-efficient for evaluating the anterior ethmoid air cells, the upper two-thirds of the nasal cavity, or the infundibular, middle meatal, and frontal recess air passages. The clinical role of plain films is thus at best limited to documenting acute maxillary or frontal sinusitis and, if baseline films are available, to following the course of infection and the response to treatment. Detection of air–fluid levels warrants evaluation by CT or nasal endoscopy.

Computed tomography. A CT scan in the coronal plane is the imaging modality of choice for confirming the anatomy and extent of pathology. It helps in understanding the functional sinus anatomy of the individual patient and supports the identification and evaluation of the frontal sinus, frontal recess, uncinate process, ethmoidal infundibulum, maxillary sinus, maxillary sinus ostia, ethmoidal bulla, lateral sinus, middle meatus, posterior ethmoidal sinus cell, sphenoid sinus, and sphenoid recess.

Indications: Patients with a history of chronic, recurrent sinusitis despite repeated medical therapy, independent of normal or pathologic findings at the lateral nasal wall (Fig. 2.24), and patients with suspicious space-occupying lesions.

Note: Nasal endoscopy should always be done before CT scanning.

Magnetic resonance imaging. MRI is not the primary imaging modality in chronic rhinosinusitis. It is usually reserved (in combination with CT) for investigating more serious conditions, such as neoplasia (Fig. 2.25a, b). It is indicated in the diagnosis of intracranial complications (e.g., epidural or subdural abscess, brain abscess) and orbital complications (e.g., orbital infection).

Magnetic resonance angiography. This helps clarify the hemodynamic activity of a tumor.

Table 2.4 Some known olfactory substances

	Stimulation of olfactory nerve	Stimulation of sensory part of trigeminal nerve	Stimulation of chorda tympani (cranial nerve VII) and glossopharyngeal nerve (cranial nerve IX)
Coffee	+		
Wax	+		
Vanilla	+		
Lavender oil	+		
Turpentine oil	+		
Birch tar	+		
Cinnamon	+		
Benzaldehyde	+	+	
Menthol	+	+	
Turpentine	+	+	
Petroleum	+	+	
Peppermint	+	+	
Camphor	+	+	
Alcohol	+	+	
Formaldehyde	+	+	
Acetic acid	+	+	
Ammonia		+	
Chloroform	+		+
Pyridine	+		+

Angiography/digital subtraction angiography. In addition to visualization of the blood vessels, this allows preoperative embolization of the arteries that supply a tumor. This is a prerequisite for endonasal endoscopic surgical resection of angiofibromas.

Ultrasound in A or B mode. This can also be used to investigate the nasal sinuses. The advantage of this method is that the patient is spared exposure to radiation. Its disadvantage is that the findings are less detailed and technical errors more likely.

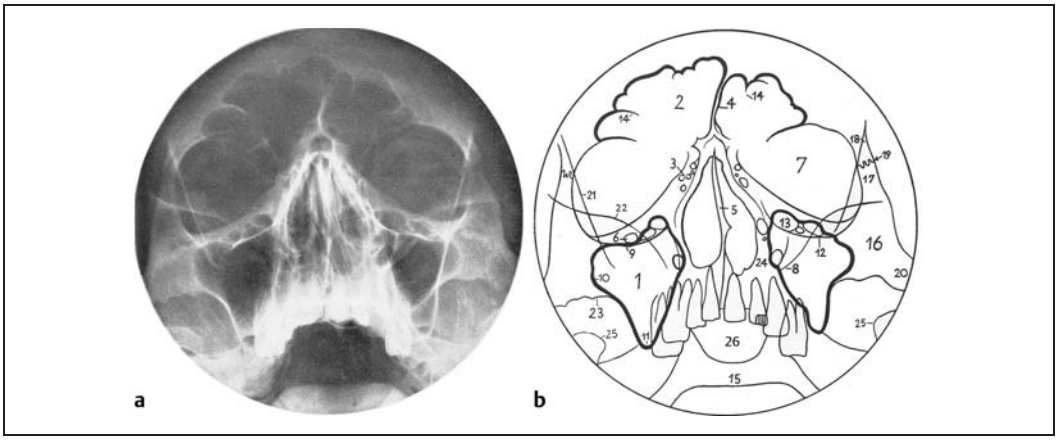


Fig. 2.23a, b Radiographic anatomy of a plain film of the paranasal sinuses. 1, Maxillary sinus; 2, frontal sinus; 3, ethmoidal air cells; 4, interfrontal septum; 5, nasal septum; 6, infraorbital foramen; 7, floor of the orbit; 8–11, medial, cranial, lateral, and caudal walls of the maxillary sinus; 12, orbital floor; 13, cranial part of maxillary sinus; 14, interfrontal septa; 15, tongue; 16, zygomatic bone; 17–19, frontozygomatic suture; 20, zygomatic arch; 21, innominate line (orthograde projection of part of the greater wing of sphenoid); 22, small sphenoid wing; 23, lower border of zygomatic bone; 24, round foramen; 25, head of the mandible; 26, sphenoid sinus.

Fig. 2.24 Coronal CT of the paranasal sinuses. On the left side of the image, there is good visualization of the bony structures of the lateral nasal wall. On the right side, there is pansinusitis, with hyperplastic mucosa in the cavities and ethmoidal air cells and destruction of septa between cells.

Lavage of the Sinuses

The maxillary sinus, frontal sinus, and sphenoid sinus can be irrigated, but not the ethmoid sinuses. There are two reasons for irrigating the sinuses:

1. For *diagnostic purposes,* to allow aspiration and lavage of abnormal secretions, bacteriologic and cytologic examination of the secretions, and possibly the introduction of a contrast medium for radiography.
2. The sinuses can be irrigated for *therapeutic purposes,* to allow drainage of abnormal secretions and introduction of locally active substances into the sinuses.

Although lavage of the sinuses was commonly used to treat maxillary empyema up until just a few decades ago, it is rarely done today. Drainage of the frontal sinus using the Kümmel and Beck method is now practically obsolete, having been replaced by endoscopic endonasal microsurgery. However, the otolaryngologist should be familiar with these techniques, to avoid inflammatory complications in case the modern approaches fail: *ubi pus, ibi evacua.*

Lavage of the maxillary antrum. Two methods are routinely used:

1. Access via the *inferior meatus* (sharp puncture)
2. Access via the *middle meatus* (blunt puncture) after fenestration during functional endoscopic surgery.

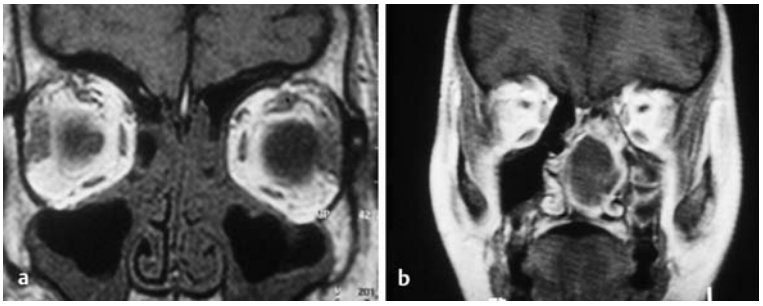


Fig. 2.25a, b Coronal MRI of the paranasal sinuses.
a Good visualization of brain, soft tissues, and mucosal pathology, poor visualization of bony structures.
b Positive enhancement on the surface of a vascularized tumor (T1-weighted with contrast medium).

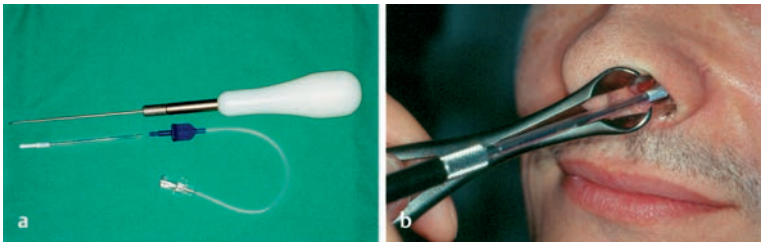


Fig. 2.26a, b A modern lavage system (Atos Medical, Hörby, Sweden; Bess, Berlin, Germany) for the paranasal sinuses. Puncture is performed in the inferior nasal meatus.

Principle of lavage via the inferior meatus: Local anesthesia of the inferior meatus is induced. A Lichtwitz trocar is placed against the lateral nasal wall beneath the origin of the inferior turbinate. After the cannula has been pushed through this part of the lateral nasal wall, which is usually thin, it can be used for aspiration, lavage, or the introduction of drugs.

A long-term drain can be introduced for conservative treatment, especially in children (see p. 180).

The SinuTroc lavage system for the maxillary sinus (Fig. 2.26a, b) is an optimized drainage system. During puncture of the maxillary sinus with a trocar in the middle nasal meatus, a 3-mm plastic tube equipped with a plastic hull (1 cm long, 1.5 mm thick) is inserted into the maxillary sinus. The trocar is then removed; two small barbs keep the self-retaining catheter from slipping away from the wall of the maxillary sinus. For repeated lavage, the self-retaining catheter is attached to an adapter tube (19 cm long, Luer-Lok or cone connection).

Complications during blunt puncture are very unlikely if it is performed expertly. During sharp puncture, the point of the cannula may be pushed inadvertently into the soft tissues of the cheek, pterygopalatine fossa, or orbit if the technique is incorrect.

Note: Air should never be used to clear the irrigated sinus, as it can lead to an air embolism. The symptoms are collapse, loss of consciousness, cyanosis, possibly hemiplegia, amaurosis, and sudden death.

Lavage of the frontal sinus. *Puncture* (as described by Kümmel and Beck) (see also p. 140) can be performed using a fine drill under local anesthesia. This allows irrigation for both diagnostic and therapeutic purposes, and also prolonged drainage of the frontal sinus for 1–2 weeks, allowing daily administration of drugs. The principle of puncture is shown in Fig. 2.27a–d.

Technique: Anteroposterior and lateral radiographs of the sinuses have to be obtained first. If the frontal sinus has not developed or is very shallow, there is a risk that the frontal lobe may be punctured.

Irrigation of the sphenoid sinus can be performed by a specialist, using special cannulas.

The ethmoidal labyrinth does not have a defined ostium and therefore cannot be irrigated. However, pathologic secretions can be evacuated using suction.

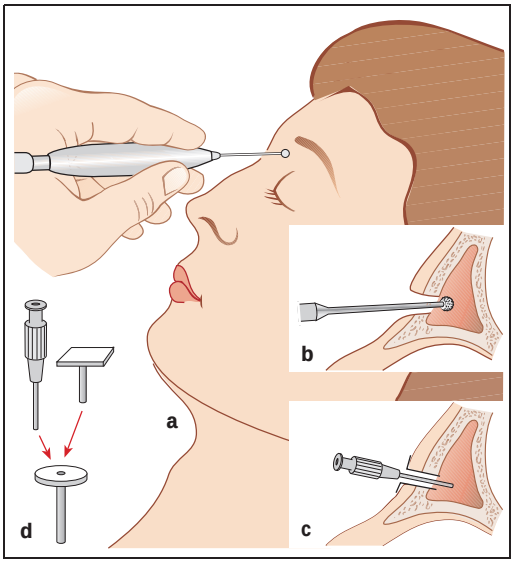


Fig. 2.27a–d The Kümmel–Beck frontal sinus trephine.
a Access and positioning of the drill.
b The bore hole in the anterior wall of the frontal sinus.
c The guide tube and lavage cannula have been placed.
d The guide tube with the lavage cannula and occlusion plate.

Specific Diagnostic Methods

Cytology. Smears of secretions or swabs taken from the mucosa can be assessed cytologically. This is useful for differentiating between catarrh, bacterial infection, allergic rhinitis, and mucosal mycoses.

Allergy studies. Investigations performed on patients include skin tests (scratch, prick, intracutaneous, or friction) and provocation tests (conjunctival and intranasal). Laboratory studies include total immunoglobulin E (IgE) in serum (paper radioimmunosorbent test, PRIST) and specific IgE in serum (radioallergosorbent test, RAST). Eosinophil counts in the blood and nasal secretions are less informative.

Biopsy. This is the method of choice whenever it is unclear whether a disease is benign or malignant.

β_2 Transferrin test. Evidence of CSF rhinorrhea is provided by a positive β_2 transferrin test. In this laboratory test, the ratio of β_1 to β_2 transferrin in the nasal secretions and blood serum is determined. As high β_2 transferrin levels occur only in CSF, they are proof of a CSF leak.

Note: Whenever the diagnosis is not clear, and particularly if a tumor is suspected, a biopsy provides the diagnosis. If the results of the biopsy are negative but a clinical suspicion of malignancy persists, the biopsy needs to be repeated several times, with specimens taken from different sites.

Dermatologic Principles for the Otolaryngologist

Otolaryngologists see dermatologic disorders in the head and neck region every day. They therefore need to understand the basics of dermatologic diagnosis and topical therapy. In some areas, such as the external ear canal, the otorhinolaryngology specialist serves as a “head and neck dermatologist.”

Before the conclusion is drawn that a skin finding is a localized change limited to the head and neck region, it needs to be considered whether it might be a regional manifestation of a systemic disease such as lupus erythematosus, systemic sclerosis, or pemphigus vulgaris, or whether it is part of a cutaneous–mucosal disease such as lichen planus or erythema multiforme, to cite but a few dramatic examples.

Skin Type

Certain disorders may be more or less common, depending on the skin type. For example, acne vulgaris with follicular comedos, papules, and pustules is more common in those with oily skin (the *seborrheic* skin type), as is rosacea, which features facial erythema, papules, pustules, and sebaceous gland hyperplasia. Rosacea appears in adult life and is occasionally called “adult acne”—an incorrect oversimplification. Atopic dermatitis is more common in those with dry skin (the *sebostatic* skin type). In our experience, healing in oily skin is poorer than in normal or dry skin, an important consideration when planning surgery. The skin type can help one decide to perform a flap rather than primary closure in an oily area.

In describing the skin, a distinction is made between the condition of the skin and its vascular supply. The *skin status* describes turgor, seborrhea, xerosis, atrophy, actinic damage, hyperhidrosis, and hypohidrosis; while the *vascular status* includes factors related to blood supply, such as cyanosis, paleness, cold, warmth, edema, and necrosis.

Types of Lesion

Just as the multiplication tables are one of the cornerstones of arithmetic, the terminology and precise description of lesions is a central part of dermatologic knowledge that is necessary for achieving a logical diagnostic approach. Even if the working diagnosis proves to be incorrect, it is easier to advance to the correct diagnosis if the lesions have been precisely and correctly described. If the initial lesions have been poorly described, then it is very difficult to reconstruct a case. The way to describe the different lesions simply has to be learned, therefore—much like learning the words of a foreign language. Describing lesions correctly is extremely important.

A distinction is made between *primary* and *secondary* lesions (Fig. 2.28a–k).

Primary lesions

- Macule (spot), urtica (hive), papule (small raised lesion), nodule (larger raised lesion), vesicle (small blister), bulla (large blister), pustule (primary pus-filled blister)

Secondary lesions

- Pustule (vesicle with secondary pus), crust (mixture of pus and scales), scales, erosion (superficial defect of epidermis), excoriation (self-induced defect in the epidermis and upper dermis), rhagades/fissure (linear tear), ulcer (deeper defect), scar, atrophy, cyst (fluid-filled lesion), and necrosis

Primary lesions are fresh lesions that appear as the first sign of a skin disease. Secondary lesions are less sharply defined. They may result from secondary changes in primary lesions (a vesicle becomes a pustule), from exogenous damage (erosion or excoriation), or over time (atrophy, scales). It is more important to describe the lesion correctly than to decide whether it is primary or secondary. It should always be possible to identify a pustule; whether it is primary or secondary can remain an open question.

Along with the type of lesion, the pattern of distribution is also of diagnostic significance. The distribution may be strikingly asymmetrical, segmental, symmetrical, or generalized, as well as fitting light-exposed, hypostatic, or other known patterns. The pattern often facilitates the diagnosis and should therefore always be documented.

Dermatologicdifferentialdiagnosisrequiresalogical approach, as one develops a sense or feeling for the process. At the start, one should consider all the possible diagnoses that fit the morphologic pattern, but always remembering that the most common disorder is the most likely diagnosis. A second important principle is always to consider the possibility of an unusual variation of a common condition; for example, many disorders are more severe or clinically atypical in patients with human immunodeficiency virus/acquired immune deficiency syndrome (HIV/AIDS).

Basics of Topical Dermatologic Therapy

Topical therapy has a special role in treatment. The medication is directly applied to the site of disease, and its effect is easily observed without complex instrumentation. A distinction is made between *vehicle, active ingredient*, and *additives*. Most topical agents contain all three components. The active ingredient is the pharmaceutical agent that is to be delivered to the skin in a vehicle. Additives can play many roles, making the vehicles more attractive (with fragrance) or stable (through preservatives). While in the past many formulations contained multiple active ingredients, today most have one or at most two such agents, reducing the likelihood of drug interactions.

Three different types or phases of vehicle are available: solid, liquid, and semisolid. They can all be combined in various ways. A favorite way of visualizing this is using a phase triangle, in which the corners represent the basic phases, while the sides of the triangle signify mixtures of two classes and the center represents a mixture of all three (Fig. 2.29).

The vehicles available for topical therapy can also be divided into two groups:

Primary vehicles (one phase): These are the corners of the phase triangle. They include:

- Solids (powders).
- Liquids (wet soaks, solutions).
- Semisolids (ointments, petrolatum, oils).

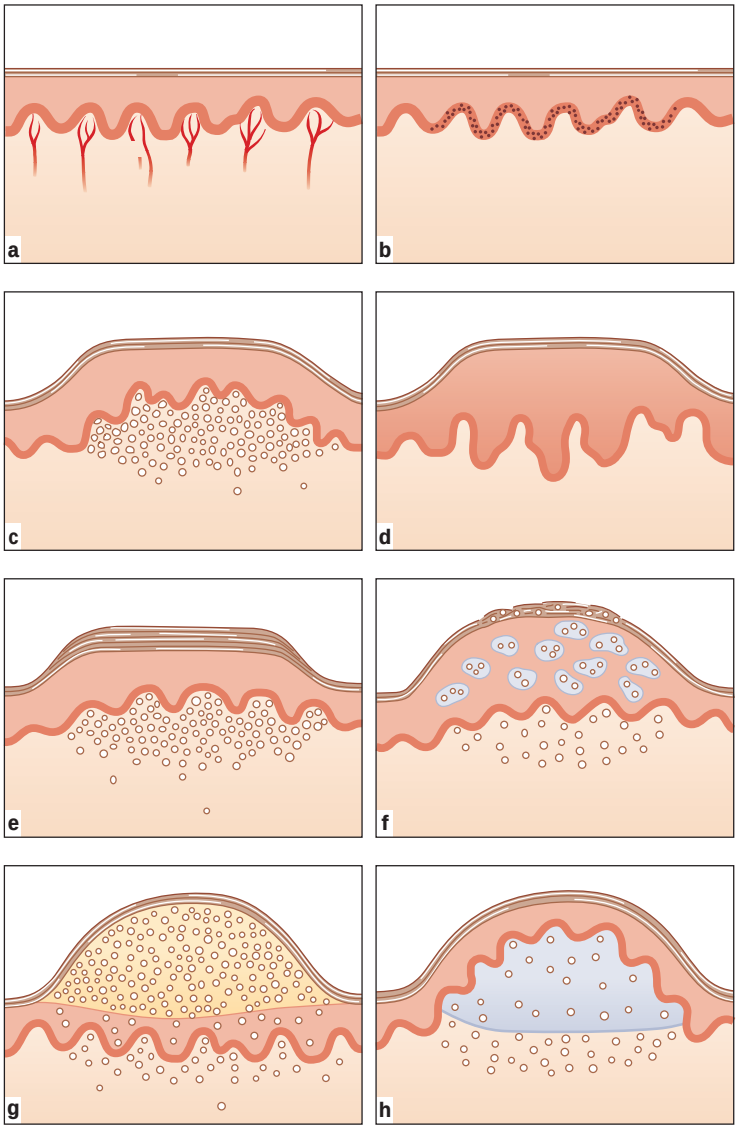


Fig. 2.28a–k The most important types of skin lesion.
a Erythema of a macule, due to circumscribed vasodilation.
b Brown discoloration of a macule, due to a circumscribed increase in pigmentation in the basal layer of the epidermis.
c Dermal papule, due to accumulation of cellular elements in the dermis.
d Epidermal papule, due to thickening of the epidermis.
e Mixed papule, due to thickening of the epidermis, including the stratum corneum, and dermal inflammatory infiltrate.
f Eczema nodules, due to thickening of the epidermis and spongiform edema, with round-cell infiltrates in both the parakeratotic stratum corneum and dermis.
g Subcorneal pustule, in which the stratum corneum is lifted from the rest of the epidermis by edema fluid and leukocytes; there are leukocytic infiltrates in both the epidermis and dermis.
h Subepithelial blister, in which the entire epidermis is lifted by an accumulation of fluid; the dermis contains an inflammatory infiltrate.

Fig. 2.28 i—k □

Combined vehicles (two or more phases) fall between the corners, depending on their composition:

- Emulsions (ointments (oil/water, water/oil), creams).
- Shake lotions.
- Pastes.

Principles for Choosing a Vehicle

The correct choice of a vehicle can greatly enhance healing, while an inappropriate vehicle can have adverse effects, even if exactly the correct active ingredient has been chosen. One should try to work with as few vehicles as possible, in order to get to know them well and acquire experience in administering them. The choice of vehicle is based on the skin type, involved area, acuteness of inflammation, and type of lesion.

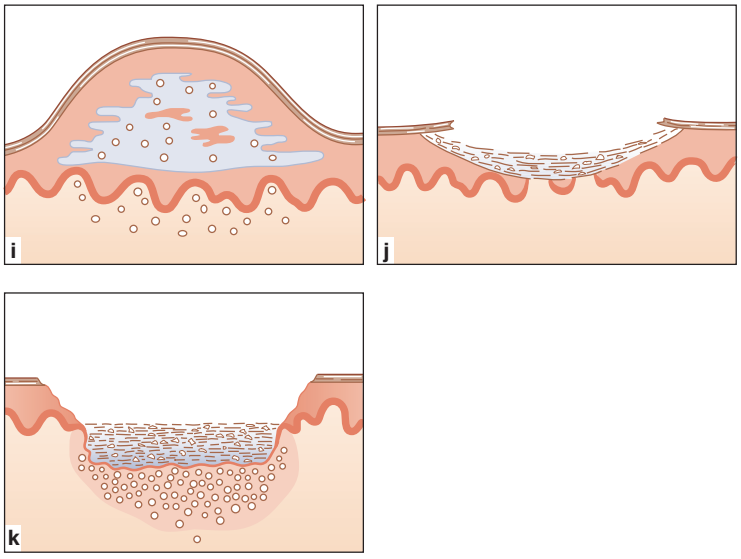


Fig. 2.28 i Intraepithelial blister; acantholysis within the epidermis has led to the formation of an irregular space containing leukocytes and desquamated scales.
j Erosion: a shallow defect in the epidermis, partially filled with serum and leukocyte detritus.
k Ulcer: a defect extending into the reticular dermis and involving destruction of the epidermis and adnexal structures (sweat glands, sebaceous glands and hair follicles), as well as papillary dermis. The defect is partially filled with serum and leukocytic detritus, and the adjacent dermis shows inflammatory infiltrates.

Skin type. *Seborrheic skin:* Powders, alcoholic solutions, shake lotions, creams; no greasy vehicles.

Sebostatic skin: Ointments, ointment emulsions, soft pastes; no drying or only minimally greasy vehicles.

Special body regions. *Scalp:* Aqueous or alcoholic solutions, easily washed emulsion creams; no pure ointments (petrolatum), pastes or shake lotions.

Intertriginous areas: Soft zinc pastes, creams, zinc oils; no shake lotions or thick ointments.

Degree of inflammation. *Acute superficial inflammation:* Dye solutions, moist dressings, powders, shake lotions, creams. Clearly weeping dermatoses should be treated with wet dressings or greasy–wet dressings (slightly greasy dressing covered with wet soaks). Occlusive dressings should not be used for weeping dermatoses, pyodermas, or fungal infections.

Chronic deeper inflammation: Vehicles are required that can ensure penetration of the active ingredients such as ointments, ointment emulsions, soft pastes, and emollient creams, but no powders, shake lotions, or wet dressings.

Morphology of skin changes. *Acute erythema:* Powders, shake lotions, creams, wet dressings.

Vesicles and bullae: Shake lotions, creams, moist dressings, greasy–wet system (greasy vehicle covered by wet dressing).

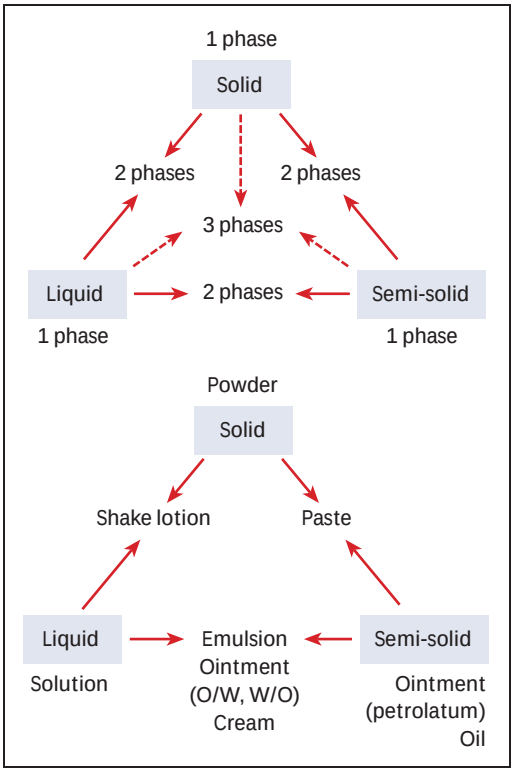


Fig. 2.29 The phase triangle for topical therapy.

Erosions: Wet dressings (wet or greasy–wet), ointments.

Crusts: Wet dressings (wet or greasy–wet), ointments, emollient ointments, soft pastes.

Scales: Ointments, emollient ointments, soft pastes, greasy–wet dressings.

Chronic infiltrates or lichenification: Ointments, emollient ointment, soft pastes.

Atrophy, scars: Ointments, emollient ointments, soft pastes.

Examples of Topical Active Ingredients

Urea: Hygroscopic (binds water), keratolytic, proteolytic, increases penetration, anti-pruritic and weakly anti-proliferative. Non-sensitizing, but irritating, depending on concentration. Indication: dry, pruritic skin.

Salicylic acid: Very keratolytic, weakly anti-inflammatory and antimicrobial, irritating in active inflammation. Usually combined with other active ingredients, such as a topical corticosteroid for pruritic otitis externa. Formulation: acid. salicyl. 3.0, hydroxyquinoline sulf. 0.25, dexamethasone 0.03, ung. alcohol. lanae aquosum ad 100.0.

Corticosteroids: Depending on choice of agent and concentration, varying degrees of effectiveness. Care is also needed on the face, with a preference for lower concentrations and less active agents; in case of infection, corticosteroids should not be used as a single agent, but only as a supplementary anti-infectious therapy. Because they are simply the best topical anti-inflammatory agents, corticosteroids are often used for otorhinolaryngology indications. Formulation (for weeping otitis externa): prednisolone 0.02, ol. zinci oxydat. ad 50.0.

Dyes and antiseptics: Dermatologists nowadays rarely use gentian violet, brilliant green, or Castellani paint (carbolfuchsin topical solution). These products are highly effective, but compliance tends to be poor. From the otorhinolaryngology point of view, these solutions are still valuable because of their good antiseptic, antimycotic, and drying effects. The main difference from dermatologic use is the employment of much smaller amounts at lower concentrations to treat limited areas such as the external ear canal. Formulation of Castellani solution (without phenol): ethanol fuchsin solution 4 % 2.0, chlorocresol 0.02, resorcin 2.0, sodium edetate 0.004, acetone 1.0, aqua purificata ad 20.0.

Our approach to acute otitis externa is to start with a nonspecific dye solution and only to use antibiotics in a targeted or at least calculated way, based on culture results or knowledge of the likely causative agents. One should always avoid indiscriminate use of multiple agents.

Antibiotics: The use of topical antibiotics is somewhat controversial in dermatology. The main reasons for this include the development of resistance and allergic sensitization, which can then leave the individual intolerant to the same or related agents administered systemically. There are many alternatives for most forms of topical antibiotic therapy. The active agents that are still in use after a long period of time and have proved their effectiveness include the following (with examples of representative products):

- *Gentamicin sulfate:* Gentamicin cream 0.1 %, gentamicin ointment 0.1 %.
- *Erythromycin:* Akne-Mycin ointment or solution, Zineryt (solution with zinc acetate), Clinofug gel 2 %/4 %.
- *Oxytetracycline HCl:* Terramycin ointment (with polymyxin B), oxytetracycline ointment 1 %.

Clinical Aspects of Diseases of the Nose, Sinuses, and Face

The main symptoms of diseases of the nose and sinuses are:

- Increased nasal secretion.
- Nasal obstruction.
- Bleeding or hemorrhagic secretion (see Tables 2.11, 2.12).
- Fetor.
- Altered or absent sense of smell (see Tables 2.1, 2.2).
- Pain in the head or face.
- Disease of neighboring organs—e. g., teeth, the lacrimal apparatus, eyes, mouth, and throat. Important symptoms of eye disease include abnormalities of refraction, limitation of the visual field, acute loss of vision, and displacement of the orbit. Diseases of the mouth and throat may be symptomatic, or a change in the quality of the voice and speech may be noted.

Inflammatory Diseases of the Nose and Paranasal Sinuses

Inflammations Confined Mainly to the External Nose

The skin of the nose and the face may be affected by the same common typical skin diseases that affect the rest of the skin, such as impetigo, acne, trichophyton, rosacea, and lupus erythematosus. These diseases are treated using the appropriate dermatologic methods.

Other skin diseases that are particularly important in the nasal area are described below.

Fig. 2.30a, b Nasal furuncle.

Nasal Eczema

Clinical Features. The disease is moist in the early phases, with vesicles and pustules. Later, crusts form, followed by painful cracks. In the chronic stage, there is itching, burning, and desquamation. The disease is localized to the external nose and the skin of the nasal vestibule and never affects the nasal mucosa.

Pathogenesis. The disease is often caused by abnormal nasal secretions, but may also be due to a contact allergy (sensitivity testing should be carried out). Promoting factors include diabetes mellitus, generalized eczema, and dietary sensitivity in children.

Treatment. The crusts should be softened with a mild greasy ointment followed by a corticosteroid (hydrocortisone). The use of stronger steroids should be avoided on facial skin. Cracks are treated with 5–10 % silver nitrate solution. The cause should be looked for and treated or eliminated.

Folliculitis of the Nasal Vestibule (Sycosis) and Nasal Furuncle

Clinical Features. Increasing pain, marked sensitivity to pressure, and a feeling of tension in the tip of the nose are followed by reddening and swelling of the tip of the nose, of the nasal ala, and of the upper lip (Fig. 2.30a, b). The area becomes edematous, and the patient may have fever. The swelling may begin to resolve before suppuration occurs. Otherwise, a typical furuncle forms, containing pus and a central necrotic core.

Diagnosis. An ascending infection via the facial vein has to be ruled out by testing the medial angle of the eye for tenderness.

Pathogenesis. A pyoderma, usually due to staphylococcal infection, arises from the hair follicles of the nasal vestibule or the upper lip, often close to the nasal tip. The disease is always limited to the skin and never affects the mucosa.

Treatment. Antibiotic creams, such as 2 % mupirocin (Bactroban) or 0.1 % chlorhexidine/0.5 % neomycin (Naseptin) cream or ethacridine lactate solution, are applied to the nasal vestibule as long as the disease remains a circumscribed folliculitis. Manipulation in the nose is contraindicated. If it is suspected that a furuncle is forming, high-dose oral or parenteral antibiotics are given, if necessary in combination with topical antibiotics. The antibiotics of choice for systemic bactericidal therapy are cefadroxil, flucloxacillin (floxacillin), or doxycycline; in severe cases, cefazolin or an aminopenicillin with a β-lactamase inhibitor. Antibiotics must be continued for several days after the symptoms have subsided; they should not be discontinued too early or used at too low a dosage. If an infection is severe, the patient will need hospital admission and intravenous antibiotic and fluid administration.

If there is tenderness in the medial angle of the eye, the facial artery and vein must be ligated.

Note: A furuncle on the nose or upper lip must never be squeezed, due to the risk of spreading the infection and causing complications such as thrombophlebitis and cavernous sinus thrombosis (see p. 117). The veins of the nose and upper lip drain into the venous system of the neck via the facial vein, but also via the angular and ophthalmic veins through the orbit to the cavernous sinus. Figure 2.2 shows the relevant anatomy, and the point at which the angular vein may be ligated if incipient thrombophlebitis is suspected.

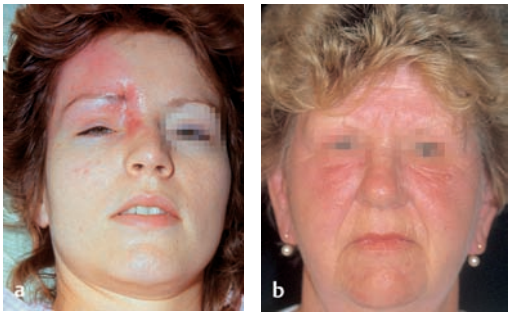


Fig. 2.31a, b a Acute erysipelas: sharply demarcated erubescence in the vicinity of a skin injury.
b Chronic erysipelas: persistent, edematous, rubescent skin with swelling.

Fig. 2.32 Rhinophyma.

Frostbite and Burns

The nose is particularly at risk for frostbite and sunburn in extremes of weather and temperature. The injury is divided into three degrees of severity, with the most severe being dry or moist tissue necrosis.

Treatment. As for thermal damage to other parts of the body.

Erysipelas

Clinical Features. The incubation period lasts several hours to 2 days. The disease starts with high fever and possibly chills. There is marked pain and sharply demarcated reddening of the edematous skin. Often there is extension on both sides of the nasal pyramid in a butterfly configuration. The disease usually resolves within 1 week with proper treatment. Treatment must be energetic and high-dose, as the disease tends to recur (Fig. 2.31a, b).

Pathogenesis. The causative organism is usually a streptococcus. The portal of entry is often a small abrasion on the skin.

Differential diagnosis. Angioneurotic edema, acute dermatitis, and herpes zoster must be considered.

Treatment. Penicillin is the first-line drug. It should be given intravenously to begin with, and continued for 10 days. In severe cases, 10 million IU per day may be required. Alternatively, erythromycin, cephalosporins, or macrolides may be given. Antibiotics are continued for 8 days after the eruptions subside, and local moist dressings are applied.

Prognosis. Good.

Rhinophyma

Clinical Features. The condition starts with coarsening of the skin over the cartilaginous part of the nose. A lumpy, bluish-red pseudotumor develops and slowly progresses to become a markedly protuberant lobular swelling of the anterior part of the nose, which may even obstruct breathing and eating. This disease usually occurs in older men (Fig. 2.32).

Pathogenesis. Rhinophyma is due to hypertrophy of the sebaceous glands. It is associated with acne rosacea.

Treatment. The disease is treated surgically. The tissue is removed in layers with a scalpel, CO_2 laser, or by dermabrasion, and is shaved down to the level of the normal nasal contour. The area is then allowed to heal spontaneously, or is covered with a split-skin graft.

Lupus Vulgaris of the Nose

Clinical Features. Fine red nodules are found on the nasal vestibule, the head of the inferior turbinate, and the septum lying opposite to it. The nodular stage is followed by central necrosis of the nodules, with ulceration and formation of granulomas, and finally scarring with deformity of the underlying cartilage. The nasal introitus becomes stenosed, and the cartilaginous framework of the nose collapses (Fig. 2.33).

Pathogenesis. The disease is due to tuberculous infection in the presence of relatively robust immunologic resistance (the proliferative form). The organism responsible is *Mycobacterium tuberculosis*, either the human or bovine type.

Diagnosis. The diagnosis is reached by demonstrating the infectious agent and by biopsy. Diagnosis of the disease is notifiable. Characteristic findings are intradermal papules that ulcerate, leaving scars.

Differential diagnosis. Eczema of the nasal introitus, anterior rhinitis sicca, perforating ulcer of the septum, syphilis, mycosis, lupus erythematosus, sarcoid, and malignancy. Tuberculosis usually affects the cartilaginous parts of the nose, whereas syphilis attacks the bony part.

Treatment. Long-term triple combinations of tuberculostatic drugs (see p. 156) and vitamin D_2 are given.

Sarcoidosis (Besnier–Boeck–Schaumann Disease)

Clinical Features. Sarcoidosis is a multisystem disorder of unknown etiology that may occur as an isolated lesion in the nose. Nasal sarcoid presents with nasal obstruction, recurrent epistaxis, crusting, and/or anosmia. Examination may show red nodules within the nasal mucosa giving the appearance of strawberry skin, nasal tenderness, and occasionally infiltration of the facial skin. There may also be a saddle deformity and septal perforation and facial swelling. There may also be associated sinusitis. Regional lymph nodes may be firm and enlarged.

Diagnosis. If nasal sarcoidosis is suspected, a vasculitic screen should be instigated. This should include a full blood count; ESR & CRP; serum biochemistry and calcium; angiotensin converting enzyme (ACE); ANCA and syphilis serology. Urine should be checked for blood and protein and a 24-hour urinary calcium assessment.

The serum ACE diagnosis is usually elevated in active sarcoidosis but is a non-specific non-diagnostic test. The diagnosis relies on obtaining a biopsy that shows non-caseating granulomata. A chest radiograph and/or chest CT should also be requested.

Differential diagnosis. This includes rhinophyma, lupus, gumma, and malignancy.

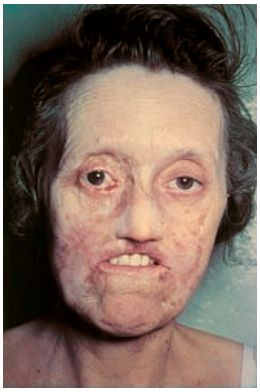


Fig. 2.33 Lupus vulgaris.

Treatment. Systemic steroids are the mainstay of treatment for active disease and should be administered under supervision of a rheumatologist. Immunosuppressive or immunomodulation medication such as methotrexate or an anti-TNF alpha blocker (infliximab) can be helpful in limiting systemic steroid use. Surgery may be considered for sinusitis or to reduce the obstruction from large inferior turbinates, but septal/reconstructive surgery can induce further inflammatory change within the nose and is best deferred until the condition becomes less active.

Syphilis

Syphilis became a rarity after the advent of penicillin, but it has become quite common again and its incidence is increasing.

Clinical Features. *Stage 1* is usually marked by the appearance of a single sore (chancre), but there may be multiple sores. *Stage 2* begins ≈ 9 weeks after the infection. It is characterized by bacteremia, generalized exanthema, mucous membrane lesions, flulike systemic signs and symptoms, and the production of antibodies. *Stage 3* occurs more than 2 years after the primary infection and shows gummatous infiltration with painful swelling of the bony part of the nose, foul-smelling secretions, formation of sequestra, sharply demarcated ulceration, and usually regional lymphadenopathy. Finally, the typical saddle nose develops, affecting the bony part of the nose, and firm, radiating scars form within the nose (Fig. 2.34).

Fig. 2.34 Syphilitic saddle nose.

Fig. 2.35 Rhinoscleroma.

Pathogenesis. This illness is due to an infection with *Treponema pallidum*. Intrauterine infection may cause early congenital syphilis within the first months of life, or late congenital syphilis beginning between the age of 3 years and puberty, but accompanied by the triad of interstitial keratitis, Hutchinson teeth, and inner ear deafness. The infection may also be acquired (extrauterine) after birth.

Diagnosis. This is based on the history, serology (which is not always positive in stage 3), Nelson test (*Treponema pallidum* immobilization test, TPI), and biopsy.

Differential diagnosis. Tuberculosis and malignancy must be considered.

Treatment. Antibiotics are administered under the supervision of a venereologist. Local treatment may be required in stage 3. Once the lesion has healed, reconstruction of the defect may be required. The antibiotic of choice is penicillin. In the early stage, benzathine penicillin should be administered intramuscularly at a dose of 2.4 million IU (1.2 million IU in each buttock) as a single dose. In case of penicillin allergy, doxycycline is the usual choice (100 mg b. i. d. p. o. for 14 days); alternatively, erythromycin (500 mg q. i. d. p. o. for 14 days) can be used. In the late stage, the same dose of benzathine penicillin is injected intramuscularly on days 1, 8, and 15. Alternatively, doxycycline (200 mg b. i. d. p. o. for 28 days) or erythromycin (2 g i. v. daily for 21 days) may be given.

Rhinoscleroma

Epidemiology. This disease occurs in eastern Europe, North Africa, Central and South America, and Asia.

Clinical Features. The disease starts with an atypical rhinitis with purulent secretion and crusts. A flat, nodular infiltrate then appears in the nasal mucosa. There is increasing coarsening of the external nose (tapir nose). The lesion heals with extensive scarring (Fig. 2.35).

Pathogenesis. The infectious organism is *Klebsiella rhinoscleromatis*.

Diagnosis. This depends on the history, with particular attention to foreign travel, and on the results of biopsy and microbiologic studies.

Differential diagnosis. This consists of tuberculosis, syphilis, sarcoid, mycoses, and Hodgkin disease.

Treatment. Antibiotics are given, as dictated by culture and sensitivity tests.

Prognosis. The outcome is uncertain and recurrence is possible.

Leprosy

Epidemiology. The disease occurs in tropical and subtropical regions.

Clinical Features. A bulbous thickening in the nasal vestibule, obstruction of the nasal cavity, extensive crusting, fetid secretion, ulceration and liquefaction of the nasal framework, and leonine facies may occur.

Pathogenesis. The disease is due to an infection with *Mycobacterium leprae* (Hansen).

Diagnosis. This is based on history of contact, symptoms of infection of other parts of the body, and neurologic lesions that are exclusively sensory. Culture and the lepromin reaction also form part of the investigations.

Treatment. Single skin lesions: rifampicin 600 mg as a single dose. Paucibacillary disease (tuberculoid leprosy): dapsone 100 mg daily and rifampicin monthly for 6 months. Multibacillary disease (lepromatous leprosy): rifampicin 600 mg and clofazimine 300 mg, both monthly for 12 months.

Acute and Chronic Inflammations Localized Mainly in the Nasal Cavity

Diseases arising primarily within the nasal cavity are discussed here separately from those arising in the nasal sinuses, although disease of the nose may originate in the sinuses and vice versa.

Acute Inflammations of the Nasal Cavity

Acute Rhinitis

Clinical Features. Since the common cold can be caused by different organisms, the symptoms are not uniform. In the common form, there is a *dry prodromal stage* with generalized symptoms, including chills and a feeling of cold alternating with a feeling of heat, headache, fatigue, loss of appetite, possibly subfebrile temperature, but often a high temperature in children, as well as itching, burning, a feeling of dryness in the nose and throat, and nasal irritation. The nasal mucosa is usually pale and dry. The *catarrhal stage* usually begins a few hours later, with watery secretions, nasal obstruction, temporary loss of smell, lacrimation, rhinolalia clausa, and worsening of the constitutional symptoms. The nasal mucosa is deep red in color, swollen, and secretes profusely. After several days, the disease changes to a *mucous phase.* The generalized symptoms begin to improve, the secretions thicken, the sense of smell improves, and local symptoms gradually regress. Resolution should be achieved within a week. The common cold, or viral rhinosinusitis, is defined as duration of symptoms for less than 10 days.

Secondary bacterial infection may occur. The secretions then turn greenish-yellow, and the disease resolves more slowly. Acute intermittent nonviral rhinosinusitis is defined as an increase of symptoms after 10 days, with a duration of less than 12 weeks (Fig. 2.36).

Initial catarrh occurs in influenza and infection with other types of viruses such as parainfluenza virus, adenovirus, reovirus, coronavirus, enterovirus, myxovirus, and respiratory syncytial virus. The symptoms are as described above, but are complicated by other manifestations such as involvement of the entire respiratory tract, the gastrointestinal tract (causing diarrhea), the meninges, the pericardium, the kidneys, and the muscles.

Pathogenesis. The infection is caused by a rhinovirus. More than 100 types belonging to the picornavirus group have been isolated. The disease may also be caused by numerous other viruses. The incubation period of the rhinovirus is from 1 to 3 days. The disease is spread by droplet infection and is exacerbated by cooling of the body.

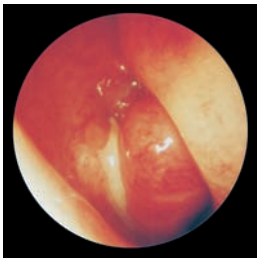


Fig. 2.36 Purulent rhinosinusitis. The mucosa is edematous and hyperemic. A track of pus is seen in the middle nasal meatus.

The most common pathogens in purulent bacterial rhinosinusitis are *Streptococcus pneumoniae, Haemophilus influenzae, Moraxella catarrhalis, Staphylococcus aureus, Streptococcus pyogenes,* and various anaerobes.

Diagnosis. Initially, it is often not clear whether a runny nose is the initial symptom or an accompanying symptom of a severe virus infection.

Differential diagnosis. This can often be made only after a few days. It includes the initial phase of an acute exanthema, allergic or vasomotor rhinitis, congenital syphilis, and nasal diphtheria (usually in children).

Treatment. There is no treatment for the basic cause. Symptomatic treatment includes decongestant nose drops or oral decongestants. Antibiotics should only be given for secondary bacterial infection, and culture and sensitivity tests should be carried out beforehand. Steam inhalations, treatment with infrared lamps, analgesics, and bed rest should be prescribed if necessary.

Prevention. While there is no scientific evidence that prevention is possible, measures to improve general health may be helpful. These include building up the patient's overall resistance using sauna baths, therapeutic regimens at health resorts, hydrotherapy, participation in sports, administration of vitamin C, and scrupulous hygiene, especially in contact with young children. Adenoidectomy may be necessary in children (see p. 243). Immunization against rhinoviruses is not yet possible, but there are vaccines against influenza.

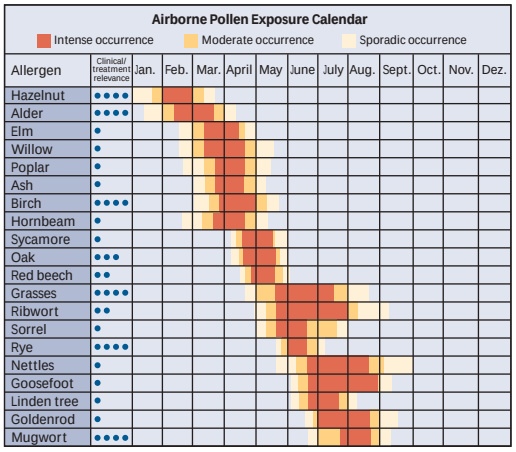


Fig. 2.37 Example of airborne pollen exposure calendar.

Allergic Rhinitis

Allergic rhinitis is mainly a type I allergic reaction. In this type of anaphylactic hypersensitivity reaction, cutaneous or mucocutaneous symptoms develop either immediately or within a few minutes after exposure. The most common form is hay fever, but other allergens may be responsible.

Clinical Features. These include itching in the nose, nasal obstruction, sneezing attacks, and a clear, watery nasal discharge. The patient may also have a feeling of stuffiness and irritation of the entire head, possibly conjunctivitis, malaise, temporary fever, loss of appetite, autonomic symptoms, possibly inability to work, and temporary hyposmia or anosmia. Secondary infection is possible.

Pathogenesis. An inhalation allergy is the cause. The shock organ is usually the nasal mucosa, but it may also be the conjunctiva or other mucous membranes. The disease is often hereditary.

Seasonal allergic rhinitis is caused by pollen (Fig. 2.37).

Perennial allergic rhinitis is due to an inhaled allergen, regardless of the season. The allergens include fungi, animal hair, house dust mites, houseplants such as primroses and roses, and also foods such as fish, strawberries, nuts, eggs, milk, and flour. There are occupational allergies—e. g., to flour for bakers, to hair and epithelial scales for hairdressers, etc.—and other allergies to bacteria and parasites.

Infection and allergy: Bacteria and viruses can act as allergens, but the practical significance of this is still controversial. Three pathogenetic mechanisms are possible:

1. An allergic reaction to bacteria or viruses without clinical infection—e. g., to nasal saprophytes.
2. An allergic reaction to bacterial or viral infection—e. g., chronic bacterial rhinitis or sinusitis with resultant sensitivity to the causative organism.
3. Secondary infection in tissue already altered by allergic reaction. In this case, the infective agent is not the same as the antigen.

The second form corresponds to the term "infection allergy." Its time course classifies it as a *late allergic reaction*.

If an infective allergy is suspected, tests should be carried out for the antigen (i. e., a positive cutaneous late reaction to a bacterial extract), and antibiotics should be given depending on the sensitivity tests. The long-term value of hyposensitization has not yet been confirmed in clinical practice.

Diagnosis. This is made primarily on the basis of the history. Investigations include cytology of the nasal secretions, intracutaneous tests, prick tests, and patch tests, intranasal challenge with rhinomanometry, olfactometry, measurement of serum and secretion IgE, and RAST.

Local findings. The nasal mucosa is livid and pale. In acute stages, the mucosa may also be deep red. The turbinates are swollen, and there are copious amounts of clear secretion.

Differential diagnosis. Nasal hyperreactivity/idiopathic rhinitis and acute rhinitis (coryza) must be considered.

Treatment. *Symptomatic:* The World Health Organization (WHO) has published guidelines on Allergic Rhinitis and its Impact on Asthma (ARIA), which make the following recommendations:

- Mild to moderate symptoms (e. g., first manifestation of allergic rhinoconjunctivitis): a third-generation oral antihistamine (e. g., levocetirizine, desloratadine) or a topical antihistamine (e. g., an azelastine nasal spray), if necessary with concomitant use of antihistamine eyedrops.

Table 2.5 Allergic Rhinitis and its Impact on Asthma (ARIA) guidelines classification of rhinitis

Infectious	Viral, bacterial or due to other pathogens
Allergic	Intermittent, persistent
Occupational (allergic and nonallergic)	Intermittent, persistent
Drug-induced	Intolerance of acetylsalicylic acid or other drugs
Hormonal	
Other causes	Nonallergic rhinitis with eosinophilia syndrome (NARES), irritants, foodstuffs, emotional factors, atrophy, gastroesophageal reflux
Idiopathic	

- Marked nasal symptoms: a combination of an oral antihistamine and a topical steroid (e.g., mometasone, fluticasone, triamcinolone).
- Severe nasal obstruction: brief application of a nasal decongestant containing an α-sympathomimetic drug. Hyposensitization: short-term titration and co-seasonal hyposensitization treatment. Causative therapy: specific immune therapy is the only treatment modality to address the underlying cause. It can prevent the development of allergic asthma.

Principle of specific immune therapy: Standardized allergen extracts of dominant single allergens (major allergens), native or polymerized (chemically modified) extracts—i. e., so-called allergoids—are used. They are bound to a carrier such as aluminum hydroxide, tyrosine, or calcium phosphate. Application may be subcutaneous (subcutaneous immune therapy, SCIT) or sublingual (sublingual immune therapy, SLIT).

Immunologic effects: T lymphocytes are influenced by the activation of regulatory $CD4^+$ T cells (IL-10, transforming growth factor β), inducing immediate hyposensitivity (lowered cytokine production) and replacement of T helper subset 2 (TH2) dominance (IL-4, IL-5, IL-13) with a TH1 response. This in turn affects the production of immunoglobulins by B lymphocytes and inhibits effector cells (e. g., mast cells, basophilic leukocytes, and eosinophilic granulocytes).

Effectiveness: Pollen allergy: symptoms decreased in 30 %; grass and birch allergies in 45 %; dust allergies in 30 %.

Surgery: Indications for surgery have to be established in septal deviation, turbinate hyperplasia, nasal polyps, rhinosinusitis, or adenoids.

Prognosis. This is generally good. The disease gradually regresses as the patients get older, but progression to bronchial asthma (or vice versa) is possible.

Complications. Involvement of the nasal sinuses and the lower respiratory tract, and polyps of the nose and sinuses are possible.

Idiopathic Rhinitis (Nasal Hyperreactivity, Vasomotor Rhinitis)

The WHO's ARIA guidelines recommend that the term "idiopathic rhinitis" should be used instead of noninfectious and vasomotor rhinitis. The synopsis of classifications in the ARIA report and the consensus report on nasal hyperreactivity defines idiopathic rhinitis as a group of nasal oversensitivity syndromes due to unknown pathomechanisms. Idiopathic rhinitis is a diagnosis of exclusion; it is considered present if the forms of rhinitis and differential diagnoses listed in Table 2.5 can be ruled out.

Clinical Features. The International Consensus Report on the Diagnosis and Management of Rhinitis (1994) requires at least two symptoms from the group *urge to sneeze, runny nose, nasal obstruction, and/or itching*, lasting for more than 1 h per day. Healthy individuals may sneeze or wipe their noses up to four times daily.

Pathogenesis. There are three pathomechanisms:

1. *Neuronal dysfunction* (Fig. 2.38*)*: Neurogenic inflammation is mediated by neurotransmitters and interacts in many ways with the immune system. Activation of *neurokinine-1 receptors* in the airways causes contraction of smooth muscles, dilation of blood vessels, glandular secretion, and extravasation of plasma proteins. Activation of *neurokinine-2 receptors* causes bronchoconstriction and stimulates afferent nerves. Hyposympathicotonia or hyperparasympathicotonia leads to obstruction and hypersecretion.
2. *Immune inflammatory disorders*: Among others, $CD3^+$, $CD25^+$, $CD8^+$, $CD45RA^+$, and T cells, eosinophils, and mast cells are increased.
3. *Mucosal damage,* with increased permeability for irritants.

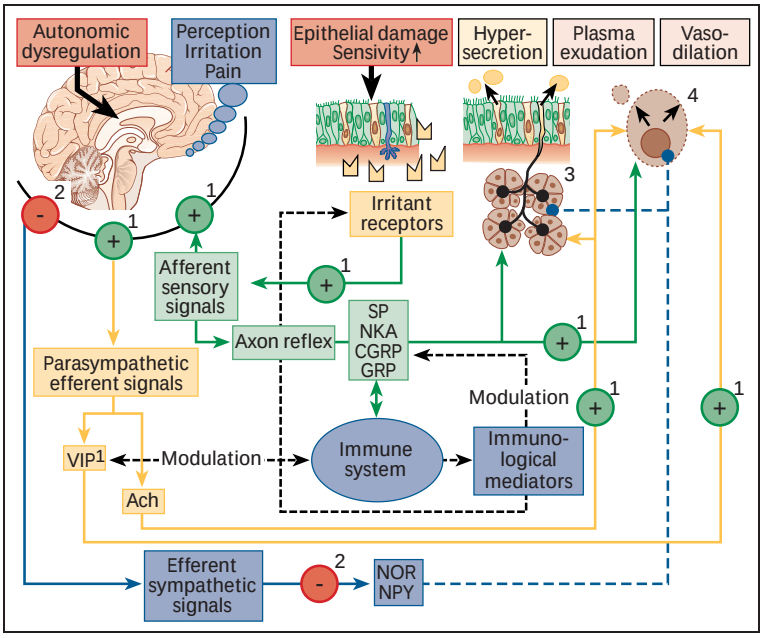


Fig. 2.38 Diagram of neural regulation of the nasal mucosa and its disorders in idiopathic rhinitis. 1, Enhanced, 2, reduced neural activity; increased density of innervation or release of neurotransmitters; 3, glands of the nasal mucosa; 4, blood vessels, vasodilation. SP, Substance P; NKA, neurokinin A; CGRP, calcitonin gene-related peptide; GRP, gastrin-releasing peptide; NOR, norepinephrine; NPY, neuropeptide Y; Ach, acetylcholine; VIP1, vasoactive interstitial peptide + VIP-like peptide (adapted from Damm M. Idiopathic Rhinitis. Laryngo-Rhino-Otol. 2006; 85:365).

Diagnosis. History, nasal endoscopy, rhinomanometry, acoustic rhinometry, allergy tests.

Treatment. *Medical:*

1. Ipratropium bromide (anticholinergic, 80–100 mg/day) is effective against rhinorrhea.
2. Topical corticosteroids are effective against symptoms such as obstruction, rhinorrhea, and postnasal drip.
3. Azelastine (antihistamine, 1.1 mg/day) works well against rhinorrhea, sneezing, obstruction, and postnasal drip.
4. An isotonic salt-water spray may be an effective treatment in mild cases.

Surgical: Invasive and surgical measures are considered only after conservative treatment has failed. Severing the nerve of the pterygoid canal (vidian nerve) used to be done to interrupt the parasympathetic innervation of the nose. This treatment option should first be simulated by reversible and repeatable blockade of the sphenopalatine ganglion. To reduce the cavernous tissue in the lower turbinate to counteract both obstruction and hypersecretion, several options such as partial excision, submucosal resection with a microdebrider, caustic agents, radiofrequency therapy (Coblation), or laser therapy, can be considered.

Foreign Bodies in the Nose

Foreign bodies are found in children in most cases and may have been retained for a very long time (Fig. 2.39). They include coins, metal fragments, peas, etc.

Clinical Features. These include unilateral nasal obstruction, worsening chronic purulent rhinitis or sinusitis, unilateral fetid secretions, and formation of a rhinolith due to deposition of calcium and magnesium salts around the foreign body.

Diagnosis. This is based on anterior rhinoscopy, nasal endoscopy, and radiology. A foreign body is often an incidental finding. The nose is inspected and probed using an endoscope after decongestion and induction of local anesthesia.

Treatment. The foreign body is removed instrumentally, if necessary under brief general anesthesia, as foreign bodies that have been present for a prolonged period are often firmly attached and provoke brisk bleeding when mobilized.

Fig. 2.39a, b In this patient, a pencil had perforated the paranasal sinus system and anterior skull base between the carotid artery and the optic nerve. It was removed 55 years later.

Note: Unilateral chronic purulent rhinorrhea in a small child should suggest the diagnosis of a foreign body, and the child must be examined by a specialist.

Chronic Inflammations of the Nasal Cavity

Rhinitis Sicca Anterior

Clinical Features. These include a feeling of dryness, irritation, formation of crusts in the nose, and also occasional mild nasal bleeding (Fig. 2.40a, b).

Pathogenesis. Several factors are responsible, such as injury to the exposed parts of the anterior nasal mucosa, dust, nose-picking, extremes of temperature, etc.

Diagnosis. The nasal mucosa on the anterior part of the nasal septum immediately posterior to the mucocutaneous junction is dry. The mucosal surface is raw, roughened, and granular. Crusts form, followed by ulceration and at times a subsequent septal perforation.

Differential diagnosis. Included here are chemical injury in chromium workers, iatrogenic septal perforation after surgery or incorrect cauterization, trauma, lupus, leprosy, and syphilis.

Treatment. Mild nasal oils that adhere well to the mucosa are applied. Saline spray is also useful. Septal perforation is discussed on p. 201.

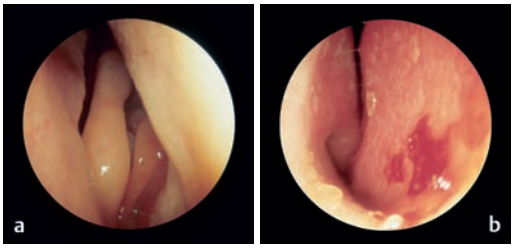


Fig. 2.40a, b The mucosa in chronic rhinitis sicca (0° lens, 4 mm).
a Normal mucosa
b Dry mucosa without a mucus layer, with bleeding at the anterior deviated septum.

Chronic Rhinitis (Nonspecific Inflammation of the Nasal Mucosa)

Chronic rhinitis is one of the most common healthcare problems, with a severe impact on lower airway disease and overall health. It is characterized by hypertrophy of the nasal mucosa, especially around the nasal turbinates, as well as by either hyperemia and edema, or true tissue hypertrophy.

Clinical Features. The main symptom is nasal obstruction, which fluctuates markedly in the early stages and also alternates from side to side. Later, it becomes continual and severe, and usually affects both sides. The secretions are viscous, stringy, colorless, and only rarely purulent. Postnasal catarrh is particularly prominent, with sniffles and compulsive throat clearing. Other symptoms include rhinolalia clausa, epiphora, secondary dacryocystitis, and secondary pharyngitis. In severe cases, fatigue, insomnia, an unsteady or woozy feeling in the head, and, occasionally, headache and a feeling of pres-

sure in the head may occur. There is a general loss of psychological and physical well-being.

Pathogenesis. The nasal mucosa is the human immunologic "front line." It is constantly forced to react to countless allergens and antigens. The most important prerequisite for this function is an intact mucociliary apparatus, which supports a wide range of reactions—e. g., absorption, secretion, transportation, cellular or humoral defenses (see p. 127). So-called inflammatory cells (macrophages, granulocytes, leukocytes, and mast cells) are always found on histopathological examination.

Chronic inflammation occurs when the normal capacity to react is overstrained, and it has many etiologies. On the one hand, recurrent inflammatory exacerbations (viral or bacterial) may cause progressive damage to the mucociliary apparatus; on the other hand, variations in anatomic shape (e. g., septal deviation, vomer spur, maxillary crest, polyps, papillomas, adenoids) may elicit chronic rhinopathy.

Cellular changes are described on p. 127.

Chronic inflammation may be due to tobacco smoke and dust, chemicals, environmental toxins, persistent extremes of temperature, abnormal humidity, pregnancy, menstruation, menopause, endocrine disorders (e. g., of the thyroid and adrenal glands, diabetes mellitus), diseases of the heart and circulation, side effects of drugs (see below), or infective allergy.

Diagnosis. The disease is long-standing, and the history often reveals one or more of the toxins named above. Examination shows a dark red and partially bluish-violet swelling, affecting the inferior turbinate especially. The nasal lumen is narrowed or obstructed. The thickened mucosa responds to decongestant nose drops in simple chronic rhinitis, but true tissue hypertrophy in chronic hyperplastic rhinitis does not.

In the later stages, the mucosa over the turbinates develops a slightly granular surface, which gradually becomes nodular and demonstrates micropolyps. These edematous processes can develop into single or multiple *nasal polyps*, especially around the inferior turbinate. Often, this true tissue hypertrophy begins at the posterior ends of the turbinates, usually of the inferior turbinate. The choanae are then blocked by mulberry-shaped masses, which can be seen only on endoscopic examination of the nasopharynx or on indirect examination with a mirror.

Differential diagnosis. This includes sinusitis, foreign bodies, specific infections of the nasal mucosa (see pp. 147, 156), adenoidal hypertrophy, allergy, Wegener granulomatosis, and tumors. Biopsy should be performed if appropriate.

Treatment. *Conservative:* Any known or suspected etiologic agents should be dealt with. Some medicines may need to be curtailed, drug overuse controlled, and the patient may need endocrinology studies by a specialist physician. Attention to the environment and occupation may prove valuable. Symptomatic medical treatment by decongestant nose drops is only of short-term benefit (see p. 159). In the long term, uncritical symptomatic treatment is not only valueless but may be damaging. Topical steroids (e. g., triamcinolone, mometasone, fluticasone) are the treatment of choice in chronic hyperplastic rhinitis. These drugs are suitable for long-term use.

Surgical treatment, in increasing order of extent:

1. *Reduction of the inferior turbinate* is performed with the use of various lasers, radiofrequency surgery (Coblation), and submucosal diathermy.
2. *Turbinectomy* includes a limiting resection of the lower edge of the inferior turbinate, occasionally of the middle turbinate, and from the enlarged posterior ends of the turbinate. The purpose is to reduce the volume of the turbinate, but atrophic rhinitis can occur if too much tissue is removed (see p. 155).

Rhinitis of Pregnancy

Increasing swelling and obstruction of the nose may occur during pregnancy, especially in the second half. The symptoms resolve after delivery.

Rhinitis Medicamentosa

Reversible or irreversible damage to the mucosa may be caused by topically or systemically applied drugs.

Mucosal swelling, potentially leading to hyperplastic rhinitis, may be caused by:

- Antihypertensives such as β-blockers, reserpine, clonidine, or angiotensin-converting enzyme inhibitors.
- Phosphodiesterase type 5 inhibitors such as sildenafil.
- Hormones such as exogenous estrogens, oral contraceptives.
- Pain relievers such as aspirin or nonsteroidal anti-inflammatory drugs (NSAIDs).

- Psychotropic drugs such as chlordiazepoxide–amitriptyline.
- Miscellaneous drugs such as cocaine or gabapentin.

Dryness of the nasal mucosa is caused by atropine, belladonna preparations, corticosteroids, imidazoline, or catecholamine derivatives.

Toxic rhinopathy may result from abuse of decongestant nose drops (see p. 159).

Fig. 2.41 Ozena. Atrophy of the nasal epithelium and the turbinates, with crusts on the epithelial surface and bacterial infection.

Atrophic Rhinitis and Ozena

Atrophic rhinitis is sometimes accompanied by a foul smell from the nose, and in these cases it is known as ozena.

Clinical Features. This disease occurs mainly (but not only) in women, often beginning at puberty. The face is typically flattened and broad. The nasal cavity usually is filled completely with greenish-yellow or brownish-black crusts. Once the crusts are removed, it can be seen that the cavity is very spacious. The mucosa is atrophic and dry due to fibrosis of the subepithelial layer, and the inferior turbinate is atrophic. In ozena, a fetid secretion and crusts are present. The repulsive smell considerably hinders social contact. The patients have anosmia, making them unaware of the unpleasant smell that they produce. However, they do have a sensation of nasal obstruction. There are often severe mucosal changes, including dryness and dry, thick crusts affecting the entire pharynx, larynx, and trachea.

Pathogenesis. This is not known with certainty, but it is probably multifactorial. Important accepted etiological factors are malnutrition, iron deficiency, vitamin A deficiency, immunodeficiency, and low IgA. The disease is more common among Asians than Caucasians, and more common in Caucasians than in people of African origin. There are geographic concentrations—e. g., in eastern Europe and India. The nasal cavity is abnormally wide due to atrophy of the mucosa and of the bony nasal skeleton. The mucosal glands and the sensory nerve fibers degenerate, the respiratory epithelium undergoes squamous metaplasia, and the mucociliary cleansing system is destroyed. The thick, gluey secretions are decomposed by bacterial proteolysis (Fig. 2.41) of *Klebsiella ozaenae*, *Staphylococcus aureus, Proteus mirabilis*.

Secondary atrophic rhinitis is caused by trauma or excessively extensive surgery of the nose and sinuses, as well as occupational exposure to glass, wood, asbestos, or extensive cocaine consumption.

Treatment. *Conservative:* The nose is cleansed by irrigating it several times a day with dilute salt water and by inserting large cotton-wool tampons impregnated with greasy ointments. Local applications include oily nose drops, emulsions, or ointments, and possibly vitamin A supplements. Steam inhalations with saline solution are given, and osmotically active powders—e. g., dextrose—are sniffed into the nose.

Surgical treatment can be used to prevent the nose from drying out by narrowing the nasal cavity. Two main procedures are used:

1. Bolstering of the nasal mucosa with autologous or homologous tissue (cartilage or bone chips)
2. Median displacement of the lateral nasal wall by mobilizing and rotating it toward the midline, then fixing it in its new position to produce narrowing of the nasal cavity.

Nasal Diphtheria

This usually occurs in children older than 6 months. The secretions may be hemorrhagic and purulent, or exclusively purulent with crusts and cracks around the nostrils. The disease is rare in adults. Nasal diphtheria can occur alone or in combination with pharyngeal diphtheria (see p. 264).

Diagnosis. Made by culture. *This is a notifiable disease.*

Treatment. With antitoxic serum (see p. 264).

Tuberculosis of the Nasal Cavity

Two forms can be distinguished: 1, lupus (see p. 147) and 2, exudative ulcerative mucosal tuberculosis. This is usually the result of hematogenous or intraluminal spread in pulmonary tuberculosis. The mucous membrane shows moth-eaten ulcerations.

Diagnosis. This is determined by bacteriology and biopsy.

Syphilis of the Nasal Cavity

Stage 1 (caused by infection from instruments) and stage 2 are very rare. In the middle of the 19th century, stage 3 was more common and was the main reason for saddle nose deformities. Nowadays, cases of syphilis are increasing in numbers again, but nasal manifestations continue to be rare. The diagnosis is made by serology and biopsy, and possibly by identification of the organism.

Sarcoidosis

See page 390.

Scleroma of the Nasal Cavity

See page 148.

Leprosy of the Nasal Cavity

See page 148.

Glanders

Epidemiology. This disease occurs mainly in Eastern Europe and North America. The causative organism is *Malleomyces mallei*. It is transmitted by ungulates.

Clinical Features. Purulent, foul-smelling nasal secretions, granulations, and ulcers are found on the nasal mucosa. The disease may extend to the external nose and surrounding areas.

Diagnosis. This is made by culture, an agglutination test, and biopsy. *The disease is notifiable on suspicion.*

Treatment. Antibiotics are administered.

Blastomycosis

Epidemiology. This disease occurs in North, Central, and South America and is caused by several types of *Blastomyces* (yeasts).

Clinical Features. Crusts and mucosal ulcers are found, which may extend to the external nose. The regional lymph nodes are usually enlarged.

Diagnosis. Culture and biopsy provide the diagnosis.

Treatment. Amphotericin B is given, and may be followed by excision of the diseased skin or mucosa.

Rhinosporidiosis

Epidemiology. The disease occurs in India, Sri Lanka, Africa, and North and South America and is caused by *Rhinosporidium seeberi*.

Clinical Features. These include polypoid, reddish, nodular granulations of the anterior part of the nasal cavity that bleed easily. The disease may extend to the nasal sinuses and pharynx.

Diagnosis. This is made by culture and biopsy.

Differential diagnoses. Mycoses that can also cause nasal symptoms include moniliasis, aspergillosis, mucormycosis, and coccidioidosis.

Treatment. The polyps are removed and the base is cauterized, but there is a danger of bleeding.

Wegener Granulomatosis

This necrotizing granulomatous inflammatory condition preferentially affects the airways and the renal parenchyma. Without treatment, it can progress to a generalized necrotizing vasculitis.

Clinical Features. This nasal disease presents with increasing nasal obstruction, epistaxis, nasal discharge, a combination of crusts, mucosal granulations, and in later stages septal perforations and saddling of the cartilaginous skeleton. There is progressive loss of tissue. In addition, the patient has pulmonary symptoms (chronic bronchitis) and generalized symptoms such as malaise, fatigue, night sweats, and intermittent limb pain (Fig. 2.42).

Pathogenesis. The cause of the disease is not clear. It may be an autoimmune condition with granulomatous arteritis, perivasculitis, and necrotizing vasculitis. There is an association with HLA-B8 and HLA-DR2 antigens, related to a family predisposition. Generalized manifestations affect other parts of the body, such as the lungs, kidneys, liver, middle ear, larynx, and trachea.

Diagnosis. The diagnosis is based on the clinical picture, the local findings, the course of the disease, and biopsy. The erythrocyte sedimentation rate (ESR) is usually markedly elevated. Electrophoresis shows reduced albumin and increased globulin. Antineutrophil cytoplasmic antibodies (ANCA) and cytoplasmic-staining antineutrophil cytoplasmic antibodies (c-ANCA; proteinase 3, PR3) are very specific and relatively sensitive tests for Wegener granulomatosis. The lungs should be investigated carefully with a chest radiograph and a CT scan if changes are present. Renal investigations and laboratory tests should also be done.

Differential diagnosis. This includes midline granuloma, leukemia, malignant lymphoma, and sarcoid.

Treatment. This depends on the extent of the disease. It includes local nasal treatment with oils and steroids. In patients with systemic disease, methotrexate 0.3 mg/kg weekly i.v., or prednisolone, and when appropriate high-dose co-trimoxazole (Septrin), or monoclonal antibody (rituximab).

Prognosis. Poor. Death is often due to renal failure.

Fig. 2.42 A young patient with Wegener granulomatosis.

Lethal Midline Granuloma/T-Cell Lymphoma

Lethal midline granuloma is a clinical term that describes some diseases with an aggressive and progressive destruction of the midface. The nasal prodromes are similar to those of Wegener granuloma. After exclusion of Wegener granuloma, almost all remaining cases that present as lethal midline granuloma are peripheral sinonasal angiocentric T cell and/or natural killer (NK) cell lymphomas. Progressive ulceration of the central part of the face also extends to the maxilla (Fig. 2.43a, b).

Fig. 2.43a, b A patient with a midline granuloma. The clinical situation 3 months after a frontal sinus operation.

Pathogenesis. The cause is unknown. The disease may be due to an immune deficiency, but may also be related to a malignancy of the reticulohistiocytic system.

Diagnosis. Typically, the disease is localized to the central part of the face. The ESR is elevated, and there is a hypochromic anemia.

Differential diagnosis. This includes Wegener granuloma, glanders, scleroma gumma, noma, blastomycosis, and histoplasmosis.

Treatment. A combination of radiotherapy, steroids, chemotherapy, immunosuppressive agents, and antibiotics is applied. Occasionally, extensive excision is required.

Prognosis. With early diagnosis and energetic treatment, the outlook is not necessarily unfavorable.

Local Conservative Treatment in the Upper Respiratory and Digestive Tracts

The following factors facilitate the treatment of diseases of the upper respiratory and digestive tracts:

- Easy accessibility for local medical treatment.
- Possibility of combining local and systemic medical treatment.
- Easy accessibility for physical methods of treatment such as inhalation, infrared or microwave thermotherapy, treatments in a health resort, change of climate, ionizing radiation for malignancy, etc.

Physical Treatment

Inhalation. Two main methods are possible:

1. *Steam inhalation:* Steam is used for its own therapeutic merits or to carry active agents. A steam kettle or tent can be used. The droplet size is greater than or equal to 30 µm, so the vapor is unstable and rapidly precipitates. Steam is often supplemented by the addition of salts, volatile oils, active agents, detergents, wetting agents, or agents that act on mucopolysaccharide secretions. Precipitation occurs mainly in the nose, mouth, pharynx, and larynx.
2. *Aerosol inhalation:* A true aerosol is used, with a droplet size of less than 30 µm in a quasi-stable suspension of liquid in air. The smaller the particles, the deeper they travel into the airways. The optimal droplet size for the nose and pharynx is 10–15 µm (up to 50 µm), for the trachea 5–10 µm, and for the bronchi and alveoli less than 5 µm. Jet nebulization is better for the upper and middle airways than ultrasound or a gas-driven steam. Drugs used in the aerosol include vasoactive substances to decongest the mucosa, secretolytic agents, antiallergic substances, steroids, antibiotics, chemotherapy, antiasthmatic drugs, and vitamin preparations.

Rules for inhalation therapy:

- The aerosol's droplet size should be sufficient to reach the principal sites of disease in the respiratory tract.
- Medications used should have a pH of around 7.0 and be water-soluble, approximately isotonic, nontoxic and nonirritating, well-retained, and capable of being atomized.
- An inhalation session should last 10–15 min.

Local thermal treatment. The nasal mucosa reacts to warmth with decongestion and swells when exposed to cold. The decongestant effect of applying external warmth is used to treat inflammatory swelling.

The currently preferred warming methods involve the use of microwaves with a wavelength of less than 1 m for the sinuses, larynx, etc. Infrared rays with a wavelength of less than 760 nm are used to treat superficial soft-tissue inflammation. They are generated by an electric lamp with a carbon filament that emits infrared-rich light, and are particularly effective in chronic catarrhal rhinosinusitis. Light in the visible spectrum, especially red light with a wavelength of between 760 and 400 nm, is used to treat superficial soft-tissue inflammations, as are moist warm dressings, mud packs, and heating pads.

Warmth elicits hyperemia by both direct and reflex action on the deep tissues, accelerating and intensifying anti-inflammatory processes in the tissues. Application of cold, on the other hand, inhibits the inflammatory process by retarding the circulation.

Spa treatment. This form of treatment is popular in Europe. In patients with chronic mucosal diseases of the upper respiratory tract, the following are indicated: exposure to the sun or bathing in sulfur springs; exposure to the dry stimulating climate of the high mountains (for chronic catarrh with increased secretions); exposure to a moist, stimulating climate (for dry chronic catarrh). Treatment in spas or health resorts should last for at least 4–6 weeks, and even several months in children.

Medical Treatment

Principles of topical drug application

- Drugs that are active only on the surface of the mucosa (e.g., drugs soluble in oil) need to be distinguished from those that are absorbed and act within the mucosa (e.g., water-soluble drugs).
- Occasionally, water-soluble solutions are absorbed through the mucosal surface as rapidly as an intravenous injection.

- The drug must not damage the ciliary action, the mucosa, or the superficial film of secretions.
- The pH of the drug should be around 7.0, and its osmotic pressure should be between 0.5 and double isotonic; hypertonic solutions dry out the nasal mucosa.
- Locally applied drugs should not have generalized effects, and should be neither antigenic nor carcinogenic.

Groups of drugs. *Vasoactive substances*—e.g., epinephrine, or imidazoline derivatives such as naphazoline and xylometazoline—cause the mucosa to shrink due to vasoconstriction. Uncritical persistent use involves a risk of habituation, leading to rhinitis medicamentosa and resistant mucosal swelling, failure of the autonomic vascular regulation, and organic mucosal damage. Local or systemic decongestants of any type should therefore be used for a brief period only—i.e., for a maximum of 1–2 weeks at a time.

Note: In infants and small children, there is a risk of acute intoxication due to the use of nose drops. Special preparations should therefore be used for children.

Antibiotics: The prerequisites for antibiotic treatment are a correct diagnosis, a clear indication for antibiotic use, and selection of the most appropriate antibiotic and duration of treatment. Swabs and cultures, white blood counts, erythrocyte sedimentation rate (ESR), C-reactive protein (CRP) assay, and in certain cases procalcitonin values are important for diagnosis and assessment of the course. Usually, several antibiotics are equally suitable. The final choice depends on the severity of the disease, the patient's immune status, age, allergies, impairment of liver or kidney function, the activity spectrum of the antibiotic, potential undesired reactions, and price.

Specific information on the most common pathogens and recommended antibiotics is discussed under the headings for the individual clinical entities.

Corticosteroids: These are a double-edged sword. Although they have strong antiallergic and antiinflammatory effects on the upper airways, they also elicit inflammation at other sites, especially in the gastrointestinal tract. Modern topical corticosteroids—e.g., mometasone, fluticasone, or triamcinolone—have marked anti-inflammatory and antiallergic effects on the nasal mucosa, but very low biological availability. Because they are absorbed only to a very slight extent, they appear in minimal concentrations in serum, thus avoiding side effects and allowing long-term administration.

Treatment is usually empirical at first. Ideally, it should be reconsidered after reexamining the patient 48–72 hours later in the light of results for cultures, resistance testing or blood cultures, and should then be continued with the most suitable drug. If possible, a narrow-spectrum antibiotic is preferable to a broad-spectrum one.

Mild bacterial rhinitis in an immunologically competent patient does not require antibiotic therapy.

Oral corticosteroids should be given in single, decreasing (tapered) doses in the morning over a period of up to 3 weeks.

Secretolytic agents: In Germany, standardized Myrtol has been shown to be effective in acute sinusitis and acute bronchitis. It has various effects on inflamed mucosa in the nose and paranasal sinuses. It alkalizes the acidic mucosal environment and has secretolytic, secretomotoric, and antimicrobial effects. Standardized Myrtol may be given for mild bacterial infections or can be used to supplement an antibiotic.

Antihistamines, leukotriene antagonists, cromons: See pp. 150, 168.

Acute and Chronic Rhinosinusitis

The European Position Paper on Rhinosinusitis and Nasal Polyps (2005) recommends use of the term "rhinosinusitis"—firstly because every infection of the nose involves the mucosa of the paranasal sinuses, which reacts in various ways and at various intensities, and secondly because the great majority of acute and chronic inflammations of the paranasal sinuses are rhinogenous.

This section is based on the following international consensus statements and position papers, and as far as possible on evidence-based criteria:

- European Position Paper on Rhinosinusitis and Nasal Polyps (2005, 2007).
- The World Health Organization (WHO) guidelines on Allergic Rhinitis and its Impact on Asthma (ARIA) (2001).

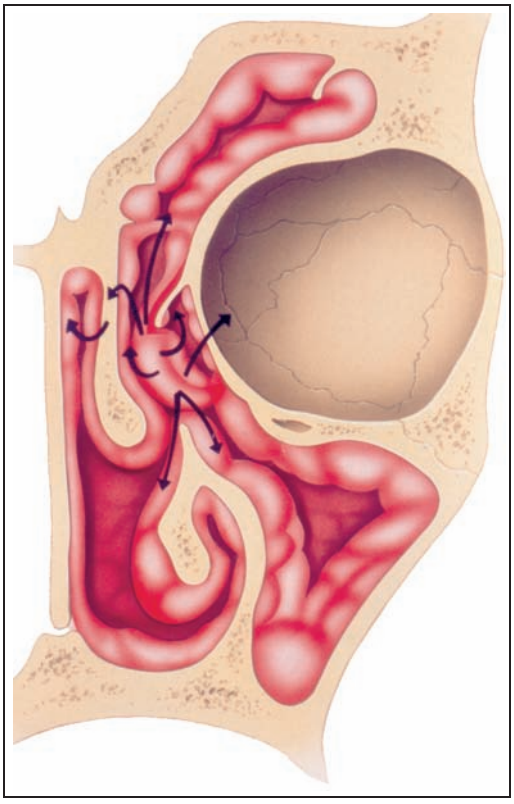


Fig. 2.44 The anterior ethmoidal labyrinth is the etiologic center for acute, recurrent, and chronic inflammations of the maxillary and frontal sinuses. The mucosal disease spreads radially from the ethmoid to the large sinuses, olfactory rim, and inferior turbinate. Inflammation may spread to the orbit through the lamina papyracea.

- International Conference on Sinus Disease: Terminology, Staging, Therapy (Princeton, 2003).
- Recommendations of the European Academy of Allergology and Clinical Immunology (EAACI).
- The European Rhinology Society (ERS) EP3OS document (2007).
- The European Position Paper on Rhinosinusitis and Nasal Polyps (EP3OS), published by the European Rhinologic Society (ERS) (2007).

Pathogens and Antibiotics (Initial Empirical Therapy)

Acute rhinosinusitis: Pathogens include *Streptococcus pneumoniae, Haemophilus influenzae, Moraxella catarrhalis, Staphylococcus aureus, Streptococcus pyogenes*, and anaerobic bacteria. Antibiotics: aminopenicillin with or without a β-lactamase inhibitor; second- and third-generation cephalosporins, macrolides, and quinolones. In severe disease, intravenous administration, otherwise orally.

Chronic rhinosinusitis: Pathogens: *Staphylococcus aureus, Streptococcus pneumoniae, Haemophilus influenzae, Pseudomonas*, anaerobic bacteria, fungi; beware of mixed infections. Antibiotics: aminopenicillin with or without a β-lactamase inhibitor; macrolides, quinolones, second- and third-generation cephalosporins, and clindamycin. In severe disease, intravenous administration.

Acute Rhinosinusitis

Acute and intermittent rhinosinusitis is defined as sudden onset of two or more of the following symptoms: *blockage/congestion, discharge, anterior/postnasal drip, facial pain/pressure, reduction/loss of smell for up to 12 weeks*.

Ethmoidal Sinusitis

The anterior ethmoid bone is the morphologic connection between the nose and the maxillary and frontal sinuses. Both paranasal sinuses drain through the anterior ethmoid to the nose (Figs. 2.44, 2.45). Viral, bacterial, or allergic infections of the nasal mucosa or nasopharynx spread immediately to the ethmoidal sinus. Mucosal swelling, edema, and subsequently hyperplasia lead to obstruction of the drainage routes (ethmoidal infundibulum, frontal recess), backup of secretions, and infection of the frontal and maxillary sinuses.

Clinical Features. Impaired nasal breathing, hyposmia or anosmia, sensation of pressure between the eye and nose, subfebrile temperatures, acute inflammation; depending on severity, a feeling of pressure and pain in frontal and maxillary sinuses (Figs. 2.46, 2.47a,b).

Diagnosis. Routine otorhinolaryngology examination; inspection (attention to edema of the upper eyelid); palpation of the medial angle of the eye, exclusion of diplopia; nasal endoscopy (critical): mucosal edema, track of pus in the middle nasal meatus; swabs for culture and resistance testing.

Diagnostic imaging: Paranasal sinuses (occipitomental and/or occipitonasal projections), coronal CT in case of recurrent exacerbations or chronic sinusitis and always before surgery.

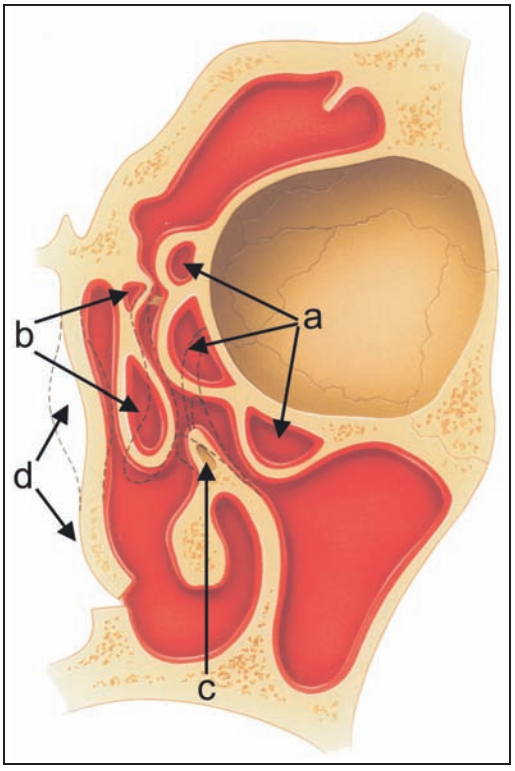


Fig. 2.45 Schematic illustration of typical cell formations causing recurrent sinusitis. a, Cell formations in the medial orbital wall, with a large ethmoidal bulla and orbitoethmoid cells (Haller cells); b, pneumatized sections of the middle turbinate; c, the uncinate process, which varies in size, length, and point of insertion; d, deviated nasal septum.

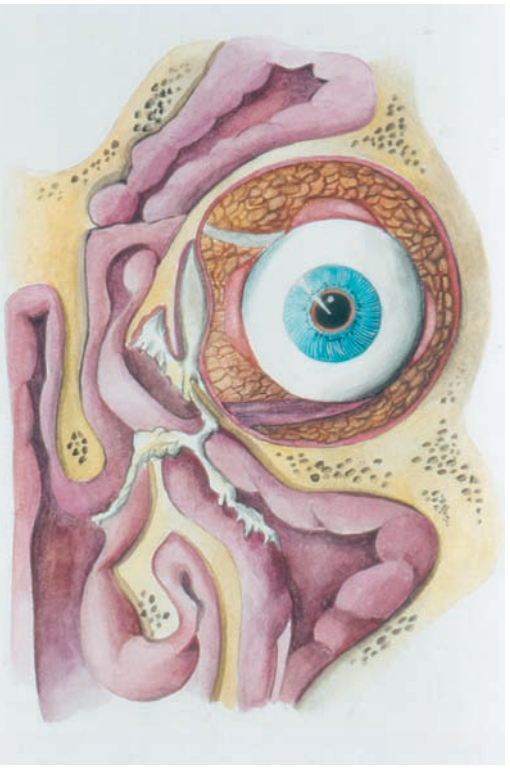


Fig. 2.46 Schematic illustration of ethmoidal sinusitis. Children are at particular risk for orbital complications, due to the thinness of the lamina papyracea. (Illustration courtesy of A. Muecke, Berlin, Germany.)

Olfactometry, allergy tests (prick test, RAST) in chronic and recurrent forms.

Differential diagnosis. Charlin neuralgia, cluster headaches.

Complications. Especially in children, rapid spread to the orbital cavity (due to thinness of the lamina papyracea) and development of orbital complications (see p. 186).

Treatment. *Acute ethmoidal sinusitis:* Decongestant nose drops, high tamponades, decongestion and aspiration of the middle nasal meatus. Application of infrared light or microwave or short-wave therapy, secretolytic agents; administration of antibiotics in case of failure of conservative therapy or purulent infection (see maxillary sinusitis). Possibly oral corticosteroids, or medialization of the medial turbinate. In some countries, supplementing antibiotics with simple antral lavage in severe acute purulent sinusitis has been found to be often effective. Surgery is indicated for recurrent sinusitis with involvement of the maxillary and/or frontal sinuses, orbital, or central complications.

Chronic ethmoidal sinusitis: Topical corticosteroids, antibiotics. Surgery is indicated if conservative therapy has failed, for recurrent sinusitis (maxillary and/or frontal), for nasal and paranasal polyposis, for chronic sinusitis with or without bronchial asthma, or for suspected tumor. The surgical techniques depend on endoscopic and CT findings: infundibulotomy; anterior or complete ethmoidectomy, possibly with supraturbinate fenestration of

Fig. 2.47a, b a A 9-year-old boy with palpebral edema as a sign of incipient orbital complications.
b The axial CT shows ethmoid sinusitis.

the maxillary cavity or enlargement of the frontal recess; pansinus operation.

Maxillary Sinusitis

Acute or chronic bacterial, fungal, or allergic rhinogenous, or (more rarely) odontogenic inflammation. The vast majority of cases originate at the lateral nasal wall and the anterior ethmoid. The maxillary and frontal sinuses are functionally downstream from the ethmoid.

Symptoms of acute maxillary sinusitis. These include acute pain in the central and ipsilateral parts of the face, hyperesthesia of the facial skin, but occasionally only a sensation of pressure or fullness. Sensitivity to tapping over the cheek, ipsilateral swelling of the turbinates, and a stream of pus in the middle meatus and on the floor of the nose are other findings (Figs. 2.48, 2.49).

Odontogenic sinusitis: Pathogens are *Streptococcus intermedius* and *Streptococcus constellatus*, often mixed with anaerobes (e.g., *Fusobacteria, Peptostreptococcus,* or *Prevotella*).

Symptoms of chronic maxillary sinusitis. Pain is often slight, and there may be only a sensation of pressure. Symptoms also include headache that increases on bending head forward, swollen cheeks, palpebral edema, chronic nasal obstruction, mucoid or purulent secretion, painful trigeminal nerve exit, and neuralgia in the distribution of the infraorbital nerve, disorders of smell including cacosmia, nasal fetor, chronic rhinitis, and hypertrophic turbinates, streams of secretion, and possibly polyps.

Diagnosis. Case history, routine otorhinolaryngology examination, inspection, palpation (cheek, facial wall of maxillary sinus), nasal endoscopy, examination for pain on bending head forward and of nerve exits, pain on tapping, percussion of the teeth, dental status.

Sonography: A-mode ultrasound reveals partial or total echo at the back wall of the maxillary sinus.

Radiography: Plain radiography shows partial or total clouding, or an air–fluid level in the maxillary sinus. CT is appropriate in severe cases, in chronic sinusitis, when complications are present, and if surgery is indicated.

Allergy testing: Prick test, RAST, intranasal provocation.

Treatment of acute rhinogenous sinusitis. *Conservative.* Nasal detumescence by use of decongestant nose drops, heat application (infrared light at a distance of 30 cm from the face and nose) or deep heat (short wave), steam inhalation with chamomile or volatile oils.

Herbal secretolytic agents (e.g., standardized Myrtol, which has bacteriostatic, mucolytic, secre-

tomotoric, and deodorizing effects); analgesics (paracetamol, ibuprofen).

Note: Acetylsalicylic acid (ASA, aspirin) must be used with caution, due to the high rate of ASA intolerance among sinusitis patients.

Antibiotics: The first choice is an aminopenicillin in combination with a β-lactamase inhibitor, or a second- or third-generation cephalosporin. Alternatives are macrolides, ketolides, or third- or fourth-generation fluoroquinolones, as indicated by resistance testing.

Lavage or drainage of the maxillary sinus is indicated in persistent empyema (swabs should be taken first).

In patients with allergies, a third-generation antihistamine (see p. 151), steroids (topical or oral), or hyposensitization are indicated.

In suspected *Aspergillus* infection, a search should be made for a foreign body (overstuffed root canal; endoscopy of the maxillary sinus). For *Aspergillus* sinusitis, see p. 182.

Treatment of acute odontogenic sinusitis. In patients with odontogenic sinusitis, the first-line treatment is dental repair, root canal treatment, or extraction.

Antibiotics: The first choice is penicillin (phenoxymethyl penicillin, aminopenicillin plus a β-lactamase inhibitor); alternatives (after resistance testing) are metronidazole, lincosamide, cephalosporins, or macrolides; reserve antibiotics are carbapenems (imipenem, meropenem).

Surgical treatment is indicated when conservative treatment fails (e.g., in chronic rhinosinusitis with or without bronchial asthma, fungal infections, recurrent empyema, or polyposis).

Treatment of chronic maxillary sinusitis. Antibiotics are used as described above.

Surgical treatment: Infundibulotomy, anterior or complete ethmoidectomy with supraturbinate fenestration of the maxillary sinus, if necessary with intracavitary repair (removal of polyps, cysts, or fungal concrements).

Frontal Sinusitis

This is an acute or chronic rhinogenous infection of the frontal sinus involving viral, bacterial, or allergic inflammation and obstruction of the anterior ethmoid (frontal recess). The spectrum of pathogens is

Fig. 2.48 A track of pus along the lateral nasal wall—a typical finding in purulent maxillary sinusitis.

Fig. 2.49 Mucosal edema in both maxillary sinuses, visualized on a plain film.

described above. It may also occur in the form of diving sinusitis or barosinusitis. Usually, anatomic variations that impede ventilation and drainage of the frontal sinus are present (e.g., high septal deviation, pneumatization variations of the ethmoid air cells; see p. 161).

Clinical Features. These include severe pain in the forehead, sensitivity of the forehead to pressure or

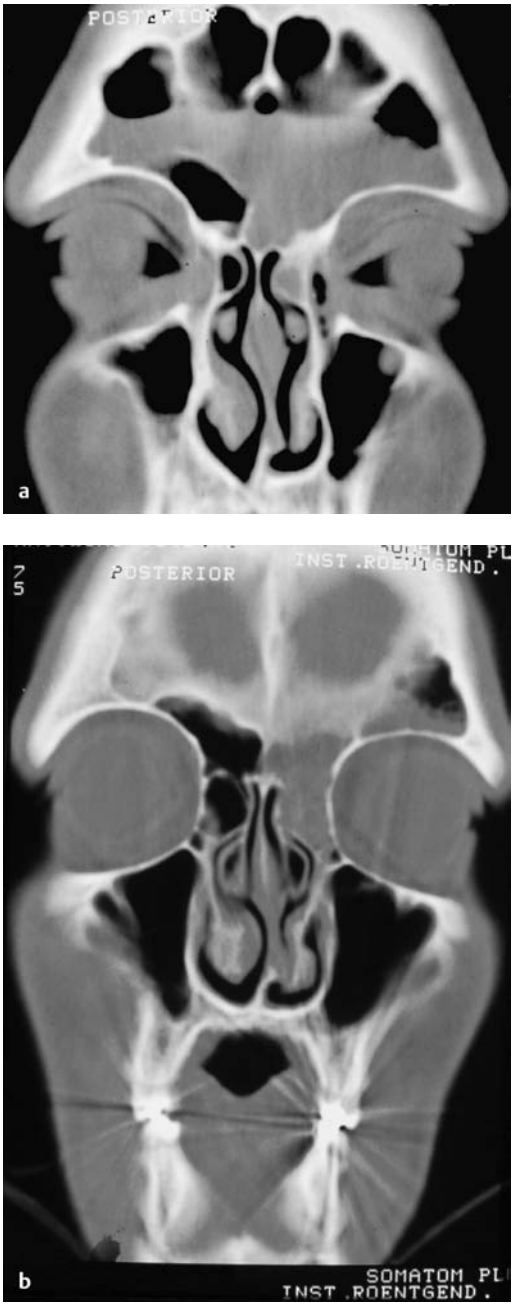


Fig. 2.50a, b a Empyema of both frontal sinuses. b Occlusion of the anterior ethmoid by hyperplastic, edematous mucosa.

tapping, tender sites of exit of the supraorbital nerve, and tracks of secretions or pus on the anterior part of the middle meatus. The mucosa of the middle meatus is often glazed, swollen, and red (Fig. 2.50a,b).

Complications. Orbital complications (see p. 186), osteomyelitis of the frontal bone with abscess formation (Pott puffy tumor), meningitis, epidural abscess, brain abscess, thrombosis of the sagittal sinus, mucocele.

Diagnosis. *Routine otorhinolaryngology examination; inspection:* edema of forehead and eyelids, conjunctivitis; palpation: floor of frontal sinus painful, exit of the first trigeminal nerve (supraorbital branch), tender pain on bending the head forward.

Nasal endoscopy: Edema, pus and mucoid pus in middle nasal meatus; inflammatory redness. Swabs: culture, resistogram. Radiography of the paranasal sinuses shows mucosal hyperplasia or empyema.

Further diagnostic measures: Coronal CT in case of recurrent exacerbations or preoperatively.

Laboratory investigations: CRP, white blood count, ESR.

Allergy tests: Prick tests, RAST.

Depending on the clinical findings, an ophthalmologist or neurologist should be consulted (lumbar puncture if meningitis is suspected).

Treatment. Decongestion and aspiration of the middle nasal meatus. Application of infrared heat or deep heat by microwave or short wave, endoscopically guided placement of high tamponade in the middle nasal meatus—e.g., using a sponge soaked with tetracaine and epinephrine. “High fillings” in the middle meatus.

Antibiotics: Start empirically, then assess effectiveness after 3 days; if necessary, adjustment according to the resistogram (see maxillary sinusitis, p. 162).

If empyema is present, the frontal sinus is drained with endoscopic surgery; in the age of endoscopic microsurgery, puncture using the Kümmel and Beck method has become less important (see p. 140).

Surgical intervention: Indications are recurrent or chronic exacerbations or orbital or central complications (e.g., subperiosteal abscess, orbital phlegmon, meningitis, epidural abscess, brain ab-

scess, sinus thrombosis, failure of conservative therapy, osteomyelitis, mucocele). The principles of endonasal and external surgery of the frontal sinus are explained on pp. 171, 177.

Sphenoid Sinusitis

Acute, chronic, rarely allergic inflammation of one or both sphenoid sinuses, usually due to morphologic or inflammatory obstruction of the ostium. Drainage of the sphenoidal cavity proceeds via the sphenoethmoidal recess, rather than through the anterior ethmoid to the nose as in the maxillary and frontal sinuses (Fig. 2.51a, b).

Pathogenesis. This disorder may develop independently and alone, or in association with other forms of sinusitis (pansinusitis). Acute and chronic sinusitis in ventilated patients (usually in intensive-care units) starts with sphenoid sinusitis, due to the patient's constantly supine position.

For the pathogen spectrum, see page 160.

Clinical Features. Signs of infection are largely lacking; overall, the symptoms are vague. The patient may report pain or a feeling of pressure within the skull, radiating to the occiput or the temple. Deep pain in the eyes is also possible. Nasal respiration is usually not obstructed. Secretions usually drain into the nasopharynx, causing postnasal drip and coughing. There is a stream of pus on the posterior wall of the pharynx or in the superior meatus.

Anatomy. Several nerves run along the lateral wall of the sphenoid sinus—e. g., the optic, abducent, and maxillary nerves. The posterior ethmoidal air cells may invade the body of the sphenoid in a cranial direction, interfering with pneumatization; they may project above the ethmoidal cavity. If they achieve a typical pyramidal configuration (i. e., with the tip pointing dorsally) with prominence of the optic nerve, they are termed Onodi cells (posterior sphenoethmoidal air cells).

Complications. Orbital complications, spread of inflammation to the optic nerve, visual impairment, paralysis of the abducent nerve; central complications: meningitis, brain abscess, thrombosis of the cavernous sinus; osteomyelitis.

Diagnosis. Routine otorhinolaryngology examination, nasal endoscopy, swabs of free pus for culture and resistance testing, coronal CT.

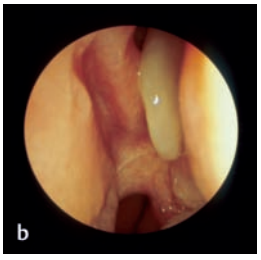


Fig. 2.51a, b Sphenoid sinusitis, caused by occlusion of the sphenoidal ostium by a polyp.

If diagnosis of the pathogen is equivocal, especially before surgery, vascularization has to be studied (e. g., by magnetic resonance angiography or angiography). Consultation with an ophthalmologist and a neurologist, and a radioisotope bone scan, are indicated if osteomyelitis is suspected. Allergy tests: prick test, RAST.

Differential diagnosis. Mucocele, benign or malignant tumors, fungal disease, pituitary tumor, meningioma of the sphenoid wing.

Treatment. *Conservative:* Localized decongestion and aspiration of the sphenoid, tamponade of the sphenoethmoidal recess with a pointed swab containing, e. g., tetracaine/epinephrine; antibiotics (see pp. 159, 160); antiinflammatory agents; in appropriate cases, antiallergic therapy (see p. 150); oral steroids.

Surgery: Indications are failure of conservative treatment and incipient or overt complications.

Principle of the operation: The method of choice is endoscopic or microscopic transnasal (via the sphenoethmoidal recess) or transethmoidal enlargement of the ethmoidal cavity ostium, or trep-

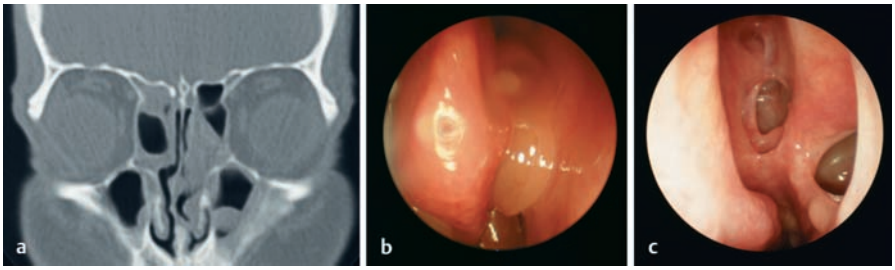


Fig. 2.52a–c Endoscopic pansinus surgery.
a The preoperative CT, showing polypoid mucosa in all of the paranasal sinuses.
b The preoperative endoscopic view, showing polyposis of the nose and paranasal sinuses.
c Endoscopic view of the cleared, spacious ethmoid, showing an open, epithelialized window to the maxillary and sphenoid sinuses (0° lens).

anation of the anterior wall. Alternatively, a transseptal or transmaxillary route can be used. For surgery of complications or tumors (see pp. 226, 288), midfacial degloving or lateral rhinotomy may be required.

Pansinusitis

Rhinogenous inflammation of all paranasal sinuses on one or (more commonly) both sides, caused by viruses, bacteria, or allergenic agents. The symptoms are those of disease of the various sinuses combined, and may be bilateral.

> **!** Note: In patients with diffuse mucosal edema or hyperplasia of all the paranasal sinuses, one should always check for acetylsalicylic acid intolerance and asthma.

Diagnosis. See Ethmoidal Sinusitis, above.

Treatment. *Conservative,* in the acute phase. Topical steroids, depending on findings; oral corticosteroids; antibiotics.

Surgery, when there is a chronic course and in severe symptoms. Ethmoidectomy with supraturbinate fenestration of the maxillary sinus, enlargement of the frontal recess, and/or removal of the floor of the frontal sinus (Draf I–III), or pansinus operation (Fig. 2.52a–c).

Chronic Rhinosinusitis

Chronic rhinosinusitis is one of the most common health problems, with a serious impact on lower airway disease and overall health. The condition is diagnosed when symptoms persist for more than 12 weeks.

Whereas acute sinusitis is an inflammatory rhinogenous process caused by obstruction of ventilation and drainage of the paranasal sinuses located functionally downstream from the ethmoid, in chronic rhinosinusitis the postulated cause involves gradual obstruction of the ostiomeatal complex by edema, induration, hyperplasia, and polyposis of the mucosa.

The role of anatomic blockage in the origin of chronic rhinosinusitis is controversial. The spectrum of pathogens involved in chronic rhinosinusitis differs from that in acute sinusitis. There has been a change in the way in which bacterial infection is assessed. In addition to biomechanical and bacterial factors, other pathomechanisms for inflammation of the nasal and sinonasal mucosa are now being considered. In the mucosa and secretions of patients with chronic rhinosinusitis, increased numbers of neutrophilic granulocytes are found, as well as eosinophils, mast cells, basophils, and mononuclear cells (T lymphocytes, $CD8^+$ suppressor cells, cytotoxic T cells, and $CD4^+$ T-helper cells). Raised concentrations of histamines and leukotrienes and prostaglandin D have been found in the nasal secretions of patients with chronic rhinosinusitis.

Chronic Hyperplastic Rhinosinusitis with Nasal Polyps

Eosinophils and fungi are considered to be of particular importance in chronic hyperplastic rhinosinusitis with nasal polyps. The granules of eosino-

philic granulocytes contain cytotoxic proteins, which are released when degranulation occurs: eosinophil cationic protein (ECP), major basis protein (MBP), eosinophil peroxidase (EPO), and eosinophil-derived neurotoxin (EDN). It has been suggested that eosinophils migrate from the epithelium into the mucus, where they dock onto T cells. When they degranulate within the supraepithelial mucus blanket, they release their cytotoxic cytokines, in particular MBP, which docks onto fungal elements to form so-called "horseshoes." This leads to a toxic–inflammatory reaction on the surface epithelium, paving the way for secondary invasion by bacteria, infection, and eventually chronic rhinosinusitis. The most common fungi involved are *Alternaria, Penicillium, Candida,* and *Aspergillus*. The term "eosinophilic fungal rhinosinusitis" has been proposed for rhinosinusitis induced by fungi.

Immune and Tissue-Specific Reactions

In contrast to the biomechanical and infectious pathomechanism involved in acute sinusitis, immune and tissue-specific inflammatory reactions play a critical role in chronic rhinosinusitis. This explains why patients and physicians face further inflammatory exacerbations and recurrences of mucosal disease, even after surgical improvement of ventilation and drainage. Surgical repair based on the principles of functional endoscopic surgery of the paranasal sinuses (the Messerklinger concept) can therefore only form part of the therapy. It is equally important to institute targeted and individualized follow-up, oriented toward the clinical findings, with long-term administration of anti-inflammatory agents (topical steroids, oral corticosteroids in appropriate cases, antibiotics for purulent inflammation; see p. 160).

Nasal Polyps

Two types of chronic rhinosinusitis can be distinguished, depending on whether neutrophilic or eosinophilic leukocytes predominate. Nasal polyps tend to occur mainly in the eosinophil-dominated type, in which they form a subgroup of chronic rhinosinusitis.

Nasal polyps appear as grape-like structures in the upper nasal cavity, originating from within the ostiomeatal complex. They are characterized by loose connective tissue, edema, inflammatory cells, and a few glands and capillaries, and are covered with various types of epithelium—mostly respiratory pseudostratified epithelium with ciliated cells and goblet cells. Eosinophils are the most common inflammatory cells in nasal polyps, but neutrophils, mast cells, plasma cells, lymphocytes, and monocytes, as well as fibroblasts, are also present. IL-5 is the predominant cytokine in nasal polyposis, reflecting activation and prolonged survival of eosinophils. The reason why polyps develop in some patients, but not in others, remains unknown. There is a definite relationship in patients who have what is known as the Sampter triad—asthma, NSAID sensitivity, and nasal polyps.

Table 2.6 Classification of polyps (adapted from Stammberger)

I	Antrochoanal polyps
II	Large single polyp
III	Polyps associated with chronic rhinosinusitis (CRS) (not eosinophil-dominated)
IV	Polyps associated with CRS (eosinophil-dominated)
	Diffuse polyposis in patients with aspirin intolerance, allergic fungal CRS, or asthma
V	Polyps associated with specific diseases (cystic fibrosis, malignancies)

In the general population, the prevalence of nasal polyps is 4%; in patients with asthma, a prevalence of 7–15% has been noted, whereas nasal polyps are found in 36–60% of patients with NSAID intolerance. Epidemiological data provide no evidence for a relationship between nasal polyps and allergy. In children, the presence of polyps should suggest the possibility of cystic fibrosis.

The clinical picture of polyps is highly variable (Table 2.6). All gradations are possible, ranging from solitary polyps to diffuse nasal and paranasal polyposis. Nasal polyps hardly ever originate within the nose itself, but rather in the anterior ethmoidal air cells, when inflamed areas of mucosa are subjected to pressure. From there, they follow gravitational pull along the path of least resistance through the middle nasal meatus to the nasal cavity (Fig. 2.53).

Nasal polyps are benign, pedicled, or sessile masses of nasal or sinus mucosa caused by inflammation.

Clinical Features. These include mechanical obstruction of nasal breathing, mechanical anosmia,

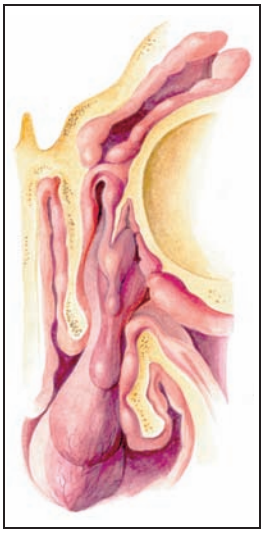


Fig. 2.53 Phases in the development of a nasal polyp from the anterior ethmoid: mucosal inflammation, edema, polyp growth, obstruction of the middle meatus and anterior ethmoid due to compression by the polyp tissue, leading to complete nasal obstruction.

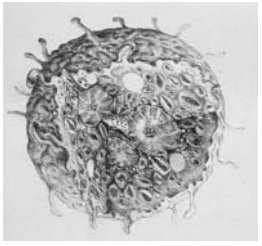


Fig. 2.54 The eosinophilic granulocyte.

epiphora, colorless stringy or purulent secretion, postnasal catarrh, headache, snoring, and rhinolalia clausa. Abnormal growth of the facial skeleton in children leads to a broad, bony nose, with an appearance known as 'frog face'. Chronic rhinosinusitis may be caused by obstruction of the sinus ostia. Polyps can also arise in one or all sinuses even if the nasal cavity is free.

Choanal polyps have a long pedicle and usually arise in the antrum. They may completely block the choana or the nasopharynx.

Pathogenesis. Immunohistochemical evidence shows that chronic rhinosinusitis with and without nasal polyps are two distinct diseases. In both types, the numbers of neutrophils, T cells, B cells, NK cells, and mast cells are increased. In polyposis, eosinophilic granulocytes, plasma cells, and macrophages are present in addition.

Biomarkers: Acute rhinosinusitis: activation of lymphocytes. Chronic rhinosinusitis: proinflammatory cytokines. Polyposis: in addition, high concentrations of nasal IgE and IL-5, derived from T cells and eosinophils and indicating a self-enhancing eosinophilic inflammatory reaction.

It has also been suggested that eosinophils migrate from the epithelium into the mucus, where they dock onto T cells. In the process of degranulation, they release cytotoxic cytokines—e. g., MBP. MBP docks onto fungal elements present in the nasal secretion, forming so-called horseshoes (Fig. 2.54). The latter cause a toxic–inflammatory reaction on the epithelial surface, which becomes infected with bacteria secondarily.

Leukotrienes: The role of leukotrienes in the pathogenesis of bronchial asthma is undisputed, and increased levels of these mediators have been detected in patients with rhinosinusitis and nasal polyps.

Patients with *ASA intolerance* are a critical group. Aspirin and other nonsteroidal anti-inflammatory drugs that inhibit the enzyme cyclooxygenase interfere with metabolism of arachidonic acid to prostaglandins. This in turn leads to relative overproduction of leukotrienes.

Staphylococcus superantigens: In nasal polyposis, but not in chronic rhinosinusitis, *Staphylococcus*-derived superantigens may at least modulate the severity and expression of disease.

United airways: The particular features of the eosinophil-dominated inflammatory reaction in nasal polyps resemble inflammation of the bronchial mucosa. Both involve an immune response in which TH2 predominates, and there is a high degree of comorbidity involving nasal polyposis and bronchial asthma or ASA intolerance. This helps explain the danger of a change in level from the upper to the lower respiratory tract in chronic hyperplastic rhinosinusitis (Fig. 2.55a–d).

Diagnosis. The typical findings on anterior rhinoscopy are solitary or multiple, glazed, transparent, smooth-walled, whitish-yellow masses, mobile to pressure with a probe, usually located in the middle meatus or the choana. They are often bilateral. Other studies include nasal endoscopy, CT, C-reactive protein, nasal cytology, and allergy tests.

C-reactive protein (CRP) is one of the acute-phase response proteins. The CRP value is useful in the diagnosis of bacterial infections. However, in patients in whom there is a suspicion of infectious disease, CRP levels of up to 100 mg/L are compatible with all types of infection (bacterial, viral, fungal, and protozoan).

Note: Polypoid sinusitis often develops in addition to nasal polyps. In patients with nasal polyps, the sinuses should therefore always be investigated by radiography and possibly by endoscopy (see pp. 132, 137).

Differential diagnosis. This includes encephalomeningocele (excluded by radiography and probing), a bleeding polyp of the septum (see p. 194), malignant nasal tumors, and a pituitary tumor—e.g., an adenoma.

Treatment. Treatment of polyposis requires a stepwise approach consisting of medical and surgical modalities. Because of the great tendency of polyps to recur, a long-term strategy involving endoscopy is necessary. The recommendations of the EAACI and European Position on Polyposis and Sinusitis (EPOS) are as follows.

Topical steroids: Owing to their strong anti-inflammatory effect, these are the first-choice drugs (e.g., mometasone, fluticasone) (Fig. 2.56a, b).

Antibiotics: Targeted antibiotic administration for inflammatory exacerbations (beware of *Staphylococcus* superantigens; empirically: amoxicillin plus clavulanic acid). Conclusive studies are still lacking for long-term treatment with antibiotics (evidence level III, level of recommendation c).

Leukotriene antagonists: These drugs have not been tested in controlled trials for rhinosinusitis and nasal polyps. A few case-controlled trials indicate that antileukotriene treatment may have a beneficial effect on nasal symptoms in patients with chronic persistent rhinosinusitis and nasal polyposis.

ASA deactivation: Adaptive deactivation by means of long-term administration of 100 mg orally has been shown to improve the chances of avoiding recurrence. A prerequisite for deactivation is in-hospital surveillance of the patient during the induction phase.

Surgery: Pretreatment with topical steroids is expedient; perioperative medication with methylprednisolone (starting at 125 mg and then gradually tapering off over 3 weeks). Functional endoscopic repair of the ethmoidal and paranasal sinuses depends on the individual endoscopic and CT findings. Procedures range from infundibulotomy to bilateral pansinus operations (shavers). An operation does not affect the cause of the mucosa's tendency toward a TH2-shifted immune response. However, it does provide a better situation for effective treatment with topical steroids. Long-term postoperative anti-inflammatory treatment with endoscopic monitoring is essential (Fig. 2.57).

Although nasal polyps shrink or disappear completely with steroids, this treatment is not appropriate, as the polyps recur when the steroids are discontinued. If an allergen can be demonstrated, it should be eliminated.

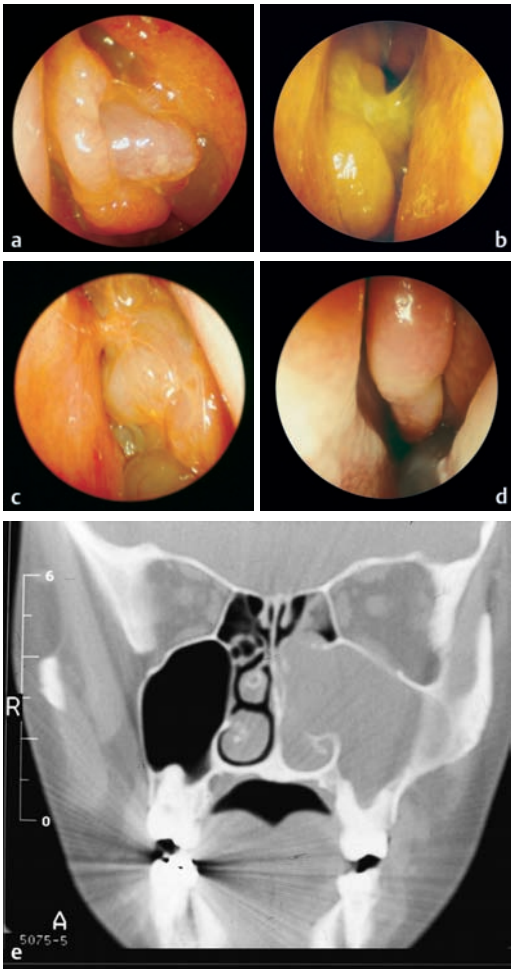


Fig. 2.55a–e Various types of nasal polyp (a–d: 0° lens).
a Chronic hyperplastic (noneosinophilic) rhinosinusitis.
b The triad of acetylsalicylic acid (ASA) intolerance, bronchial asthma, and nasal polyposis: typical color of the mucosa, rheologic changes, and thickening of the mucus.
c Induration and scarring after repeated polypectomies.
d, e An antrochoanal polyp; this is the only polyp that originates in the maxillary sinus.

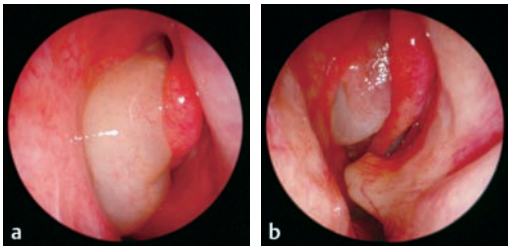


Fig. 2.56a, b The effect of topical corticosteroids on a large polyp in the middle nasal meatus, a before and b after steroid therapy.

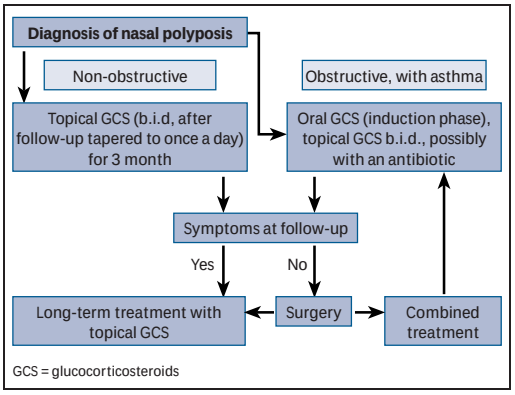


Fig. 2.57 Decision algorithm for the treatment of polyposis nasi (adapted from C. Bachert, *Allergologie* 2004;12: 484–94).

Polypectomy: So-called blind polypectomy is no longer indicated. The method of choice is functional endoscopic surgery of the ethmoid (ethmoidectomy) and the paranasal sinuses. Microdebriders have the advantage of allowing efficient and atraumatic surgery with little bleeding, preventing postoperative adhesions (Fig. 2.58a, b).

Churg–Strauss Syndrome

The Churg–Strauss syndrome is a necrotizing vasculitis classically characterized by hypereosinophilia and bronchial asthma.

Clinical Features. Otorhinolaryngology manifestations include sinusitis initially, and later asthma.

Diagnosis. Required diagnostic criteria are asthma, eosinophilia, sinusitis, pulmonary infiltrate, histologic proof of vasculitis, and mononeuritis multiplex.

Treatment. Corticosteroids alone or with cyclophosphamide and/or plasma exchange.

Prognosis. Favorable in the long term.

Rhinosinusitis and HIV

Rhinosinusitis is very common in HIV patients. Nasal obstruction may be due to rhinosinusitis, allergy, neoplasm or adenoid hypertrophy.

Clinical Features. Kaposi sarcoma or lymphoma present with nasal obstruction, epistaxis, and rhinorrhea. Adenoid hypertrophy in adults raises a concern regarding HIV.

Pathogenesis. Acute sinusitis pathogens are similar to those in normal patients. *Staphylococcus, Pseudomonas*, anaerobes, and fungi are found in chronic rhinosinusitis in HIV patients.

Diagnosis. Swabs from the middle nasal meatus, and determination of the current viral load.

Treatment. CD4 counts > 200/mm^3: amoxicillin plus clavulanic acid, systemic decongestants, guaifenesin 1200 mg b. i. d. for 3 weeks; topical decongestant spray for 1 week. CD4 counts < 200/mm^3: add coverage for *Staphylococcus, Pseudomonas,* and anaerobes—e. g., with ciprofloxacin plus clindamycin; severe infections require hospitalization, intravenous antibiotics, and endoscopic drainage; in selected patients with HIV, endoscopic sinus surgery is effective, although less so than in immunologically competent patients.

■ Rhinogenous Headache

The quality of rhinogenous headache depends on the underlying cause. Sinus inflammation is characterized by a dull, nagging, position-dependent headache that is associated with a sensation of pressure over the affected sinus (Fig. 2.59). Headache is a late symptom of tumors of the nose and paranasal sinuses. Adenoid cystic carcinoma grows

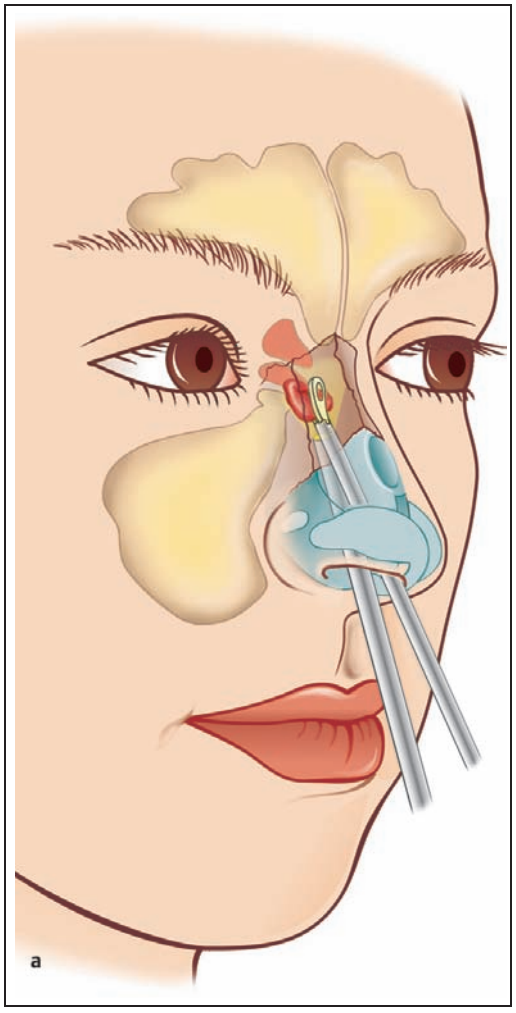


Fig. 2.58a, b a Endoscopic microsurgery with paraendoscopic preparation is now the standard surgical treatment for polyposis of the nose and paranasal sinuses.
b Microdebriders ensure smooth wound surfaces, little bleeding, and rapid healing.

along nerve fibers and is associated with pain. A neoplasm that reaches the dura mater will produce intense, unremitting pain. Mucoceles, which almost always occur in surgical or posttraumatic cavities, lead to pressure erosion of the adjacent bone. Typically, the pain subsides when the mucocele can expand by eroding through the lamina papyracea or orbital roof toward the globe.

■ Facial Neuralgias

Trigeminal nerve: Idiopathic trigeminal neuralgia is marked by paroxysms of intense, stabbing pain on one side of the face (*tic douloureux*).

Nasociliary nerve: Severe, unilateral, paroxysmal pain that is maximal at the medial canthus of the eye, epiphora with marked conjunctival injection, and edematous swelling of the ipsilateral nasal mucosa are features of nasociliary neuralgia (Charlin neuralgia).

Pterygopalatine ganglion: Unilateral, aching nocturnal pain centered in the lower half of the face ("lower-half headache"), combined with variable rhinorrhea and sneezing attacks, may be symptomatic of pterygopalatine ganglion neuralgia. It is caused by tumors and inflammations of the nose, sinuses, orbit, or pterygopalatine fossa.

Post-Caldwell–Luc syndrome: Inflammatory exacerbations in a previously operated maxillary sinus, scar traction on the infraorbital nerve, severe maxillary deformity, or scar-related infiltrates and abscesses can cause an aching or stabbing pain of variable and sometimes agonizing intensity. Anesthetic blockades may provide clues as to the nasal or sinogenic origin of the head and facial pain. If the pain is relieved by local mucosal anesthesia or conduction anesthesia of the trigeminal nerve branch and recurs after the anesthesia subsides, this confirms the origin of the pain.

■ Principles of Surgery of the Paranasal Sinuses

Endoscopic Endonasal Microsurgery of the Paranasal Sinuses and the Anterior Base of Skull

Endonasal surgery of the paranasal sinuses developed at the beginning of the 20th century. During the past 30 years, thanks to the introduction of optical systems and endoscopes in particular, it has become the international state of the art. Today, radical procedures on the paranasal sinuses (e. g., the Caldwell–Luc, Riedel, and Ritter–Jansen methods) are indicated only in selected cases or for revision surgery.

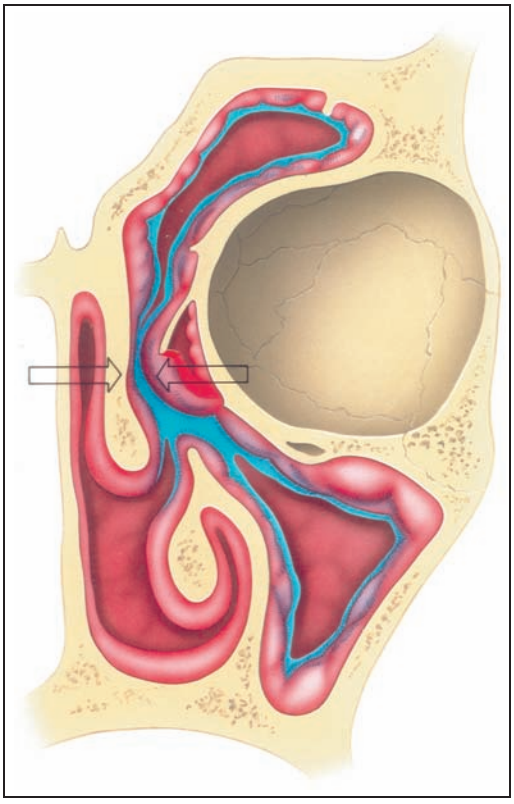


Fig. 2.59 Rhinogenic headache often starts when drainage and ventilation of the ethmoid sinus are obstructed.

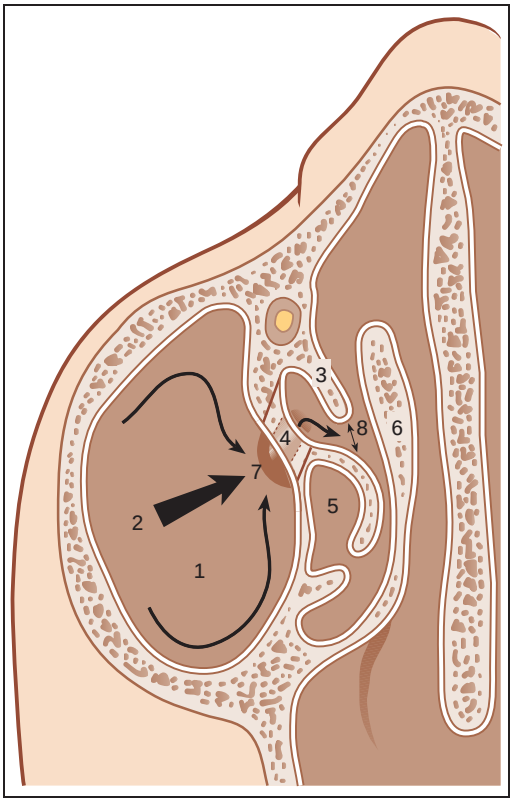


Fig. 2.60 Mucociliary transportation of secretions through the lateral nasal wall, illustrated with the maxillary sinus as an example. 1, Maxillary sinus; 2, secretion tracks in the maxillary sinus; 3, uncinate process; 4, ethmoidal infundibulum; 5, ethmoidal bulla; 6, medial concha; 7, natural ostium of the maxillary sinus; 8, semilunar hiatus.

The philosophy and details of endonasal surgery are described in numerous publications representing various schools of thought (Messerklinger, Stammberger, Wigand, Kennedy, Draf, Simmen, and others).

Concept. The concept of *functional endoscopic sinus surgery (FESS)*, the Messerklinger technique—spread throughout the world through the efforts of Stammberger and Kennedy—was broadly accepted in the 1980s and has been evaluated in numerous prospective and retrospective case-controlled studies and nonrandomized clinical trials. The original idea involved a concept of endoscopic diagnosis for the nose and lateral nasal wall; the development of an endoscopic surgical strategy was secondary. One of the essential aims of this strategy is to restore the mucociliary apparatus (Figs. 2.60, 2.61).

In the functional approach to rhinosinusitis, it was hypothesized that recovery of the diseased sinus mucosa could be achieved by allowing ventilation through the natural ostia and restoring mucociliary clearance, using a minimally invasive endoscopic technique (Fig. 2.62a–c).

Depending on the findings, endoscopic surgery can be limited to the removal of the uncinate process for wider access to the ethmoidal infundibulum (infundibulotomy), or used for anterior or complete ethmoidectomy, with or without supraturbinate fenestration of the maxillary cavity and enlargement of the access to the frontal sinus (Draf I–III), or extended into a bilateral pansinus operation.

The range of indications for endonasal microsurgery in the hands of specialized surgeons has expanded enormously

as a result of several decades' experience with endoscopic microsurgery and constant advances in endoscope and instrument technology, the incorporation of navigation systems, and completely new interdisciplinary developments (such as interventional radiology, endocrinology, and ophthalmology).

Indications. The *main indications* for endonasal microsurgery are acute (frontal sinus empyema), recurrent, and chronic sinusitis; nasal and sinus polyposis; and supportive measures (at the nasal septum, in surgery of the turbinates).

Extended indications include postsaccal tearduct stenosis, CSF leaks, decompression of the optic nerve, tumor operations (e.g., pituitary tumors, meningiomas, osteomas, and certain angiofibromas).

Mucosa-sparing microtherapy is based on the following pathophysiologic and histopathologic presuppositions:

- The transportation of secretions into and out of the paranasal sinuses proceeds along defined pathways.
- The maxillary and frontal sinuses are functionally downstream from the ethmoidal sinus.
- Chronically inflamed mucosa has a strong capacity for self-repair.
- Restoration of ventilation and drainage are the most important prerequisites for recovery of the epithelium.

Preoperative diagnostic requirements. Nasal endoscopy, coronal CT, allergy tests.

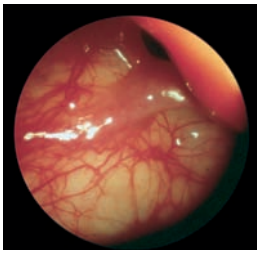


Fig. 2.61 The confluence of the secretion tracks in front of the ostium of the right-sided maxillary sinus (0° scope).

Balloon Sinuplasty and Biostatic Endoscopic Ethmoid Surgery (BESS)

Catheter-based balloon sinuplasty is a new procedure to restore ostium patency without removing tissue. For selected indications, it seems especially suited to open blocked ostia: acute recurrent sinusitis, barosinusitis, sinus empyema, primary or secondary stenosis. *Biostatic endoscopic ethmoid surgery (BESS)* is a new surgical strategy. The surgeon has to distinguish between biostatic and nonbiostatic cell formations to keep the ethmoidal space wide and open in the long term and to avoid shrinking by scarification. Both procedures can also be combined in a *hybrid operation*.

Maxillary Sinus Surgery

Infundibulotomy

The three-dimensional space of the ethmoid infundibulum, which runs in a sagittal direction through the lateral nasal wall, is approached using a crescent-shaped incision and resection of the uncinate process (Fig. 2.64). After this, the natural ostium in the caudal part of the infundibulum can be entered endoscopically and enlarged (supraturbinate fenestration). Intracavitary manipulations (e.g., removal of cysts or polyps, aspiration of secretions) can now be performed under vision using a 30°, 45°, or 70°

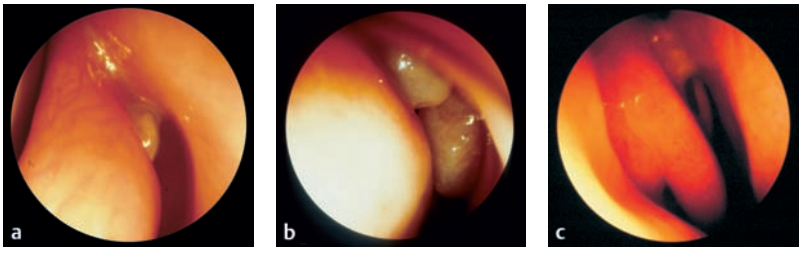


Fig. 2.62a–c With the aid of nasal endoscopy, even minimal mucosal pathology can be identified (0° scopes).
a Edema of the middle nasal meatus.
b Polyps in the ethmoidal infundibulum.
c Prolapse of the medial infundibular wall in chronic inflammation.

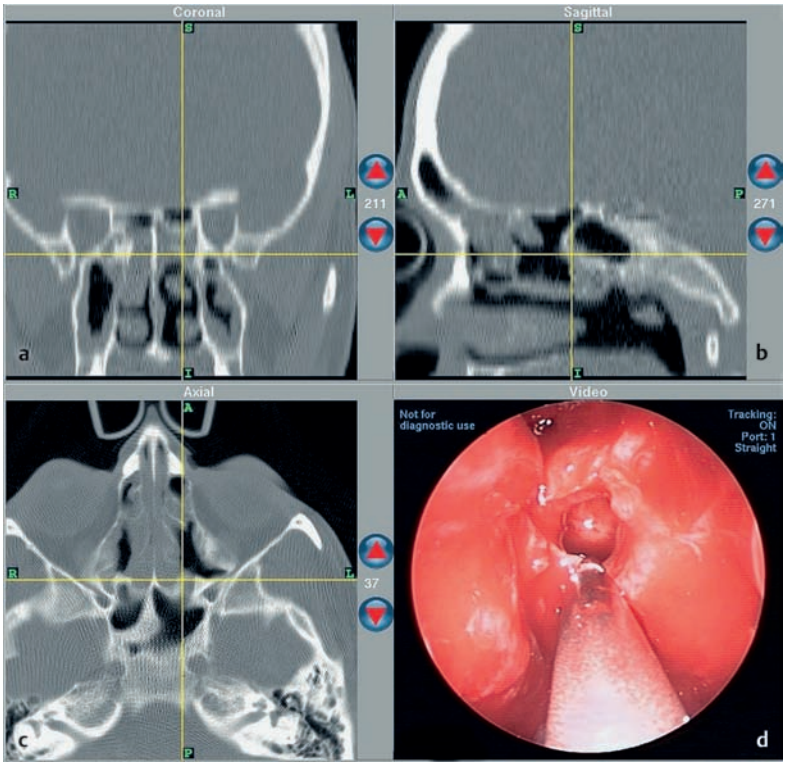


Fig. 2.63a–d Navigation-supported endoscopic surgery of the sphenoid sinus.
a Coronal view.
b Sagittal view.
c Axial view.
d Intraoperative endoscopic view of the anterior wall of the sphenoid sinus.

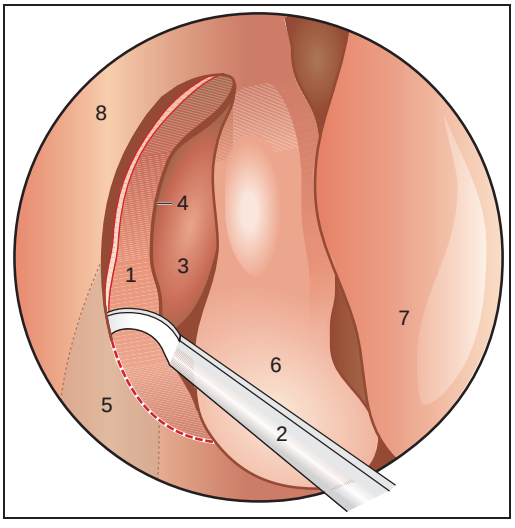


Fig. 2.64 Resection and removal of the uncinate process for access to the ethmoidal infundibulum of the nose (ethmoidal infundibulotomy). 1, Uncinate process; 2, sickle knife; 3, ethmoidal bulla; 4, semilunar hiatus; 5, ethmoidal infundibulum; 6, middle turbinate; 7, nasal septum; 8, agger nasi.

lens (Fig. 2.65a–d). However, there are still a few blind spots, so that in some cases access to the maxillary sinus cavity has to be completed using an infraturbinate trepanation or puncture of the canine fossa (Fig. 2.66).

Note: Infraturbinate fenestration for therapeutic purposes is inappropriate, as the secretions are transported past it toward the natural ostium.

Anatomical definitions: The ethmoidal infundibulum is the space enclosed by the uncinate process (medially), the lamina papyracea, and possibly the frontal process of the maxilla and lacrimal bone (laterally), and the ethmoid bulla (dorsally). The infundibulum can be accessed via the inferior semilunar hiatus.

The inferior semilunar hiatus (the gateway to the infundibulum; see Fig. 2.62) is a sagittal, crescent-shaped slit between the free posterior border of the uncinate process and the anterior surface of the bulla.

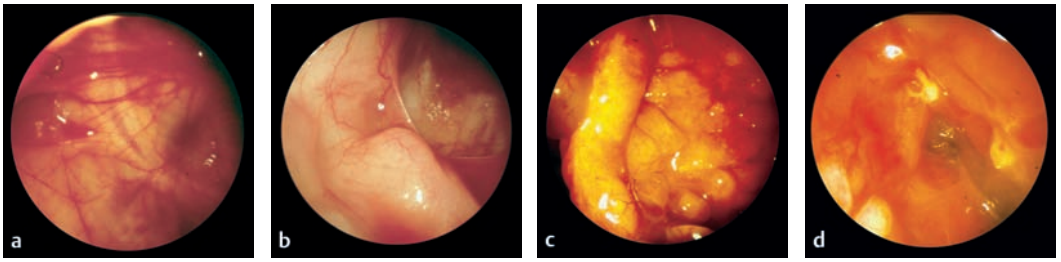


Fig. 2.65a–d Mucosal findings in the maxillary sinus (0° and 30° scopes).
a Normal mucosa in the maxillary sinus.
b Dental root in the maxillary sinus.
c What is known as a "cobblestone relief" is left to heal by itself.
d Edematous mucosa in acute inflammation, with obstruction of the natural ostium.

Caldwell–Luc Radical Antrostomy

Radical surgery is nowadays no longer a primary procedure, and it is indicated only for revision surgery (Fig. 2.67).

Local or general endotracheal anesthesia is used. Access is obtained via the oral cavity. The soft tissues of the cheek are elevated from the canine fossa, and a window is created in the anterior wall of the antrum. After the diseased antral mucosa is removed, a large window is created in the lateral nasal wall, leading from the antrum into the inferior meatus.

Intranasal Antrostomy

This is a palliative procedure for recurrent catarrhal maxillary sinusitis. Local or general anesthesia may be used. A large window is created between the inferior meatus and the antral cavity, with the physician working from the nasal cavity.

Frontal Sinus Surgery

Endonasal Methods

The types of endonasal drainage of the frontal sinuses as defined by Draf (1991) are as follows (Fig. 2.68a–c):

- Type I. Simple drainage: opening of the frontal recess by removal of the anterior ethmoid and dissection to the anterior base of the skull (Fig. 2.69a).
- Type IIa. Extended drainage: enlargement of the route of access without touching the middle turbinate (Fig. 2.69b).
- Type IIb. Expanded drainage: removal of the medial floor of the frontal sinus from the lamina papyracea to the nasal septum, including the anterior vertical lamella of the middle turbinate.
- Type III. Endonasal median drainage: removal of both the floor of the frontal sinus, as in type II,

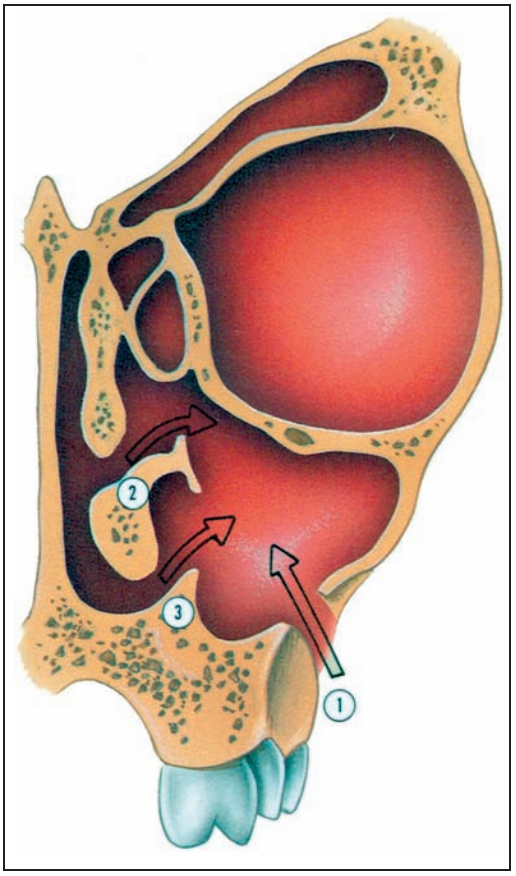


Fig. 2.66 Surgical approaches to the maxillary sinus. 1, Supraturbinal access route; 2, infraturbinal route; 3, canine fossa route.

and of the nasal septa and frontal sinus septa adjacent to the floor of the frontal sinus (Fig. 2.69c).

Table 2.7 shows the indications for endonasal frontal sinus drainage types I–III using the Draf methods.

External Methods

Bicoronal incision with osteoplastic sinusotomy. The bicoronal incision divides the skin and epicranial aponeurosis behind the frontal hairline (Fig. 2.70). The plane of dissection is supraperiosteal and suprafascial and extends past the supraorbital margins. A bone flap is elevated and fractured inferiorly, leaving it attached on a hinge of periosteum. This technique provides good exposure of all recesses in well-pneumatized frontal sinuses. The surgeon can dissect through the frontal infundibulum into the cranial part of the ethmoid bone (Fig. 2.71a, b).

Indications: Large osteomas; mucoceles in large sinus; central rhinogenous complications (brain abscess); treatment of defects, fractures, and CSF leaks involving the posterior wall of the frontal sinus; osteomyelitis (Pott puffy tumor). The bone should be burred away to a level that extends past the frontal sinus boundaries. Defect reconstruction is carried out with bioceramic glass implants made using computer-aided design/computer-aided manufacturing (CAD/CAM), or with autologous bone.

Fig. 2.67 Radical surgery of the maxillary sinus. This used to be one of the most common operations, but today it is done only when very strictly indicated, or as a revision procedure.

Contralateral, ipsilateral, and median (CIM) drainage concept for external revision surgery. Patients with a history of persistent frontal sinus disorders and often several previous operations, or of skull base fractures, often suffer from mucociliary drainage disorders as well as morphological problems in the lateral sinus (Fig. 2.72). Unfortunately, frontal sinuses that are less well pneumatized are especially susceptible to inflammatory disease, making the osteoplastic approach problematic in these cases. Typical problem areas in previously operated frontal sinuses are the lateral recess, the frontal sinus roof, the orbital roof, and the frontal infundibulum.

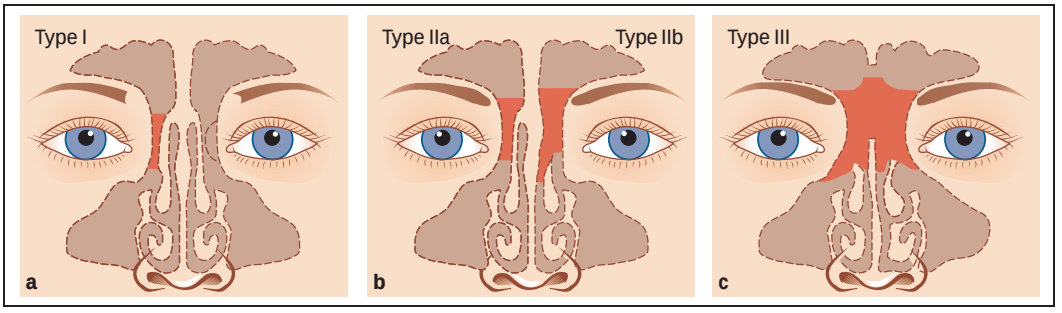


Fig. 2.68a–c Drainage pathways from the frontal sinus according to Draf.

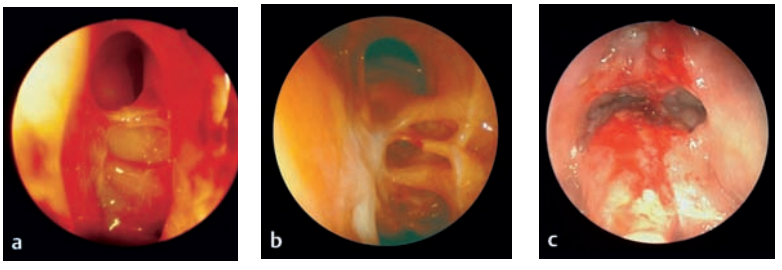


Fig. 2.69a–c Endoscopic drainage of the frontal sinus.
a Draf type I.
b Draf type II.
c Draf type III.

Table 2.7 Indications for endonasal frontal sinus drainage (Draf types I–III)

Indications for type I drainage
Acute sinusitis: failure of conservative surgery, orbital and endocranial complications
Chronic sinusitis: first-time surgery, absence of risk factors (e. g., aspirin intolerance, asthma triad), revision after incomplete ethmoidectomy
Indications for type IIa drainage
Serious complications of acute sinusitis, medial mucopyocele, tumor surgery (benign tumors), healthy mucosa
Indications for type IIb drainage
All indications for type IIa if the resulting drainage is small, when drilling is necessary
Indications for type III drainage
Primary procedure in patients with prognostic risk factors (asthma triad, cystic fibrosis), Kartagener syndrome, ciliary immotility syndrome, benign and malignant tumors

CIM is a surgical strategy that uses an open approach. Depending on the local situation, an ipsilateral, contralateral, or median drainage pathway to the nose can be reconstructed. In many cases, it is necessary to involve all of the pathways at once. It is very important to rebuild the bony defect of the sinus floor using ear cartilage. To achieve an inconspicuous scar and a good esthetic outcome, a special dermis-everting suture is used (Figs. 2.73, 2.74a, b).

Beck trephine. The principle of the *Beck trephine* is shown in Fig. 2.27a–d and described on page 140. A burr hole is made in the anterior wall of the sinus in the supraorbital region. A short blunt cannula is introduced, through which the cavity of the frontal sinus can be aspirated, irrigated, and filled with antibiotics. The cannula should not be retained for more than 8 days, due to the foreign-body response of the sinus mucosa.

The use of the Kümmel and Beck technique for frontal sinus trepanation has now declined to the point of near-obsolescence in the age of endoscopic microsurgery of the paranasal sinuses. The appropriate approach to relieve frontal sinus empyema is endoscopic surgery.

Principle of the Traditional Frontal Sinus Operations

The following surgical procedures are described here because they were the most commonly used otorhinolaryngology operations for several decades. Even today, patients present to otorhinolaryngologists with operations of this type in their medical history, or with late complications of these types of surgery. The operations are no longer indicated as primary procedures (Fig. 2.75).

1. Jansen–Ritter method. A curved incision is made near the medial canthus and extended into the eyebrow. Through a window created in the floor of the frontal sinus, the mucosa is cleared completely from the sinus. Next, a wide drainage opening is created into the nasal cavity. A permanent communication between the nose and the frontal sinus is secured using an Uffenorde mucosal flap or a large synthetic stent for at least

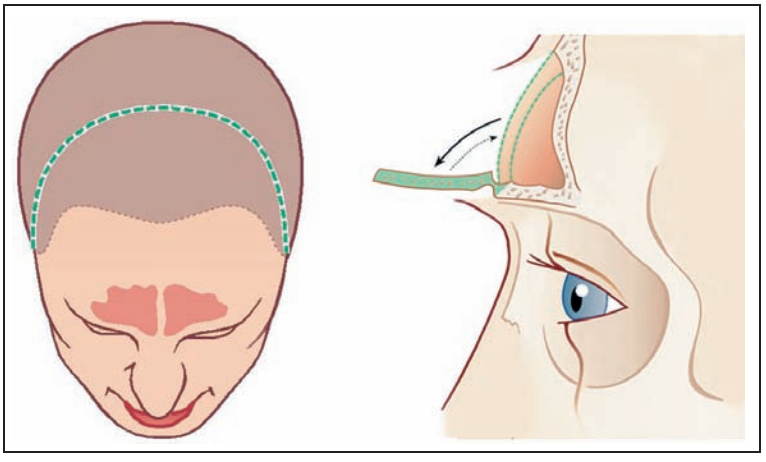


Fig. 2.70 Osteoplastic surgery in the frontal sinus.

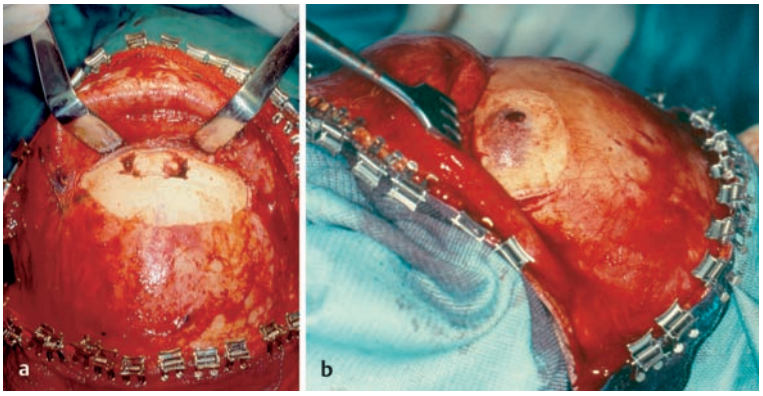


Fig. 2.71a, b Surgical approaches to the frontal sinus and frontal bone.
a For a large osteoma.
b For an intraosseous hemangioma.

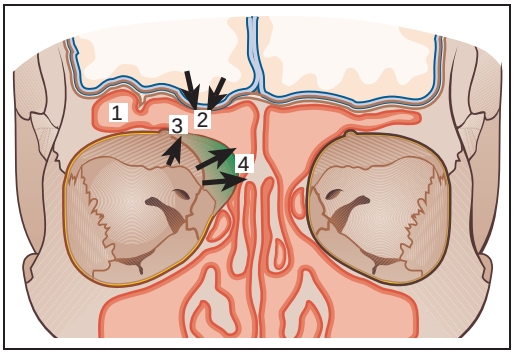


Fig. 2.72 Typical problematic regions in the frontal sinus following trauma or previous surgery. 1, Lateral recess; 2, roof of the frontal sinus; 3, floor of the frontal sinus; 4, frontal infundibulum/frontal recess.

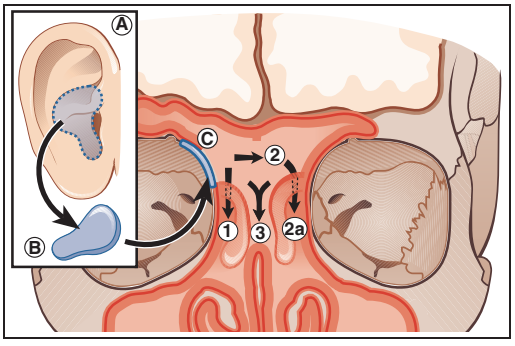


Fig. 2.73 The contralateral, ipsilateral, and median (CIM) concept of drainage and reconstruction of the frontal sinus floor (according to Behrbohm). A, Cartilage is harvested from the auricular concha; B, a suitable cartilage graft is prepared; C, the frontal sinus floor or lamina papyracea is reconstructed. 1, Ipsilateral transethmoidal route; 2, contralateral route through the interfrontal septum; 2a, adjunctive endoscopic clearance or enlargement of the frontal recess; 3, median drainage.

8 days. The advantage of this procedure is that it allows a cosmetically acceptable external incision. The disadvantage is that it can only be used for relatively small frontal sinuses. The procedure is not suitable for dealing with trauma to the anterior base of the skull.

2. Killian method. Access is obtained as in method 1 above. In addition, a window is created in the anterior wall of the frontal sinus, with retention of the bony orbital arch to preserve the profile. The operation then proceeds as in method 1. This procedure is indicated for very large frontal sinuses, as it guarantees access to all the loculi of the sinuses and thus allows removal of all parts of the mucosa. The contour of the forehead is preserved.
3. Riedel method. Access is obtained using method 1 above. The floor and anterior wall of the frontal sinus are removed, and the operation continues as in method 1. The advantage of this procedure is that it ensures clearance of the sinus and postoperative obliteration of the frontal sinus area. Its disadvantage is the resulting cosmetic deformity in the face due to sinking in of the forehead, requiring subsequent correction using a synthetic implant.

Sphenoid Sinus Surgery

The method of choice is endoscopic sphenoid sinus surgery. Depending on the indications—e.g., large, well-vascularized tumors—access may be via a transmaxillary or transseptal route, lateral rhinotomy, or midfacial degloving.

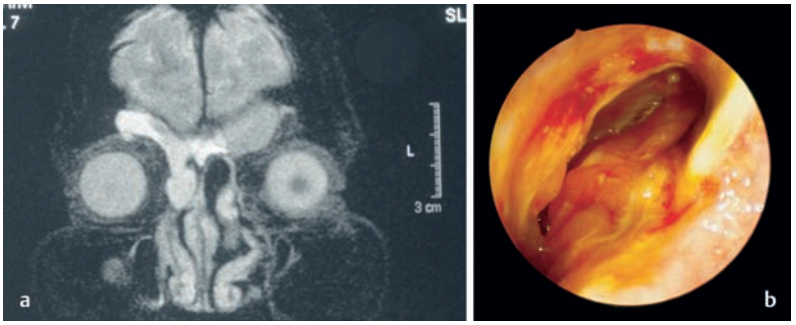


Fig. 2.74a, b a MRI showing mucoceles in both frontal sinuses following several previous operations.
b Endoscopic view of the frontal sinus after repair (45° lens).

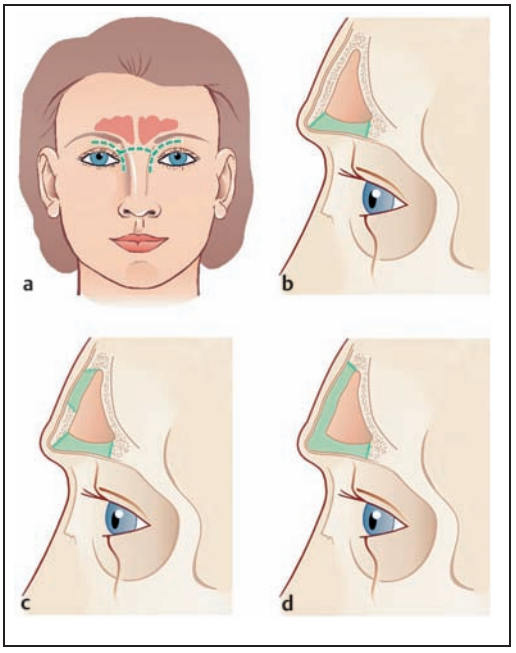


Fig. 2.75a–d The principle of external frontal sinus operations.
a Incisions.
b Bone removal with the Jansen–Ritter method.
c Killian operation.
d Riedel operation.

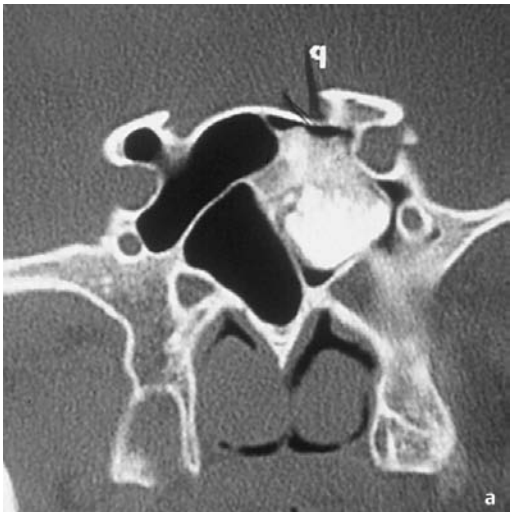


Fig. 2.76a, b a Osteoma of the left-sided sphenoid.
b Mucocele of the sphenoid sinus.

Endoscopic Sphenoid Sinus Operations

The ostium of the sphenoid sinus is identified in the sphenoethmoidal recess, ≈ 10 mm above the vertex of the choanae (nasal part) in the anterior wall of the sphenoid; or the anterior wall of the sphenoid sinus is identified and trephined in the ethmoidal part via posterior ethmoidectomy (Fig. 2.76a, b).

Fig. 2.77 View into an operated sphenoid sinus. s, Intersphenoid septum; oc, optic canal; ica, internal carotid artery (0° lens).

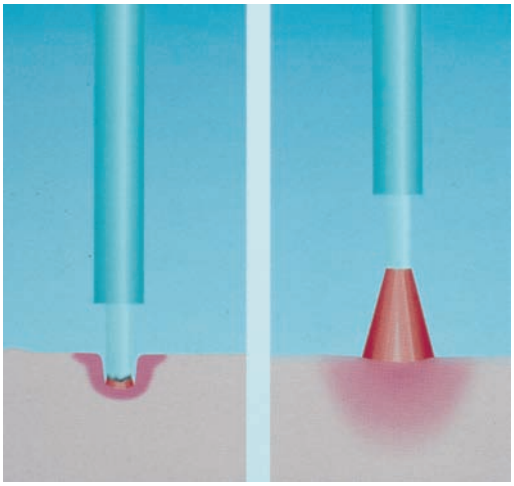


Fig. 2.78 The principle of contact and noncontact techniques in laser application.

The anterior wall of the sphenoid sinus (sphenoid bone) is thicker and harder than the septa separating the air cells of the ethmoid bone. The possibility of anatomic variations (e. g., Onodi cells) means that secure orientation is needed in this region.

Note: If the nature of a tumor is uncertain, preoperative exclusion of a vascular tumor using MRI or magnetic resonance angiography (MRA) is always required. Blind use of sharp instruments in the sphenoid sinus is strictly prohibited, due to the risk of injury to the carotid artery and the optic nerve (Fig. 2.77)

Lasers in Rhinology

The various lasers differ in the way they are generated and in their wavelengths, their specific absorptions, the depth to which they penetrate biological tissues, and thus in their range of surgical applications (Fig. 2.78).

Physical properties (Table 2.8)

- *CO_2 lasers:* The rays are absorbed in the outermost layers of tissue. These work very well for ablation and cutting, but not for coagulating.
- *Neodymium:yttrium-aluminum-garnet (Nd:YAG) and diode lasers:* The effects are practically the converse of the CO_2 laser. Depending on the power, the rays can penetrate deeper than 4 mm into tissue and are strongly diffracted. The coagulation effects are marked.
- *Holmium:yttrium-aluminum-garnet (Ho:YAG) lasers:* Generated in pulses. Coagulation and ablation are possible.
- *Potassium titanyl phosphate (KTP) lasers:* These are basically Nd:YAG lasers that work at double frequency.
- *Nd:YAG and argon lasers:* A unique feature of this type is absorption by hemoglobin, allowing special effects in well-vascularized tissue (e. g., "optical purse-string suture").

Rhinosinusitis in Children

Rhinosinusitis in childhood is anatomically, immunologically, and clinically different from that in adults.

Pathogens of acute infections. Rhinoviruses, coronaviruses, adenoviruses, myxoviruses, and respiratory syncytial viruses may be implicated. The most important pathogens for acute bacterial rhinitis in children are *Pneumococcus, Haemophilus influenzae, Moraxella catarrhalis,* and *Staphylococcus aureus.*

Anatomy. The child's skull is different from that of an adult in many respects. In a toddler, the floor of the maxillary sinus is at a higher level than in an adult, never extending below that of the floor of the nose; the alveolar process is not yet developed. Thus, the ethmoid bone is longer than the maxillary sinus. The infundibulum is short and narrow. The distance between the medial wall of the infundibulum and the lamina papyracea is only a few millimeters. The lamina papyracea is particularly thin, facilitating the spread of rhinogenous infections to the orbit. Diseases of the maxillary sinuses may occur at any age. Pneumatization of the frontal sinus does not take place until the fourth year of life, and frontal sinusitis seldom occurs before the age of 6 years. Approximately the same age data apply for the sphenoid sinus (Fig. 2.79).

Table 2.8 Characteristics and biological effects of some important laser types

Type of laser	Wavelength	Operating mode	Depth of tissue penetration (mm)	Effects	Applications
CO_2	10.6 µm	CW	≈ 0.15	Cutting, ablation coagulation (to a limited extent)	No contact
Argon	488/514 nm	CW	≈ 2.0	Cutting, coagulation	No contact
KTP	532 nm	CW	≈ 2.0	Cutting, coagulation	No contact
Ho:YAG	2.1 µm	Pulsed	≈ 0.4	Cutting, coagulation	No contact
Nd:YAG	1064 µm	CW	1.5–4.0	Cutting, coagulation	No contact, in contact
Diodes	805, 950, 980 nm	CW	1.5–4.5	Cutting, coagulation	No contact, in contact

CW, continuous wave; Ho:YAG, holmium:yttrium-aluminum-garnet; KTP, potassium titanyl phosphate; Nd:YAG, neodymium:yttrium-aluminum-garnet.

Immunology. An infant has immunity, acquired through transplacental transfer of maternal immunoglobulin G (IgG), allowing it to ward off respiratory infections. This immunity is gradually replaced by the infant's own production of IgG. The production of IgA contributes additionally to the defense against infections. With its ability to achieve rapid production of IgM, the lymphatic tissue in the nasopharynx further supports specific immune defenses against respiratory infections.

Clinical Features. In infants and toddlers, nonspecific, generalized symptoms such as fever and lymphadenitis predominate (Table 2.9). Younger children react with generalized mucosal disease of the airways. Only as the children grow do the symptoms become limited to various levels of the airways. The main symptoms of acute sinusitis include dry cough and nasal congestion and discharge. Nocturnal coughing may indicate sinusitis. Chronic sinusitis may remain concealed behind vague symptoms such as loss of appetite, malaise, low-grade fever, recurrent coughing, and pharyngitis.

Diagnosis

Note: The history usually depends on statements made by the parents, who are likely to have already interpreted the symptoms.

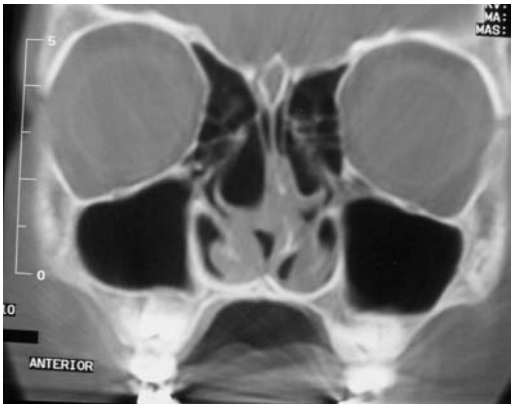


Fig. 2.79 Coronal CT in a child with bilateral concha bullosa, missing alveolar recesses, and narrow ethmoid infundibula.

The diagnosis is based on the history, inspection, and nasal endoscopy. Allergy test, nasal cytology, immune status (IgG, IgA, IgM), A-mode sonography, and nasal secretion studies (bacteriology, mycology, cytology) may also be useful investigations. If chronic rhinosinusitis is suspected, a CT examination is necessary.

Therapy. *Acute rhinosinusitis:* Supportive measures consist of decongestant nose drops, heat or shortwave application, aspiration of the nose with a simple suction apparatus, administration of an iso-

Table 2.9 Symptoms of acute and chronic rhinosinusitis in children

Acute rhinosinusitis	Acute rhinitis, purulent rhinorrhea, nasal obstruction, fever, lymphadenitis
Chronic rhinosinusitis	Coughing, mouth breathing, pharyngitis, serous otitis media

Fig. 2.80 A child with Thornwald drains in both maxillary sinuses. This treatment option is now more or less obsolete in the era of targeted antibiotic therapy and endoscopic microsurgery.

tonic salt solution as nose drops or inhaled, and secretolytic agents.

Antibiotics: The first-line agent is amoxicillin, 50 mg/kg/day, in three divided doses. Children who have been started on amoxicillin (e.g., in case of infection with *Moraxella catarrhalis* or *Haemophilus influenzae*) should be given amoxicillin plus clavulanic acid, 50 mg/12.5 mg/kg/day, in two divided doses; alternatively, clarithromycin, 15 mg/kg/day in two divided doses, may be given. Treatment should be continued for 10–14 days (Fig. 2.80).

Chronic rhinosinusitis: Supportive measures such as decongestant nose drops (beware of long-term use), secretolytic agents, adenoid removal with or without antral lavage, and possibly FESS are useful.

Antibiotics: Amoxicillin plus clavulanic acid, 50 mg/kg/day in three divided doses, or clindamycin, 40 mg/kg/day in three divided doses. Children older than 14 years of age may be given doxycycline, 5 mg/kg/day in one dose.

Mycotic infections: Amphotericin B, ketoconazole, or 5-flucytosine.

Complications. Orbital complications are more common in children than in adults, because of the thinness of the lamina papyracea (for staging, see p. 186). Intracranial complications are less common; they may consist of subdural or epidural abscesses, meningitis, brain abscesses, cavernous sinus thrombosis, or osteomyelitis (see the individual disease entities). Pathogens and antibiotics are described given on page 188 (Fig. 2.81a, b).

■ Fungal Diseases of the Paranasal Sinuses

Fungi are ubiquitous and are frequently being inhaled. Detection of fungi within the nose is therefore a normal finding in the healthy nose. However, in the pathological sense, fungal disease can be considered as:

- Noninvasive—the most common group of disorders.
- Invasive—rarely seen.

Fungi have also been implicated in the pathogenesis of eosinophilic fungal rhinosinusitis (EFRS), characterized by extensive polyps with eosinophilic mucus. The current understanding is that high numbers of eosinophils migrate to the nasal mucosa and pass into the overlying mucus, where they cluster around fungal hyphae and spores. The eosinophils try to destroy the fungi by degranulating and releasing major basic protein (MBP), which is highly toxic to the mucosal cellular surface. As more fungi are inhaled, the cycle continues and MBP increases.

Some patients also have an IgE-mediated allergy to fungi, but these patients do not always have MBP within the mucus. It has recently been noted that the IgE component is neither causative nor a major factor in the pathogenesis of this disease, and the previously accepted diagnosis of “allergic fungal sinusitis” is a misconception (Stammberger, personal communication, 2008).

Noninvasive Forms

Saprophytic Forms

Fungi can colonize the surface of mucus crusts, inspissated debris, foreign bodies, internal splints, stents and nasogastric tubes. The treatment consists of removing the mucus, crusts, or foreign body. Localized unilateral crusts may form over abnormal mucosa, and malignant change has to be excluded.

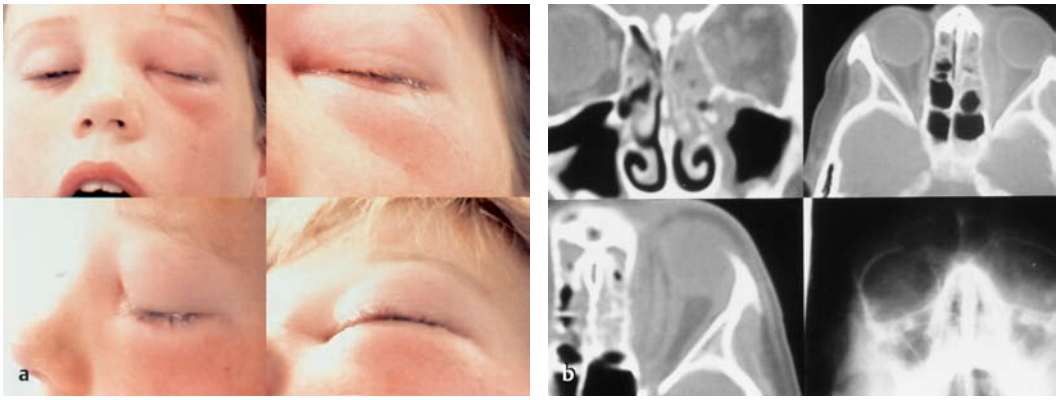


Fig. 2.81a, b a A 10-year-old boy with marked soft edema of the upper and lower eyelids. b Subperiosteal abscess, visualized on coronal and axial CTs.

Fungus Ball

Fungal masses occur within a sinus air cell or sinus lumen and act as an irritant foreign body.

Clinical Features. These resemble those of chronic sinusitis. Portions of the fungal conglomerate may reach the nose, causing bloody secretions and a fetid odor. The most frequently affected sites are the maxillary and frontal sinuses.

Diagnosis. Nasal endoscopy reveals chronic rhinosinusitis. MRI and CT show characteristic opacification. CT often shows hyperdense central areas that look calcified due to high concentrations of calcium sulfate, phosphate, and metals, which are metabolized by the fungi.

Treatment. This consists of endoscopic cleansing of the affected paranasal sinus and establishment of unobstructed ventilation. The fungal concretions must be removed completely, but mucosa can be preserved.

Samples should be sent for fungal culture and histology. Antimycotic therapy is not indicated (Fig. 2.82).

Invasive Forms

Acute Fulminant Form

Clinical Features. Acute illness with fever, abnormal fatigue, and facial pain, followed later by visual impairment and somnolence to the point of coma.

Pathogenesis. Usually occurs in patients receiving immune suppression or individuals with impaired immunity (e. g., those with diabetes mellitus, AIDS, or wasting diseases). Fungi such as *Mucor, Absidia,* or *Aspergillus* invade the tissues, including blood vessels, leading to hematogenous spread to the central nervous system or the orbit.

Treatment. Immediate, complete excision of all affected tissue, following the principles of cancer surgery. Systemic administration of amphotericin B, ketoconazole, itraconazole, or the more recent voriconazole for long periods is needed. There is recent evidence that long-term itraconazole is effective for invasive *Aspergillus* infection. The prognosis is serious.

Chronic Indolent Granulomatous Form

This is rarely seen in industrialized countries, but is more common in north Africa. In contrast to the acute form, patients are not usually immunocompromised.

Clinical Features. A slowly progressive disease over many years. Invasion beyond the paranasal sinuses may affect the orbit, with subsequent orbital swelling. The infratemporal fossa or infratemporal fossa may also be affected. Cranial nerve defects may occur.

Diagnosis. Samples should be sent for both fungal culture and histology. CT and MRI scans should be obtained.

Treatment. The diseased area should be debulked and long-term antimycotic therapy instigated. The prognosis with this regimen is good.

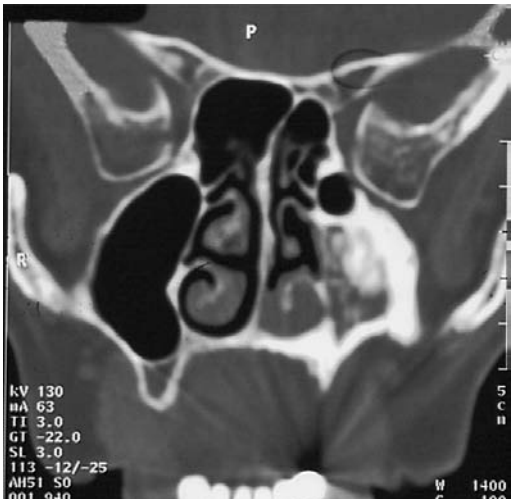


Fig. 2.82 A left-sided hypoplastic maxillary sinus, with fungal conglomeration and appositional growth of bone in reaction to the inflammation.

■ Pathophysiologic Relationship between the Sinuses and the Rest of the Body

United Airways Syndrome (Sinobronchial Syndrome)

The upper and lower airways form a functional unit. The mucosa, submucosa, and vascularization are similar in the two regions, and biochemical regulatory mechanisms involve the same mediators. The mucosa of both the upper and lower airways reacts to stimuli from allergic, physical, or chemical irritants or microbial infection with eosinophilic infiltration, mucosal edema, and endocrine malfunction.

The WHO's Allergic Rhinitis and its Impact on Asthma (ARIA) guidelines emphasize the importance of consistent treatment of allergic rhinitis (see above) in preventing allergic bronchial asthma.

> **!** Note: One child in three with allergic rhinitis develops asthma by the age of 7 years.

Cystic Fibrosis

This is the most common congenital disorder in children. The function of the exocrine glands is abnormal, causing a constant increase in chloride and sodium in the sweat and saliva, and abnormally viscous secretions. The main symptoms are pulmonary (progressive bronchial obstruction) and abdominal (due to deficiency of pancreatic enzymes), rectal prolapse, and liver cirrhosis. Nasal manifestations include sinusitis and polyps of the nose and sinuses.

Primary Ciliary Dyskinesia

Immotile cilia syndrome (ICS) is an autosomal-recessive disease with genetic heterogeneity, characterized by abnormal ciliary motion and impaired mucociliary clearance. In 1933, Kartagener described a syndrome characterized by the triad of situs inversus, chronic sinusitis, and bronchiectasis. Later, it was noted that these patients have defects in the ultrastructure of the cilia.

■ Mucoceles and Cysts

A *mucocele* is characterized clinically as a benign but locally destructive epithelial space-occupying lesion that arises within an anatomical cavity, but almost extends beyond its boundaries, compressing and eroding neighboring structures (Fig. 2.83a, b).

Whereas the classic signs of inflammation, such as redness, heat, and pain are absent with mucoceles, the clinical presentation of which depends entirely on space-occupying effects, a *pyocele* usually presents as an inflammatory condition with conspicuous symptoms such as eyelid swelling, pain, fever, and generalized manifestations (Fig. 2.84).

Mucoceles and pyoceles are caused by obstruction of sinus drainage and the resulting retention of secretions in the sinus. The frontal, ethmoid, sphenoid, and maxillary sinuses are affected, in decreasing order of incidence. The cause may be inflammation, trauma, surgery, or tumor. In the absence of spontaneous drainage, the increasing internal pressure gradually converts the bony sinus wall into a fibrous capsule extending in the direction of least resistance, such as the floor of the frontal sinus or the lamina papyracea.

Frontal sinus mucoceles: Three factors are critical in the pathogenesis of frontal sinus mucoceles: damage to the mucoperiosteum, displacement of the drainage zone, and retention of secretions. The rising intrasinus pressure leads to erosion of the bony sinus wall and formation of a fibrous capsule. The pressure may act in all directions, affecting the orbit, the anterior cranial fossa, and the ethmoid bone.

Solitary mucoceles can often be treated by *marsupialization*. This involves partial removal of the lower part of the mucocele sac, allowing the mucocele to drain to the nose while preserving the epithelial lining of the frontal sinus. This procedure is often facilitated by the fact that years of mucocele growth and expansion into the ethmoid bone will have caused widening of the frontal infundibulum. The endonasal endoscopic approach using the 45° scope is very advantageous in these cases.

Clinical Features. The main symptoms include displacement of the orbit externally or interiorly and limitation of movement of the bulb, leading to disorders of vision and double vision. Optic atrophy and amaurosis can occur in extreme cases. Mucoceles of the posterior sinuses can cause an orbital apex syndrome (see p. 187) or simulate a hypophyseal or cerebral tumor. Lesions arising in the maxillary sinus cause increasing distension of the cheek, but ocular symptoms are unusual.

Diagnosis. This is by coronal or axial computed tomography.

Differential diagnosis. Early inflammatory complications, malignancy, and meningocele must be considered.

Treatment. Clearance of the affected sinus and creation of a wide communication with the nasal cavity must be attained.

Fig. 2.83a, b A patient with extensive bilateral frontal sinus mucoceles and mild swelling at the medial canthus of the left eye.

Fig. 2.84 Pyocele of the right frontal sinus.

Fig. 2.85 A cyst in the maxillary sinus (0° scope).

Cysts

Cysts are relatively common in the paranasal sinuses, especially in the maxillary sinus. They may produce clinical symptoms or may be noted as incidental findings. Unlike a mucocele or a pyocele, a cyst does not extend beyond the anatomical boundaries of its cavity. A cyst forming a tense, convex mass impinging on the apposed bony walls of the sinus may cause a pressure sensation or headache.

Mucous retention cysts (small and yellowish, with thick walls) can be distinguished from true cysts (larger, with thin walls and fine vascular markings) by endoscopic examination (Fig. 2.85).

Cysts arising in the maxillary sinus may be *dental radicular* (arising from a tooth root) or *follicular* (arising from a displaced tooth germ). They produce typical radiologic findings.

Treatment. This is by removal of the cyst, either via the antral cavity or via the mouth; radicular cysts are removed by a maxillofacial surgeon.

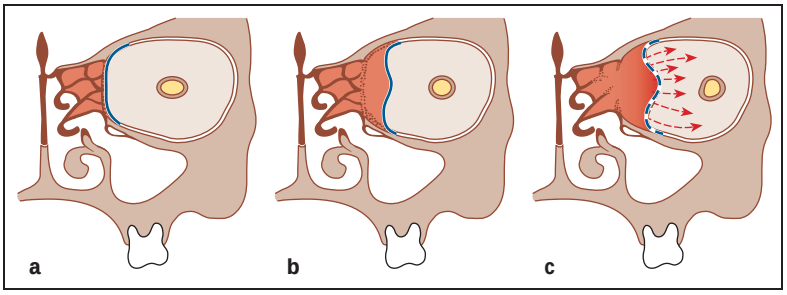


Fig. 2.86a–c The clinically most important orbital complications arising from the ethmoid: a, periorbital cellulitis; b, subperiosteal abscess; c, orbital abscess.

Complications of Sinus Infections

Extension to the External Soft Tissues

This may be due to frontal sinusitis (causing swelling of the soft tissues of the forehead or the upper eyelid), ethmoiditis (causing swelling of the eyelids, especially the lower one), or maxillary sinusitis (causing swelling of the cheek and edema of the lower eyelid). The cause is usually acute sinusitis or an acute exacerbation of chronic sinusitis. The soft tissues over the point of rupture show a boggy swelling that is usually reddened and painful on pressure.

Diagnosis. This is based on the history, clinical findings, and CT or sometimes MRI.

Treatment. Appropriate antibiotic therapy and surgical drainage of the affected sinus in suitable cases are essential.

Orbital Complications

These are relatively common and potentially very serious. They usually originate in the ethmoidal or frontal sinuses, and less frequently from the maxillary sinus. The term "periorbital cellulitis" is often used to refer to all degrees of orbital infection and is imprecise. However, a system of grouping the various stages of infection according to severity has been described (Chandler 1970) (Fig. 2.86a–c):

- Periorbital cellulitis (preseptal edema).
- Orbital cellulitis.
- Subperiosteal abscess.
- Orbital abscess or phlegmon.
- Cavernous sinus thrombosis (see p. 189).

Clinical Features. Orbital complications, especially in children, often occur without pain. Orbital involvement is initially manifested by edema and erythema of the medial aspects of the eyelid, and later by swelling, exophthalmos, and impaired extraocular eye movements. Periorbital or orbital cellulitis may result from direct or vascular spread of the sinus infection.

Periorbital Cellulitis

This is the most common complication of rhinosinusitis in children. The inflammation involves tissue anterior to the orbital septum.

Clinical Features. Orbital pain, blepharal edema, and high fever are typical; eye movement is not impaired. The sclera will be white and not red.

Diagnosis. Nasal endoscopy and/or axial CT reveal soft-tissue swelling anterior to the orbital septum.

Therapy. This should be started with empirical antibiotic therapy, which should be as targeted as possible and cover upper respiratory tract anaerobes.

Orbital Cellulitis

This implies spread of the inflammation to the orbital septum, leading to limitation of ocular movements and possibly exophthalmos.

Clinical Features. Chemosis, exophthalmos, ocular pain, and tenderness are accompanied by decreased action of the extraocular muscles.

Diagnosis. Axial CT can distinguish between an orbital and a subperiosteal abscess.

Therapy. This is based on ophthalmologic assessment of visual acuity and intravenous administration of antibiotics. Conservative therapy is appropriate if an abscess can be excluded. Supportive

measures—e.g., so-called "high tamponade" or spreading of the middle turbinate, depend on the endoscopic and CT findings.

Subperiosteal Abscess

Clinical Features. Swelling of the eyelids, pain, and lateral and inferior bulb displacement are noted. Protrusion of the bulb may be present. Depending on the extent, diplopia or limitation of eyeball movement, chemosis, or fever may occur in children. If the abscess spreads to the eyelid there is pain, redness, and firm swelling of the eyelid.

Diagnosis. This requires inspection, nasal endoscopy, consultation with an ophthalmologist, and axial CT.

Treatment. At first calculated high-dose intravenous antibiotic therapy with ampillicin and sulbactam, or clindamycin, or ceftazidim; later targeted antibiotic therapy depending on the resistogram.

Absolute indication for endoscopic surgical drainage of the abscess and sinus of origin.

Orbital Abscess or Phlegmon

An orbital abscess or phlegmon is the most serious immediate threat to the eye. The most important symptom distinguishing it from other orbital complications is the hard swelling that surrounds the bulb, visibly limiting its range of motion (Fig. 2.87).

Clinical Features. Marked edema and discoloration of the lids, chemosis, marked protrusion of the bulb, rapid loss of visual acuity, severe pain that increases when pressure is applied to the bulb or on movement of the eye, and limitation of movement of the eye due to damage to the ocular muscles and their nerves. Later features include complete paralysis of the orbit, congestion of the retinal veins, papilledema, and possibly panophthalmitis. Secondary intracranial extension to the cavernous sinus is possible.

Treatment. In addition to the treatment as for a subperiostal abscess, wide fenestration of the lamina papyracea is important to drain the abscess or phlegmon from the orbit to the nose.

Differential diagnosis. This depends on radiographic, CT, and ophthalmologic findings. Malignancy of the orbit, mucoceles, benign tumors (e.g., osteoma), inflammation of the lacrimal drainage system, cavernous sinus thrombosis, noninflammatory orbital diseases, and erysipelas must be excluded.

Fig. 2.87 Right orbital abscess with "board-like" swelling, visual impairment, and loss of eyeball mobility.

Orbital apex syndrome occurs when a lesion of the orbital apex affects the nerves and vessels that cross the optic foramen and the superior orbital fissure. The main symptoms are loss of vision, ptosis, double vision, severe temporoparietal headache, and exophthalmos due to compression of cranial nerves II, III, IV, V, and VI. Causes include trauma, neoplasms, and extension of ethmoidal or sphenoidal sinusitis. Treatment is by immediate decompression, because of the danger of amaurosis.

Rhinogenous retrobulbar optic neuritis is due to extension of inflammation of the sphenoidal air cells or posterior ethmoidal sinuses into the retrobulbar space. It is rare. Symptoms are primarily ophthalmologic. Treatment is by surgical drainage of the appropriate sinus, provided there is clear evidence of the presence of infection of that sinus.

Malignant exophthalmos (dysthyroid eye disease) due to thyrotoxicosis can be managed using a nasal operation (see p. 171). Edema of the soft tissues of the orbit, with resulting effects on the circulation and an actual increase in tissue, leads to increased intraorbital pressure and consequent injury to the bulb and the optic nerve. Decompression can be achieved by removing the bony floor of the orbit, the ethmoidal air cells, and the lamina papyracea, while preserving the infraorbital nerve.

Note: Any unilateral or bilateral loss of vision or oculomotor disorder causing double vision requires thorough rhinologic investigation.

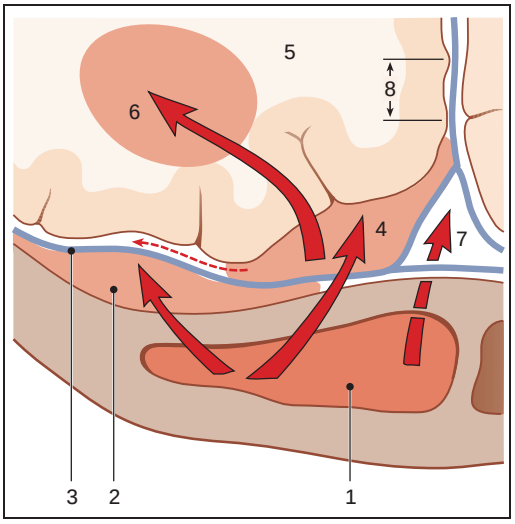


Fig. 2.88 Genesis of the intracranial complications of sinusitis. 1, Frontal sinus with empyema; 2, epidural abscess; 3, dura mater; 4, subdural abscess with marginal adhesion (8) or with extension as meningitis in the direction shown by the small dashed arrow; 5, brain; 6, abscess; 7, extension to the sagittal sinus.

Intracranial Complications

Intracranial complications include epidural or subdural abscesses, brain abscess, meningitis, cerebritis, and cavernosus sinus thrombosis. All endocranial complications begin as cerebritis, but as necrosis and liquefaction of brain tissue progress, a capsule develops, resulting in a brain abscess (Fig. 2.88).

Diagnosis. A CT scan is essential for diagnosis, as it allows accurate assessment of bony involvement. MRI is essential when there is any soft-tissue involvement—e.g., in cavernous sinus thrombosis. Abscesses and tumors are best visualized using MRI or CT scans with contrast administration. Lumbar puncture may be useful if meningitis is suspected.

Figure 2.88 shows the principal pathways of spread of rhinogenous (usually sinus) infections into the intracranial cavity. These can be explained anatomically as follows:

- Direct extension after destruction of the bone by circumscribed osteitis, and necrosis of the bony wall of the sinus, possibly due to trauma.
- Extension via osteomyelitis (see p. 190).
- Extension via blood vessels, usually the veins in the bone, which communicate between the sinus and the intracranial cavity.
- Extension via the general circulation (hematogenous metastasis).

Epidural Abscess

Clinical Features. There are no typical symptoms of this disease. Symptoms may include subfebrile temperatures, headache and pressure in the head, and fatigue. Usually there are no signs clearly indicating the local disease. The findings in cerebrospinal fluid are unremarkable. Epidural abscess is often only discovered incidentally during surgery on the sinus of origin, in which osteitis or bony necrosis—e.g., of the posterior wall of the frontal sinus—may be found.

Supplementary diagnostic procedures. MRI; CT scans with contrast medium may be useful.

Differential diagnosis. Epidural hematoma.

Treatment. This consists of drainage of the sinus of origin with wide exposure of the inflamed dura until healthy tissue is exposed on all sides, and drainage into the nasal cavity.

Subdural Abscess

Clinical Features. As with epidural abscess, there are no typical symptoms. The patient may occasionally report headache and show increasing signs of meningeal irritation—e.g., pleocytosis of the CSF. The symptoms of meningitis or brain abscess—e.g., clouding of consciousness, neurologic irritation, neurologic signs, convulsions, or hemiparesis—develop gradually. The CSF may be normal or may show inflammatory changes, and CSF pressure may be raised. Meningitis with an unfavorable prognosis may develop.

Diagnostic methods. These include CT scans of the sinuses, MRI, or cerebral CT (with contrast medium); neurologic examination; electroencephalography; and carotid angiography.

Differential diagnosis. Subdural hematoma, in which there is blood in the CSF.

Treatment. Drainage of the sinus of origin and of the subdural abscess (via the pathway of spread) must be carried out, supported by administration of high-dose antibiotics.

Rhinogenous Brain Abscess

This usually arises from the frontal sinus, and only rarely from the ethmoid air cells. Even rarer is a hematogenous *frontal lobe abscess.*

Clinical Features. Frontal lobe abscesses cause relatively few localizing symptoms. Four stages of a rhinogenous frontal lobe abscess can be distinguished: initial, latent, manifest, and terminal. Generalized symptoms, symptoms typical of increased intracranial pressure, and focal symptoms are usually pronounced. Confirmatory signs include a deteriorating overall condition, occasionally fever, increasing boring headache and sensitivity to pressure on the vault of the skull, nausea, vomiting, bradycardia, papilledema, unilateral anosmia, confusion, increasing somnolence, decreasing awareness of the surroundings, loss of concentration, overall mental torpor, altered behavior such as inappropriate joking and excessive euphoria, restlessness, coma, and cranial nerve paralysis (particularly of cranial nerves I, III, and VI).

Supplementary diagnostic methods. These include MRI or CT scans (with contrast medium) and assessment by a neurosurgeon.

Treatment. Therapy depends on the stage of the disease and the size and site of the abscess. The mainstay of treatment of abscesses arising from the nose or sinuses is combined neurosurgery and nasal surgery to excise the abscess, accompanied or followed immediately by drainage of the appropriate sinus.

Sinus Thrombosis

Cavernous Sinus Thrombosis

Clinical Features. These include edema of the upper and lower lid, oculomotor disorders, proptosis, papilledema, increasing loss of vision, possibly leading to blindness, chemosis, a septic temperature course with chills, a high continuous or remitting irregular fever, headache, variable pulse rate, and increasing clouding of consciousness. Examination of the CSF shows an increased cell count and increased protein content. The generalized symptoms of sepsis, with splenomegaly and a typical blood picture, may also be found.

Pathogenesis. Causes include propagated thrombophlebitis arising from a furuncle of the upper lip or nose and spreading via the angular vein, septal abscess, sphenoiditis with adjacent osteomyelitis, acute osteomyelitis of the frontal bone, orbital phlegmon, or petrositis with extension to the cavernous sinus.

Differential diagnosis. This covers orbital abscess, dental or tonsillar phlegmons and septicemia, otogenic sinus thrombosis, and hematogenous spread from elsewhere (i. e., of staphylococcal sepsis).

Treatment. Broad-spectrum antibiotics should be given at *high dosages* for a *prolonged period* at the earliest suspicion of thrombophlebitis, if possible selected according to culture and sensitivity tests after blood cultures. Treatment also includes bed rest, fluid balance, and the use of heparin anticoagulation. If there is a furuncle of the nose or upper lip, the angular vein should be divided with electrocautery or partially resected, depending on the local findings.

In some advanced cases, an attempt to drain the cavernous sinus surgically may be justified.

Prognosis. Very poor.

Complications. These include meningitis and septic metastases in the pulmonary and general circulation (Fig. 2.89).

Rhinogenous Meningitis

Symptoms of typical purulent meningitis. High fever, hyperesthesia, photophobia, variable pulse rate, nuchal stiffness, headache, vomiting, Jacksonian attacks, and symptoms of motor irritability (floccillation) are present. The Kernig and Lasègue sign and Brudzinski signs are all positive, and later there is opisthotonus and scaphoid abdomen.

The cranial nerves, especially III and IV, are affected. Examination of the CSF shows high-grade pleocytosis (normal: ≤ 12/3 cells/mL), markedly elevated pressure (normal: 7–12 cm H_2O), increased protein (normal: 25–40 mL/100 mL), and decreased sugar (normal: 40–90 mg/100 mL). The causative organism may also be isolated.

Fig. 2.89 Severe purulent leptomeningitis following frontal sinusitis. There are waxy, layered deposits of pus, hypervascularization, and ectasia of the cerebral blood vessels.

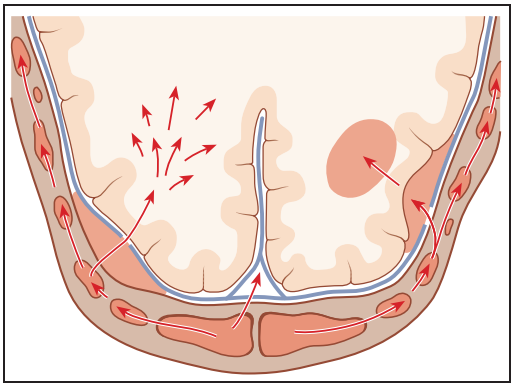


Fig. 2.90 Genesis of osteomyelitis of the flat bones of the skull. There is extension from a frontal sinus empyema to the diploë and marrow spaces of the vault, followed by an epidural or subdural abscess, sinus thrombosis (sagittal sinus), diffuse or circumscribed encephalitis, and brain abscess.

Supplementary diagnostic procedures. These include lumbar or suboccipital puncture, neurologic investigation, bacteriologic examination of the CSF, MRI, or CT (with contrast medium) to rule out a brain abscess.

Differential diagnosis. Nonrhinogenous meningitis—e.g., epidemic meningitis, viral meningitis, subarachnoid hemorrhage, and other cerebrospinal diseases—must be considered (Fig. 2.89).

Treatment. Therapy consists of immediate surgical drainage of the sinus of origin and exposure and closure of any defects found in the dura mater. High-dose antibiotics are given, and lumbar puncture is repeated frequently until the cell count falls below 100/mL. Recurrent episodes of meningitis may be due to an anterior skull base defect, which needs to be excluded or identified and repaired.

Bony Complications

Osteomyelitis of the Flat Bones of the Skull (Pott Puffy Tumor)

This life-threatening disease may spread directly from frontal sinusitis due to inflammatory disease or trauma, but may also be due to hematogenous spread (Fig. 2.90).

The course is often fulminating, because the diploë of the vault of the skull (spongiosa, medullary space, diploic veins) does not have any anatomic barriers. Infection that has broken through into the diploë thus spreads rapidly in all directions and then breaks out of this layer into the intracranial cavity, allowing the development of typical intracranial complications (see p. 188). External rupture, giving rise to a subperiosteal abscess, is also possible.

Clinical Features. Adolescence is the most susceptible age. The course may be stormy or indolent.

Main symptoms. High fever, chills, deteriorated overall condition and fatigue, severe headache, clouding of consciousness, tenderness of the frontal region and the vault of the skull, and a boggy soft-tissue swelling over the diseased part of the bone may occur. In addition, there may be clear symptoms of the development of an intracranial complication, over a longer or shorter period (see p. 188).

Diagnosis. In a patient with a painful, boggy swelling on the forehead, the physician must consider a Pott puffy tumor if the swelling exceeds the boundaries of the frontal sinus and if its peripheral borders are difficult to define. Important information is provided by CT and radioisotope bone scans, and by alkaline phosphatase as a laboratory parameter (Fig. 2.91a, b).

Differential diagnosis. Erysipelas and other septic diseases have to be excluded.

Treatment. The critical steps in the management of frontal bone osteomyelitis with a subperiosteal abscess (Pott puffy tumor) are early diagnosis and initiation of radical surgical therapy, supported by targeted antibiotic therapy with an antibiotic that can penetrate bone (e.g., fosfomycin).

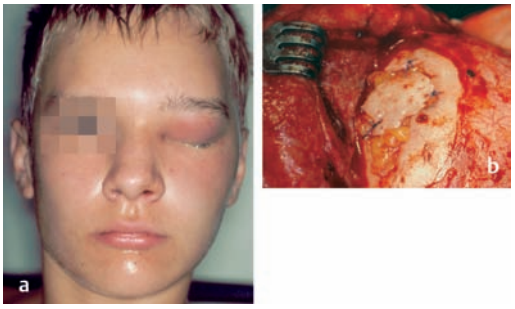


Fig. 2.91a, b a A young patient with osteomyelitis of the frontal bone. b After removal of the entire osteitic bone via an osteoplastic access route, the defect is covered with bone from the cranial vault.

The bone of the vault of the skull should be exposed widely with a healthy margin, with removal of the external table, decortication, and removal of the diploë and, occasionally, the internal table of the vault of the skull. If necessary, an already manifest intracranial complication has to be managed surgically.

Osteomyelitis of the Upper Jaw

This condition is not common. It is usually caused by dental disease; more rarely, it may be due to hematogenous spread, or may originate from the sinuses—e. g., in trauma. It may also arise from bony necrosis after infectious fevers—e. g., typhus, measles, or scarlet fever, or from radionecrosis following radiotherapy.

Clinical Features. These include swelling of the cheek, pain in the face, abscesses pointing into the mouth, the antrum, or externally.

Treatment. Removal of the (dental) cause is indicated. Antibiotics are administered, and the diseased bone is removed.

Osteomyelitis of the Upper Jaw in Infants

This condition is accompanied by acute swelling of the cheek, signs of local infection, and an abscess pointing either into the mouth or externally.

Cause. Possibly infection of the antral cavity, which is still very small in infants.

Treatment. This consists of high-dose antibiotics, and possibly careful drainage from the vestibule of the mouth, with care being taken not to damage the follicles of the teeth.

The causes of swelling of the face are shown in Table 2.10.

■ Epistaxis

Epistaxis is one of the most frequent emergencies in otolaryngology, and other specialty disciplines may be involved in the clinical management.

Although the course is usually innocuous and the cause often banal and nonspecific, epistaxis may be a serious disease that is extremely difficult to treat, with a cause that may not be remediable, and that may lead to death. Epistaxis should therefore not be regarded as harmless, either from the diagnostic or therapeutic point of view. The most common sources of bleeding are shown in Fig. 2.10. Depending on the etiology, epistaxis due to local causes is distinguished from symptomatic nosebleeds with a generalized cause. The most important etiologies are summarized in Table 2.11. Two important causes that should be considered are:

1. By far the most common source of bleeding (in ≈ 90 % of cases) is the Kiesselbach plexus (see Fig. 2.10), located on the anterior portion of the septum. The mucosa in this area is very fragile and adheres tightly to the underlying cartilage, thus offering little resistance to mechanical or functional stress.
2. Capillary hemangioma is another source of bleeding that is found occasionally on the anterior third of the septum. This type of circumscribed, dark red, benign angiomatous neoplasm is thought to result from mechanical irritation.

Diagnosis. Table 2.12 shows the necessary diagnostic steps. In some patients, it may be very difficult or even impossible to localize the source of bleeding with certainty. Bleeding from the posterior part of the nasal cavity or from the middle and superior meatus (see Fig. 2.10) is never innocuous, and requires immediate expert investigation and treatment. It may arise from the anterior or posterior ethmoidal artery or the sphenopalatine artery.

Table 2.10 Facial swelling

Site	Cause	Typical symptoms
Forehead and upper lid	Frontal sinusitis and complications	Soft-tissue edema, sensitivity to pressure, reddening
	Osteomyelitis of the frontal bone	Boggy soft tissue swelling, sensitivity to pressure on the vault of the skull, rapidly progressive edema, chemosis, swelling of the lids, swinging temperature chart. Typically, the edema is not confined to the borders of the frontal sinus
	Mucocele of the frontal sinus or the ethmoidal sinus	Resilient, nontender, noninflamed swelling that grows slowly
	Encephalocele	Noninflammatory, pulsating swelling at the root of the nose
	Tumors	Noninflammatory, firm, usually immobile swelling; maybe edema and pain on pressure
	Trauma	Edema and often hematoma of the upper lids, signs of fracture, pain on pressure
Upper jaw, cheek, and lower lid	Maxillary and ethmoidal sinusitis	Soft tissue edema of the cheek, lower lid, and medial canthus; pain on pressure; reddening
	Mucocele of the antrum or ethmoids	Resilient, noninflammatory swelling of the cheek or the medial canthus, not tender and not inflamed
	Osteomyelitis of the upper jaw due to sinusitis, dental causes, or radiotherapy	Painful inflammatory swelling and redness
	Zygomatic abscess due to otitis media	Painful inflammatory swelling of the temple and zygoma with edema of the lid and ear symptoms
	Parotitis	Inflammatory swelling of the preauricular area and cheek, severe pain, and tenderness
	Sialadenosis of the parotid gland	Painless, occasionally fluctuating, marked preauricular swelling
	Parotid tumors	Benign: slow-growing, no pain, no facial nerve paralysis Malignant: rapid growth, pain, and possibly facial paralysis
	Inflammatory dental disease	Painful swelling of the cheek, edema of the lids, and possibly trismus, disappearance of the mucosal reflection between alveolar process and cheek
	Odontogenic cysts	Painless swelling of the cheek and slow growth
	Dacryocystitis	Very sensitive to pressure; inflammatory swelling, possibly fluctuation in the medial canthus, drainage of pus from the lacrimal duct
	Tumors	Firm, usually poorly mobile swelling, usually not very tender
	Trauma	Edema, often hematoma of the lower lids, possibly signs of fracture, sensitivity to pressure
	Angioedema	Allergic edema with a sensation of tension and pruritus, facial swelling, particularly of the lip and the lid, sudden onset and tendency to recur

Table 2.10 Facial swelling (continued)

Site	Cause	Typical symptoms
Mandible	Osteomyelitis due to dental diseases or radiation	Inflammatory painful swelling, possibly with trismus
	Inflammatory dental disease	Painful swelling and possibly trismus
	Odontogenic cysts	Painless, slow-growing swelling
	Masseteric hypertrophy	Painless thickening of the masseter muscle on mastication
	Sialadenitis of the submandibular gland	Inflammatory swelling in the submandibular region
	Sialadenosis of a major salivary gland	Painless swelling, slow growth
	Tumors	Firm, usually poorly mobile swelling
	Trauma	Swelling, often a hematoma on the injured area and possibly signs of a fracture

Diffuse facial swellings may also occur in nephrosis, chronic nephritis, endocrine disorders (e. g., myxedema), and Cushing disease

Differential diagnosis. This includes bleeding that does not arise in the nose, but in which the blood escapes through the nose—e. g., due to tumors of the nasopharynx or the larynx, hemoptysis, bleeding esophageal varices, and escape of blood due to injury to the vessels around the base of the skull (e. g., the internal carotid artery) via the sphenoid sinus or the eustachian tube.

Treatment of Epistaxis (Table 2.13)

Supportive measures in treatment of severe nosebleeds

- Maintain a calm atmosphere.
- The patient should sit with the upper part of the body tilted forward and the mouth open so that blood can be spit out rather than swallowed.
- Cold compresses are applied to the nape of the neck and to the dorsum of the nose.
- Mild pressure is applied to both nasal alae for several minutes.

Local Procedures

Coagulation and chemical cauterization of the Kiesselbach plexus. Several methods can be used to stop bleeding that has been localized to the Kiesselbach plexus. In everyday practice, bipolar coagulation after local anesthesia has become the favored method for obliterating a bleeding vessel. Traditionally, chemical cauterization with silver nitrate ($AgNO_3$) or chromic acid beads was widely used; application of either of these chemicals is still an acceptable alternative.

Lasers offer another modality for managing recurrent epistaxis, but they are not suitable for treating acute epistaxis. Their method of action is determined according to the wavelength: argon and Nd:YAG lasers are absorbed selectively by hemoglobin, resulting in a photothermolytic effect that makes them suitable for the so-called "optical purse-string sutures" used to treat recurrent hemorrhage in areas of the skin or mucosa—e. g., in Rendu–Osler–Weber disease. Other lasers (CO_2 lasers, diode lasers) produce coagulation.

Anterior nasal packing and balloons. *Technique:* First, topical anesthesia of the nasal mucosa is induced (see p. 195). Strips of ointment-saturated gauze, 2–4 cm in width, are introduced into the nasal cavity in layers from above downward, or from behind forward. The pack should produce sufficient pressure to compress the bleeding source. Pneumatic packing with an inflatable balloon can be used as an alternative.

The following procedures may be required to achieve hemostasis if anterior packing fails to stop profuse hemorrhage—e. g., following trauma or vascular rupture in hypertension, or if the source of bleeding is concealed and lies far posteriorly.

Table 2.11 Epistaxis

Local causes of epistaxis	
Idiopathic	This refers to mild, recurrent nosebleeds in children and adolescents
Vascular	Microtrauma to Kiesselbach plexus
Anterior rhinitis sicca	Due to chemical or thermal injury to the nasal mucosa or to septal perforation, this condition frequently involves minor bleeding or blood-stained nasal discharge, accompanied by a feeling of dryness or crusting in the nose
Environmental influences	E.g., living at high altitudes, reduced air pressure, or drying due to air-conditioning may be implicated
Trauma	E.g., fractures of the nasal bone or the nasal septum, injuries to the facial skeleton or the anterior base of the skull. There is usually severe, profuse hemorrhage as a direct result of the trauma. Injury to the internal carotid artery may pose an immediately life-threatening problem, or there may be an interval before the formation of an aneurysm and attacks of bleeding
Foreign body in the nose, or rhinolith	This may cause unilateral mild bleeding and fetor, and purulent secretion in the long term
Bleeding polyp of the septum	This refers to a histologically telangiectatic granuloma or hemangioma, with a marked tendency to bleed when it is disturbed
Tumors	Malignant tumors of the nose or sinuses in particular often cause blood-tinged secretions only Lesions in the nasopharynx, especially nasopharyngeal angiofibroma, may cause massive life-threatening hemorrhage
Causes of secondary epistaxis	
Infection	Acute contagious diseases such as influenza, measles, typhus, and catarrh, may be involved. The epistaxis usually involves minor, brief bleeding episodes, principally in children and adolescents
Vascular and circulatory diseases	E.g., arteriosclerosis and hypertension. The bleeding is arterial, often pulsating and spurting. It affects middle-aged and elderly individuals and tends to recur
Blood diseases and coagulation diseases	Thrombopathy, e. g., in thrombocytopenic purpura or idiopathic thrombocytopenic purpura (Werlhof disease), sickle-cell anemia, leukemia, thromboasthenia (Glanzmann disease), and von Willebrand–Jürgens constitutional thrombopathy, or myeloproliferative diseases—e. g., essential thrombocythemia
Coagulopathy	E.g., hemophilia, Waldenström disease, prothrombin deficiency or overdosage with anticoagulants, fibrinogen deficiency, and deficiency of vitamins K and C
Vasculopathy	E.g., scurvy, infantile scurvy (Möller–Barlow disease), and Henoch–Schönlein purpura. The bleeding in this group is usually a superficial trickle of relatively dark blood
Uremia and liver cell failure	
Endocrine causes	E.g., vicarious menstruation due to endometriosis, epistaxis during pregnancy, and pheochromocytoma, which causes hypertensive crises due to circulating catecholamines
Hereditary hemorrhagic telangiectasia with typical mucosal lesions (Osler–Rendu–Weber disease)	This causes recurrent, mild to moderate, treatment-resistant, and often multifocal bleeding, principally in the anterior and posterior part of the septum

Gauze or a swab soaked with an ointment containing panthenol, an antibiotic, tetracaine with epinephrine or thrombin solution are used to stanch nosebleeds. In the United Kingdom, bismuth–iodoform–paraffin packs are widely used. An alternative is a tamponade consisting of a fingerstall filled with polyvinyl alcohol, which is a compressed foam polymer (Merocel). Hygroscopic tamponades containing oxidized cellulose (Oxycel), cellulose, or synthetic material—e. g., hydrocolloid fabric (Rapid Rhino)—that expand when moistened can also be applied. Other options are gelatin sponges, hyaluronic acid packings, or packings containing hemostyptic additives—e. g., thrombin. Fibrin glue can also be used. In the inflatable balloon designed by Masing for tamponing the nasal cavity and the nasopharynx, the pressure is fully controllable. Other products that work on a similar principle include epistaxis catheters (e. g., Epi-Max), choanal balloon catheters, epistaxis balloon catheters, and Foley catheters.

Table 2.12 Epistaxis: diagnostic steps

1	History
2	Localization of the source of bleeding and determination of its cause *Anterior bleeding:* nose-picking, idiopathic epistaxis, rhinitis anterior, infectious diseases *Posterior or middle bleeding:* hypertension, arteriosclerosis, fractures, tumors *Superficial bleeding:* hemorrhagic diatheses, coagulation disorders, Osler–Rendu–Weber disease
3	Measurement of blood pressure and assessment of the circulation
4	Analysis of blood coagulation
In appropriate cases:	
5	Computed tomography of the nose and paranasal sinuses (e. g., if a tumor is suspected)
6	Examination by a general physician to exclude generalized causes (see Table 2.8)

Posterior nasal packing. This procedure is very painful and requires, if possible, general anesthesia and intubation, or at least very good topical anesthesia.

Principle: A gauze or sponge pack attached to a string is used to close off the choana and is fixed in the nasopharynx to prevent the blood escaping from the nose into the nasopharynx. Anterior packing is then performed. The original packing method described by Bellocq was quite stressful for the patient and required skill on the part of the surgeon, as well as satisfactory anesthesia (Fig. 2.92). The posterior part of the nasal cavity can be closed off from the nasopharynx more easily and efficiently using a catheter with an inflatable cuff.

Technique: A catheter with an inflatable cuff is passed into the nasopharynx through the side of the nose that is bleeding, with the patient under general anesthesia or, if necessary, with topical anesthesia. The cuff is then filled with water until the nasopharynx is occluded securely and the flow of blood posteriorly has been stopped. The anterior part of the nose then is packed, and the end of the catheter emerging from the nose is fixed.

An inflatable catheter of this type should always be at hand in every hospital and otorhinolaryngology practice.

A severe septal deviation or prominent spur may necessitate surgical correction of the septum.

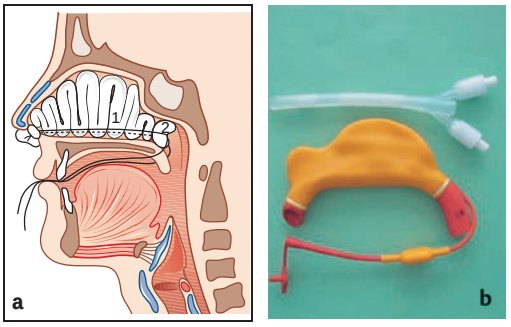


Fig. 2.92a, b a Nasal packing. 1, Anterior nasal packing in vertical layers. 2, Completion of the packing (Bellocq tamponade).
b Two inflatable nasal balloons for tamponading the nasal cavity and nasopharynx.

Complications: Although balloon tamponades are effective for hemostasis, they may—depending on the cuff pressure—occlude circulation, leading to mucosal necrosis and consequent infection, ulcerations, and finally scars and adhesions.

Table 2.13 Epistaxis: treatment

Supportive measures
Calm patient (if necessary with medication)
Have the patient sit upright
Apply cold to the nape of the neck
Lower the blood pressure if hypertension is present
Administer fluid and plasma expanders
Blood transfusions are needed if hemoglobin (hematocrit) falls below 50 %
Do coagulation studies and check levels if the patient is on anticoagulants
Local measures
Occlusion of the bleeding source by:
Local application of hemostatic substances, e. g., thrombin, gelatin tampons, oxycellulose, or fibrin
Injection of vasoconstricting agents into the bleeding area
Cautery, galvanocautery, or laser applied to a small bleeding point
Endoscopic ligation/coagulation of the sphenopalatine artery and the anterior and posterior ethmoidal artery is now commonly performed and is the surgical procedure of first choice
Anterior packing or inflatable tampons
Posterior packing (balloon catheter)
Selective embolism during angiography or digital subtraction angiography; alternatively, vascular ligation may be performed
Depending on the source of the bleeding, and provided the bleeding cannot be arrested by any other means, the following blood vessels may be divided:
Internal maxillary artery
Anterior and posterior ethmoidal arteries
External carotid artery
Replacement therapy in bleeding disorders:
Fresh blood transfusions, vitamin C, and systemic hemostatic agents in thrombopathies
Coagulation-promoting plasma, transfusion of fresh blood, Cohn fraction, in appropriate cases administration of vitamin K for coagulopathy or ACTH, steroids, calcium, vitamin C, and estrogens for vasculopathy.
For Osler–Rendu–Weber disease: “optical purse-string suture” using photocoagulation with an argon or Nd:YAG laser
Septal dermoplasty (Saunder plasty)
Another effective means of controlling recurrent bleeding in this condition is closure of the nasal airway using the Young procedure

ACTH, adrenocorticotropic hormone; Nd:YAG, neodymium: yttrium-aluminum-garnet.

A catheter left in the nose or a string attached to a postnasal pack must not exert pressure on the nasal alae or the columella. Necrosis can occur rapidly, causing scars at the tip of the nose and in the anterior part of the nasal cavity. A posterior nasal pack should not be left in for longer than necessary, and never for more than 5–7 days. Antibiotic coverage should be given while the pack is in place, as there is a risk of sinusitis or otitis media due to blockage of the pharyngeal ostium of the eustachian tube if the pack is in the correct position.

> **!** Note: All nasal packing must be visibly secured to prevent aspiration. Caution must be exercised in the use of ointments and oils, as disfiguring lipogranulomas or paraffinomas that require surgical removal may develop even years later.

Endoscopic location and control of bleeding. This is done either under local anesthesia or in theater under general anesthesia, combined when necessary with septal surgery and occasionally ethmoid dissection.

Endoscopic ethmoidal and sphenopalatine artery occlusion can be carried out using bipolar diathermy.

Embolization. Interventional radiologic embolization is indicated for recurrent bleeding that cannot be stanched using conservative methods. The most recent method for obliterating an actively or recurrently hemorrhaging artery is selective embolization during digital subtraction angiography. Selective embolization can also be achieved with less technical apparatus using conventional angiography. Examples of target vessels are the maxillary and facial arteries.

Vascular ligation. This procedure has to be performed in cases of uncontrolled, life-threatening epistaxis if the methods described above fail. Depending on the source of the bleeding, it may be necessary to ligate the internal maxillary artery in the pterygopalatine fossa, the anterior and posterior ethmoidal arteries (Fig. 2.93), or the external carotid artery at the anterior border of the sternocleidomastoid muscle above the origin of the lingual artery.

However, it should be noted that these techniques were developed at a time when endoscopic location and cautery and endoscopic sphenopalatine artery occlusion were not yet possible.

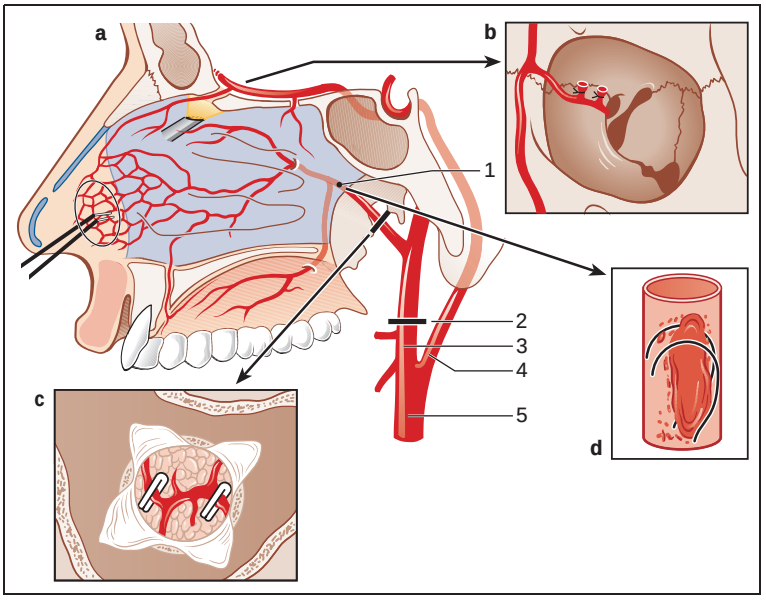


Fig. 2.93a–d Sites for ligation in severe epistaxis.
a Overview. 1, Internal maxillary artery; 2, ligation of the external carotid artery; 3, external carotid artery; 4, internal carotid artery; 5, common carotid artery.
b Ligation or embolization of the ethmoidal arteries.
c Ligation or embolization of the internal maxillary artery or sphenopalatine artery in the pterygopalatine fossa.
d Embolization with a coil.

Hereditary telangiectasia (Osler–Rendu–Weber disease) (Fig. 2.94). Homozygotic carriers of this trait suffer from recurrent hemorrhaging even during childhood, in part precipitated by transfusions. Trauma to the mucosa must be avoided as much as possible, and the mucosa should be cared for with oils and isotonic solutions or loose, mildly antibiotic packing.

A modern method of symptomatic treatment for hereditary telangiectasia is known as the "optical purse-string suture." A noncontact technique is used to irradiate the submucous venous plexus in a given area of skin or mucosa using an argon or Nd:YAG laser, until a delicate, whitish discoloration—not coagulation—of the spot appears. At first, there is a marked tissue reaction, with some renewed bleeding. As scar formation and obliteration proceed, the incidence and intensity of the bleeding decreases.

In Saunders dermoplasty, the mucosa of the anterior septum is removed and replaced with split skin from a distant site.

Complete occlusion of a nasal cavity using flaps in the region of the internal nasal valve (Young procedure) has been used effectively in patients with severe, frequent bleeding in whom the above techniques have not been successful.

Fig. 2.94 A patient with Rendu–Osler–Weber disease. Hereditary telangiectasia of the lips and nasal mucosa.

Diseases of the Septum

Few people have a perfectly straight, perpendicular nasal septum. The septum usually shows slight bends and spurs. Provided these do not obstruct nasal respiration, they should not be regarded as abnormal.

Deviation of the Nasal Septum

This may be *developmental,* due to unequal growth of the cartilage and bone of the nasal septum; or it may be *traumatic,* due to facial fracture, fracture of the nose or septum, or possibly due to injury at birth. The parts of the septum may either be too large for the available skeletal space, or may have

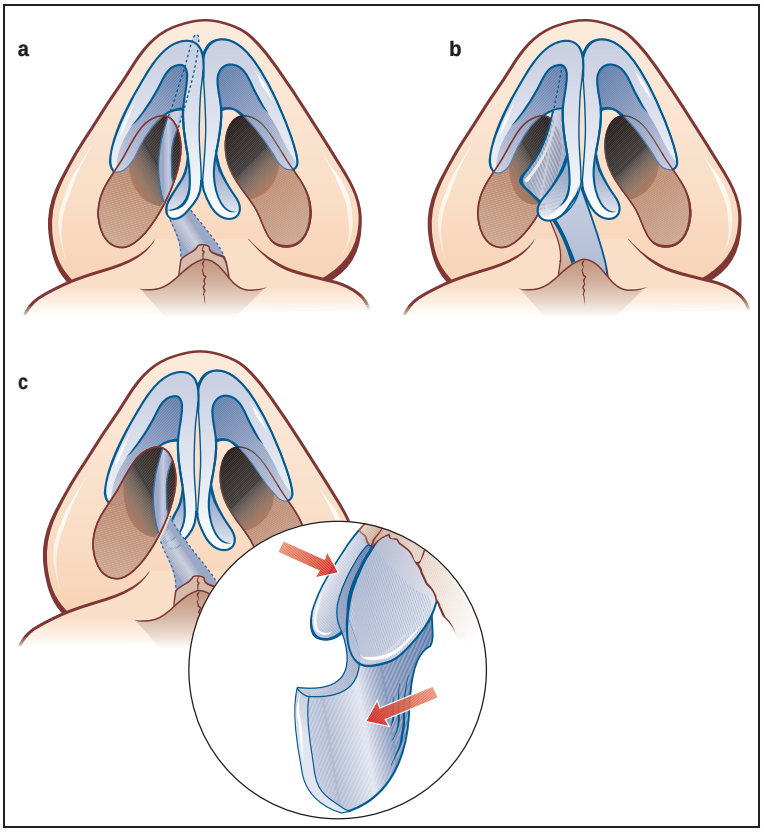


Fig. 2.95a–c The most common types of septal deviation. a The dorsobasal extension of the septum is too long. b Vomeral crest. c The craniocaudal extension of the septum is too long. *Inset:* A septal deviation causes stenosis on both sides of the septum.

dislocated and healed in an incorrect position. These situations lead to deviations, spurs, and crests that reduce the patency of the nasal cavity (Fig. 2.95)

Clinical Features. These include nasal obstruction, which is often unilateral and may be intermittent, hyposmia or anosmia, and headaches that may vary depending on the condition of the nasal cavity. Subluxation of the septum—i. e., displacement of the ventral edge of the septum and obstruction of the nasal introitus to one side and septal deviation with obstruction of the nasal cavity on the other side—is especially likely after trauma. This combination of factors can lead to complete bilateral obstruction of the nasal cavity

Tension septum: The septum is too large for the available space, subjecting it to tension and constricting the nasal valve to less than 15°. The nares are often unrounded and slit-like, potentially causing nasal obstruction and headaches.

Diagnosis. The diagnosis is based on rhinoscopy, nasal endoscopy, and computed rhinomanometry.

Treatment. *Surgery:* Figure 2.96 illustrates the principles involved in septoplasty. It is possible to remove all parts of the septum and reimplant them if necessary after shifting and remodeling. However, the function of the nasal cavity may often only be restored satisfactorily with simultaneous correction of the external nasal pyramid (see the section on septorhinoplasty, p. 212).

Principles of submucous septoplasty: Maurice H. Cottle (1896–1981) introduced his cartilage-preserving operation as an alternative to the Killian septal resection. The Cottle operation strives to preserve the supporting function of the septal cartilage and the physiological function of the nasal mucosa. After the margin of the septum has been exposed, the mucoperichondrium is undermined to create a superior tunnel on the left side and an inferior tunnel on both sides. The mucosa remains adherent to the septal cartilage on the right side.

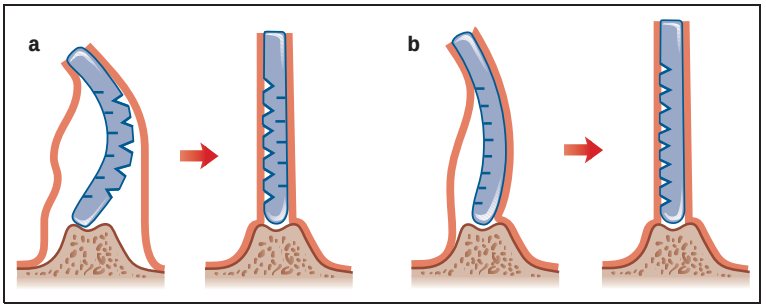


Fig. 2.96a, b The aim in modern submucosal septal surgery is to preserve as much cartilage as possible, using incisions and excisions to trim it into an optimum shape. Tunneling under the mucoperichondrium is carried out to allow work on the cartilage.

The classic Cottle operation has been continually modified over the years. One modification involves the "swinging door" technique, in which the septal cartilage is detached just anterior to the perpendicular plate to increase the mobility of the cartilage during the operation. The anterior septal cartilage can be trimmed using incomplete and complete incisions until it can be positioned in the midline without tension (Figs. 2.97, 2.98)

Complications of septoplasty include septal perforations (see below). If too much cartilage is resected, the cartilaginous part of the nose may collapse, causing an anterior or inferior saddle, or duckbill nose (see p. 213). In both cases, reconstructive procedures are available and indicated. Patients should also be warned about possible loss of sensation in the incisor teeth.

Fig. 2.97 Exposure of the border of the anterior septum via a hemi-transfixion incision.

The Pediatric Nasal Septum

The septum, maxilla, and premaxilla develop independently of each other. Strict criteria have to be applied in selecting children for septal operations. Nevertheless, even small children can successfully undergo surgery for traumatic deformities or malformations of the septum that cause significant nasal obstruction. Deformities of the anteroinferior septum are the most common problem. It is important that the surgery should preserve the perichondrium, the growth zones (e.g., the caudal septum), the premaxilla, and the sutural junctions with the perpendicular plate and vomer. The aim in pediatric septal operations should be a chondroplastic mode of surgery.

Septal Hematoma and Septal Abscess

These are usually caused by trauma, and usually occur in children. Blunt nasal trauma with a shearing effect leads to elevation of the mucoperiosteum from its cartilaginous or bony attachment. A hematoma forms in the newly created perichondrial—periosteal space, and it is often bilateral. Infection leads to a septal abscess.

Clinical Features. These include increasing nasal obstruction, tenderness, and pain. If an abscess forms, local pain increases, and the patient complains of headaches, fever, pain on pressure, and reddening of the bridge of the nose. In the long term, the cartilaginous part of the nose undergoes necrosis and sinks in.

Diagnosis. The history reveals an incident of trauma. Examination shows that the hematoma is usually located to the most anterior part of the septum. There is also evidence of swelling of the septum, which occludes the lumen of both nostrils and feels cystic when palpated with a probe.

Treatment. Wide incision and drainage of the hematoma is performed, followed by splinting (with Doyle splints) of the nostrils to encourage the mucoperichondrium and mucoperiosteum to re-adhere. If infection is found, the incision must be held open with a drain to prevent cartilaginous necrosis, which would lead to deformity of the ex-

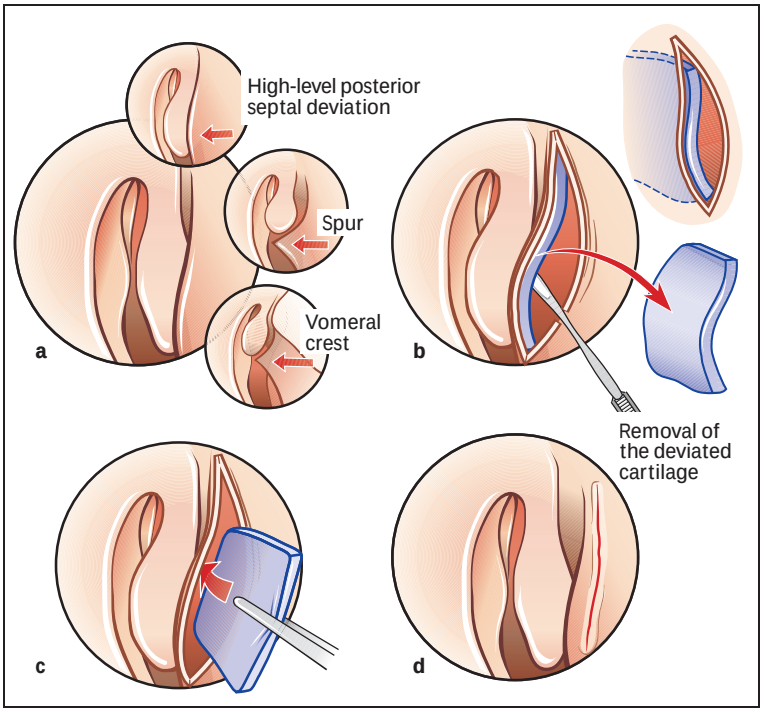


Fig. 2.98a–d Surgical steps in adjunctive endoscopic septal surgery.
a Indications for adjunctive septal surgery.
b Removal of the deviated posterior septum, external straightening with cross-hatching, morselization or incomplete cartilage incisions.
c Reimplantation of the cartilage.
d Straightening of the septum, with decompression of the middle turbinate.

ternal nose or septal thickening due to fibrosis. Antibiotic coverage is given. If necrosis occurs, the nonviable fragments are removed, and it may be necessary to implant preserved cartilage to support the nose.

Complications. If an infection is not drained, it may extend to the meninges or the cavernous sinus.

Septal Perforation

This condition is usually the result of trauma, nasal surgery, rhinitis sicca anterior, frequent nose picking, cocaine sniffing, infection (e.g., lupus, syphilis, leprosy, rhinoscleroma), or an occupational injury.

Clinical Features. These include crusting, fetor (possible but rare), slight recurrent nosebleeds, and a whistling noise on inspiration and expiration if the perforation is small. Large perforations located posteriorly may cause no symptoms whatsoever.

Diagnosis. A biopsy of the edge of the defect may be helpful to rule out generalized disease (e.g., Wegener granulomatosis). The consensus in the United Kingdom is that biopsy is not necessary in most patients. However, if the perforation edge is obviously abnormal or granulomatous, then a biopsy may be helpful.

Treatment. *Surgical treatment:* Various methods have been proposed, based on reconstruction of the cartilage defect (e.g., with turbinate, cartilage, or occasionally prosthetic material such as porcine intestinal submucosa or bioglass), covering it with mucosal flaps, and inducing epithelization. Figure 2.99 shows the principle of bilateral bridge flaps developed by Schulz-Coulon. More important than the size of the perforation is the extent of the remaining septum and the mucosa surrounding the defect.

Conservative treatment: The local symptoms of crusting and epistaxis may be alleviated by daily nasal douches and mild ointments. Occlusion of the defect with a Silastic collar stud–shaped button may be effective, but patients may remain symptomatic. An obturator is a foreign body, and plastic reconstruction is therefore preferable.

Trauma to the Nose, Paranasal Sinuses, and Facial Skeleton

Trauma to the Nose

Clinical Features. These include a visible deformity, lateral dislocation or depression of the nasal pyramid, hematoma of the soft tissues, orbital hematoma, swelling of the overlying soft tissue, septal hematoma, pain on application of pressure to the nasal pyramid, possibly headaches, epistaxis, nasal obstruction, and disorders of olfaction.

Fractures of the nasal bone are among the most common midfacial fractures.

Pathogenesis. This is circumscribed trauma to the nose from the front or from the side. Nasal injuries may also be part of a severe trauma to the face and skull. Depending on the intensity of the impact, the cartilaginous and/or bony nasal skeleton may be broken. In addition to the nasal bones, the frontal process of the maxilla as well as the nasal septum are often fractured.

Causes include traffic accidents, occupational accidents, blows, brawls, falls, etc. The injuries can be divided into closed and open. In *closed* injuries due to blunt trauma, the injury is not penetrating and the nasal skeleton is covered completely by soft tissues, with no external communication. In *open* injuries due to abrasions, cuts, or stab wounds, the cartilaginous or bony part of the nasal skeleton is exposed.

Diagnosis. This begins with the history and an examination, which reveals deformity and abnormal mobility of the external nose, soft-tissue injuries across the nose, and crepitation of the fragments on lateral pressure to the nose. Radiography of the nasal pyramid should be done (Fig. 2.100), and rhinomanometry and olfactometry may be useful after a few days. Involvement of the paranasal sinuses and the anterior base of the skull must be excluded.

> Note: A greenstick fracture in young patients, or an apparently harmless soft-tissue swelling, may initially conceal a fracture and severe skeletal injury. If fracture of the nasal bone is suspected, thorough investigation should always be carried out for functional, cosmetic, and medicolegal reasons.

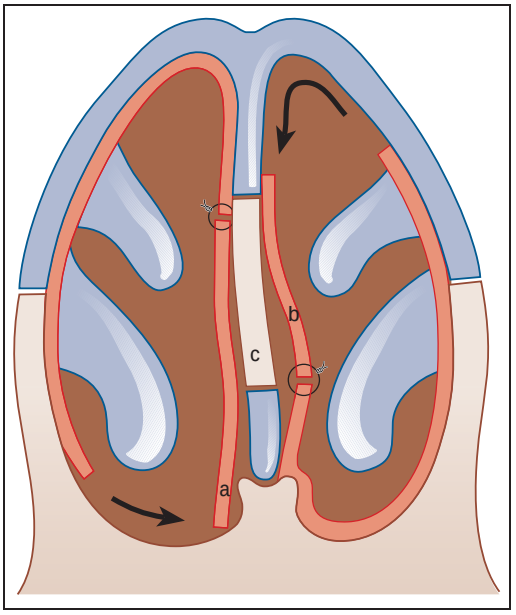


Fig. 2.99 Closure of a septal perforation using the Schulz-Coulon bridge-flap technique. a, Caudal flap; b, cranial flap; c, septum perforation.

Treatment. An open injury is first carefully cleaned. The wound edges and the nasal bones are checked, the displaced fragments are repositioned, and the soft tissues are repaired with atraumatic sutures. Antibiotic cover is necessary.

Repositioning of Fractures of the Nasal Bone

Simple, Closed Fracture

Repositioning should be carried out *within the first 48 h*, as the fracture can be most easily reduced in the early stages. However, in some cases it may be advantageous to wait for resolution of severe edema of soft facial tissue, before reducing the fracture on the fourth or fifth day. In theory, reduction is possible up to 10 days after an injury, but delay may have considerable cosmetic and functional sequelae if the fragments heal in a displaced position.

Technique. Short-acting intravenous or intubation anesthesia is used. The deviated part of the skeleton is repositioned, and the depressed part of the skeleton elevated as shown in Fig. 2.101a. If the fragments are already fixed or wedged, they must be refractured in the direction opposite

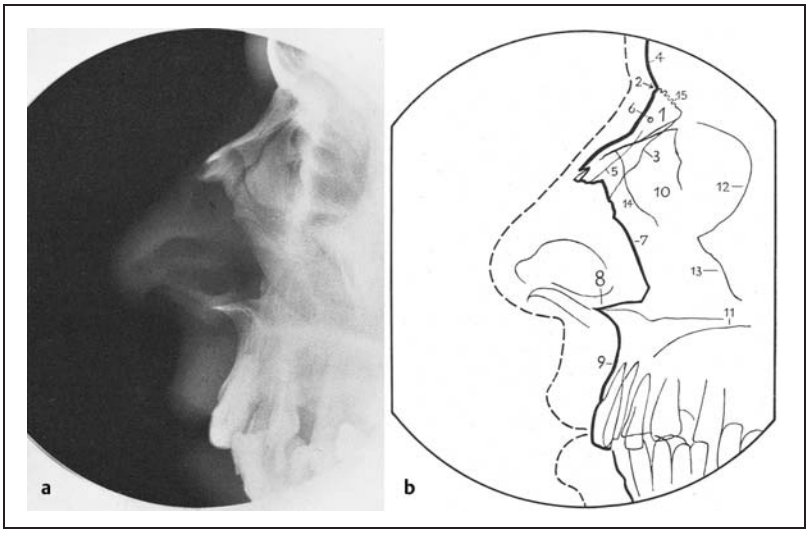


Fig. 2.100a, b Radiographic anatomy of a plain film of the nasal bone. 1, Nasal bone; 2, root of nose; 3, nasomaxillary suture; 4, frontal bone; 5, ethmoidal sulcus; 6, nasal foramen; 7, piriform aperture; 8, anterior nasal spine; 9, alveolar process; 10, frontal process of maxilla; 11, hard palate; 12, lateral border of orbit: 13, zygomatic bone; 14, anterior border of ethmoidal perpendicular plate; 15, nasofrontal suture.

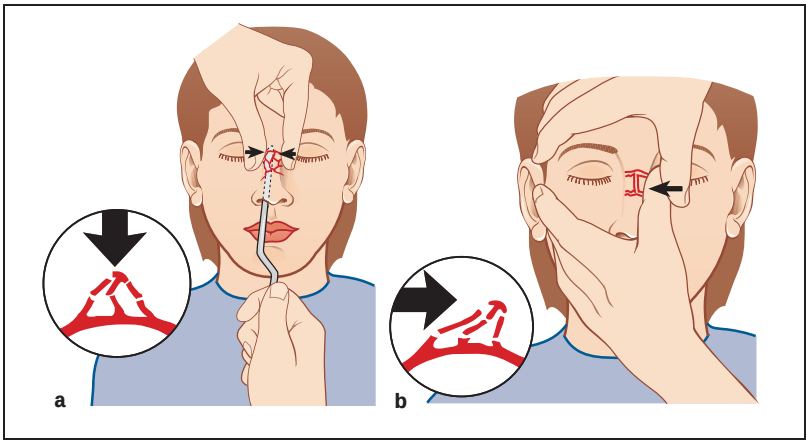


Fig. 2.101a, b Fracture of the nasal bone.
a By a blow from the front, and reposition by elevation of the fragments
b By a blow from the side, and subsequent reposition.

that of the original trauma (Fig. 2.101b), or even repositioned by osteotomies. The septum must be checked and corrected if necessary. Anterior nasal packing is introduced, and an external splint is worn for 8 days to ensure fixation.

Management of a Compound Fracture of the Nasal Bone or Nasal Pyramid

The affected part of the skeleton is examined and debrided carefully through the soft-tissue wound, which may need to be extended. The fragments are replaced and fixed, if necessary, with wires. The soft tissues are sutured, packing is introduced, and a splint is applied. Antibiotic cover is given.

If the soft tissues have been extensively damaged and show evidence of contamination, swelling, or necrosis, treatment of a compound fracture of the nasal bone may have to be delayed for 8–14 days until the acute tissue reaction has settled. The decision about the timing of surgery for a fracture of the nasal bone must be made by the rhinologic surgeon (Fig. 2.102).

Open repositioning of fractures of the nasal bone and/or septum is indicated in the following cases:

- Fractures that cannot be repositioned without open surgery.
- Penetration of the skin by bony fragments or perforation of soft tissues.
- Fractures requiring immediate reconstruction.
- Severe simultaneous injuries with a vital or absolute indication for surgery.

Supportive treatment includes antibiotics (if indicated), analgesia, prevention of nose blowing, internal or external splinting, and hemostyptic packing.

Complications. These include septal deviation (see p. 197), septal hematoma, and abscess (see p. 199). Long-term cosmetic defects may be caused by poor initial management or secondary infection. Elaborate and difficult plastic surgery is then needed, often in several stages. For a discussion of injuries to the nasolacrimal system, see p. 210.

Fig. 2.102 A fracture of the nasal bone is immobilized with a splint or cast for a total of 10 days.

Trauma of the Middle Third of the Face and the Sinuses

Figure 2.103 shows typical examples of the mechanism.

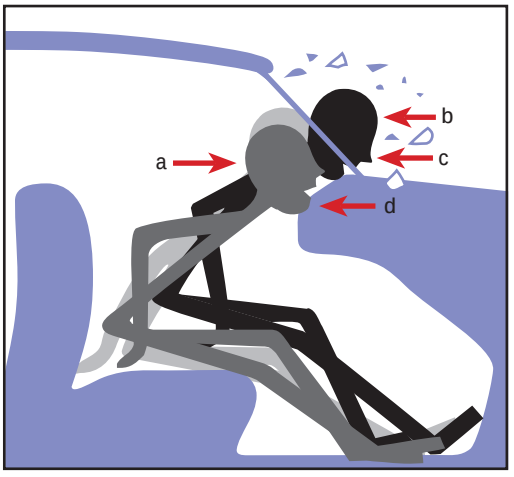


Fig. 2.103 The mechanism of trauma to the facial skeleton and base of the skull in a traffic accident. The following regions are frequently affected: a, cervical spine (whiplash injury); b, forehead and anterior base of the skull; c, midfacial structures; d, chin.

Maxillary Fracture

Sagittal, vertical fractures are unusual, whereas transverse fracture involving both sides of the face and in all cases the antral cavity is most common. It may also involve the other nasal sinuses and the anterior base of the skull. Figure 2.104 shows the three typical horizontal fractures of the middle third of the face. These are usually classified using the Le Fort system.

Symptoms

1. In *Le Fort class I* (low maxillary horizontal fractures), the alveolar process is detached. The patient has abnormal occlusion and a hematoma or fracture of the wall of the antrum.
2. In *Le Fort class II* (pyramidal fractures), the upper jaw is detached, and the fracture passes through the nasal bone, the frontal process of the maxilla, the medial orbital floor, and the zygomaticomaxillary suture. In this form of fracture, there is often considerable dislocation and depression of the central part of the face, with involvement of the ethmoids, the orbital contents, and the lacrimal apparatus, and an increase in the interpupillary distance, causing hypertelorism.
3. In *Le Fort class III* fractures, the facial skeleton is separated from the base of the skull. The fracture usually runs along the zygomaticofrontal, maxillofrontal, and nasofrontal sutures. The ethmoids, sphenoids, and often the frontal sinus and the orbit and its contents are involved, as are all the structures of the central part of the facial skeleton, often with massive depression of the middle third of the face and multiple fractures of the bones of the face, producing a so-called "dish face." This type of fracture also usually extends to the anterior part of the base of the skull (Fig. 2.104).

The typical symptoms of shock, concussion, and cerebral contusion almost always occur immediately after the injury.

For facial swelling as a symptom, see Table 2.10.

Pathogenesis. Central middle-third fractures are typical high-speed injuries, mainly due to traffic accidents and more rarely to occupational injuries.

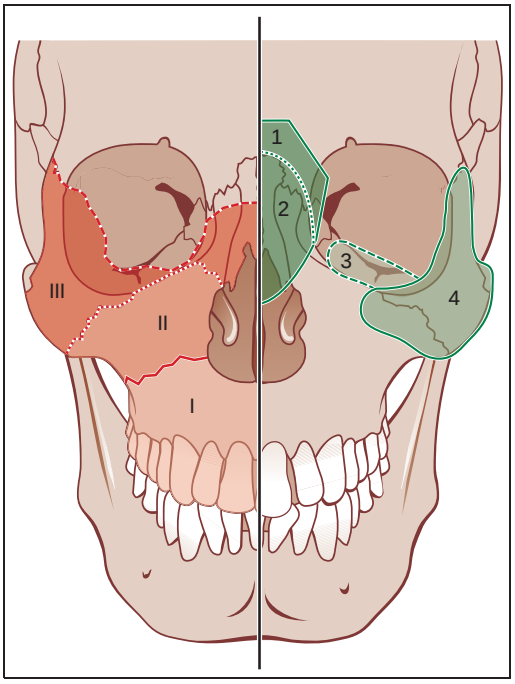


Fig. 2.104 Types of middle-third fracture. Le Fort classes I, II, III; 1, naso-orbito-ethmoid complex; 2, nasomaxillary complex; 3, orbital floor; 4, zygomatic bone.

Diagnosis. A history is taken to obtain information about the type, direction, and force of the trauma. Inspection often shows massive skeletal fractures, dislocations, and depression, with little soft-tissue injury. The symmetry of the central middle third of the face and frontal area should be noted. The inside *and* outside of the nose must be inspected. The orbit and its contents also have to be examined, with attention being given to unilateral or bilateral hematoma, eye movements, and the possibility of double vision. Vision is assessed. Palpation is used to assess tenderness on pressure, abnormal movement of the upper jaw, a break in the normal facial contour, steps and defects in the bony skeleton, especially of the orbital rim, trismus, and crepitation of the root of the nose (Fig. 2.105). The movement of the mandible in all directions, occlusion, and the condition of the teeth are assessed, and defects of sensory or motor innervation, any CSF rhinorrhea, and prolapse of brain tissue are looked for.

Radiography: CT scans are best for diagnosis of bony injury and dislocations. In certain cases, MRI, angiography, or functional diagnostic studies (e. g., olfactometry) may also be needed.

A maxillofacial surgeon, an ophthalmologist, a neurologist, or a neurosurgeon may need to be consulted. If multiple injuries are present, advice from a general or orthopedic surgeon may be necessary before treatment is started. The timing sequence of the various procedures should be agreed.

> **!** Note: Skeletal asymmetry or deformity in middle-third fractures is often concealed by rapid soft-tissue swelling or bloody effusion. In addition, harmless-looking soft-tissue injuries in the region of the base of the skull may conceal serious life-threatening skeletal injuries.

Treatment. Before a patient is removed from the setting in which the injury occurred, the ABC of traumatology has to be observed:

A: The airway is secured and aspiration is prevented.

B: Bleeding must be controlled.

C: Circulatory shock must be treated.

> **!** Note: Every patient with a fracture of the middle third of the face must be hospitalized. The department to which the patient is to be admitted is decided on the basis of the extent and type of the injury to the skull and other injuries to the extremities, thorax, and abdomen. Since patients with head injuries very often require immediate treatment by special trauma teams consisting of a neurosurgeon, a rhinosurgeon, a maxillofacial surgeon, and an ophthalmic surgeon, immediate admission to a suitably equipped large trauma center is indicated.

The purpose of definitive surgery is to restore normal anatomy and function by debridement, ventilation, and drainage. The specific duties of the rhinologic surgeon are:

1. Treatment of the soft-tissue injury and management of the nose and sinuses by debridement, drainage, and ventilation.
2. Assessment of the base of the skull (because of the frequent combination with a fracture of the anterior cranial fossa and the pterygopalatine fossa).
3. Correction of the facial skeleton and bony orbit (see p. 206).

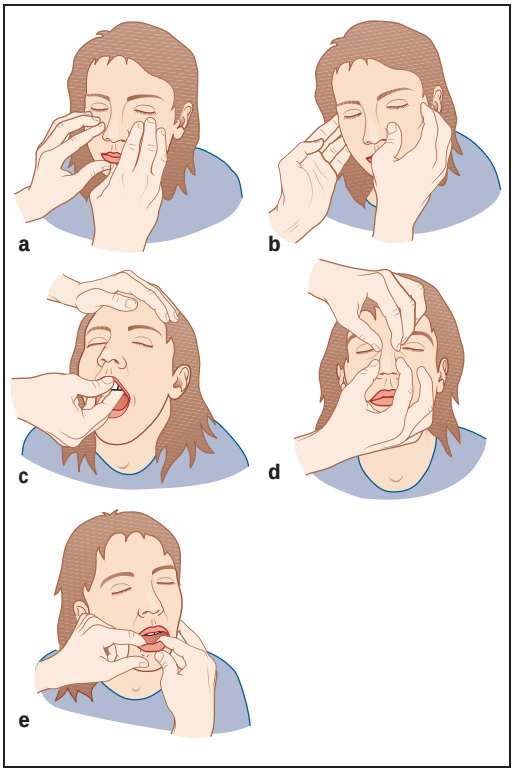


Fig. 2.105a–e Palpation of central middle-third fractures.
a Bony orbital rim.
b Zygomatic bone.
c Maxilla.
d Nasal pyramid.
e Mandible.

Definitive surgery for middle-third fractures should be performed as quickly as possible, as the fracture may heal very rapidly in the wrong position due to the callus formation. The management of simultaneous frontobasal injuries is described on p. 207.

Fractures of the Zygoma and Bony Orbit

Combined fractures involving the trifurcate zygoma with the zygomatic arch, and also the orbital rim with the orbital floor (lateral midface fracture), are frequent. Isolated blow-out fractures of the orbital floor and isolated fracture of the zygomatic arch may also occur. Fractures of the zygoma and orbit may also form part of a more severe midfacial fracture or frontobasal fracture. The antrum is almost always involved (Fig. 2.106a, b). The mechanism of fracture is usually blunt violence to the lateral part of the face by a blow from a fist, a traffic accident, or a fall down stairs. The fracture is almost always depressed. Dislocation of the fragments may be minimal, but there may be extensive comminution, with multiple fragments that can only be repositioned and fixed with great difficulty (Fig. 2.106a, b).

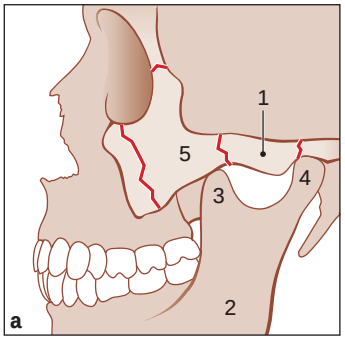
Clinical Features. These include orbital hematoma, swelling of the eyelids, asymmetry of the middle third of the face with a sunken contour of the cheek on the side of the fracture, inferior displacement and possibly enophthalmos on the side of the fracture, a step in the bony wall of the orbit inferiorly or laterally, rarely of the upper rim, and possibly trismus. The soft tissues in the zygomatic area swell initially, but the underlying bony outline is flattened. Sensation may be lost in the infraorbital nerve. In a blow-out fracture, there is partial limitation of the movements of the eye, with double vision due to trapping of the inferior rectus or inferior oblique muscles.

Diagnosis. This is based on the history of the type and direction of trauma, inspection, and bimanual palpation, which shows asymmetry of the facial skeleton, a step in the orbital wall, or limitation of movement of the lower jaw. Radiography includes standard views of the nasal sinuses, special views for the zygomatic arch, and tomograms. An ophthalmologic examination should be performed.

> **!** Note: Fractures of the zygoma are relatively commonplace. They are often overlooked initially because of the soft-tissue swelling of the cheek and of the lateral part of the face. They are therefore recognized late, after bony consolidation in an incorrect position. Even after relatively mild trauma to the middle third of the face, from in front or from the side, asymmetry of the facial skeleton or a step in the orbital wall or loss of sensation of the infraorbital nerve should always be looked for using bimanual palpation, to allow comparison of the two sides.

Treatment. Several operations are available for repositioning and stabilizing fractures of the zygoma (Fig. 2.107a, b):

1. Access from the vestibule of the mouth and from the maxillary antrum.
2. Access from the temporal region.
3. Access directly through the overlying soft tissues.



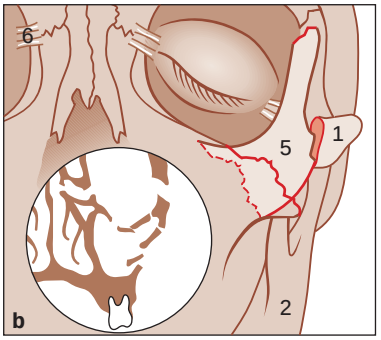
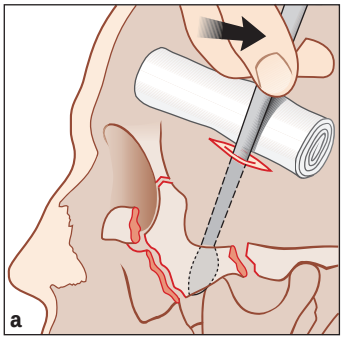


Fig. 2.106a, b Zygomatic fracture.
a Lateral view. 1, Zygomatic arch; 2, mandible; 3, coronoid process; 4, head of mandible; 5, body of zygomatic bone.
b Anterior view. 1, Zygomatic arch; 2, mandible; 5, body of zygomatic bone; 6, medial palpebral ligament. The inset shows involvement of the orbit, antral cavity, and ethmoids.



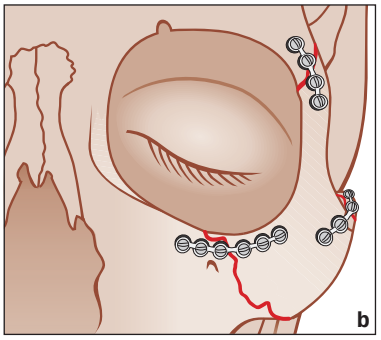


Fig. 2.107a, b Management of a typical zygomatic fracture.
a Soft-tissue incision to allow repositioning of the fragments and access for elevation.
b The condition after repositioning and osteosynthesis with microplates.

The use and combination of repositioning and methods of stabilization—e.g., single-pronged retractors, osteosynthesis with microplates, or wiring—depend on the type and severity of the fracture. If there is loss of sensation of the infraorbital nerve, the nerve must be exposed and decompressed.

Isolated Blow-out Fracture

Fracture of the orbital floor with descent of the orbital contents into the maxillary cavity, caused by blunt force acting on the ocular bulb in an axial direction; it may be accompanied by fracture of the medial zygoma or the infraorbital rim.

Such fractures are caused by violence localized to the orbital contents by a blow—e.g., from a fist, a tennis ball, or a champagne bottle cork. The thin bony floor of the orbit breaks and prolapses into the antral cavity (Fig. 2.108a–c). This can lead to trapping of the orbital contents (fat, ocular muscles such as the inferior rectus and inferior oblique muscles) by the fragments and to prolapse of the fractured floor of the orbit into the antral cavity. A blow-out fracture can also occur in the ethmoid, caused by a defect in the lamina papyracea.

Clinical Features. These include enophthalmos, double vision, limitation of movement of the eye (most obvious when the patient is looking upward, due to trapping of the lower ocular muscles), and disorders of sensation of the infraorbital nerve.

Diagnosis. Precise documentation of the course of the accident, routine otorhinolaryngology examination, inspection (enophthalmos, abnormal position of the bulb), palpation (step formation at the infraorbital rim, cutaneous emphysema), bulbar movement (double vision, especially on looking upward), comparison of sensory perception on both sides (especially of the infraorbital nerve), nasal endoscopy, CT, and ophthalmologic examination.

Indications for surgery are dislocated fractures (double vision, sensory deficiencies) or orbital herniation, with or without impaired motility of the bulb (Fig. 2.109).

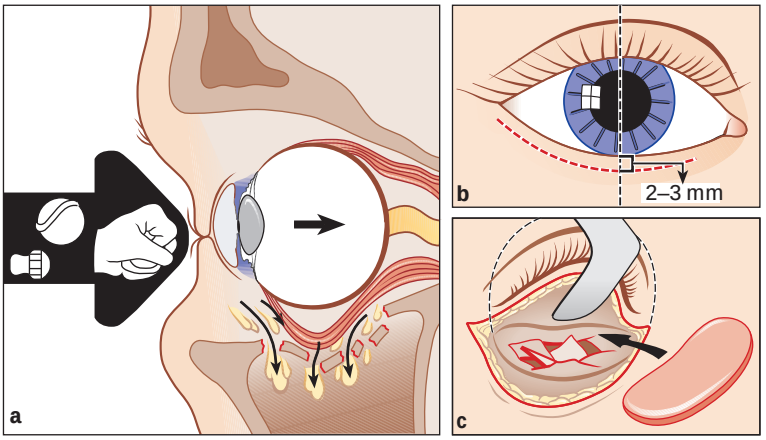


Fig. 2.108a–c Blow-out fracture.
a Mechanism.
b Subciliary approach to the orbital floor
c Stabilization of the orbit with an implant (septal cartilage, titanium).

Treatment. The antral cavity must be explored as early as possible, preferably using the endoscopic endonasal supraturbinal approach. (For differentiation between a mucosal hematoma or edema and an orbital hernia, see p. 208). The bony fragments are exposed and the prolapsed part is replaced, possibly combined with bridging or stabilization of the bony defect with lyophilized dura, fascia, or autologous septal cartilage. Alternative or supplementary measures include access via the orbit through an incision in the lid and introduction of a septal cartilage transplant, lyophilized dura, or fascia to support the orbital contents. If the fracture heals in an incorrect position, the resulting enophthalmos can be corrected by supporting the orbital contents with an implant, preferably using autologous rather than synthetic material. Decompression of the infraorbital nerve and possibly osteosynthesis of the orbital limbus may be required.

Supportive treatment: Prohibition of nose blowing; administration of analgesics, antibiotics, and nose drops; application of cold.

Barotrauma of the Sinuses

This is caused by the considerable difference between the air pressure in the sinus and that of the environment if rapid equalization of pressure is not possible due to anatomic abnormalities. Aviators, divers, and parachute jumpers are most often affected.

Clinical Features. These include sudden, severe, stabbing pain in the region of the nasal sinuses, usually the frontal and rarely the antrum, during and after a temporary marked pressure difference. The result may be a nosebleed or sinusitis due to severe mucosal damage to the affected cavity.

Treatment. This includes decongestion of the nasal mucosa, treatment of symptoms, possibly antibiotics, and surgery to the nose and sinuses, such as septal correction, conchotomy, or functional endoscopic sinus surgery (FESS) as a preventive measure.

Frontobasal Injury to the Anterior Cranial Fossa and the Neighboring Sinuses

Trauma to the frontal area and the root of the nose usually occurs in traffic accidents, but occasionally also in occupational injuries. It causes a fracture that first affects the upper sinuses (the frontal, the ethmoid, and the sphenoid sinuses), and from there extends into the anterior cranial fossa. Alternatively, the fracture may primarily affect the upper part of the frontal bone, together with the dura mater and the intracranial structures, and the fracture line may extend from there into the nasal sinuses. Frontobasal fractures occur in 70% of all fractures of the skull base. There are typical fracture lines in the anterior cranial fossa and the rhinobase, similar to the fracture lines in laterobasal fractures (see pp. 208, 209, Fig. 2.110a–d).

Dural tears and brain injuries, which may be open or closed, thus often arise in the anterior cranial fossa. Infection may ascend to the intracranial cavity through the fracture line from the nose or the sinuses, either immediately after the acci-

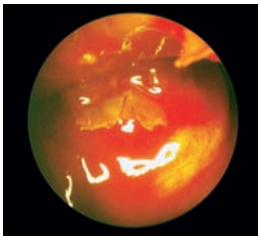


Fig. 2.109 Endoscopic view of a blow-out fracture through a supraturbinal window (45° lens).

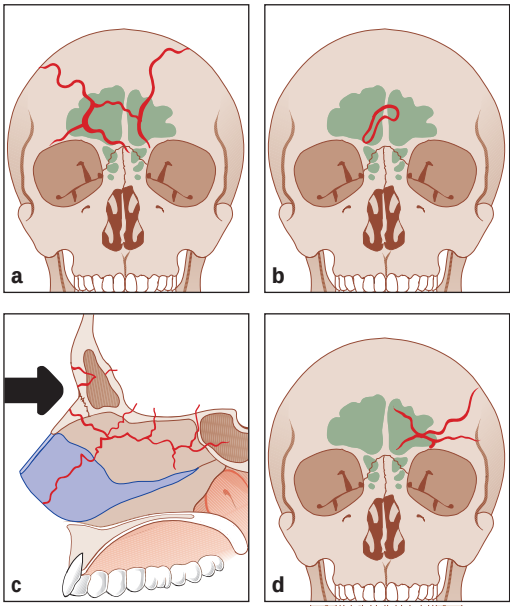


Fig. 2.110a–d Frontobasal fractures.
a High fracture (Escher type I).
b Middle fracture (Escher type II).
c Deep fracture (Escher type III).
d Latero-orbital fracture (Escher type IV).

dent (early infection) or years later (late infection), leading to meningitis or a brain abscess.

Clinical Features. Cardinal symptoms include CSF rhinorrhea, brain tissue prolapsing from the nose or from the external wound of the nasofrontal area, extensive facial hematoma, possibly surgical emphysema, proptosis with or without loss of vision, and frontal pneumoencephalocele.

Symptoms also include cerebral concussion or contusion, unilateral or bilateral ocular hematoma, which is *not* conclusive evidence of frontobasal injury, and occasionally severe bleeding from the pharynx, mouth, and nose. Cerebrospinal fluid rhinorrhea is a definite sign of a dural tear, but dural tears are possible without a CSF leak. Anosmia occurs in 75% of patients, and cranial nerve II and, more rarely, cranial nerves III IV, V, and VI may be injured. External soft-tissue injuries are minor or absent in 20% of all patients. Occasionally, signs of incipient increased intracranial pressure are present—e. g., bleeding, pulse rate alteration, homolateral dilation of a pupil, and fixed pupil.

Diagnosis. This is based on the history and type of accident, radiographs of the skull in two planes, radiographs of the sinuses in several different projections (occipitomental, occipitofrontal, axial, and overextended axial), and computed tomography (Fig. 2.111).

Evidence of CSF rhinorrhea is provided by a positive β_2 transferrin test. In this laboratory test, the ratio of β_1 to β_2 transferrin in the nasal secretions and blood serum is determined. As high β_2 transferrin levels occur only in CSF, they are proof of a CSF leak. Sodium fluorescein can be used to locate a dura lesion with a CSF leak. Although this drug is not approved for intrathecal use, it has proved useful for locating CSF fistulas in thousands of cases. It is applied via lumbar puncture at least 10 min before surgical dissection of the base of the skull. It is very important to use a highly purified solution; the recommended dose is 1.5 mL of a 0.5% solution.

Use of indicator paper to determine the glucose content of nasal secretions, which are elevated in the presence of CSF, does not offer conclusive proof of a CSF leak.

Another option is radioisotope scanning of the cerebrospinal space.

Treatment. As soon as the patient has been stabilized by treatment for shock, the anterior cranial fossa and the affected nasal sinus should be exposed to allow debridement and watertight closure of the intracranial cavity with dural repair. The procedure is performed via the nasal sinuses. Injuries to the nasal sinuses are repositioned.

Three groups of indication for surgical intervention include:

1. *Vital indications* requiring immediate surgical intervention:
   - A life-threatening increase in intracranial pressure due to intracranial hemorrhage.
   - Life-threatening hemorrhage into the sinuses, nasopharynx, or base of the skull.

2. *Absolute indications* mandating surgical intervention as soon as possible:
   - Evidence of a dural tear—e. g., CSF rhinorrhea or pneumoencephalocele.
   - Open brain injury.
   - Early or late intracranial complications—e. g., meningitis, extradural abscess, subdural abscess, or cerebral abscess.
   - Impacted foreign body.
   - Orbital complications.
   - Osteomyelitis of the frontal bone.
   - Depressed fracture with a suspected dural tear.
   - Cranial nerve lesions requiring decompression.
   - Impalement injuries.
3. *Relative indications* requiring surgical intervention within 1–2 weeks:
   - Fractures affecting the frontal, ethmoid, or sphenoid sinus in which dura involvement is suspected, but not proved with certainty.
   - Depressed fractures and fractures with obvious displacement of fragments, with or without suspicion of a dural tear.
   - Injuries to the nasal sinuses with penetrating soft-tissue injuries.
   - Infection of the nasal sinuses already present at the time of injury.
   - Posttraumatic sinusitis and mucopyocele.

The purpose of the operation is to achieve wide exposure of the injured area together with the dura, to allow removal of fragments and management of injury to the brain and the base of the skull. This is followed by closure of the dura with fascia or galea and establishment of free drainage of the affected sinus using one of the typical rhinologic sinus operations (see pp. 171 ff.).

The four typical routes of access are shown schematically in Fig. 2.112:

1. Endoscopic, endonasal.
2. Fronto-orbital.
3. Transfrontal–extradural.
4. Transfrontal–intradural.

The choice of procedure and the sequence of the surgical steps depend on the individual patient and on joint treatment planning by a neurosurgeon, rhinologist, and maxillofacial surgeon, possibly with the addition of an ophthalmic surgeon, and a general surgeon or traumatologist in patients with multiple injuries.

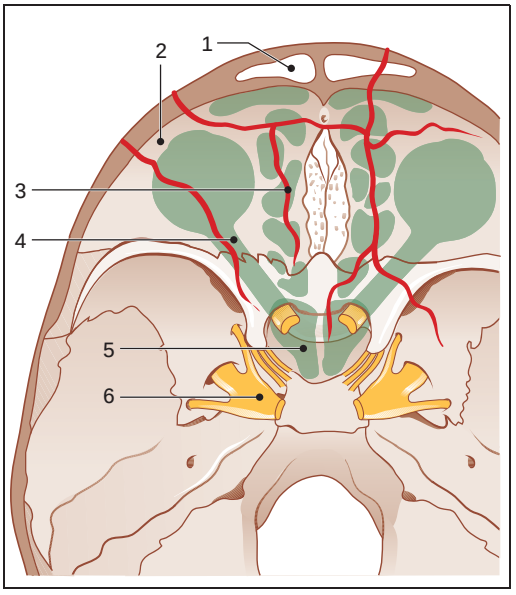


Fig. 2.111 Typical fracture lines in the anterior base of the skull. 1, Frontal sinus; 2, orbit; 3, ethmoid; 4, optic nerve; 5, sphenoid sinus; 6, Gasserian ganglion.

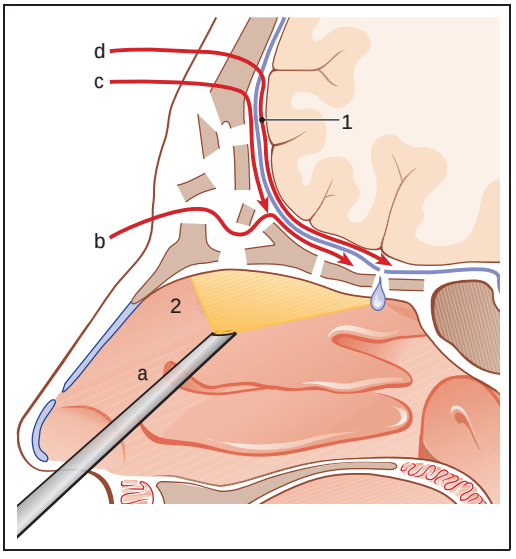


Fig. 2.112 Routes of access for management of frontobasal fractures. a, Endoscopic, endonasal approach; b, fronto-orbital approach; c, transfrontal–extradural approach; d, transfrontal–intradural approach. 1, Dura mater; 2, nasal cavity.

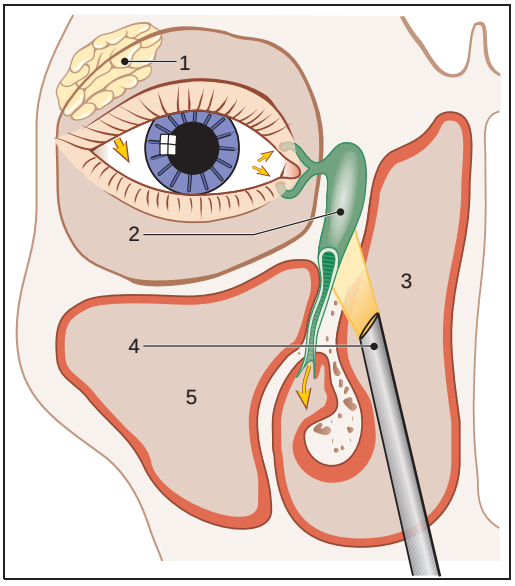


Fig. 2.113 The eye and nose. 1, Lacrimal gland; 2, lacrimal sac with lacrimal ducts; 3, nasal cavity; 4, site for dacryocystorhinostomy; 5, antral cavity.

Complications. These include CSF fistula, recurrent late meningitis, early or late brain abscess, osteomyelitis of the flat bones of the skull (see p. 190), and formation of mucoceles or pyoceles (see p. 184).

Other Possible Injuries to the Facial Skeleton

Injury to the facial nerve. See page 112.

Injuries to the lacrimal system (Fig. 2.113). These are quite commonly combined with injuries to the sinuses and the middle of the face. If possible, they should be repaired in the same session as the injuries to the sinuses and face. If this is not possible, or is not successful, stenosis or complete obstruction of the lacrimal sac or nasolacrimal duct may occur (see Fig. 2.113), requiring correction (dacryocystorhinostomy; see Fig. 2.113) either by an ophthalmic or rhinologic surgeon.

Injuries to the mandible and temporomandibular joint. These injuries are managed by a maxillofacial surgeon, who is also responsible for restoring correct occlusion.

The main symptoms of mandibular fractures include swelling of the lower part of the face, abnormal movement or deformity of the mandible, anomalies of occlusion, pain on movement, compression or torsion of the mandible, and possibly trismus.

Immediate first aid measures should be undertaken for comminuted fractures, particularly fractures of the chin with extensive soft-tissue injury. The patient should be intubated, or a tracheotomy should be performed, due to the danger of respiratory obstruction. Profuse bleeding should be arrested, if necessary with a pressure dressing. Soft-tissue defects or scars are dealt with in the usual way using plastic surgery reconstructive procedures.

Congenital Anomalies and Deformities of the Nose

Congenital Anomalies of the Nose

Development of the embryonic face involves nine processes. Due to the complexity of these processes, congenital anomalies are relatively common, although many of them are only slight. Anomalies incompatible with life—e. g., arhinia—are extremely rare.

Cleft Face and Nose

Oblique facial clefts, even in a rudimentary form, are rare, as are *transverse facial clefts,* in which the angle of the mouth is located near the tragus, causing macrostomia.

Median clefts are more common, but are usually only rudimentary. They range in extent from hypertelorism, with or without a true median facial cleft and with or without meningoencephalocele, to dog nose, proboscis, or even double nose.

Treatment. Plastic surgery.

Cleft Lip, Jaw, and Palate

These anomalies are relatively common, affecting 1 % of Caucasians.

Treatment. Reconstructive surgery, occasionally in stages, and orthodontic and prosthetic procedures. In addition, speech therapy may be needed. Treatment planning is described on page 281.

Cleft lip and palate present the rhinologist with several problems: firstly, anomalies of the external nose, such as a flattened nasal ala, and anomalies

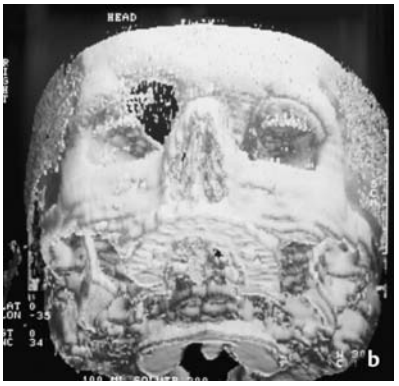


Fig. 2.114a, b Post-traumatic meningoencephalocele following frontobasal fracture with a large frontal bone defect.
a Caudal displacement of the eyeball.
b Bony defect in the anterior base of the skull.

within the nose, such as dislocation of the septum; secondly, speech disorders, such as rhinolalia aperta of varying degrees of severity; and thirdly, abnormalities of tubal aeration which are regularly present, causing seromucotympanum or chronic otitis media.

Nasal Fistulas, Nasal Cysts, Dermoid Cysts, and Gliomas

Median nasal fistulas, from which a cloudy secretion may drain, usually end blindly at the level of the glabella or in the ethmoidal–septum region. This is also true of *congenital nasal cysts,* which develop at the same site, but also may arise in the nasal vestibule or nasal septum.

Dermoid cysts are more common. They often contain hair and accessory structures, such as ectodermal inclusions, which lie above, below, or within the frontal bone.

Gliomas are tumors of the frontal region or the root of the nose in the midline; they consist of solid glial tissue.

All of these anomalies must be removed surgically.

Meningoencephalocele

Dural and brain herniations (hernias and celes) may be found in the nose, where they are easily mistaken for nasal polyps. They may also be located extranasally and be related to the frontal bone, ethmoid, or nasal septum. The causes are incomplete closure of the neuropore during the third week of embryonic development, and trauma (Fig. 2.114a, b). Meningoencephaloceles and gliomas can often be removed and the defect can nowadays be repaired endoscopically.

Diagnosis. MRI with contrast medium and CT.

Treatment. Surgical removal, closure of the dura, and osteoplastic correction of any existing hypertelorism, if required.

Stenosis and Atresia of the Nostrils

These are usually congenital, but may also be acquired—e.g., due to trauma or destructive infections.

Treatment. Plastic surgery.

Choanal Atresia

This is a bony or membranous occlusion of the posterior nasal opening, and may be bilateral. Girls are more often affected than boys. Hereditary factors have been demonstrated. The disorder is usually congenital, but it may also be acquired due to trauma. Incomplete atresia (stenosis) also occurs.

Clinical Features. Newborn babies with bilateral choanal atresia present with intermittent cyanosis, especially during feeding. Other symptoms include chronic purulent nasal discharge, inability to breathe through the nose or to sneeze, and anosmia.

Note: Bilateral atresia in the newborn is a life-threatening condition. As the infant is unable to breathe satisfactorily through the mouth due to the relatively high position of the larynx, it has to rely on nasal respiration during feeding. This poses a risk, in bilateral atresia at least, of asphyxia, cyanosis, atelectasis of the lungs, or aspiration pneumonia. Spontaneous feeding is difficult or impossible.

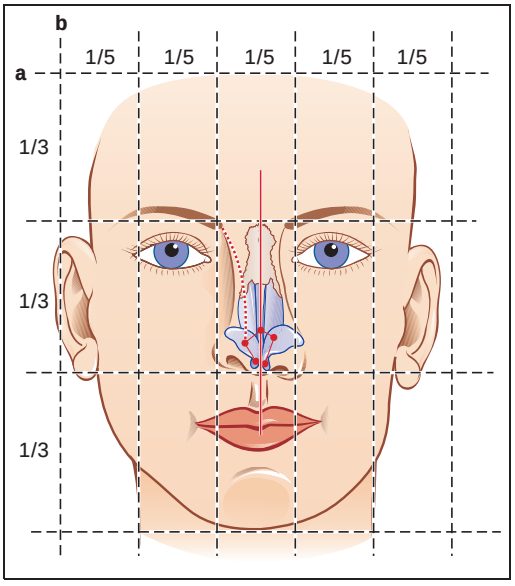


Fig. 2.115a, b Facial proportions and symmetry.
a Lines dividing the face into vertical thirds (Leonardo da Vinci).
b Lines dividing the face into horizontal fifths (Powell and Humphreys). Right half of face, features of symmetry: esthetic eyebrow-tip line, facial midline, nasal tip rhomboid. Left half of face, major causes of asymmetry: asymmetrical eyebrow-tip line, hypoplasia of maxilla, midface or mandible (usually with a crooked mouth), slanted nasal base (cleft lip and palate, asymmetry of specific structural elements such as the upper lateral or alar cartilage).

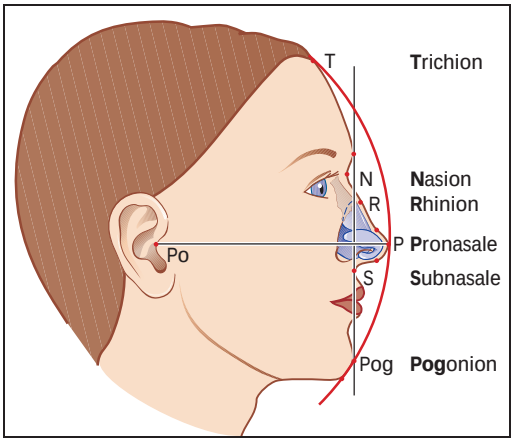


Fig. 2.116 Geometric points and lines used in profile analysis.

Diagnosis. The most reliable and simple test is to hold a cold metal spatula just beneath the nose and look for misting. Further tests include probing the nose with a smooth catheter and endoscopy of the nose and nasopharynx.

Differential diagnosis. This includes any disorders that cause nasal obstruction, especially polyps, foreign bodies, encephalocele, and tumors.

Treatment. When *bilateral atresia* has been recognized, the airway obstruction can be relieved by inserting an oropharyngeal airway until surgical correction can be arranged. Transpalatal surgery has now been largely superseded by dilation under direct vision of the posterior choana using a 120° endoscope in the pharynx.

In *unilateral atresia*, surgical correction can be deferred to a later date.

Disorders of Shape of the External Nose

Anomalies of the shape of the external nose often require rhinoplasty, both for esthetic reasons and because of impaired function.

Anomalies of the shape of the nose may be due to the cartilaginous or bony parts of the internal or external nose skeleton being too large or too small, or being improperly positioned in relation to each other or to surrounding structures. Analysis of the deformities is based on certain parameters, as illustrated in Figs. 2.115 and 2.116, on standardized photographs. Certain angles in the facial contour also have to be taken into account.

Fundamentals of Rhinoplasty

Rhinoplasty has two purposes:

1. It should restore a normal shape to the nose, so that it harmonizes with the rest of the face.
2. The function of the nose and nasal sinuses with regard to respiration, olfaction, etc., should be maintained, improved, and returned to normal.

This dual character of modern nasal surgery often requires correction of both the outside and inside of the nose—i. e., *septorhinoplasty*. These operations must be performed by a suitably trained rhinologist.

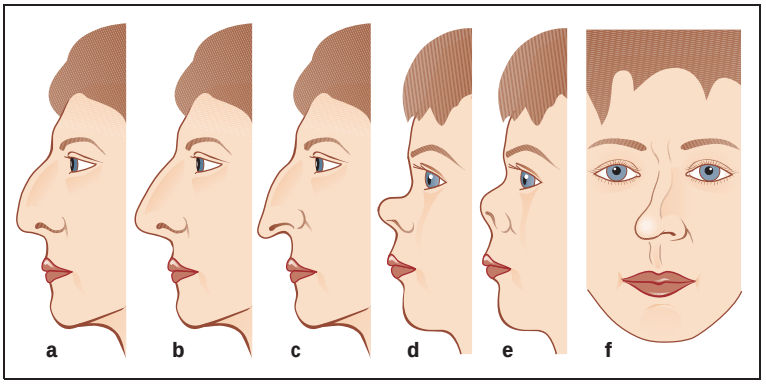


Fig. 2.117a–f Different nasal types.
a Hump nose.
b Overprojecting nose, functional tension nose.
c Drooping nasal tip.
d Saddle nose.
e Short nose.
f Deviated nose.

A distinction is made between *corrective* and *reconstructive* operations. In corrective rhinoplasty, and particularly for correction of the supporting framework of the nose, all of the surgical steps are performed from inside the nose, without an external incision (Fig. 2.117).

The principal steps in rhinoplasty are:

- An incision in the nasal vestibule or at the medial columella.
- Elevation of the soft tissue covering the nasal skeleton.
- Exposure, mobilization, and correction of all skeletal elements.
- Union of the mobilized and corrected skeletal parts to form a pleasing nasal framework and a functionally competent nasal cavity.
- Fixation of the mobilized, corrected skeletal elements in the required position.

Anomalies of the cartilaginous part of the nose include anomalies of the shape of the base of the nose—e.g., a hanging nasal tip, or a tip that is too flat or too wide, cleft, too long, or too short; nasal alae that are flaccid, too arched, or asymmetrical; a nasal columella that projects too much, is too retracted, too thick, too short, or too bent. The entire nose may also be too long or too short.

Anomalies of the bony part of the nose include anomalies of the shape of the bridge of the nose—e.g., hump nose, saddle nose, twisted nose, broad nose, or narrow nose; of the root of the nose; and of the nasal septum. The bony and cartilaginous parts of the nose usually need to be corrected together.

Fig. 2.118 The splitting approach for cranial volume reduction of the alar cartilage.

Approaches. Three approaches have proved valuable for rhinoplasty:

- The splitting approach (Fig. 2.118).
- The delivery approach (Fig. 2.119).
- The open approach (Fig. 2.120).

Which approach is used depends on the specific morphologic problems. Other decisive factors are types of skin and connective tissue. Thick skin predisposes to healing problems, whereas thin skin limits the selection of surgical techniques—e.g., using tip or shield grafts that might remain visible through the skin.

Reconstructive rhinoplasty deals with partial loss of the soft tissue, of the skeleton of the nose, or total loss of the nose, and reconstruction of a complete nose. Local or distant flaps are used, depending on the type and extent of the defect (see p. 217).

Hump Nose

This is due to an excess of the bony or cartilaginous nasal skeleton. Usually, it does not cause any marked functional disturbance.

Treatment. Rhinoplasty is performed, with removal of the hump and narrowing of the nasal bridge. The principles of the procedure are shown in Figs. 2.121a,b and 2.122.

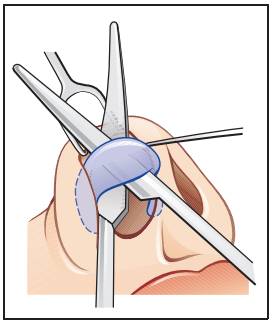


Fig. 2.119 The delivery approach for shaping the nasal tip.

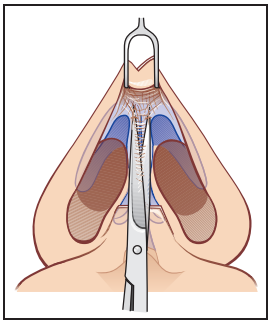


Fig. 2.120 The open approach for correction of difficult asymmetries of the tip and nasal dorsum.

Fig. 2.121a, b a A young woman with dorsal hump, overprojecting nasal tip, recessed chin, and effaced nasolabial angle.
b The appearance 3 years after septorhinoplasty. The dorsum has been lowered and projection decreased, the chin appears normal, and the upper lip and nasolabial angle are relaxed.

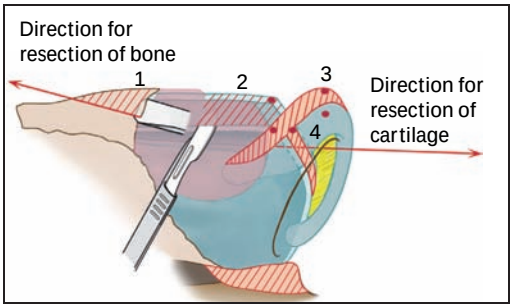


Fig. 2.122 The principle of nasal hump correction. 1, Bony resection; 2, cartilaginous resection; 3, shaping of the nasal tip; 4, correction of the anterior nasal septum.

Saddle Nose

The term "saddle nose" denotes a multifactorial condition that is associated with destabilization or destruction of the bony or cartilaginous structures of the nose. Osseous forms of saddle nose are nowadays rare and usually result from dysplasia of the nasal bones or from nasal midfacial trauma.

Cartilaginous saddle nose is a more frequent concern for rhinologists. The central problem in this condition is the serious structural compromise caused by a loss of anterior septal cartilage between the rhinion (keystone area) and what is known as the septal pedestal at the level of the premaxilla and the anterior nasal spine.

Pathomechanism. The septal cartilage is partly or completely absent due to trauma, an incorrectly executed septoplasty, a septal abscess, or an infection; the condition may also be congenital.

There is considerable functional disturbance due to ballooning—i. e., expansion of the nasal valve caused by collapse of the ala (Figs. 2.123a, b and 2.124a–c).

Clinical Features. The profile of the nose is typical. The patient may also report nasal obstruction due to absence of support for the nasal valve and introitus.

Treatment. A reconstructive procedure is needed for surgical treatment of a saddle nose. Rhinoplasty is performed, with implantation of several struts and implants to restore the cartilaginous framework of the nose, particularly the important central structure of the anterior nasal septum.

Deviated Nose

The cartilaginous *and* bony nasal skeleton and nasal septum are most often involved here. The cause is usually trauma, but a deviant nose may be congen-

ital. Deviation usually causes considerable interference with function (Fig. 2.125).

Analysis. Assessment of the axial deviation is important in order to determine whether the deviation is due to cartilaginous or bony factors, whether it has a C or an S shape, and whether the condition is a pseudodeviation caused by differences in the esthetic eyebrow lines.

The *esthetic eyebrow line* is a theoretical line connecting the medial end of the eyebrow with the point that defines the tip of the nose (the pronasal or tip-defining point).

Treatment. This consists of septorhinoplasty with correction of the skeletal parts of the external nose and of the septum. The principles are shown in Figs. 2.119, 2.120, and 2.122 and the septal operation in Fig. 2.96.

Reconstruction of the bony nasal pyramid requires osteotomy. Spreader grafts are slender implants taken from the septal cartilage and used subcutaneously for various indications, one of which is correction of a deviant nose.

Other Deformities of the Nose

A *broad nose* is usually due to trauma, particularly if initial treatment has been incorrect, whereas a *narrow nose, large nose,* or *long nose* are all congenital. Interference with function can be expected, especially in cases of narrow nose and posttraumatic wide nose. A large nose and a long nose usually only cause esthetic problems.

Treatment. Septorhinoplasty is performed.

Anomalies of the Nasal Alae

Nasal alae that are too weak and prolapse on inspiration, or that are too flared, are corrected by strengthening or reducing the nasal cartilage. The lateral alae can be stabilized by autologous transplantation of cartilage taken from the septum or a turbinate (Fig. 2.126).

The profile of the forehead and the chin have to be included in an esthetic analysis of the face as a whole, in addition to the shape of the nose. A profile plasty may sometimes be required, with reduction or building up of the chin or forehead.

Fig. 2.123a, b a A young woman with a broad, saddled nasal dorsum. b Three years after septorhinoplasty.

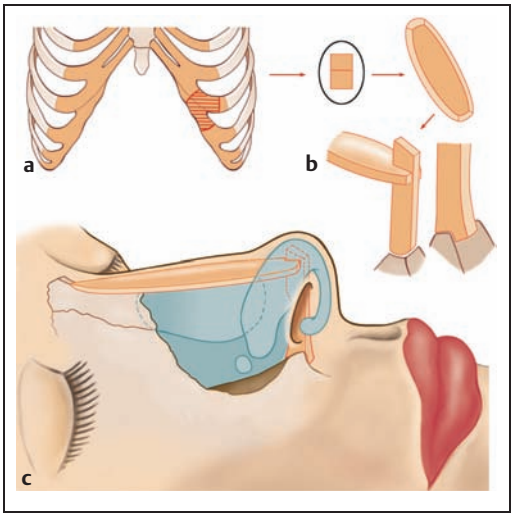


Fig. 2.124a–c Reconstruction of saddle nose with costal cartilage.
a Harvesting of the transplant.
b Shaping of a balanced costal graft.
c Tongue-and-groove connection of the grafts.

Basic Plastic Reconstruction Procedures in the Head and Neck

Soft-tissue and skeletal defects in the head and neck due to trauma, mutilating tumor operations, or congenital anomalies are encountered frequently; they require reconstruction for functional and esthetic reasons. This type of operation requires special surgical techniques and a thorough understanding of the complex anatomy and physiology of the structures of the head and neck. Such oper-

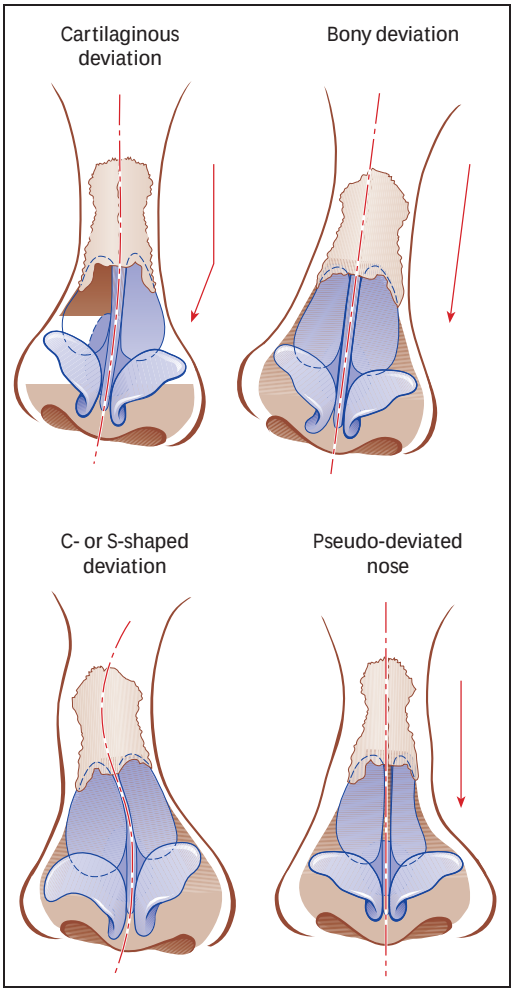


Fig. 2.125 Types of nasal deviation.

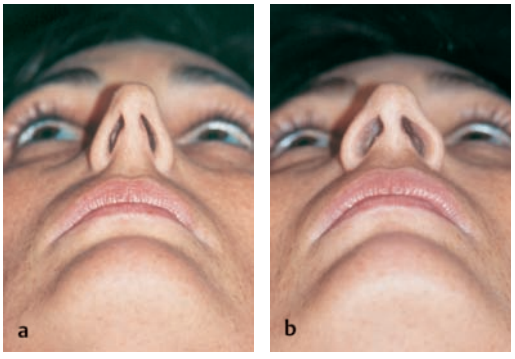


Fig. 2.126a, b a Functional tension nose, with obstruction of the nasal valve.
b Enlargement of the nasal valve angle by shortening the height of the nasal septum.

ations therefore have to be performed by surgeons who are well trained in this area and who are able to combine preservation of function with reconstruction. The principles of regional plastic surgery also have to be taken into account in preoperative planning of reconstruction when a mutilating resection has to be performed.

Priority in treatment of defects in the head and neck regions is given to reconstructing them as soon as possible. The British plastic surgeon Sir Harold Gillies (1882–1960) stated the principle "Replace like with like." Although that is not always feasible, the principle emphasizes that autologous tissue should be used to the greatest possible extent for closure of defects.

Numerous reconstructive procedures are available, but they can be reduced to a few basic ones that can be modified and combined, depending on the type and site of the defect. This applies to planning and execution of the incision, wound closure, and wound care. Defects in overlying soft tissue can be closed using several different operations, depending on the site and size. A fundamental principle is that the relaxed skin tension (RST) lines need to be observed when making incisions, including incisions for taking flaps; in the face, the direction of most of these lines is perpendicular to that of the muscles.

Simple Incisions

Z-plasty can alter the direction of tension on a scar, pulling it more in the direction of the main RST lines. Continuous Z-plasty can be used to achieve "optical resolution" of visible scars (Fig. 2.127).

VY-plasty can be used to achieve elongation or slackening of an area of skin.

Free Transplants

In principle, autologous grafting is the method of choice for the face, particularly the nose. Free skin grafts of various thicknesses are used to cover superficial defects in the skin at different sites. A distinction is made between full-thickness and split-skin grafts (Fig. 2.128).

Composite grafts have the advantage that they are able to replace two layers of skin and one of cartilage (Fig. 2.129). They are taken from the auricle and are used for reconstruction of the nasal alae or the columella, for example (Fig. 2.130a–c).

Grafting of autologous or homologous tissue can be used for structural reconstruction—of a saddle

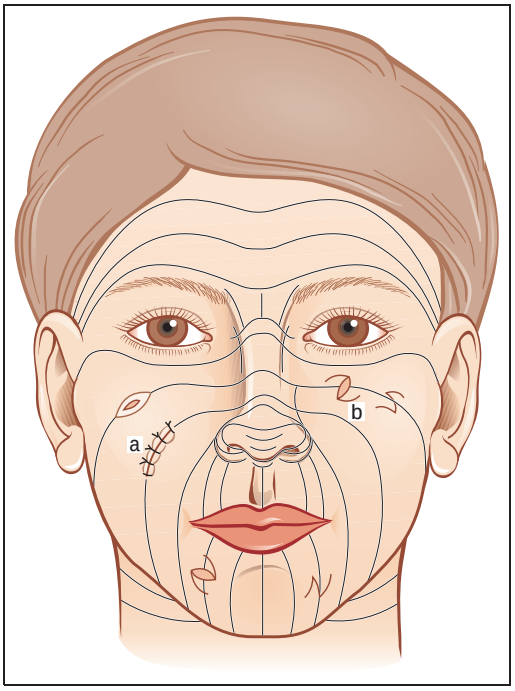


Fig. 2.127 Relaxed skin tension lines and the principles of scar correction (a) and Z-plasty (b).

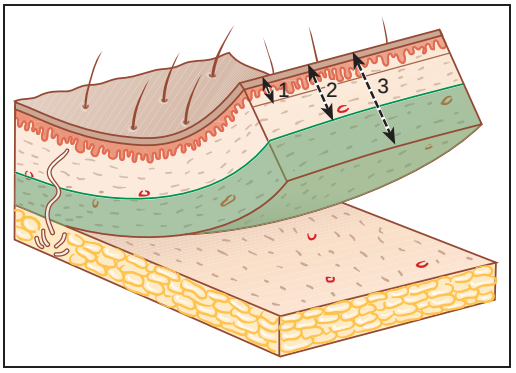


Fig. 2.128 Different types of free skin graft. 1, Thiersch graft; 2, split skin graft; 3, full-thickness graft.

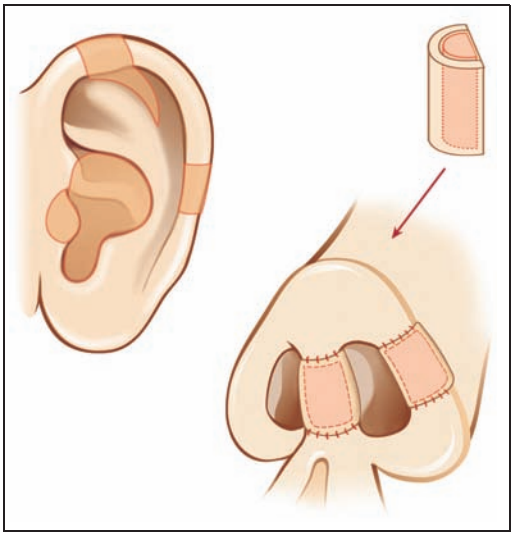


Fig. 2.129 Composite graft from the auricle for reconstruction of an alar nasal defect.

nose, for example—as a *structural graft*, or to fill contour defects as so-called *contour grafts*. Auricular cartilage is particularly well suited as a biomaterial for this, as it is available in a wide variety of shapes. Alternatives include rib cartilage and fascia. Alloplastic implants may either be enclosed in fibrin or overgrown by connective tissue; silicone remains biologically inert, whereas porous Gore-Tex is incorporated into the body tissues.

Flaps

Local flaps: As they are developed from the tissue immediately adjacent to the defect, the circulation in local flaps is intact, so that they can be set into their final position using advancement or rotation techniques.

Advancement flaps: These must run parallel to the relaxed skin tension lines. Good elasticity of the skin is a prerequisite. Excision of *Burow triangles* at the base of the flap prevents buckling due to excess tissue. With regard to the blood supply, a distinction is made between *random-pattern* and *axial-pattern flaps* (Fig. 2.131a, b). Examples are subcutaneous pedicled gliding flaps and island flaps. Advancement flaps have the drawback that they develop tension in a longitudinal direction; they must not be used if there is tension, particularly on the eyelid, lip, or nasal ala.

Transposition flap: The base of the flap is located immediately adjacent to the defect. This type of graft is particularly suitable for covering defects in the glabella, canthus, or lower third of the nose. Examples are bilobed flaps and the Limberg flap, in which the primary defect is trimmed to form an equilateral rhombus (Figs. 2.132a–d, 2.133a, b, 2.134a–d).

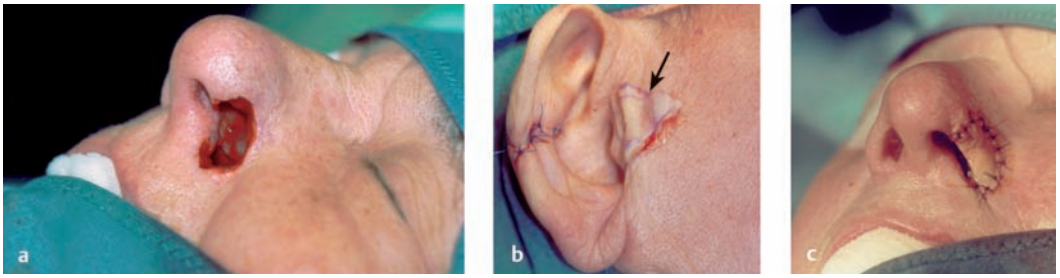


Fig. 2.130a–c Composite graft (arrow in b) for defect repair.

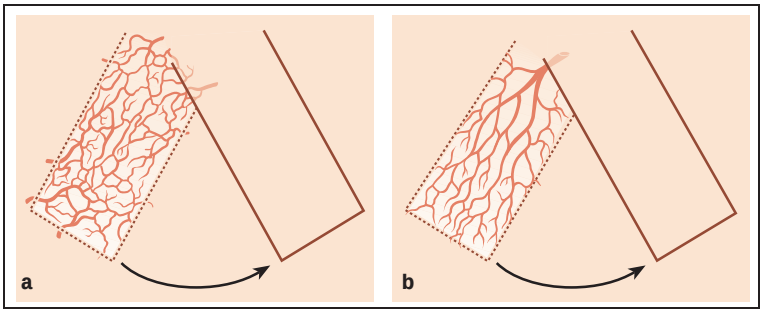


Fig. 2.131a, b a Random-pattern flap (with random blood supply from the area). b Axial-pattern flap (with blood supply from a single longitudinal artery).

Rotation flap or sliding flap: The advantage of this type of flap is its broad base, guaranteed blood supply, and great variability of design. It is suitable for covering defects of the cheek, lateral neck region, and forehead, for example (Fig. 2.135a–c).

Pedicled regional flaps: By virtue of their axial blood supply, these flaps can bridge long distances for covering defects. An example is the paramedial forehead flap (Fig. 2.136a–d).

Distant flaps: In the form of pedicled distant flaps or tunnel grafts (also called tube flaps or rope flaps), this type of graft is an important technique for covering defects caused by oncologic operations. Examples are the deltopectoral graft (Fig. 2.137) and the latissimus dorsi flap.

Microvascular grafts: The radial forearm graft is the prototype of a free fasciocutaneous neurovascular graft with microvascular anastomosis to cover defects of the face or neck. The advantages of this transplanted flap are that it has large blood vessels and good vascularization (Fig. 2.138).

Customized flaps: These flaps are not available in normal anatomy. They range from enlargement of the skin surface (possibly including several tissue appendages) using an expander to embedding of cartilage for reconstruction of deformities of the auricle.

Prostheses

In certain cases, it may be more reasonable to replace a defect with a prosthesis rather than embarking on protracted multistage reconstructive procedures—e. g., after total loss of the auricle or the orbital contents.

Tumors of the Nose and Sinuses

Benign Tumors

These are relatively unusual in the nasal cavity and sinuses. In addition to rare tumors, benign tumors include osteoma, ossifying fibroma, papilloma, hemangioma, lymphangioma, chondroma, fibroma, and giant cell tumor.

Osteoma

This condition is relatively common in the frontal and ethmoid sinus, but rare in the maxilla and sphenoid bones. Its etiology is unknown; there is an association with Gardner syndrome.

Clinical Features. Headache and pressure in the head predominate. Recurrent sinusitis or mucocele may occur due to obstruction of drainage. Displacement of the eye and later intracranial complications may arise. Many osteomas are asymptomatic and are found incidentally.

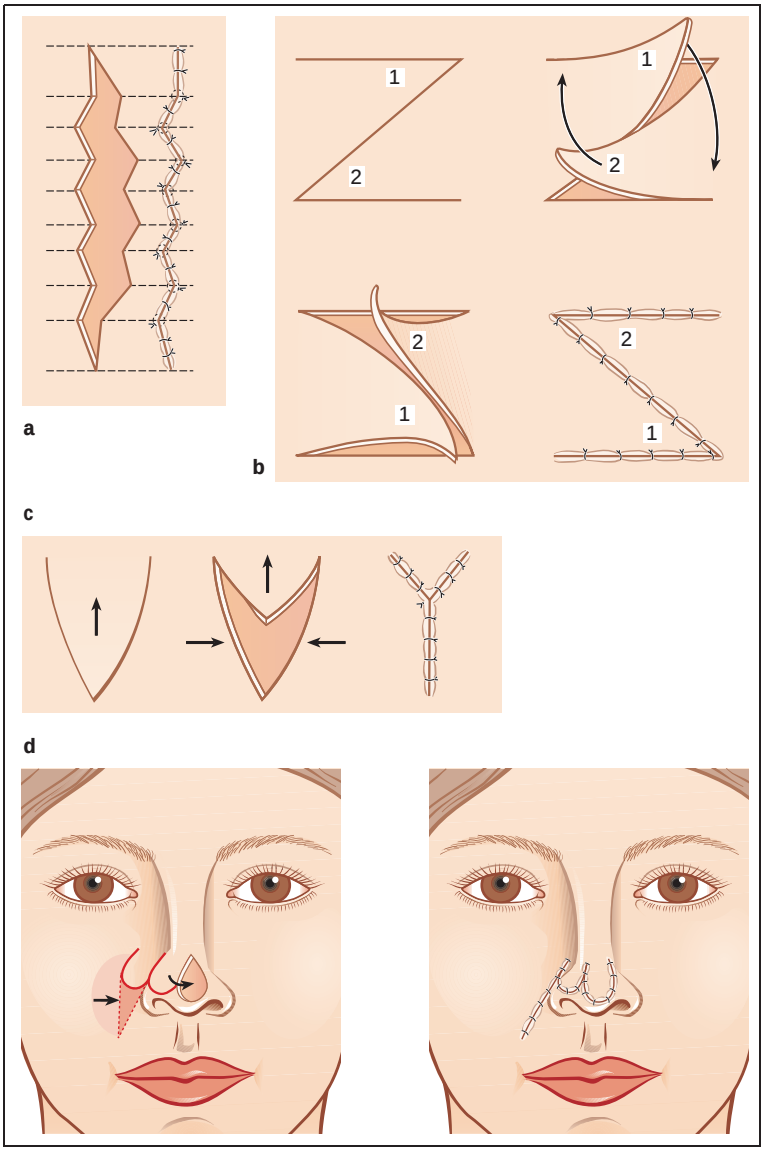


Fig. 2.132a–d Techniques used in plastic surgery of the face.
a Continuous W-plasty for correction of a scar.
b Tissue transposition with a Z-plasty.
c Tissue elongation or release of tension with a VY-plasty.
d Coverage plasty using a bilobed flap.

Diagnosis. CT scans of the paranasal sinuses typically show a calcified, well-demarcated tumor, which can be diagnosed clinically and then treated by surgery if necessary.

The differential diagnosis needs to distinguish between osteomas (which often have a slender pedicle) and ossified fibromas (which involve extensive laminar bony metaplasia). Whereas removing an osteoma using appropriate methods is uncomplicated, ossified fibromas tend to recur (Fig. 2.139).

Treatment. Small asymptomatic tumors may not require intervention. Small medial tumors can be removed endoscopically. Larger osteomas of the frontal sinus invariably require external surgery. The tumor fragments should be sent for histology (see Fig. 2.71a).

Metaplastic Osteogenesis

Bone is formed not only by osteoblasts normally present in bony tissue, but also by cells derived from pluripotent precursors (usually fibroblasts)

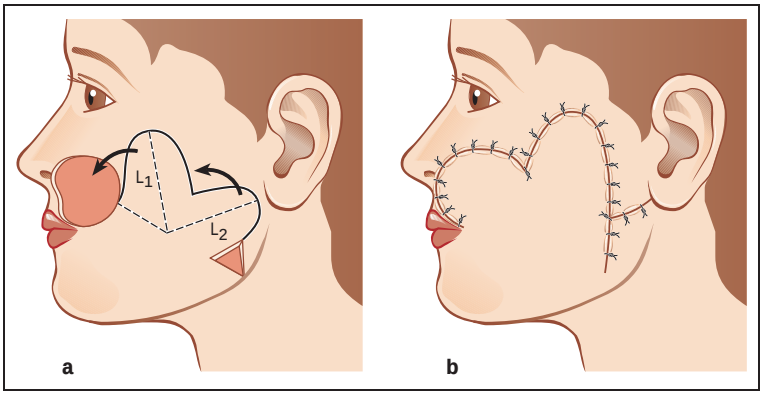


Fig. 2.133a, b Coverage of a defect with a bilobed flap.

Fig. 2.134a–d Reconstruction of a large defect with a bilobed flap following resection of a malignant skin tumor.
a Defect.
b Dermoplasty with alloplastic material (after histologic control of wound margins).
c Three days postoperatively.
d Six months later.

that have undergone transformation (metaplasia) into osteoid-producing osteoblasts. Bony metaplasia occurs in numerous tumors of bony and soft tissues—e.g., fibrous dysplasia, ossifying fibroma of the facial skeleton, or malignant fibrous histiocytoma.

Ossifying Fibroma

A more comprehensive term is "benign fibro-osseous lesion." This includes fibrous dysplasia, ossifying fibroma, and cementifying fibroma.

Ossifying fibromas are considered to be a localized form of fibrous dysplasia of the facial skeleton. They occur principally in the middle third of the face and at the base of the skull in children and adolescents.

Clinical Features. These include unilateral expansion of the facial skeleton, headaches, and possibly visual field loss and lesions of other cranial nerves.

Diagnosis. CT scans show a fairly radiopaque tumor arising from normal skeletal structures. A biopsy can also be performed.

Treatment. Usually only partial removal is possible. There is a tendency for the lesions to recur.

Papilloma

Inverted papillomas are the most common sinonasal papillomas, accounting for ≈ 70% of such tumors and ≈ 3% of all neoplasias of this region. Because of their strong tendency to recur, destructive growth, the possibility of multicentric lesions, and the danger of malignant transformation, surgical treatment of inverted papillomas is very challenging. A radical surgical procedure involving transfacial block resection via lateral rhinotomy or midfacial degloving was therefore favored for many years. Today, the high standard of endonasal endoscopic microsurgery makes it possible to remove papillomas with comparable results. Even lower rates of recurrence than after transfacial operations

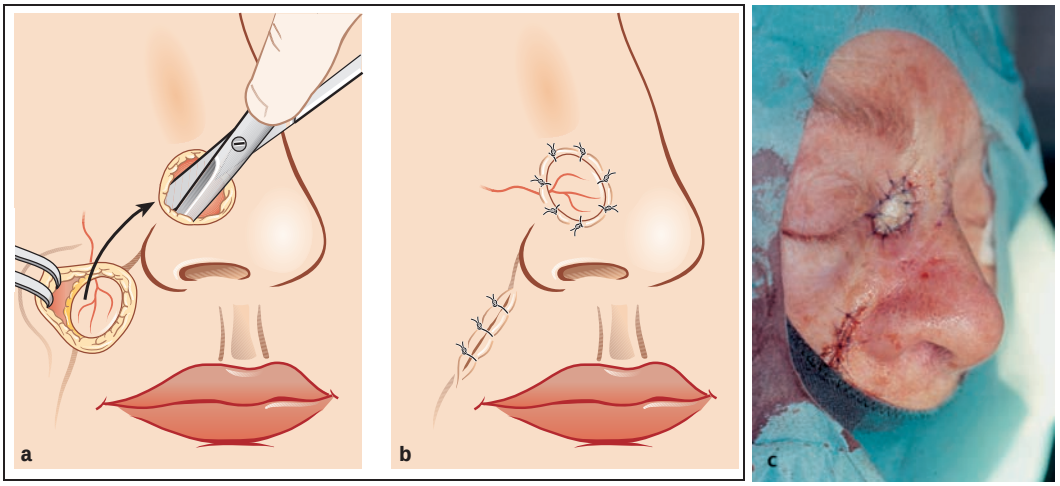


Fig. 2.135a–c Regional island flap transplantation.

Fig. 2.136a–d Reconstruction of the tip of nose using a forehead flap.
a Elevation of the flap.
b Temporary wound closure.
c Four weeks later, before severing the pedicle.
d Six months after the operation.

have been reported several times in the recent literature.

Clinical Features. Nasal obstruction and epistaxis are typical. Endoscopically, papillomas appear as solid tumors with a lobed surface. On CT scans, they are seen unilaterally more frequently than polyposis. Macroscopically, inverted papillomas are often indistinguishable from nasal polyps (Fig. 2.140).

Diagnosis. This is established by histological examination of biopsy material.

Note: Tumors of the nose and paranasal sinuses are always masked by mucosal edema and inflammation. Complete and careful inspection of all regions of the nose and lateral nasal wall is therefore essential.

Treatment. Because of the tendency to malignant degeneration and recurrence, surgical excision is indicated. Papillomas do not respond to radiotherapy.

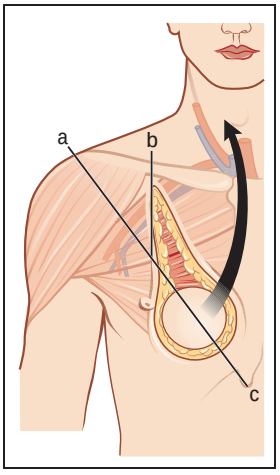


Fig. 2.137 Myocutaneous pectoralis major island flap. a–c, Acromion–xiphoid process line; b, midclavicular line.

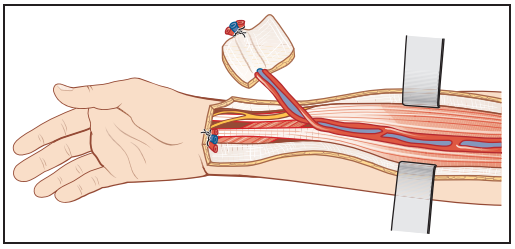


Fig. 2.138 Radial forearm flap for tissue grafting with microvascular anastomosis.

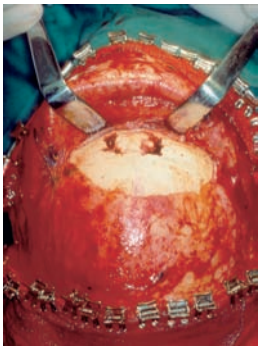


Fig. 2.139 A large osteoma of the frontal sinus; open approach, osteoplastic flap.

Fig. 2.140 Inverted papilloma in the nasal cavity—distinguishable from a polyp due to its solid, lobulated structure.

Glioma

This condition is described on page 288.

Nasopharyngeal Angiofibroma

This condition is described on page 288.

Malignant Tumors of the External Nose

Primary tumors of the external nose include *basal cell carcinoma, squamous cell carcinoma,* and *malignant melanoma. Senile keratoma* and *xeroderma pigmentosum* are included in the group of *precancerous lesions. Keratoacanthoma* is actually benign, but may be very difficult to differentiate from a squamous carcinoma.

Basal Cell Carcinoma

Basal cell carcinoma is a malignant epithelial skin tumor with a locally destructive growth pattern, but with a very low tendency to metastasize. It is the most common malignant cutaneous tumor worldwide in white populations, and its incidence is increasing. The most common exogenous risk factors are lifetime cumulative exposure to ultraviolet light and frequent sunburns. Further risk factors are chemical carcinogens, ionizing radiation, genetic predisposition, and immunosuppression. The tumor is predominantly located in the central face. Eighty-six percent of these tumors are localized on the head: 26 % on the nose, 19 % in the frontal or temporal area, 16 % on the cheek, 14 % near the eye, especially the medial canthus, and 11 % on the auricle. Basal cell carcinoma is the most common malignant tumor of the external nose (Fig. 2.141).

Clinical Features. Initially, there is a small single nodule with telangiectasia. The nodule often has a dimpled center, which grows slowly. Later, it ulcerates and infiltrates superficially into the underlying tissue. There is no predilection for either sex, and the tumor usually first appears between 60 and 70 years of age. The superficially recognizable skin lesion has a diameter of less than 1 cm in 70 % of patients, but extension into the surrounding tissue is very often considerable, with the tumor often occupying an area of more than 20 cm^2. A true basal cell carcinoma does not metastasize, and transition to squamous cell carcinoma is very rare. However, malignant degeneration may occur in tumors that have previously been irradiated or insufficiently

excised, producing a very aggressive malignancy that may seed to other sites (e.g., the lung) via the bloodstream.

Clinical types of basal cell carcinoma:

- Nodular type: convex, shiny nodules, often with central ulceration.
- Rodent ulcer: ulcerating basal cell carcinoma; convex tumor with flat ulcerations.
- Invasive basal cell carcinoma: rapid, deep invasion over a wide area.
- Sclerodermiform basal cell carcinoma: atrophic lesions, spreading to the corium, with a bead-like limbus.

Note: The extent of spread is always greater than it appears clinically.

- Superficial basal cell carcinoma: flat, spreading, often light brown, squamous plaques with a finely beaded edge.

Diagnosis. The most common site is the center of the face and the auricle. Diagnosis should be confirmed by biopsy (punch biopsy) if there is any doubt as to the diagnosis.

Differential diagnosis. Squamous carcinoma and precancerous lesions must be considered.

Treatment. The treatment of first choice is complete, histologically controlled excision.

The initial treatment should consist of complete excision with a histologically controlled margin. The defect is then closed with an alloplastic skin graft (e.g., an alloplastic synthetic skin substitute for temporary wound coverage). Reconstruction using regional rotational or transposition flaps or a free graft is performed after complete tumor resection has been confirmed histologically. In selected cases of superficial basal cell carcinoma, radiotherapy, topical immunotherapy with imiquimod (5% cream), or photodynamic therapy may be therapeutic alternatives.

Mohs surgery: Frederic E. Mohs (1910–2002) described a special technique of skin tumor surgery in 1941. The Mohs technique consists of stepwise removal of small layers of skin along with immediate microscopic control of the skin cancer margins. The advantage of this technique is that it allows a controlled tumor resection with minimal resection volume and wound surfaces. Frozen skin sections are used for microscopic analysis.

Fig. 2.141 Basal cell carcinoma: a centrally ulcerated, raised nodule.

Prognosis. This is good if the initial resection is complete, with histologic checking of the margins and immediate further resection if necessary. Basal cell carcinoma does not metastasize unless it undergoes malignant degeneration.

Squamous Cell Carcinoma

This is the second most common tumor of the external nose.

Clinical Features. After beginning as a precancerous lesion or as a nodular, firm, nonhealing lesion of the skin, squamous cell carcinoma then grows rapidly, soon ulcerating to form a crater. Regional lymph-node metastases occur relatively early.

Diagnosis. The most common site is the lower third of the face. The diagnosis is confirmed by biopsy and lymph-node staging.

Differential diagnosis. This includes precancerous lesions, keratoacanthoma, and basal cell carcinoma.

Treatment. Histologically controlled excision of the tumor is the treatment of choice. Other treatment options are radiotherapy and cryotherapy. Laser or photodynamic therapy and polychemotherapy are only indicated in inoperable tumors.

Prognosis. This is favorable if the tumor is removed immediately with histologic control, and depending on the lymph-node stage.

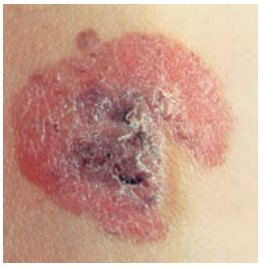


Fig. 2.142 A superficially spreading melanoma, 6 mm in diameter.

Malignant Melanoma

The most common malignant skin tumor, malignant melanoma affects females, with a 2 : 1 preponderance over males. The maximum incidence is between the twentieth and sixtieth years of life. Ten percent of malignant melanomas occur on the head and neck, affecting the face and the scalp.

Clinical Features. The tumor often arises from a pigmented mole, but also may arise from apparently normal skin.

Suspicious symptoms include an increase in surface area, increased prominence, and darkening of color of a pigmented mole, possibly with a blackberry type of surface, formation of satellites in the neighboring tissue, or enlargement of the regional lymph nodes. *Clinical forms* include:

- Superficial spreading melanoma (60%): irregularly pigmented spots and plaques with blurred and polycyclic borders.
- Primary nodular malignant melanoma (20%): regular, dark brown, rarely pink or skin-colored nodules lacking a halo.
- Malignant lentigo (≈ 10%) (Fig. 2.142).
- Acrolentiginous malignant melanoma (≈ 5%), which is rarely found on the head and neck.
- Primary nodular malignant melanoma (≈ 40%).

The most unfavorable type is nodular melanoma, which grows deeply and metastasizes rapidly from the start, whereas superficial spreading melanoma, malignant lentigo, and acrolentiginous melanoma initially spread horizontally on the surface. The site is often the face, or scalp and head, or the ear and neck. It is rare on the external nose, but more common on the anterior part of the septum, where it leads to epistaxis, nasal obstruction, and formation of crusts. The tumor can also occur on the oral and pharyngeal mucosa.

Diagnosis. A dermatologic consultation is necessary. Biopsy should be avoided, as it may activate and spread the tumor. Total removal with a wide margin of healthy tissue is indicated immediately on well-founded suspicion of malignant melanoma. Amelanotic melanomas also may occur. There are also numerous pigmented tumors that are not malignant melanoma, and an absolute diagnosis without biopsy therefore is very difficult.

Sentinel node: If the tumor is more than 1 mm thick, excision of the sentinel node is recommended. The sentinel node is a regional lymph node that is fed directly by the lymphatic vessel draining the tumor. Radioisotope scans of lymphatic drainage and the histologic findings are of critical importance for prognosis and treatment.

Differential diagnosis. This includes juvenile melanoma, papilloma, telangiectatic granuloma, pigmented basal cell carcinoma, and blue nevus.

> **!** Note: Manipulation of a malignant melanoma should be avoided if at all possible, and immediate surgery is indicated.

Treatment. The mainstay of treatment is rapid and extensive excision by three-dimensional surgery with a macroscopic margin of 5 cm, although this cannot always be obtained on the face. Frozen sections of the margins should be examined immediately so that further resection can be undertaken if necessary. The defects should be reconstructed immediately with advancement flaps or free skin grafts. Conservative lymph-node surgery or radical neck dissection is indicated, depending on the histology of the sentinel node, at least for high-risk malignant melanomas of the head and neck. However, the value of this is controversial, because of occasional widespread early metastasis. Supportive immunotherapy, chemotherapy, or endolymphatic radioisotope therapy by a dermatologist or radiotherapist may be helpful. Convincing benefit has not been shown with conventional radiotherapy, immunotherapy with bacillus Calmette–Guérin (BCG), or currently available forms of chemotherapy.

Prognosis. This depends on early diagnosis and is determined by Clark levels (Fig. 2.143) of growth depth and the presence or absence of metastases. With adequate early surgery in stage I, the 5-year survival rate is 70%, and in

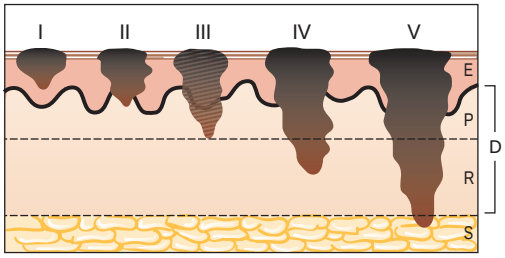


Fig. 2.143 Invasion levels of primary malignant melanoma of the skin as defined by Clark. Level I, limited to the epidermis; level II, into the underlying papillary dermis; level III, to the junction of the papillary and reticular dermis; level IV, into the reticular dermis; level V, into the subcutaneous fat. E, epidermis; D, dermis; P, papillary dermis; R, reticular dermis; S, subcutaneous tissue.

stage II, 30–40 % lower. The poor prognosis depends less on the superficial extent of the lesion than on its total thickness, depth, and metastatic spread. A depth of extension of 0.76 mm marks the boundary between high-risk and low-risk malignant melanoma.

■ Malignant Tumors of the Nasal Cavity and Nasal Sinuses

The most common tumors of the nasal cavity and nasal sinuses are *squamous cell carcinoma, adenoid cystic carcinoma,* and *adenocarcinoma.*

Mesenchymal tumors such as spindle cell sarcoma, round cell sarcoma, chondrosarcoma, osteosarcoma, and malignant lymphoma, are rare. Malignancies arising in the nasal cavity or sinuses represent less than 1 % of all malignant tumors. Histologically, the relative incidence is as follows: squamous cell carcinoma, ≈ 57 %; adenocarcinoma and similar forms, ≈ 18 %; undifferentiated carcinomas, ≈ 10 %; and mesenchymal tumors (malignant lymphoma, sarcoma, and melanoma), ≈ 15 %.

In children, histiocytosis X (eosinophilic granuloma) and rhabdomyosarcoma are relatively common.

Olfactory Neuroblastoma

This is the most typical olfactory tumor, originating from deep inside the olfactory epithelium. This tumor is malignant, aggressive, and forms metastases. The incidence curve of the tumor shows two peaks, one in the second and one in the sixth decade of life. Tumors in patients between 20 and 30 years of age have a stronger tendency to metastasize.

Clinical Features. Loss of olfaction and taste, obstruction of nasal breathing, bloody or blood-tinged secretions, and—depending on the extent of the tumor— headache or CSF leakage may occur.

Therapy. The therapy of choice is radical surgery combined with irradiation. One of the main factors affecting the prognosis is the histological grade (types I–IV). Tumors with an intracranial localization—i. e., mainly meningiomas—or tumors of the olfactory sulcus may also affect the olfactory system, as may tumors of the medial and posterior sections of the frontal base. Surgical treatment of such tumors requires a combination of otolaryngologic procedures and neurosurgical techniques, to provide optimal access to the cribrous lamina and ensure a high degree of radical functional surgery.

Symptoms of Malignant Tumors of the Nose or Sinuses

Tumors of the nasal cavity are often clinically silent for a long period, making early diagnosis often difficult.

Suspicious symptoms, especially in persons over 50 years of age, include unilateral nasal obstruction, unilateral chronic nasal discharge, hemorrhagic nasal secretion, fetid secretion, loss of sensation of one of the branches of the trigeminal nerve, swelling of the cheek or of the medial canthus, pressure in the head or face, and a feeling of fullness in the nose. Marked headache only occurs in later stages, when the tumor extends to the dura, causing treatment-resistant pain. Signs of invasion of neighboring tissue include restriction of eye movements, displacement of the eye or proptosis, swelling of the soft tissue of the medial canthus, the eyelids, the cheek, the palate, and the alveolus, loosening of the teeth, interference with mastication, cranial nerve palsies, epiphora, and regional lymphadenopathy.

Prognosis. The further cranially and dorsally a tumor of the paranasal sinuses is located (Fig. 2.144), the poorer the prognosis. This is the implication of the levels defined by Sebileau and the plane of Ohngren, although these are rarely used today.

Diagnosis. This is based on the history and on inspection and palpation of the oral cavity, nose, and

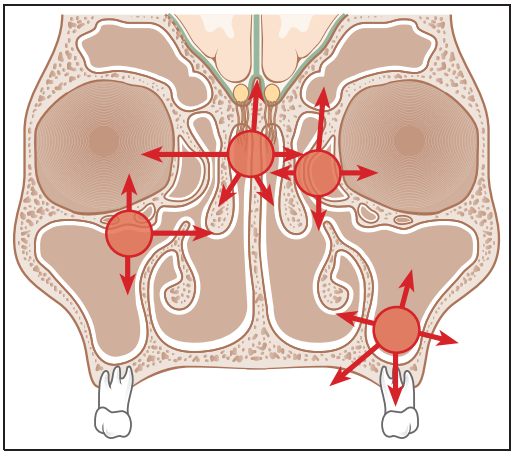


Fig. 2.144 Typical directions of extension of tumors of the nose and sinuses.

oropharynx. Endoscopy can also be used. Axial radiographs of the sinuses and of the base of the skull show a diffuse opacity destroying and obscuring the normal bony contour. CT scans are also obtained. The patient should be examined by a neurologist and an ophthalmologist. If necessary, a biopsy is taken after the suspect sinus has been opened. If invasion of the base of the skull is suspected, angiography should be performed and the CSF examined. Regional lymph-node metastases and distant metastases to the lung, skeleton, brain, and liver are looked for.

Differential diagnosis. This includes infection of the sinuses and the associated complications, and benign tumors (see pp. 186–193).

The TNM system is used for staging to determine treatment and prognosis and to allow recording and analysis of the results (Table 2.14).

Table 2.14 TNM classification: nasal cavity and paranasal sinuses

Maxillary sinus	
T1	Mucosa
T2	Bone erosion or destruction, hard palate, middle nasal meatus
T3	Posterior bony wall maxillary sinus, subcutaneous tissues, floor or medial wall of orbit, pterygoid fossa, ethmoid sinuses
T4a	Anterior orbit, cheek, pterygoid plates, infratemporal fossa, cribriform plate, sphenoid or frontal sinuses
T4b	Orbital apex, dura, brain, middle cranial fossa, cranial nerves other than V2, nasopharynx, clivus
Nasal cavity and ethmoid sinus	
T1	One subsite
T2	Two subsites or adjacent nasoethmoidal
T3	Medial wall or floor orbit, maxillary sinus, palate, cribriform plate
T4a	Anterior orbit, skin of nose or cheek, anterior cranial fossa (minimal), pterygoid plates, sphenoid or frontal sinuses
T4b	Orbital apex, dura, brain, middle cranial fossa, cranial nerves other than V2, nasopharynx, clivus
All sites	
N1	Ipsilateral single ≤ 3 cm
N2	a. Ipsilateral single ≥ 3 cm to 6 cm b. Ipsilateral multiple ≤ 6 cm c. Bilateral or contralateral ≤ 6 cm
N3	>6 cm
Distant metastasis	
MX	Distant metastasis cannot be assessed
M0	No distant metastasis
M1	Distant metastasis

Principles of Management of Malignant Tumors of the Nose and Paranasal Sinuses

Three methods of treatment are available: surgery, radiotherapy, and chemotherapy (cytotoxic drugs, administered intra-arterially if required). In some situations, combinations of the three methods may be appropriate.

Surgery. Three standard methods of removing the tumor are in use: 1, partial maxillectomy and, if necessary, resection of the frontal bone; 2, total maxillectomy; and 3, exenteration of the orbit, combined with procedures 1 or 2. The principles are shown in Fig. 2.145a–c. The operation is determined by the type, site, and size of the tumor (Fig. 2.146).

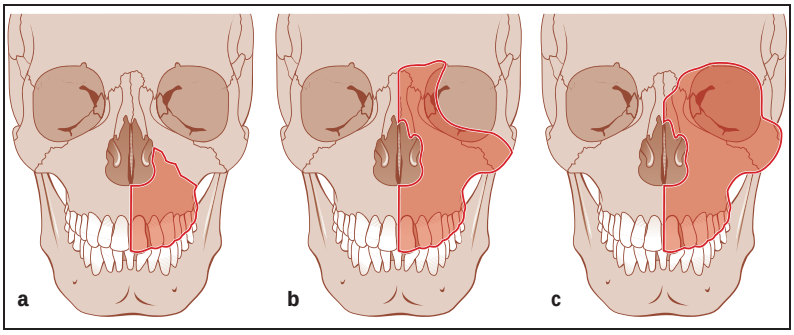


Fig. 2.145a–c Resection of the upper jaw.
a Partial resection.
b Total resection.
c Total resection with exenteration of the orbit.

Since the operation almost always needs to be extensive, secondary reconstruction or provision of a prosthesis to cover both external and palatal defects is necessary.

Only ≈ 10 % of these patients have lymph-node metastasis, and only 5 % have hematogenous metastasis. Suspicious lymph nodes must be sought out, using MRI and sonographic monitoring. Neck dissection is usually performed when lymph-node metastases are suspected, or for certain other oncologic considerations.

Radiotherapy. This can be administered alone or following surgery. The survival rate following radiotherapy of the nasal cavity and the sinuses as the sole treatment for squamous carcinoma is not as good as that for surgery, depending on the stage and site of the disease. Radiotherapy is therefore usually reserved for highly radiosensitive tumors—e. g., mesenchymal tumors—and for inoperable tumors. For all other tumors, a combination of surgery and radiotherapy, possibly supplemented by chemotherapy, is preferred. As a rule, the operation precedes radiotherapy, but preoperative radiotherapy can also be used, particularly for very large tumors. What is known as sandwich treatment—i. e., half the dose of radiotherapy, then surgery, followed by the second half of the radiotherapy—is also practiced. Radiotherapy is usually administered with cobalt-60 or supervoltage therapy (e. g., with the betatron) to deliver a tumor dose of 58–70 Gy. Radiotherapy may be the treatment of choice for certain small tumors of the external nose with no invasion of the skeleton.

The dose and volume of postoperative radiotherapy and the decision to combine radiotherapy with chemotherapy (chemoradiotherapy) depend on whether the resection is complete. If the tumor has been completely resected, all of its margins are histologically free of tumor (R0 resection, low risk). In an R1 resection (high risk), microscopic residual disease is left behind.

Fig. 2.146 Coronal CT, showing destructive tumor growth with invasion of the orbit. The orbital floor and parts of the orbital lamina have been destroyed by destructive tumor growth.

In brachytherapy, small radioactive rods are implanted directly into the tumor or placed next to the area requiring treatment. This ensures a high and selective tumor dose.

Chemotherapy. At present, chemotherapy still does not have an established role as a primary curative treatment for epithelial malignancies of the face, nose, and sinuses—especially with regard to the long-term results. Depending on the tumor stage and the histology of the tumor, an interdisciplinary, individual therapeutic strategy needs to be determined, including surgery, radiotherapy, chemotherapy, and combinations of these.

Prognosis. With suitable treatment, 5-year average survival rates of 40 % can be achieved. This is only an overall figure, and it may be more or less, depending on numerous oncologic factors.

3 Mouth and Pharynx

Applied Anatomy and Physiology

Basic Anatomy

Oral Cavity

The oral cavity is bounded anteriorly by the lips, posteriorly by the anterior faucial arch, inferiorly by the floor of the mouth, and superiorly by the hard and soft palates. It is continuous with the oropharynx through the anterior faucial arch (Figs. 3.1, 3.2a).

The faucial arch and the base of the tongue form the *faucial isthmus.* The oral cavity is divided into two parts by the upper and lower alveolar process and the teeth: first, the *vestibule of the mouth,* lying between the lip and cheek on one side and the teeth and the alveolar process on the other and second, the *oral cavity proper,* limited externally by the alveolar process and the teeth.

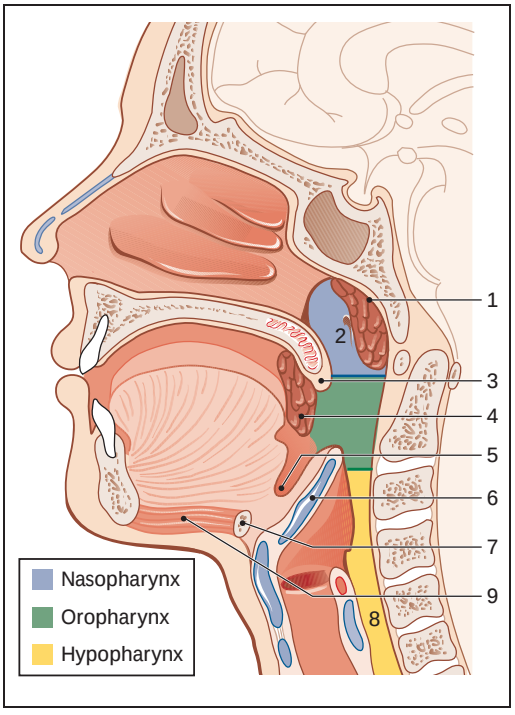


Fig. 3.1 Anatomy of the mouth and pharynx. 1, Roof of the nasopharynx; 2, ostium of the eustachian tube; 3, soft palate; 4, palatal tonsil; 5, vallecula; 6, epiglottis; 7, hyoid bone; 8, hypopharynx; 9, floor of the mouth.

The *vestibule of the mouth* communicates directly with the oral cavity proper on both sides, even when the teeth are in apposition, between the ascending ramus of the mandible and the last molar tooth.

This is of practical importance: in intermaxillary wiring, the patient is able to ingest a fluid diet using this route, even when the teeth are fixed in occlusion.

The *tongue* fills the oral cavity almost completely when the mouth is closed (Fig. 3.2b, c). The surface tension force ensures that the tongue adheres to the hard and soft palate, thus maintaining closure of the mouth.

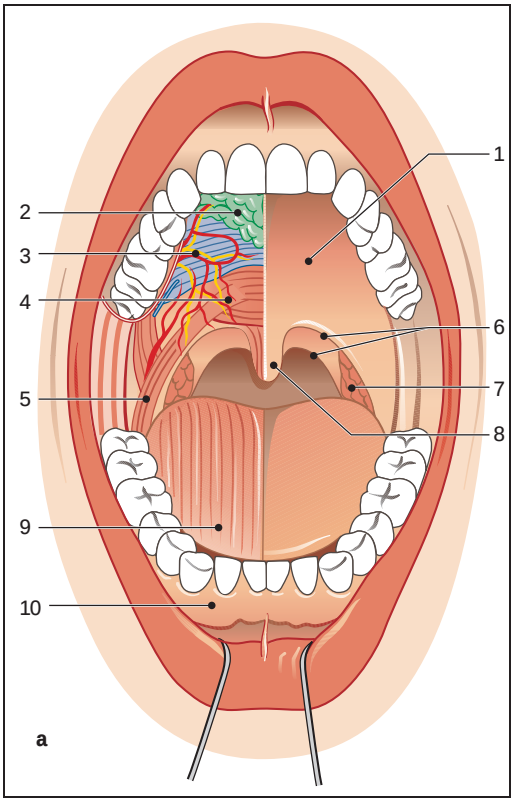
The following parts of the tongue are distinguished: the *tip,* the *margins,* the *body,* the *base,* the *dorsum,* and the *ventral surface*.

The dorsum of the tongue is covered with a modified epithelium containing the filiform papillae at the tip, the fungiform papillae at the tip and margins, the foliate papillae on the posterolateral part of the tongue, and the vallate papillae on the dorsum. The boundary between the body of the tongue and the base of the tongue is formed by the V-shaped *terminal sulcus,* the central point of which is the foramen cecum, a remnant of the thyroglossal duct.

The base of the tongue contains the *lingual tonsil,* which can be a site of inflammation and abscess due to an impacted foreign body and can cause mechanical difficulty in swallowing when it is hypertrophic. The base of the tongue is limited inferiorly by the edge of the *epiglottis.* The two *valleculae* lie in the angle between the epiglottis and the base of the tongue. They are sometimes difficult to inspect and may be a site for cysts, foreign bodies, and malignant tumors. In supine unconscious patients or patients under general anesthesia, the base of the tongue may fall backward to occlude the entrance to the larynx, and together with the epiglottis may lead to respiratory obstruction. This is prevented by

pulling the tongue forward and by introducing an oropharyngeal airway.

The arrangement of the musculature of the tongue provides it with extreme mobility. Two groups of muscles can be distinguished: firstly, those without any bony attachments, running free in the body of the tongue—i. e., the transverse, superior, and inferior longitudinal muscles, and the vertical muscles; and secondly, muscles that are attached to fixed points—i. e., the styloglossus, the genioglossus, the hyoglossus, and the palatoglossus muscles (Fig. 3.3).



The *floor of the mouth* is formed mainly by the mylohyoid muscle, which is stretched between the U-shaped mandible like a diaphragm and which is inserted into the hyoid bone and the median raphe. On the *oral surface,* with the tip of the tongue elevated, the plica sublingualis with the sublingual caruncle can be found on both sides of the *lingual frenulum*.

In the caruncle and the immediate neighborhood lie the efferent ducts of the submandibular gland, the submandibular duct (Wharton duct), and the sublingual gland and sublingual duct (Bartholin duct). The efferent duct of the parotid gland (Stensen duct) opens into the cheek at the level of the second upper molar, and that of the anterior lingual gland (Blandin gland) in the region of the fimbriated fold on the ventral surface of the tongue.

The *mandible* consists of two separate bones at birth, but these consolidate to form one bone during the first year of life. Figure 3.4a shows the most important anatomic details and the typical sites of fracture. The third branch of the trigeminal nerve runs in the body of the mandible, together with the blood vessels that supply the lower teeth. The mandibular nerve enters the mandible at the mandibular foramen and exits at the mental foramen.

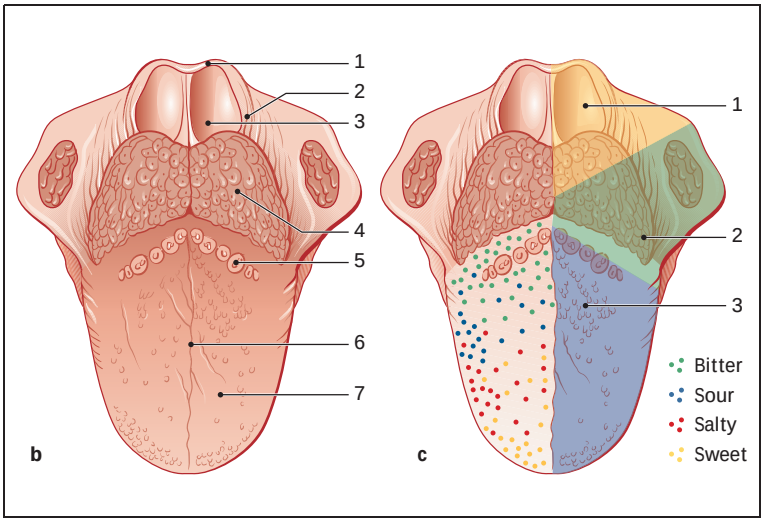


Fig. 3.2a–c Oral cavity and tongue.
a Topographic anatomy of the oral cavity. 1, Hard palate; 2, palatine glands; 3, palatine arteries and nerves; 4, veli palatine muscles; 5, palatoglossus; 6, palatoglossal arches; 7, palatine tonsil; 8, uvula; 9, tongue; 10, gingiva.
b Anatomy of the tongue. 1, epiglottis; 2, lateral glossoepiglottic fold; 3, epiglottic vallecula 4, lingual tonsil; 5, vallate papillae; 6, median sulcus of the tongue; 7, body of the tongue.
c Innervation and distribution of qualities of taste on the tongue. 1, Vagus nerve (X); 2, glossopharyngeal nerve (IX); 3, lingual nerve (mandibular nerve, V3).

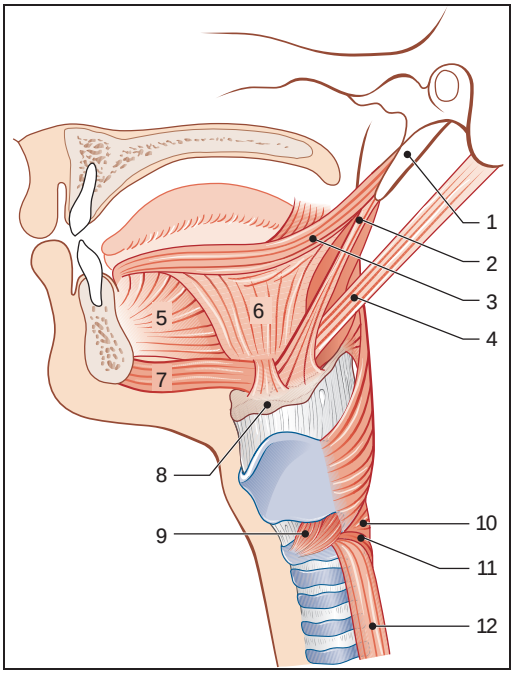
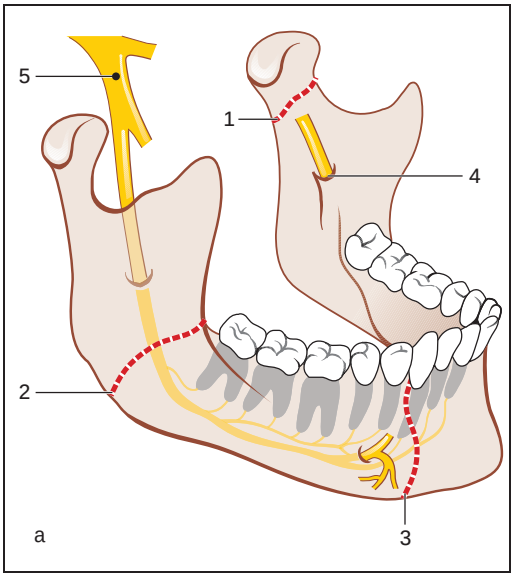


Fig. 3.3 Muscles of the tongue and pharynx. 1, Styloid process; 2, stylohyoid muscle; 3, styloglossus muscle; 4, digastric muscle; 5, genioglossus muscle; 6, hyoglossus muscle; 7, geniohyoid muscle; 8, hyoid bone; 9, cricothyroid muscle; 10, Killian triangle; 11, inferior part of the cricopharyngeus; 12, esophagus.

The *temporomandibular joint* is of considerable clinical interest, as it may be involved in dental diseases, in trauma of the facial skeleton and skull, in otologic diseases, and in generalized arthropathies. In addition, it may be the cause of headache in Costen syndrome. Figure 3.4b shows the anatomic relationships. Its proximity to the auditory meatus and mastoid, the lateral part of the base of the skull, the parotid gland, and the lateral wall of the oropharynx and nasopharynx should be noted.

The *epithelial lining of the oral cavity* consists of nonkeratinized stratified squamous epithelium that is thickened in certain points such as the alveolar edges and hard palate, where it unites with the underlying periosteum to form a mucoperiosteum. Subepithelial collections of minor salivary glands are found all over the oral cavity, and are more common in some parts than others (see Fig. 7.2).

Vascular supply. The external carotid artery supplies the tongue via the lingual artery; the floor of the mouth via the sublingual artery; the cheek via the facial artery; and the palate via the ascending pharyngeal artery and descending palatine artery. The latter arises from the internal maxillary artery. Venous drainage is through the veins of the same names to the facial vein, the pterygoid venous plexus, and the internal jugular vein. There is also



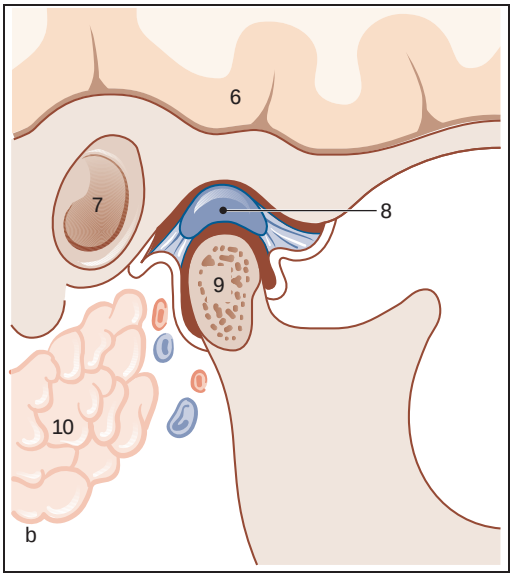


Fig. 3.4a, b a Mandible with typical fracture lines. 1, Fracture of the neck; 2, fracture of the angle of the jaw; 3, fracture of the chin; 4, mandibular foramen; 5, mandibular nerve.

b Temporomandibular joint and surrounding structures. 6, Middle cranial fossa; 7, external auditory meatus; 8, articular disk; 9, head of the mandible; 10, parotid gland.

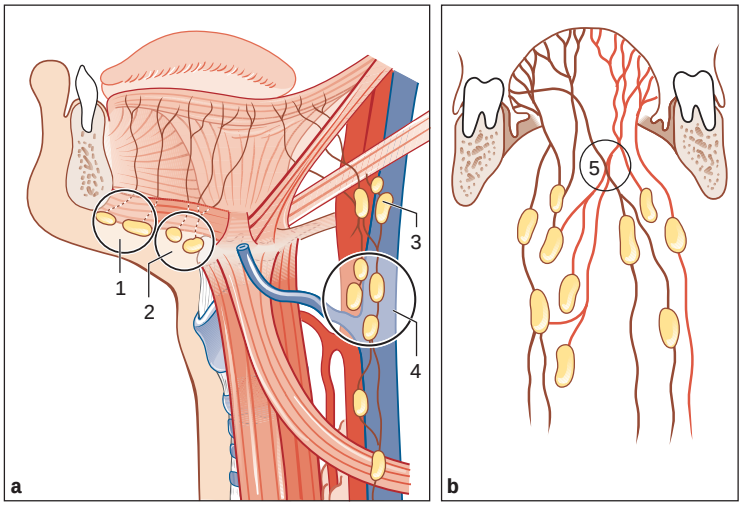


Fig. 3.5a, b Lymph drainage of the tongue.
a Groups of lymph nodes. 1, Submental; 2, submandibular; 3, upper deep cervical, with lymph nodes at the superior venous angle (4).
b Contralateral lymphatic drainage of the tongue. 5, Crossing of the lymphatic drainage.

a connection to the cavernous sinuses via the pterygoid plexus.

The lymph drains via the regional submental, submandibular, and parotid nodes to the internal jugular chain. The lymph drainage of the base of the tongue and the floor of the mouth is to the same side *and* to the opposite side (Fig. 3.5a, b). This is important for the development of contralateral lymph-node metastases (see pp. 285, 399).

Nerve supply. The tongue derives its motor supply from the hypoglossal nerve. Its sensory supply comes from the lingual nerve and the vagus nerve, which innervates the posterior part of the base of the tongue. Taste fibers come from the glossopharyngeal nerve to the base of the tongue and from the chorda tympani fibers of cranial nerve VII, accompanying the lingual nerve for the anterior two-thirds of the tongue (see Fig. 3.2c).

The floor of the mouth derives its motor supply from the mylohyoid branch of the mandibular nerve and its sensory supply from the trigeminal nerve. Parasympathetic secretory fibers from the salivary glands are supplied from the chorda tympani and branches of the submandibular ganglion. Sympathetic fibers for blood vessels of the glands come from the carotid plexus.

The masticatory muscles derive their motor supply from the mandibular branch of the trigeminal nerve, but the buccinator muscle is supplied from the facial nerve.

The teeth of the upper jaw receive their sensory supply from the maxillary nerve and those of the lower jaw from the mandibular. These are both branches of cranial nerve V.

The temporomandibular joint derives its nerve supply from the auriculotemporal branch of the mandibular nerve.

The soft palate derives its motor innervation from the glossopharyngeal, vagus, and trigeminal nerves, and probably the facial nerve as well.

■ Nasopharynx, Oropharynx, and Hypopharynx

The pharynx is a muscular tube, 12–13 cm long in the adult; it narrows from above downward, is covered with mucosa, and is divided into three compartments, each of which has an anterior opening (Fig. 3.6).

The *nasopharynx* is limited superiorly by the base of the skull and inferiorly by an imaginary plane through the soft palate, and it opens into the nasal cavity. The most important anatomic structures are as follows: anteriorly, the choanae; superiorly, the floor of the sphenoid sinus; posterosuperiorly, the adenoid; laterally, the pharyngeal ostium of the eustachian tube and the cartilaginous torus tubarius, immediately posterior to which is the Rosenmüller fossa (pharyngeal recess) and the tubal tonsil; and anteriorly and inferiorly, the soft palate. The embryonic pharyngeal bursa (Fig. 3.7) may persist in the posterior wall of the nasopharynx, causing chronic inflammation and retention of secretions. The posterior wall of the nasopharynx is

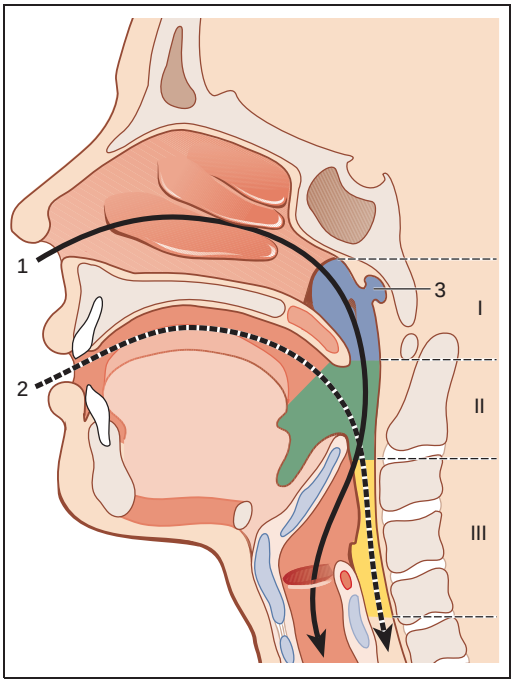


Fig. 3.6 Divisions of the pharynx. I, Nasopharynx; II, oropharynx; III, hypopharynx. Crossing of the upper airway (1) and the upper food passage (2). Site of the pharyngeal bursa (3).

separated from the spinal column by the tough prevertebral fascia, which lies on the longus capitis muscles, the deep muscles of the neck, and the arch of the first cervical vertebra.

The shape and width of the nasopharynx show marked individual variation. The epithelial lining consists of respiratory ciliated and stratified squamous epithelium, with transitional epithelium at the junction with the oropharynx.

The *oropharynx* extends from the horizontal plane through the soft palate described above to the superior edge of the epiglottis (Fig. 3.7) and is continuous with the oral cavity through the faucial isthmus. It contains the following important structures: the posterior wall, consisting of the prevertebral fascia and the bodies of the second and third cervical vertebrae; the lateral wall, containing the palatine tonsil with the anterior and posterior faucial pillars; and the supratonsillar fossa, lying above the tonsil between the anterior and posterior faucial arches.

The valleculae (see Fig. 3.1), the base of the tongue, the anterior surface of the soft palate, and the lingual surface of the epiglottis are usually described as being part of the oropharynx.

The epithelial lining consists of nonkeratinizing stratified squamous epithelium.

The *hypopharynx* extends from the upper edge of the epiglottis superiorly to the inferior edge of the cricoid cartilage (see Fig. 3.6). It opens anteriorly into the larynx. On each side of the larynx lie the funnel-shaped piriform sinuses. Important anatomic structures and relationships include: on the anterior wall, the marginal structures of the laryngeal inlet and the posterior surface of the larynx; on the lateral wall, the inferior constrictor muscle and the piriform sinus, the latter being bounded medially by the aryepiglottic fold and laterally by the internal surface of the thyroid cartilage and the thyrohyoid membrane. Immediate relationships of the hypopharynx at the level of the larynx include the common carotid artery, the internal jugular vein, and the vagus nerve. Relationships of the posterior wall, apart from the pharyngeal constrictor muscle (see below), include the prevertebral fascia and the bodies of the third to the sixth cervical vertebrae. Inferiorly, the hypopharynx opens into the esophagus, the boundary being the superior sphincter of the esophagus. The epithelial lining consists of nonkeratinized stratified squamous epithelium.

The muscular tube of the entire pharynx consists of two layers with different functions:

1. A circular muscle layer consisting of the three pharyngeal constrictor muscles: the superior constrictor, inserted into the base of the skull; the middle constrictor, inserted into the hyoid bone; and the inferior constrictor, inserted into the cricoid cartilage (see Figs. 3.3, 3.7). Each of these funnel-shaped muscular segments is overlapped at its lower end by the segment below. All of the segments are inserted posteriorly into a tendinous median raphe.

   The inferior constrictor muscle is of particular clinical importance. It is divided into a superior thyropharyngeal part and an inferior cricopharyngeal part. Figure 3.7 shows how the triangular dehiscence *(Killian triangle)* is formed from the posterior wall of the hypopharynx between the superior oblique and the inferior horizontal fibers. A pharyngoesophageal pouch (Zenker diverticulum) may develop at this weak point in the hypopharyngeal wall.

2. Raising and lowering of the pharynx are also carried out by three paired muscles radiating into the pharyngeal wall from outside. These are the stylopharyngeus, the salpingopharyngeus, and the palatopharyngeus muscles. The stylohyoid and styloglossus muscles are also responsible for elevation. A true longitudinal muscle does not occur in the pharynx and only begins at the mouth of the esophagus. The ability of the pharynx to slide over a distance of several centimeters is due to the existence of fascial spaces (parapharyngeal and retropharyngeal) filled with loose connective tissue. The importance of these tissue spaces in the spread of infection is described on p. 270 and in Fig. 3.45a, b.

Vascular supply of the pharynx. The *arterial supply* is provided by the ascending pharyngeal artery, the ascending palatine artery, the tonsillar branches of the facial artery, branches of the maxillary artery (i.e., the descending palatine artery), and branches of the lingual artery. These all arise from the external carotid artery. *Venous drainage* is via the facial vein and the pterygoid plexus to the internal jugular vein.

The *lymphatic drainage* is either via an inconstant retropharyngeal lymph node and then to the deep jugular lymph nodes, or directly to the latter group. The inferior part of the pharynx also drains to the paratracheal lymph nodes and is thus connected to the lymphatic system of the thorax. See also page 231.

Nerve supply of the pharynx. The individual pharyngeal muscles gain their motor supply from the glossopharyngeal (IX), vagus (X), hypoglossal (XII), and facial (VII) nerves. The nasopharynx derives its sensory nerve supply from the maxillary division of the trigeminal nerve (V), the oropharynx from the glossopharyngeal nerve (IX), and the hypopharynx from the vagus nerve (X) (see p. 231).

Lymphoepithelial System of the Pharynx

Note: The term “lymphoepithelial tissue” is used to indicate the close symbiosis of epithelial and lymphatic cells on the surface of a mucosa.

The epithelial and subepithelial tissue is loosely arranged so that lymphatic cells can enter it in large numbers (“reticulated epithelium”). The reticulohistiocytic system, more commonly known as the reticuloendothelial system (RES), with its storage cells, is strongly represented in lymphoepithelial tissue. Figure 3.8 shows the principle of a lymphoepithelial unit. Solitary units of this type, solitary follicles, are found in all parts of the mucosa. The epithelium is also diffusely interspersed with lymphocytes.

A very distinct collection of lymphoepithelial tissue, the Waldeyer ring, lies at the opening of the upper aerodigestive tracts.

The lymphoepithelial organs are called *tonsils.* From above downward, the following are distinguished:

1. The *pharyngeal tonsil,* the adenoids, which is single and lies on the roof and posterior wall of the nasopharynx

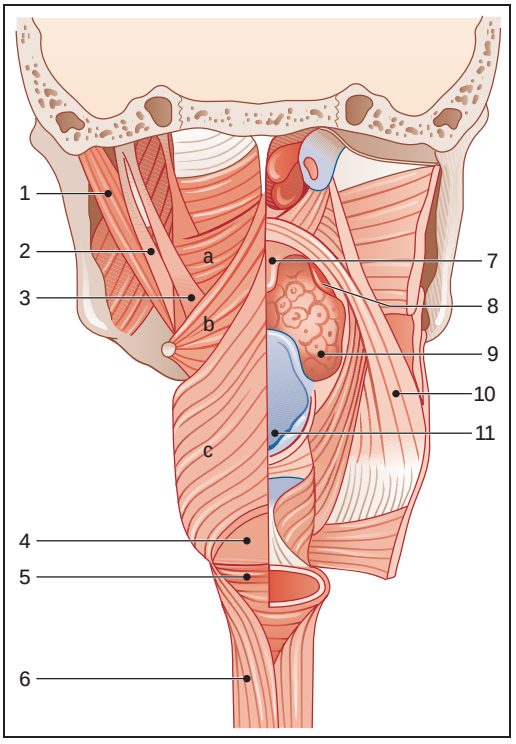


Fig. 3.7 Pharyngeal musculature. 1, Digastric muscle; 2, stylohyoid muscle; 3, stylopharyngeus muscle; 4, Killian triangle; 5, inferior part of the cricopharyngeus muscle; 6, esophagus; 7, uvula; 8, palatine tonsil; 9, tongue; 10, palatopharyngeus muscle; 11, epiglottis; a, superior pharyngeal constrictor muscle, b, middle pharyngeal constrictor muscle, c, inferior pharyngeal constrictor muscle.

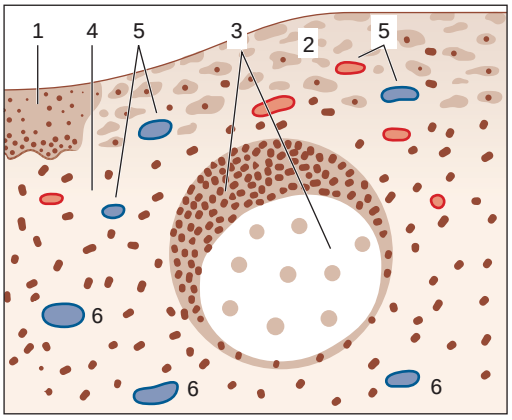


Fig. 3.8 Lymphoepithelial tissue. 1, Continuous squamous epithelium; 2, reticular epithelium; 3, secondary nodes with light centers and a dark zone of small lymphocytes; 4, basic lymphoid tissue; 5, arterioles and venules; 6, postcapillary veins.

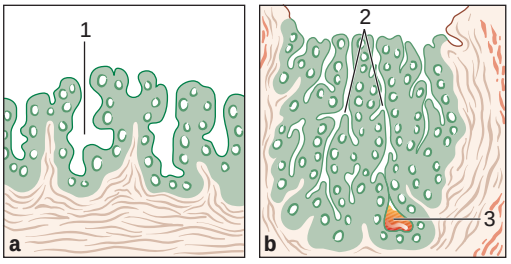


Fig. 3.9a, b The nasopharyngeal tonsil and adenoids (a), and the palatine tonsil (b). 1, Tonsillar lacunae; 2, tonsillar crypts; 3, cryptic abscess.

2. The *tubal tonsil,* which is paired and lies around the ostium of the eustachian tube in the Rosenmüller fossa
3. The paired *palatine tonsil,* lying between the anterior and posterior faucial pillars
4. The *lingual tonsil,* which is single and lies in the base of the tongue.

Less constant and obvious are:

5. The *tubopharyngeal plicae,* lateral bands, which run almost vertically at the junction of the lateral and posterior walls of the oropharynx and nasopharynx
6. Lymphoepithelial collections in the laryngeal ventricle

Unlike lymph nodes, lymphoepithelial organs only have efferent lymph vessels and do not have afferent vessels. Differences in the pathology and physiology of the individual collections of lymphoid tissue are due to their different structures. Figure 3.9 shows the structure of a palatine tonsil and of the adenoids.

The fine structure of a tonsil (Figs. 3.8, 3.9a, b) is in principle as follows: the soft-tissue lamellae or septa arise from a basal connective tissue capsule. These serve as a supporting framework in which blood vessels, lymphatics, and nerves run. This fan-shaped supporting framework considerably increases the active surface of the tonsil, as it holds the actual lymphoepithelial parenchyma. In the palatine tonsil, the active surface is sunk into the mucosa, whereas in the adenoids, it projects above the surface. The broad flat niches opening into the oral cavity caused by infolding are called *lacunae,* and the branching clefts running throughout the entire substance of the tonsil are called *crypts.* The actual tonsil tissue consists of a collection of a very large number of the lymphoepithelial units described above (see Fig. 3.8). The crypts usually contain cell debris and round cells, but may also contain bacteria and colonies of fungi, collections of pus, and encapsulated microabscesses. (See p. 266 for the description of chronic tonsillitis.)

The tonsils of the Waldeyer ring are present at the embryonic stage, but they only acquire their typical structure with secondary nodes during the postnatal period—i. e., after direct contact with environmental pathogens. They begin increasing rapidly in size between the first and third years of life, with peaks in the third and seventh years.

They involute slowly as of early puberty. Like the rest of the lymphatic system, they atrophy with increasing age.

The arterial blood supply to the pharyngeal tonsil is provided by various branches of the external carotid artery, including the facial artery and/or the ascending palatine artery, the ascending pharyngeal and lingual arteries, and possibly direct tonsillar branches.

The veins of the pharyngeal tonsil usually drain via the palatal vein to the facial vein, and from there to the jugulofacial venous angle of the internal jugular vein. There is also drainage via the pterygoid venous plexus to the internal jugular vein. This route provides a possible pathway of spread for infection from the tonsils to the cavernous sinus (see Figs. 3.45a, b; 3.46).

Physiologic and Pathophysiologic Principles

Several functional systems are collected in the mouth and pharynx, including the masticatory system, the swallowing apparatus, the taste organs, the lymphoepithelial ring, pregastric digestion, and articulation. In addition, the respiratory and digestive tracts cross in this area (see Fig. 3.6). This requires a reliable reflex protective system. An important prerequisite for this is a well-functioning autonomic and voluntary nervous supply to this region, and a mucosa adapted to this dual function. The mouth is only involved in respiration as a supplementary measure (see p. 232); continuous mouth breathing causes considerable local damage and can also affect the entire body. Typical functional disturbances due to defects of the nervous supply are summarized in Table 3.1.

Eating, Preparation of Food, and Swallowing

Normal feeding requires a normal masticatory apparatus and teeth, masticatory muscles, and temporomandibular joint. The function of the cranial nerves also has to be normal (see Table 3.1). The preparation of food serves to reduce the size of the food bolus by chewing and to moisten the food with saliva, of which 1.0–1.5 L are produced daily. The saliva lubricates the mucosa and makes the food capable of being swallowed. In addition, the enzymes contained in saliva prepare the food by partial chemical decomposition for further digestion in the gastrointestinal tract.

The results of quantitative or qualitative defects of saliva are summarized on p. 416. Satisfactory moistening of the mucosa of the oral cavity and pharynx by saliva is also necessary for normal speech and for normal taste (see discussion of salivary function on p. 415).

The act of swallowing. This an extremely complex sequence of events that is controlled by several cranial nerves (trigeminal nerve, facial nerve with chorda tympani, glossopharyngeal nerve, vagus nerve, superior and inferior laryngeal nerves, hypoglossal nerve) and cervical nerves (cervical plexus, C1–C4). Central coordination is performed in the motoric cortex of the precentral gyrus, the amygdaloid body, nuclear regions of the hypothalamus, and the ventral tegmentum of the midbrain. The deglutition centers are located in the rhombencephalon of the reticular formation and in the medulla oblongata. The "pattern generator" coordinating the interactions of the nerves and muscles is inborn.

Phases of swallowing. *Oral preparation phase:* Food is chewed and mixed with saliva. The lips close, round off, and are retracted. The mandible and the tongue move in all directions, the cheeks tauten, and the soft palate faces anteriorly.

Oral phase: The bolus is pressed along the back of the tongue into the oropharynx, eliciting the swallowing reflex. The corresponding receptors are located in the palatoglossal arch and the base of the tongue.

Pharyngeal phase: This phase of the swallowing reflex consists of a succession of movements that include closure of the larynx and further transportation of the bolus through the pharynx to the esophagus.

Esophageal phase: This phase comprises transportation of the bolus by peristaltic waves through the esophagus and into the stomach.

Pathophysiologic aspects. Closure of the laryngeal inlet by the epiglottis is not strictly necessary for normal swallowing. A patient whose epiglottis has been removed—e.g., in tumor surgery—usually learns to swallow without difficulty. However, the sensory nerve supply to the hypopharynx and the laryngeal inlet from the superior laryngeal branch of the vagus nerve has to be intact, as well as the pharyngeal constrictor for reflex protection of the laryngeal inlet. Increased tone or spasm of the cricopharyngeus is a common autonomic disorder—e.g., in thyrotoxicosis or Plummer–Vinson (Paterson–Kelly) syndrome. This can cause the globus symptom of discomfort on swallowing at the level of the cricoid cartilage, true dysphagia (see Table 3.1), and possibly the development of a hypopharyngeal diverticulum.

Disturbances of swallowing may also occur in paralyses of one or more cranial nerves (especially cranial nerve X, but also nerves IX and XII), which affect the tongue, soft palate, or pharyngeal musculature.

Table 3.1 Differential diagnoses of dysphagia

Site	Causal disease
Oropharyngeal	*Inflammatory:* glossitis, abscess of the floor of the mouth, specific and nonspecific pharyngitis, tonsillitis, peritonsillar abscess, retropharyngeal and hypopharyngeal abscess, edema of the uvula and angioneurotic edema
	Neural: lesions of vagus, hypoglossal and glossopharyngeal nerves, glossopharyngeal and vagus neuralgia
	Mechanical obstruction: foreign body, hypopharyngeal diverticulum, pharyngeal stricture, styloid process syndrome, benign and malignant tumors
	Congenital anomalies: macroglossia, clefts of the lip and jaw and palate, cysts of the base of the tongue, lingual thyroid, hyoid anomalies, cervical cysts and fistulas (median and lateral)
	Miscellaneous: results of radiotherapy, xerostomia, humidifying disorders, sialadenitis, fractures of the upper and lower jaw and the hyoid, lesions of the masticatory muscles, burns and scalds, results of surgery
Laryngeal	*Inflammatory:* epiglottitis, specific and nonspecific laryngitis, perichondritis of the larynx
	Neural: neuralgia of the superior laryngeal nerve, paralysis of the superior laryngeal nerve
	Miscellaneous: laryngeal trauma (contusion, distortion, fracture), results of radiotherapy, laryngocele, benign and malignant tumor, foreign bodies, and results of surgery
Esophageal	*Inflammatory or traumatic:* esophagitis including reflux esophagitis, esophageal mycosis, esophageal trauma, caustic burns, results of trauma and surgery, strictures and stenoses, esophageal foreign bodies and esophageal perforation
	Disorders of motility: esophageal spasm, “upper achalasia” (spasm of the cricopharyngeus muscle), external compression by goiter, aortic aneurysm or tumors of the lung and mediastinum, achalasia (cardiospasm), scleroderma, results of vagotomy, esophageal varices, presbyesophagus
	Neighboring organs: goiter
	Congenital anomalies: diverticulum, megaesophagus, hiatus hernia, congenital esophageal stenosis, vascular abnormalities
	Tumors: benign or malignant esophageal tumors (the latter are more common)
Diseases of the cervical spine	Arthritis of the cervical spine, subluxation of the cervical spine, dislocation of the cervical spine, prolapsed disk, spondylolisthesis, cervical rib
Neurologic disorders	Amyotrophic lateral sclerosis, bulbar paralysis, pseudobulbar paralysis, bulbar poliomyelitis, polyneuritis, multiple sclerosis, syringomyelia, cerebral and cerebellar ischemia, thrombosis of the posterior inferior cerebellar artery or basilar artery, brain tumors and brain stem tumors, disorders of cerebrospinal fluid circulation, vertebrobasilar insufficiency, myasthenia gravis, chorea (Huntington–Sydenham disease), Parkinson disease, tabes dorsalis, diabetic and alcoholic neuropathy, Wallenberg syndrome, lead intoxication
General diseases	Infections, botulism, iron-deficiency anemia (Plummer–Vinson or Paterson–Kelly syndrome), megaloblastic anemia, agranulocytosis, tetany, tetanus, goiter, thyroiditis, hypocalcemia, leukemia, vitamin A and B_1 deficiency, mitral valve disease, aortic aneurysms
Skin diseases	Scleroderma, urticaria, lupus erythematosus, erythema multiforme, pemphigus, recurrent aphthous ulcers, hereditary epidermolysis bullosa, dermatomyositis
Autonomic dysphagia	Autonomic dysfunction, psychogenic overlay (globus hystericus)

Taste

The basic taste sensations are *sweet, salty, sour,* and bitter. A proposed fifth taste sensation is *umami* (a sensation of savoriness), which applies to specific glutamic acid receptors. All other tastes are mixed sensations in which the sense of smell is also integrated. Many foods are "tasted" by the olfactory nerve. Pure sensory nerve fibers to the tongue and oral mucosa may also be stimulated by sour or spicy foods.

The sensory organs for taste are the taste buds lying in the vallate papillae, foliate papillae, and fungiform papillae on the tongue and on the hard palate (Fig. 3.10), the anterior faucial pillar, the tonsil, the posterior pharyngeal wall, the esophageal orifice, and the buccal mucosa. The fine gustatory hair cells have to be bathed in saliva or other fluids to allow the sense of taste to be evoked. Figure 3.2c shows the topical arrangement on the tongue of the sites where the different taste qualities are recognized. The sensory nerve supply is provided peripherally by two nerves in particular: the chorda tympani, arising from cranial nerve VII and accompanying the lingual nerve from cranial nerve V, and the glossopharyngeal nerve, arising from cranial nerve IX.

The boundary of the area supplied by each individual nerve in the mouth and pharynx is still not fully agreed upon, but the following is generally accepted (see Fig. 1.20). The anterior half of the tongue is supplied by the ipsilateral chorda tympani via the nervus intermedius, with synapses in the sensory geniculate ganglion. The posterior third of the tongue and the walls of the oral pharynx receive their sensory supply from the glossopharyngeal nerve. It is probable that the vagus nerve also has sensory contributions to the epiglottis, the laryngeal inlet (aditus), the upper part of the esophagus, and possibly also a small part of the center of the base of the tongue. Gustatory sensations from the soft palate are transmitted by the palatine nerves via the pterygopalatine ganglion, the greater petrosal nerve, the geniculate ganglion, and the nervus intermedius to the medulla oblongata.

The boundary between the area supplied by the lingual nerve and the accompanying chorda tympani and the glossopharyngeal nerve is still not generally agreed on.

Reflex reactions may affect the sense of taste, as they do the sense of smell. These include alterations in the quantity and quality of salivary secretion, the production of gastric juice, and interference with the course of the swallowing act.

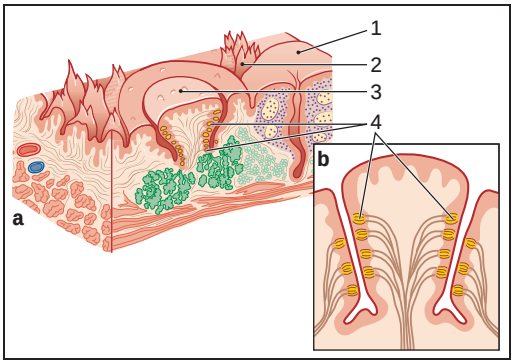


Fig. 3.10a, b Structure of the sense of taste on the tongue. 1, Lingual tonsil; 2, filiform and fungiform papillae; 3, vallate papilla; 4, taste buds.

Basic pathophysiology. Although sensory disorders of taste are less common than those of smell, they occur more commonly than is generally thought. Apart from direct, accidental, inflammatory, viral, or iatrogenic causes, they may also be due to neural lesions, certain drugs (see below), endocrine diseases, and deficiency diseases (Table 3.2). Disorders of taste may vary in degree and involve different qualities of taste (Table 3.3). Usually the threshold is increased—i. e., gustatory function is reduced. Reduction of the threshold is rare, but may occur in mucoviscidosis. The relation of the four senses of taste to each other may be disassociated.

Drugs that have side effects on the sense of taste include acetylsalicylic acid, biguanidine, carbamazepine, lithium, methylthiouracil, oxyfedrine, penicillamine, and phenylbutazone. Side effects may also occur after local use of ether oils, chlorhexidine, and hexetidine.

Function of the Tonsils

The tonsils have an immune-specific defense function. They send immunologically marked lymphocytes and immunoglobulins from the organism's "front line" to the various stations in the immune system. This is made possible by the way in which the tonsils are structured. In the crypts and reticular epithelium, intensive contact with the widest variety of antigens in the oral cavity is ensured. The tonsils form lymphocytes and plasma cells; T lymphocytes serve specifically for cellular immune defense, while immunoglobulins (e. g., IgA, IgE, IgM) and B lymphocytes are formed for humoral immunological reactions. At the same time, lymphocytes and immunoglobulins are released directly from

Table 3.2 Classification of disorders of taste

Quantitative taste disorders	
Hypogeusia	Reduced sensitivity—e. g., due to radiotherapy
Hypergeusia	Increased sensitivity—e. g., in glossopharyngeal neuralgia
Ageusia	Absence of the sense of taste. This may be partial, due to a lesion in the chorda tympani; total, due to toxins; or selective, as in "taste blindness" for certain substances
Qualitative taste disorders	
Parageusia	Faulty taste; may be due to virus infection
Cacogeusia	Unpleasant taste, occurring typically in cerebral sclerosis
Phantogeusia	Perception of taste sensations in the absence of a source
Gustatory hallucinations	May occur due to drug abuse, psychoses, and disorders of the central nervous system

Table 3.3 Definition of taste disorders

Epithelial origin	Damage to taste buds as a sequela of infection or radiation exposure, deficient oral hygiene, fungal infections, diabetes mellitus, Sjögren disease, adverse drug effects, hepatic and renal diseases, atrophic glossitis, salivary deficiency (e. g., due to antihypertensive drugs, antihistamines, antidepressants), burning mouth syndrome, hyperthyroidism, Cushing syndrome
Neural origin	Damage to cranial nerves VII, IX, X (e. g., following tonsillectomy, basal skull fractures, neurodegenerative disorders)
Central origin	Central nervous disorders of the taste fibers (e. g., posttraumatic anosmia/ageusia syndrome, brain tumors, brain stem lesions, neurodegenerative disease, temporal lobe epilepsy)

the tonsillar crypts into the oral cavity. The tonsils thus have both local and systemic immunological functions.

Basic pathophysiology. The increase in lymphoepithelial tissue during the early years of childhood development is explained by immunobiologic requirements. This increase in size is primarily only an expression of an active defensive function of the infant's organism to antigenic substances in the environment. During this period, tonsillar hyperplasia is therefore a welcome condition and in no way suggests excess inflammation. Since the tonsils lie at a narrow point in the respiratory and digestive tract, the nasopharynx and the faucial isthmus, an increase in their volume beyond a certain point leads to increased narrowing of the diameter of this essential pathway, to the detriment of the rest of the body (Fig. 3.11a, b). Obstructive sleep apnea may develop in extreme cases. Removal of the tonsils and adenoids is therefore justified in these circumstances, despite potential immunologic disadvantages. The palatine tonsils alone have slitlike, branching, poorly drained crypts permeating their entire substance. As long as these clefts drain freely into the oral cavity, the function of the tonsil is not endangered. However, if the physiologic content of the crypt stagnates due to

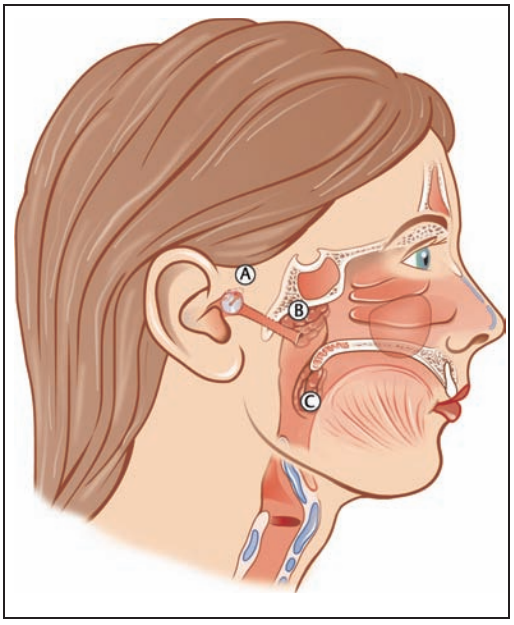


Fig. 3.11 Key points for clinical diagnosis in children. A, Otoscopy. Depending on their size and direction of growth, the adenoids (B) can block the nasopharynx and the pharyngeal ostium of the eustachian tube. Both the size and the extension of the tonsils (C) toward the posterior wall of the pharynx should be assessed. The easiest way of doing this is with an endoscope (4 mm, wide angle).

anatomic or infective stenosis, an ideal culture medium is set up for microorganisms. Colonies of bacteria or fungi become established, leading to chronic suppuration (cryptitis), small abscesses in the crypts, and superficial ulceration of the surface of the crypts. In anatomicopathologic terms, this represents *chronic tonsillitis*. This is in no way related to the size of the tonsil. Figure 3.8 shows how the superficial tonsillar capillaries are unprotected and course close to the lumen of the crypt, allowing relatively unhindered access for infective or toxic materials to the general circulation.

Formation of Sound and Speech

The oral cavity and pharynx make an important contribution to the timbre of the speech and voice, due to their action as a variable resonating space. In addition, the tongue, along with the palate, is necessary for the formation of consonants and vowels. Despite this, experience with tumor surgery shows that large parts of the tongue can be removed without loss of comprehensible speech.

With regard to nasality, it is still unclear whether closure of the nasopharynx during the articulation of consonants functions according to an "all or nothing" rule or whether it involves a graded mechanism of muscular obstruction by the soft palate.

Methods of Investigation

Inspection, Palpation, and Examination with the Mirror

The examination is carried out, with good illumination from a head lamp or head mirror, using two tongue depressors (see Fig. 2.14b, c; Figs. 3.12, 3.13). The following should be observed:

- The color and the normal symmetrical mobility of the lips, the condition of the skin and mucosa, and changes in the surface, ulcerations, induration, and tenderness of the lips are inspected.
- The arrangement of the teeth and the occlusion are examined with the lips open. The symmetry of the contour of the jaws, the mobility of the mandible, and the function of the temporomandibular joint are also examined.
- The shape and mobility of the tongue is examined with the mouth open. In hypoglossal paresis, the tongue deviates slightly to the paralyzed side. The floor of the mouth and the two caruncles are examined using a tongue depressor, with the tongue elevated. The surface and consistency of the tongue and articulation are also assessed.
- The properties of the mucosa of the mouth and cheeks are assessed, with particular attention to color, moisture, dryness, membranes, ulceration, tumors, and disorders of sensation.
- The condition of the hard and soft palate is examined. The innervation of the two sides is compared. In paralysis of the palate, the uvula deviates to the healthy side. The innervation of the pharyngeal musculature is tested.
- The upper and lower vestibule of the oral cavity are examined with a tongue depressor.
- The parotid duct in the cheek opposite the upper second molar tooth is inspected.
- The palatine tonsils, lingual tonsil, and mucosa of the posterior wall of the pharynx are examined using two tongue depressors (Fig. 3.13e). Normally, these structures should be pale yellow to pale pink, moist, and shiny. Dryness, coating, glazed crusts, and yellow streams of pus may be noted.

Fig. 3.12 Some traditional instruments for examining the mouth and pharynx. 1, Bruening tongue depressor; 2a, b, suction tubes for mouth and oropharynx; 3, Reichert hook; 4, long curved cotton applicator; 5, angled tongue depressor for the base of the tongue.

Examination of the tonsils: A tongue depressor is laid carefully with the left hand on the lateral part of the posterior part of the tongue, and the tongue is pressed gently downward. The spatula should not be placed on the base of the tongue, since this elicits the gag reflex. As soon as the tonsillar cleft

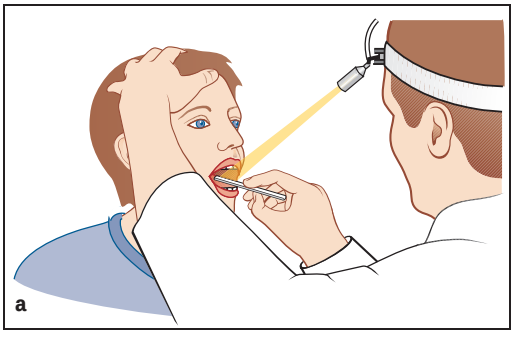
can be seen, the other hand is used to introduce the second tongue depressor between the ascending ramus of the mandible and the tonsil, and the edge of the tongue depressor is placed gently on the anterior faucial pillar, lateral to the tonsil, to dislocate it from its fossa into the oral cavity. An attempt is made to press material out of the visible crypt opening. The size of the tonsil and its connective tissue fixation to the tonsillar fossa, the color and properties of the surrounding mucosa on the faucial pillars, and the color and properties of the surface of the tonsil, including any exudate and the expressed contents of the crypt, are noted. Differences between the two sides are also looked for. Palpation of the lymph nodes now follows, with particular attention being paid to the nodes at the angle of the jaw and in the submandibular and submental areas (see Figs. 6.11, 6.16a–e).

Suspect areas in the oral cavity and base of the tongue should always also be palpated. The index finger, enclosed in a finger cot or glove, is used to palpate the suspicious area carefully for induration, infiltration, ulceration, and tender areas. Most patients tolerate careful examination. In patients with an exaggerated gag reflex, the oral and pharyngeal mucosa—particularly that of the soft palate, the base of the tongue, and the posterior wall of the pharynx—can be rendered insensitive first by applying a spray or a cotton applicator saturated with 1% tetracaine or lidocaine (Xylocaine). Local anesthesia of the pharynx is also recommended if satisfactory examination of the nasopharynx, hypopharynx, or larynx is not possible due to a marked gag reflex.

Note: Examination of the mouth with a tongue depressor or with the finger must be performed gently after the patient has received an explanation of the procedure. This is the only way to prevent gagging and allow a satisfactory view of the whole of the mouth and pharynx (Fig. 3.13).

The nasopharynx, the hypopharynx, and the larynx are now examined with the mirror (Fig. 3.14a). The technique of posterior rhinoscopy is described on page 132 and the technique of indirect laryngoscopy on page 298. The introduction of the loupe endoscope is described on page 298 (Fig. 3.14b).

Indirect mirror examination of the hypopharynx may be very difficult in patients with a protruding or infiltrated tongue base, if it is tender, or in those with a sensitive gag reflex. These patients are best examined using transnasal hypopharyngoscopy with a flexible endoscope. Flexible nasendoscopy (FNE) is the standard method now used by speech and language therapists.



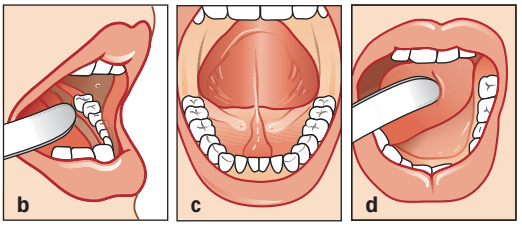


Fig. 3.13a–e Examination of the oral cavity.
a Situation during the clinical examination.
b Inspection of the buccal mucosa and parotid duct orifice, opposite the second upper molar.
c Inspection of the floor of the mouth and submandibular duct orifices.
d Evaluation of the lateral oral floor.
e Examination of the palatine tonsil with two tongue depressors.

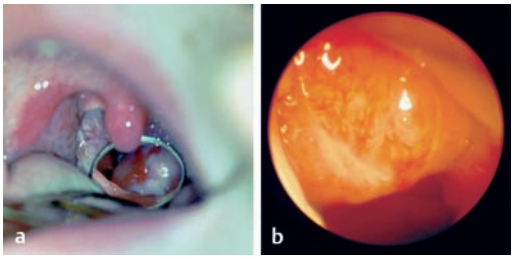


Fig. 3.14a, b a Examination of the nasopharynx with a mirror.
b Endoscopy of the nasopharynx in a child with adenoids.

Note: When there is clinical suspicion of a tumor, all lymph-node fields in the head and neck have to be carefully palpated (see p. 385) and appropriate imaging studies performed (B-mode ultrasonography, computed tomography, magnetic resonance imaging). During palpation of the neck, the head should be flexed toward the examination side, to relax the muscles and fascia (see Fig. 8.13).

Endoscopy

Various endoscopic techniques are available for examining the pharynx and hypopharynx. These include flexible transnasal endoscopy with local anesthesia, and rigid endoscopy in the clinical setting using suspension laryngohypopharyngoscopy (see Fig. 4.8) with the patient under general anesthesia.

Rigid endoscopy is an important diagnostic method in oncologic patients for evaluating the size, extension, and precise location of a tumor and for obtaining biopsies from margins of the lesion. The results are important for decision-making regarding the operability of a tumor, the surgical approach, and defect repair.

Imaging Studies

Tumors, enlarged adenoids, and other conditions in the *nasopharynx* are well displayed in a lateral view of the skull. This projection is also suitable for imaging the soft tissues of the nasopharynx.

Computed tomography (CT), magnetic resonance imaging (MRI; Fig. 3.15), and occasionally radionuclide scanning can be used for demonstrating the limits of a tumor or destruction caused by a nasopharyngeal tumor. Bilateral carotid angiography and superselective angiography to identify individual branches of the carotid artery are used to investigate nasopharyngeal angiofibromas, which are highly vascular, in preparation for embolization (Fig. 3.16; see also p. 242).

The *hypopharynx* is best demonstrated after administration of a contrast agent such as Gastrografin or barium. The swallow with contrast is very useful for diagnosing a pharyngeal pouch, stenoses, and swallowing disorders.

Lateral views of the neck and the upper thoracic region can be used to localize the site of radiopaque foreign bodies in the hypopharynx and upper esophagus. This projection is also of great value in inflammatory soft-tissue swelling and surgical emphysema of the parapharyngeal tissues due to pharyngeal injuries, pharyngeal abscess, mediastinal abscess, etc.

Radiographic demonstration of the salivary glands using sialography and scanning is described on page 418.

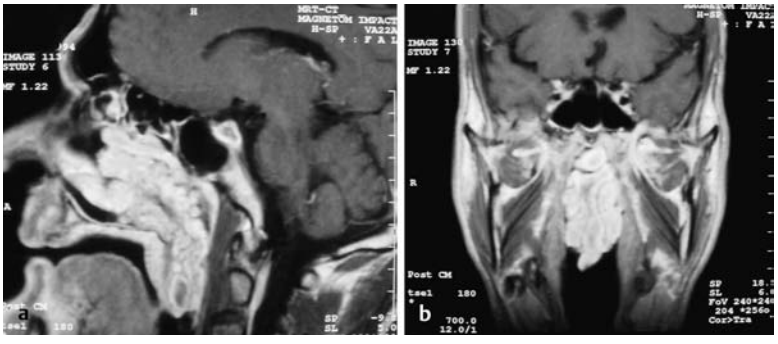


Fig. 3.15a, b A large nasopharyngeal angiofibroma (magnetic resonance images with contrast medium).

Fig. 3.16a, b Angiography of a nasopharyngeal angiofibroma. Before (a) and after (b) embolization of the tumor artery.

Examination of the Saliva

See page 416.

Gustometry

Taste can be tested by applying substances to the tongue that represent the four taste qualities—sweet, salty, sour, and bitter—at increasing concentrations, to assess the lowest concentration that can be recognized.

Examination of overall taste sensation. The three-drop method makes it possible to evaluate the threshold of recognition for the qualities sweet, sour, salty, and bitter. Börnstein's concentrations are used for testing. These are as follows: glucose 4%, 10%, 40%; sodium chloride 2.5%, 7.5%, 15%; citric acid 1%, 5%, 10%; quinine 0.075%, 0.5%, 1% (stale solutions should not be used). The sensation of taste is observed 0.5–4.0 s later, depending on the site tested, the temperature of the solution, and the size of the area tested. The test solution is applied alternately to the right and left sides of the tongue with a pipette, or better with a small piece of blotting paper ≈ 1 cm^2 in size. Confirming the threshold of recognition is usually satisfactory in ordinary practice.

Alternatively, test solutions of substances at concentrations slightly below or above the taste threshold can be administered in a defined sequence as one-drop tests. The sum of the recognized concentration levels of all four taste qualities is taken as a gustatory index. This test has high test–retest reliability—i. e., it is highly reproducible.

Assessing the ability to identify taste qualities. Oral application of sweet, sour, salty, and bitter test solutions, each at a concentration above the threshold that the patient is supposed to identify, is suitable for screening purposes. The test substances can be applied in solid form (known as tasties, taste strips, or wafers) or as fluids (drops, sprays). The patient is asked to identify each taste quality immediately after application.

Electrogustometry. Electric current can also be used to stimulate the taste receptors instead of test solutions. An anode current is used, with a normal threshold in adults between 2 and 7 µA. Electrogustometry has numerous advantages, but it is usually only used in specialist practices or hospital departments.

Objective gustometry can also be used in university hospitals. Reflex changes in the respiratory resistance of the nose or the electrical resistance of the skin are recorded simultaneously in response to a taste stimulus. Table 3.2 shows the most frequent causes of disorders of taste.

Specific Diagnostic Procedures

Bacteriologic, mycologic, and virologic culture. Culture is still the basis of treatment with anti-inflammatory drug agents. The diagnostic methods provided by microbiology, such as culture testing and sensitivity testing, can be frequently used.

Biopsy. Tissue needs to be taken for biopsy if a tumor is suspected and if abnormal findings remain unexplained. Tumors and lesions in the mouth and pharynx are easily accessible, and biopsy in this area is preferable to aspiration or cytology.

Clinical Aspects of Diseases of the Mouth and Pharynx

The main symptoms and disorders that indicate disease of the mouth and pharynx include:

- Pain on eating, chewing, or swallowing.
- Dysphagia (see Table 3.1).
- Globus symptoms (see p. 277 and Table 3.1).
- Burning of the tongue (see Table 3.7).
- Blood in the sputum.
- Oral fetor (see Table 3.5).
- Disorders of salivary secretion (see p. 416).
- Disorders of taste (see Table 3.2).
- Respiratory obstruction (see Table 6.2).
- Disorders of speech.
- Swellings on the head, neck, mouth, floor of the mouth, and swelling of the lymph nodes at the angle of the jaw (see p. 401).

Hyperplasia of the Lymphoepithelial Organs

The adenoid, the tonsil, and occasionally the lingual tonsil cause symptoms due to their size.

Hyperplasia of these organs is not in itself a disease, but only the morphologic expression of marked immunobiologic activity. A marked increase in the size of the tonsil produces primary mechanical obstruction of the respiratory or digestive tract and has detrimental effects on the entire body. Inflammation of neighboring organs is secondary. For this reason, tonsillar hyperplasia is here discussed separately from inflammation.

Adenoid Hyperplasia

Clinical Features. These include nasal obstruction, leading to mouth breathing; difficulty in feeding, especially in small children; noisy respiration; snoring; a typical adenoid face—i. e., dull facial expression, open mouth, dilated and flattened nasolabial folds, indrawn nasal alae, protruding upper incisor teeth; enlarged lymph nodes at the angle of the jaw or in the nuchal area; adenoid habitus; and rhinolalia clausa.

Obstruction of the nasopharynx may be responsible for:

1. *Aural diseases,* including obstruction of the eustachian tube, chronic tubal and middle ear catarrh, serous effusion, recurrent acute otitis media, formation of adhesions, and also progression of chronic otitis media and conductive deafness (see p. 58).
2. Diseases of the *nose and paranasal sinuses,* including chronic purulent rhinitis or sinusitis, and even pansinusitis.
3. *Disorders of the masticatory apparatus, including:* maldevelopment of the upper jaw—i. e., arched or "gothic" palate due to absence of the pressure of the tongue on the hard palate, and absence of lateral pressure on the upper jaw and alveolus by the tension of the buccinator muscle and the masticatory muscles because of the open mouth. Also including anomalies of position of the teeth, such as incorrect contact and orientation of the mandibular occlusion, and gingivitis.
4. *Disorders of the lower respiratory system*—i. e., chronic laryngitis, tracheitis, and bronchitis.
5. *Other somatic effects,* including a flat chest, round shoulders, thirst, loss of appetite, poor general development, and sensitivity to attacks of infection.
6. Effects on *intelligence and mental development* due to chronic respiratory obstruction and hypoxia during sleep; increased levels of CO_2 in the blood leading to restless, broken sleep and causing tiredness during the day, apathy, dullness, poor school performance, and "pseudodementia."

Pathogenesis. The disease is caused by above-average hypertrophy of the lymphoepithelial tissue of the pharyn-

geal ring, which is immunobiologically active during childhood. There is probably a hereditary disposition. Endocrine and constitutional factors and an influence of diet, particularly carbohydrates, have been suggested.

Diagnosis. The main symptoms include chronic mouth breathing, snoring, and proneness to infection. Examination by transnasal endoscopy or posterior rhinoscopy shows the enlarged adenoid (Fig. 3.11).

Differential diagnosis. This includes choanal atresia, foreign bodies in the nose, and other causes of nasal obstruction such as nasopharyngeal angiofibroma and malignant tumors of the nasopharynx, possibly of mesenchymal origin, especially in children. Dental causes should be looked for to explain positional anomalies of the teeth and malocclusion.

Treatment. Conservative treatment, with a change of climate, diet, drugs, and so forth is *not* satisfactory.

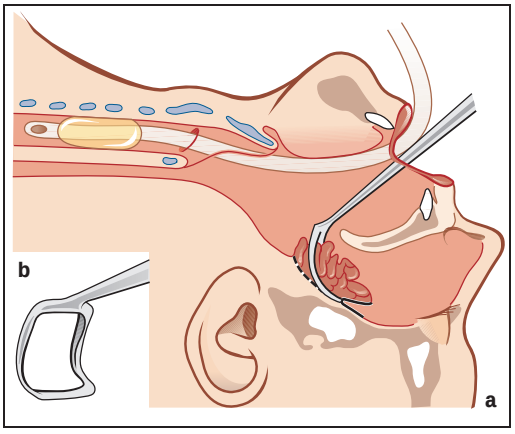


Fig. 3.17a, b Adenoidectomy with the head extended (a), using a Beckmann ring curette (b).

Surgical treatment consists of adenoidectomy (Figs. 3.17a, b, 3.18a–c). Endotracheal anesthesia should be used to prevent aspiration and to ensure optimal operating conditions. However, the operation can be performed under a brief anesthetic without a tube, provided that the neck is extended. Adenoidectomy is usually performed with the head in the hanging position. A Beckman ring adenotome is usually used to remove the adenoid. The instrument separates the adenoid at its base.

Tonsillar Hyperplasia

Clinical Features. This is usually combined with hypertrophy of the adenoid (see above). In addition, there is increased difficulty in swallowing and eating because of obstruction of the faucial isthmus. Considerable respiratory obstruction may also occur when only the tonsils are hyperplastic.

Diagnosis. See the section on adenoid hyperplasia. The local findings are obvious.

Differential diagnosis. This is similar to that for adenoid hypertrophy. It is important to determine whether the tonsils alone are hypertrophic, or whether there is coexisting adenoid hypertrophy.

> Note: Unilateral hyperplasia of the tonsil in an adult must always lead to a suspicion of malignancy. Rapid hyperplasia of the lymphatic pharyngeal ring indicates a disease of the entire lymphatic system.

Treatment. The modern practice for the treatment of tonsillar hyperplasia is tonsillotomy. This can be performed using different types of laser, radiofrequency surgery, or Ultracision. In patients with a history of recurrent or chronic inflammation, a tonsillectomy is indicated.

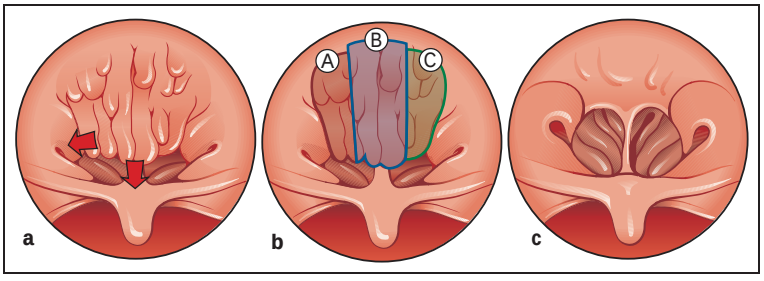


Fig. 3.18a–c a Obstruction of the pharyngeal ostium of the eustachian tube.
b Typical adenoidectomy in three portions.
c Decompression of the ostia.

Note: Not every enlargement of the tonsil or adenoid in a child is an indication for removal. There must be considerable hyperplasia with obvious mechanical obstruction of the nasopharynx or oropharynx, and the appropriate clinical effects and disorders must be present.

Course and prognosis. The symptoms usually resolve rapidly after removal of the mechanical obstruction. The child usually returns surprisingly rapidly to normal physical, psychological, and intellectual health. The prognosis is very good; recurrence after correctly performed adenoidectomy is unusual. The main complications are postoperative bleeding and aspiration. These are only to be feared if hemostasis is not achieved during surgery, if postoperative care is inadequate, or if tissue has been left behind.

Note: To avoid overlooking a pathologic bleeding tendency, the following investigations should be performed *before* adenoidectomy or tonsillectomy:

1. A history and family history relative to bleeding and coagulation disorders should be taken. This is the most important diagnostic step for avoiding pathologic bleeding.
2. The bleeding time should be determined if the history is consistent with a bleeding problem.
3. Further tests include the partial thromboplastin time (PTT) and the thrombocyte count.
4. The individual coagulation factors and the thrombocyte function should be investigated if the history and the routine tests indicate a disorder of hemostasis. In addition, analgesics such as salicylates should not be given for at least 10 days before the operation, as they inhibit the function of thrombocytes.

Adenoidectomy or tonsillectomy may still be performed in patients with manifest coagulation disorders if there are convincing indications. However, the operation should be performed with the appropriate substitution therapy, in a special unit.

Other postoperative complications include a change in the sound of the voice, which is usually only temporary, although occasionally rhinolalia aperta may persist. Rare complications include adhesions in the nasopharynx, injuries to the ostium of the eustachian tube, and, very rarely, injuries to the cervical spine.

Relative contraindications include cleft palate, either corrected or not. A speech therapist consultation must be obtained *before* a decision is made for surgery.

Hyperplasia of the lingual tonsil rarely occurs in children, but may occasionally be seen in adults. Symptoms include a feeling of pressure in the throat, especially on swallowing, and occasionally recurrent inflammation of the base of the tongue. If necessary, the lymphoepithelial tissue can be partially removed. The cryoprobe or the laser are particularly suitable for this.

Dysphagia

Diagnosis. The two mainstays of diagnosis are radiographic videography and video endoscopy. Radiographic evaluation of swallowing using fluoroscopy requires the administration of a contrast medium bolus (barium sulfate, provided there is no risk of aspiration; otherwise nonionic, low-osmolar contrast medium containing iodine). Phases and functional disorders can be visualized that elude identification by endoscopy—e.g., pumping movements of the base of the tongue, passage into the larynx, or peristaltic waves.

Etiology. A distinction is made between *neurogenic* and *structural* etiologies. Neurogenic dysphagias occur as sequelae to apoplexy, cerebrocranial trauma, hypoxia, neurosurgical operations, or degenerative disease. Structural dysphagias result from tumors or tumor resection in the aerodigestive tract (see also Table 3.1, p. 236).

Pathophysiology. Dysphagia is defined as difficulty in swallowing and may be accompanied by the following symptoms (Fig. 3.19):

- *Drooling:* The bolus leaves the mouth through incompletely closed lips (e.g., due to lip defects or facial nerve paralysis).
- *Leaking:* The bolus enters the pharynx during the oral preparation phase (e.g., due to tongue paralysis or soft palate impairments such as clefts or paralysis).
- *Larynx penetration:* The bolus arrives at the entrance to the larynx. Aspiration is prevented only by the vestibular folds and the vocal folds.
- *Regurgitation:* Larynx elevation, which plays an important role in opening the superior sphincter, may be impaired (e.g., after tracheotomy or excision of goiter), causing reflux into the mouth if the closing pressure in the

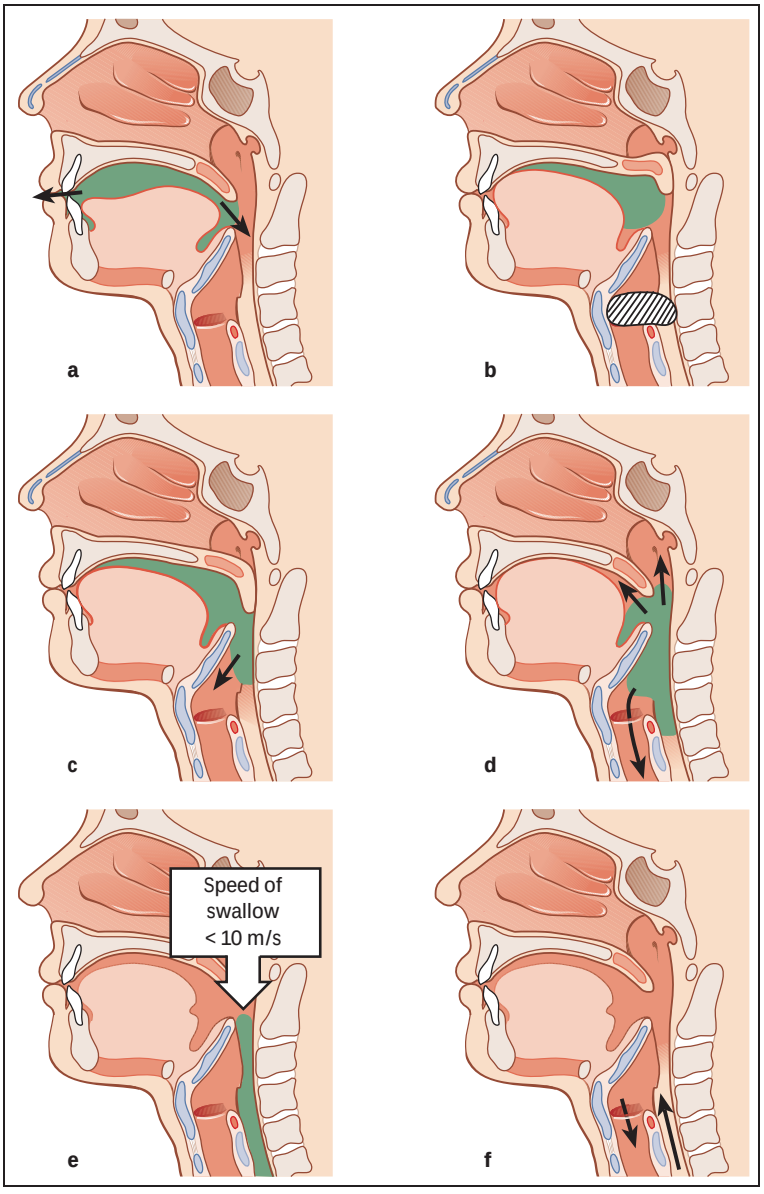


Fig. 3.19a–f Typical swallowing disorders.
a Drooling.
b Globus pharyngeus.
c Laryngeal aspiration.
d Regurgitation.
e Odynophagia.
f Postdeglutitive aspiration.

superior sphincter is too high. Reflux to the epipharynx—e. g., in velopharyngeal insufficiency—is known as nasal regurgitation.

- *Odynophagia:* Pain on swallowing—e. g., due to inflammation.
- *Bolus retention*—e. g., in an epiglottic vallecula, the piriform sinus, or a diverticulum—is possible. Gastroesophageal reflux leads to aspiration at the end of the swallowing act (post deglutition).

Functional treatment. The goal of logopedics or speech therapy is to improve swallowing efficiency and prevent aspiration.

Exercises and facilitation techniques: Dysphagia is treated using techniques that facilitate the sequence of movements and inhibit undesired elevation of muscle tone. Sensibility is stimulated, and the coordination, force, and endurance of motor activity are improved by providing training in target-oriented movements and patterns of motion.

Pathologic reflexes are reduced and pharyngeal contractions improved.

Compensatory techniques: Instead of abolishing the causes, these techniques compensate for deglutition disorders. Their purpose is to facilitate swallowing and prevent aspiration. They include changing the posture of the head by inclination and rotation.

Adaptation methods: This term refers to adjustment of food consistency, positioning the food if there are transportation disorders within the oral cavity, and using special eating and drinking aids.

Inflammatory Diseases

Labial and Oral Mucosa

Internal or dermatologic diseases often present on the lips, oral mucosa, gingiva, and tongue. Table 3.4 provides an overview of the most common and most important of these disorders. Since changes in the mucosa of the lips, mouth, or pharynx can occur in many disorders, the following description will be confined to the most common ones.

Rhagades of the commissures, along with slight bleeding and pain in the commissures accompanying opening of the mouth. Causes include ill-fitting false teeth, mycotic infection, poor general resistance, diabetes, iron-deficiency anemia, nonspecific pyogenic infections, and syphilis. If possible, the cause should be confirmed and dealt with before treatment is attempted. Carcinoma of the commissure may also simulate rhagades in the early phases. Nonspecific local treatment includes topical corticosteroids, either as a cream or in a special base designed to adhere to mucosal surfaces. In addition, a long-established favorite is gentian violet solution, also known as crystal violet, methyl violet, or pyoktanin. A standardized version is available in Europe using one active ingredient— methylrosaniline chloride 0.1 % aqueous solution (crystal violet). It is recommended to use this form, which is safe and inexpensive.

Cheilitis may be solitary and acute due to trauma, thermal injury (hot food), chemical injury (smoke), actinic damage (sunburn), or exposure to radiation.

Cheilitis granulomatosa, Miescher disease, is a chronic recurring disease which is usually ushered in with a complete *Melkersson–Rosenthal syndrome* of cheilitis, granulomatous glossitis, and facial paralysis. The pathogenesis of this triad is unknown, and the treatment is the same as that of idiopathic facial paralysis (see p. 111).

Although *tuberculosis* or *syphilis,* primary or secondary, may occur on the lips, a chronic or recurrent erosive or hyperkeratotic lesion of the labial mucosa must always be suspected of being premalignant (leukoplakia, Bowen disease). Numerous diseases affecting the oral mucosa also affect the lips.

Stomatitis, often combined with *gingivitis* or *inflammation of the buccal mucosa,* may be a primary disease of many different causes or may be secondary to other diseases. The clinical symptoms and prognosis are thus extremely variable.

Ulceromembranous Stomatitis

Clinical Features. The disease usually begins on the gingival margins with redness, swelling, and sensitivity to pressure. Swelling of the buccal and lingual mucosa, stomatitis simplex, may also occur. The disease often progresses to ulceration with severe pain, presenting with superficial, and occasionally deep, mucosal ulcers with a dirty-gray fibrinous membrane. There are marked constitutional symptoms: oral fetor, sialorrhea, possibly cloudy or purulent saliva, loss of taste, difficulty in eating, and high fever in the initial stages. The disease may spread to the pharynx, and the regional lymph nodes may be enlarged and painful.

Pathogenesis. This includes poor oral hygiene, reduced general resistance, infections from cutlery, dental damage, virus infections with possible secondary bacterial infection, mucosal rhagades, gingival pockets, and dental calculus (Fig. 3.20).

Diagnosis. Bacteriologic culture is necessary, and often shows spirochetes and fusiform rods as in Vincent angina (see p. 265).

Differential diagnosis. This consists of mucosal mycosis, excluded by culture, virus infection (herpes simplex, aphthous stomatitis, and herpes zoster), syphilis, tuberculosis, acquired immune deficiency syndrome (AIDS), hematologic diseases including agranulocytosis and leukemia, which can be excluded using a differential white blood count, and carcinoma, which requires a biopsy.

Treatment. This includes appropriate oral and dental hygiene or 1–2 % gentian violet solution. Anti-

Table 3.4 Common lesions of the oral mucosa in generalized and dermatologic diseases

	Cause
Dryness	Febrile infectious diseases, uremia, polyglobulinemia, cachexia, atropine poisoning, Sjögren syndrome and other sialadenoses, vitamin A deficiency, occasionally diabetes mellitus and hyperthyroidism, Plummer–Vinson (Paterson–Kelly) syndrome (iron deficiency), hypertension, prolonged use of certain drugs such as phenothiazines, belladonna and psychotropic drugs
Alterations of pigmentation	
Pallid	Anemia
Cyanotic	Pulmonary congestion
Intense red	Polycythemia rubra vera, reactive polyglobulinemia
Reddish-violet	Right heart insufficiency
Yellow	Jaundice, often as an initial symptom, hepatic congestion, megaloblastic anemia
Red, like lipstick	Hepatic insufficiency
Whitish patches like leukoplakia with dry mucosa	Vitamin A deficiency
Grayish-violet staining of the gingival mucosa	Argyrosis
Grayish-blue to brownish discoloration of the gingiva	Bismuth and lead intoxication
Spotted hyperpigmentation	Oral contraceptives
Punctate or striated, occasionally diffuse pigmentation of the lips, cheeks, gingiva, tongue and palate	Addison disease, of which it is often the first symptom
Bleeding	
From the gingiva, with a dark-red discoloration and swelling of the interdental papillae	Scurvy
Bleeding from cavernous angiectasia on the vermilion border and on the oral mucosa	Hereditary hemorrhagic telangiectasia (Rendu–Osler–Weber disease)

Fig. 3.20 Ulceromembranous stomatitis. Extremely severe ulcerous destruction in the region of the right side of the palate and the gingiva. Swabs often reveal spirochetes and fusiform rods, as in trench mouth.

biotics are given if indicated by culture and sensitivity tests. Local and general antimycotic therapy is given for fungal infections.

Course and prognosis. Both are good if the cause is treated appropriately.

Herpes Simplex Stomatitis and Gingivitis

Clinical Features. These include a burning sensation in the mouth, difficulty in eating, a feeling of being unwell, fever in the early stages, and lentil-sized, clear vesicles at the mucocutaneous junctions of the lip and the nasal introitus, and also in the entire mouth. The vesicles may progress to superficial circular or oval ulcers with a red center. The disease often occurs in conjunction with febrile general infections or overexposure to sunlight. There is marked oral tenderness, oral fetor, sialorrhea, and painful regional lymphadenopathy. The disease is contagious. Serial crops of fresh vesicles may occur. Children are most at risk.

Table 3.4 Common lesions of the oral mucosa in generalized and dermatologic diseases (continued)

	Cause
Punctate lesions	
White spots surrounded by an erythematous zone. The site of predilection is the buccal mucosa opposite the molar teeth, Koplik spots	Measles
Reticular or striated bluish-white membrane and edematous red spots mainly on the lips, but also on the tongue	Lupus erythematosus
Opalescent plaques, often with superficial ulceration	Secondary syphilis
Membranes on the oral mucosa	
Whitish, striated, nonadherent membrane brilliant white punctate spots in infants	Candidiasis
Vesicles, erosions, and cysts	Varicella (the vesicles are the size of hempseed and lie mainly on the palate), erythema multiforme, herpes simplex, herpes zoster, hereditary epidermolysis bullosa dystrophica, pemphigus vulgaris, mucosal pemphigus, AIDS
Aphthous ulcers	Aphthosis and Behçet disease
Stomatitis and necrotic ulcers	Pellagra, agranulocytosis, thrombocytopenia, panmyelophthisis, leukemia, mercury intoxication
Gingival hyperplasia	Pregnancy, possibly the contraceptive pill and hydantoin
Atrophic lesions	
Induration, sclerosis, narrow pale lips, shortened frenulum, microglossia	Progressive scleroderma

Pathogenesis. The cause is infection with the herpes simplex virus and usually occurs first in childhood. The first infection often causes no symptoms. The disease is very infectious: 90 % of the population are said to be carriers of the virus, but clinical manifestations in the form of herpes labialis (Fig. 3.21) or stomatitis herpetiformis only occur in 1 %.

Fig. 3.21 Herpes labialis. Small blisters at the border between lip and skin.

Diagnosis. This is made by exclusion. An attempt can be made to isolate the virus from the contents of the vesicle, if possible within the first 24 h of the vesicular stage. The material is inoculated in the rabbit cornea.

Differential diagnosis. Chronic recurrent aphthous stomatitis, varicella, the acute infectious exanthemas, herpangina, foot and mouth disease, Behçet disease, pemphigus, and mycoses must all be considered.

Treatment. Systemic antiviral therapy (acyclovir, valaciclovir, famciclovir) is most effective. It should be used for 72 h for initial infections. Recurrent cases can often be controlled with topical measures including topical anesthetics, 0.1% methylrosaniline chloride aqueous solution, or topical antiviral agents (acyclovir, penciclovir). If the patient is treatment-resistant, systemic treatment or prophylaxis can be tried. A bland liquid diet and gentle rinsing should also be advised. Steroids must *not* be given.

Course and prognosis. This disease is usually harmless and lasts 1–2 weeks. The vesicles heal to form crusts, but do not form scars. Recurrence is frequent, but herpetic sepsis and herpetic encephalitis are very rare.

Viruses of the Picornaviridae family (coxsackievirus, echovirus) and rarely of the variola group may also cause small oral ulcers.

Metal stomatitis. A stomatitis with discoloration of the gingiva can be caused by either medical or occupational exposure to mercury or bismuth. The same also applies to workers handling lead. The use of gold in the treatment of arthritis can cause a gingival stomatitis. Finally, the mucosa can be damaged by arsenic, chlorine, chromium, fluoride, copper, manganese, nickel, sulfur, thallium, and zinc; by organic substances such as benzol, dimethyl sulfate, tetrachlorocarbons, tetrachloroethylene; and by mixed substances such as corrosive agents, synthetic resins, synthetic materials, enamels, etc., as well as by wood, dyes, hops, wool, and insecticides.

Stomatitis due to drugs. This may be observed particularly after the use of bromides, iodides, salicylates, antibiotics, and sulfonamides, psychoactive drugs that dry the mouth, and antiepileptic agents, and after pyramidone, barbiturates, laxatives such as phenolphthalein, and the contraceptive pill.

Allergic stomatitis. Hypersensitivity reactions on the oral mucosa and the lips, with varying severity, with or without angioneurotic edema, may be observed in response to almost all drugs, dental material, mouthwashes, toothpaste, cosmetics, chewing gum, and also to some foods such as fruit, fish, protein, and milk. The diagnosis can be established by testing, and if the allergen is found it should be withdrawn. Otherwise, antiallergic or local symptomatic treatment are given.

Candidal Stomatitis

Clinical Features. Burning in the mouth and tongue, superficial white foci, and exudates on the mucosa are symptomatic. The exudate can be wiped off with mild pressure.

Pathogenesis. In normal individuals, almost all cases of oral yeast infections are caused by *Candida albicans;* rarely, other agents may be involved in severely immunosuppressed hosts. Candidiasis affect individuals with diabetes mellitus, reduced resistance, and after prolonged administration of antibiotics, chemotherapy, steroids, oral contraceptives, and after radiotherapy. Acute severe infection with ulceration can be a presenting sign of immune deficiency, human immunodeficiency virus (HIV) infection, or acute leukemia.

Diagnosis. This is made on the basis of the characteristic membranous white or gray exudates and very inflamed mucosa. Superficial ulceration occurs. A specimen is taken for culture for fungi.

Differential diagnosis Diphtheria.

Treatment. The standard therapy is nystatin or imidazole solutions or rinses. Options include 0.1% methylrosaniline chloride aqueous solution or borax–glycerin solution. Good oral hygiene is also crucial.

Course. The prognosis is good if the patient is relatively healthy. Prompt treatment for several weeks, with monitoring by culture, is advised in more severe cases. In immunosuppressed individuals, there is a risk of systemic spread if treatment is not adequate.

Herpes Zoster

Clinical Features. Unilateral rapidly progressive vesicles are quickly followed by fibrinous superficial epithelial defects, affecting the segments of the face innervated by the second and third divisions of the trigeminal nerve. The disease is very painful. Mucosal lesions may occur in the same stage and may be partially confluent and arranged in groups.

Pathogenesis. This is a neurotropic infection with the varicella-zoster virus, which is morphologically identical to the herpes simplex virus.

Diagnosis. The diagnosis is established on the basis of the typical segmental arrangement of the vesicles, severe pain, and culture of the contents of the vesicle (Fig. 3.22).

Differential diagnosis. This includes herpes simplex and recurrent aphthous stomatitis.

Treatment. Prompt institution of high-dose regimens of systemic antiviral agents (acyclovir, valaciclovir, famciclovir) within 48 h has been shown to reduce the duration and severity of the acute disease and somewhat reduce the risk of postherpetic neuralgia. In severe cases or immunosuppressed patients, intravenous aciclovir should be considered. A prophylactic vaccine has recently been approved. Good oral hygiene, a bland liquid diet, and topical anesthetics or 0.1% methylrosaniline chloride aqueous solution, can be used for symptomatic relief.

The disease is often followed by severe, therapy-resistant neuralgias, which may persist for months after resolution of the mucosal lesions. Occasionally, other regions and internal organs may be involved simultaneously. The generalized form in older patients suggests a systemic malignancy or immunosuppression.

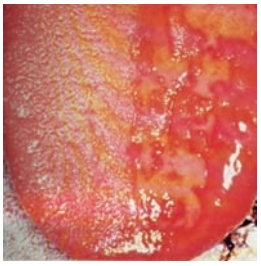


Fig. 3.22 Herpes zoster of the tongue. The oral symptoms are due to involvement of the second or third branch of the trigeminal nerve. The incidence increases in older age groups.

Fig. 3.23 Angular cheilitis, an early sign of human immunodeficiency virus (HIV) infection. The condition is due to *Candida* infection at the corners of the mouth.

Fig. 3.24 Kaposi sarcoma. The gingiva is the second most common location for oral Kaposi sarcomas. Proceeding from uncharacteristic swelling of the gums, blue-black changes with bleeding develop.

Acquired Immune Deficiency Syndrome (AIDS)

Clinical Features. Approximately 35–40% of HIV infections are associated with otorhinolaryngologic symptoms, including early symptoms such as angular cheilitis and Kaposi sarcoma (Figs. 3.23, 3.24). HIV infection has a relatively high association with cervical lymphadenopathies, candidiasis, herpes simplex, and herpes zoster. Other potential manifestations of HIV infection include sinusitis, tonsillitis, gingivitis, pharyngitis, esophagitis, tracheitis, sudden hearing loss, facial paralysis, and facial pain. General accompanying features are fever, anorexia, headache, muscle and joint pain, transient or persistent lymph-node enlargement, diarrhea, and profound weight loss.

Pathogenesis. HIV infection.

Diagnosis. HIV infection is diagnosed by the detection of HIV antibodies in the serum in a screening test (e.g., enzyme-linked immunosorbent assay, ELISA) followed by a confirmatory blot test.

Differential diagnosis. See the list of symptoms above. HIV infection should be suspected whenever a "classic" disease has an unusual location and presentation, runs an atypical course, and presents in an atypical age group, especially if the patient is in a high-risk group (male homosexuals, intravenous drug users, etc.).

Treatment. The HIV virus cannot be eliminated by any of the current treatment regimens. In addition, no vaccine is available. However, highly active antiretroviral therapy (HAART) using multiple drugs in different therapeutic classes designed to inhibit HIV

in different ways has radically improved the outlook for infected patients and altered the spectrum of oral diseases seen. Oral Kaposi sarcoma has become uncommon again, and severe oral candidiasis, oral hairy leukoplakia, and recurrent herpetic infections are all suppressed with HAART.

Chronic Recurrent Aphthae

Aphthae are recurrent oral ulcerations, generally small with an erythematous base. Three types are observed:

1. Minor type (Mikulicz): only two to six oval aphthae in the oral mucosa; also known as canker sores.
2. Major type (Sutton): large mucosal aphtha (> 1 cm), usually occurring on the soft palate, cheek mucosa, tongue or lips.
3. Herpetiform type (Cooke): many very small and painful herpetiform ulcerations on the lateral border of the tongue.

Clinical Features. Aphthae occur intermittently, affecting the buccal mucosa, the tongue, the palate, and the gingiva. They are very painful. The regional lymph nodes may be swollen, and concomitant stomatitis is possible.

Pathogenesis. The cause is unknown. The disease is not thought to be infectious, but instead to represent activation of intrinsic inflammation in susceptible individuals. Aphthae can be triggered by infections, hormonal factors such as menstruation, and certain foods.

Diagnosis. This is made on the basis of the long history of the condition and its tendency to recur. There is no sialorrhea, no oral fetor, and no fever.

Differential diagnosis. Herpes simplex, which occurs with fever, fetor, sialorrhea, a general malaise, and a large number of vesicles which are confluent and arranged in groups must be considered. Behçet disease can present with recurrent multiple aphthous ulcers.

Treatment. Causal or prophylactic treatment is not possible. Symptomatic treatment includes 0.1% methylrosaniline chloride aqueous solution, borax solution, chlorhexidine, and topical corticosteroids. The major type sometimes responds to antibiotics and oral tetracycline solution may be helpful.

Course. The lesions heal without scarring in 1–3 weeks, but early recurrence is possible. The course may extend over decades, and familial occurrences are known.

Behçet Disease

Clinical Features. Aphthae occur in crops in the mouth and on the genitals. *Eye symptoms* are often monocular and typically wax and wane. They include hypopyon iritis, which is often fleeting, and later papilledema, involvement of the retina, and blindness. Rheumatic symptoms and renal involvement may also occur.

Pathogenesis. The cause is unknown. It may be generalized vasculitis, an autoimmune event, or a virus infection.

Diagnosis. The main, and often the first, symptom is involvement of the eye. Acute cochleovestibular disturbances may also occur.

Treatment. Treatment relies on immunosuppressive medication (cyclophosphamide, cyclosporine) und long-term steroids. Oral mucosal lesions can be treated locally with topical corticosteroids, 0.1% methylrosaniline chloride aqueous solution, and gentle rinses.

Course. With treatment, the symptoms can be eased, the mucosal infections can be reduced, and the disease can be controlled over a lifetime.

Tuberculosis

Clinical Features. Mucosal lesions may take the form of a mucosal lupus or exudative ulcerative *mucosal tuberculosis.* Round nodules occur in groups in mucosal lupus, and they demonstrate yellowish-brown flecks in the oral mucosa on pressure with a glass spatula. They are not painful. Flat, dirty, exudative, painful ulceration with undermined edges and lymph-node involvement is found in ulcerative mucosal tuberculosis (see p. 265).

Pathogenesis. The oral cavity is nowadays almost never a primary site of manifestation of tuberculosis. The disease is usually due to hematogenous or intraluminal spread from the primary site, usually the lung.

Diagnosis. This is made by biopsy and culture, and chest radiography. Tuberculosis is a notifiable disease.

Differential diagnosis. Syphilis, mycoses, and carcinoma must be excluded.

Treatment. The original focus in the lung is treated by tuberculostatic drugs. Triple therapy consisting of isoniazid, rifampicin, and ethambutol is now usually given, supervised by a chest physician.

Course. This depends on the outcome of the primary lesion. However, the prognosis for mucosal lesions is good with general antituberculous therapy.

Syphilis

Clinical Features. All stages of syphilis can occur in the mouth.

Stage I (primary syphilis): primary chancres occur on the lips, tonsil, anterior part of the tongue, commissure, gingiva, and buccal mucosa (Fig. 3.25). A sharply delimited nodule 2–3 mm in diameter grows to the size of a penny. After a few days, a painless ulcer forms, with a very hard edge, and there is painless regional lymphadenopathy in the submandibular or jugulodigastric area. The primary lesion regresses spontaneously after 3–6 weeks.

Stage 2 (secondary syphilis): Eight to 10 weeks after the infection—i. e., 5–7 weeks after the appearance of the primary chancre and at the same time as or before the skin lesions—a superficial exanthema develops in the entire oral cavity. Dark-red mucosal spots a few millimeters in size form, with a tendency to merge. The lesions are of varying severity and last for several weeks. Dark-red papules form gradually, and also flat areas with a cloudy epithelial surface. The surface of the tongue looks like sugar icing with areas of loss of papillae. A very firm indolent lymphadenopathy develops.

Stage 3 (tertiary syphilis): A gumma develops on average 15 years after the primary infection. The sites of predilection in the mouth are the lips, the hard palate, the tongue, and the tonsils. There is a diffuse nodular infiltrate with liquefaction of the center, fetor, a sharp punched-out ulcer, and radiating scars.

Pathogenesis. This infection is caused by *Treponema pallidum.* The pathway of infection is either genital or extragenital. The incubation period is on average 3.5 weeks.

Diagnosis. Demonstration of the organism by culture and dark-ground illumination is used in stages 1 and 2. Serologic tests become positive from the fourth week. The *Treponema* immobilization test (Nelson test) only becomes positive in the ninth week. In stage 3, serologic reactions are positive and the disease can be demonstrated by histology. This is a notifiable disease. (See Syphilitic Tonsillitis for further details.)

Fig. 3.25 Primary chancre of the tongue.

Differential diagnosis. In stage 1 lesions, tuberculosis, mycoses, and herpes should be considered; in stage 2, erythema multiforme and tuberculosis; in stage 3, malignant tumors and leukemia.

Treatment. Penicillin should be administered by a dermatologist or a specialist in venereal diseases (see also p. 265).

Hyperkeratosis and Leukoplakia

Clinical Features. These include a velvety or nodular, usually sharply circumscribed epithelial lesion, hyperkeratosis, a flat epithelial plaque or white thickening that cannot be wiped off, and leukoplakia usually occurring on the lips, floor of the mouth, or buccal mucosa.

Pathogenesis. This is an epithelial disease with many different causes, including exogenous irritative factors such as chronic mechanical irritation by the irregular edge of teeth, pressure from a denture, smoking, excess alcohol consumption, lichen planus, syphilis, and lupus erythematosus. There may be no recognizable cause.

Two groups of lesions can be distinguished in leukoplakia, depending on the color and surface:

1. *Simple leukoplakia* with sharp edges, occurring in ≈ 50 % of cases. This is only rarely premalignant.
2. *Patchy leukoplakia,* which can be divided into: a) *verrucous leukoplakia* (≈ 25 % of cases), which shows an irregular, wrinkled grayish-red speckled surface and which may be premalignant; and b) *erosive leukoplakia* (≈ 25 % of cases), with a reddish erosive lesion and often an irregular nodular surface—this form becomes malignant in ≈ 35 % of cases and is very similar to Bowen disease.

The probability of malignant degeneration depends largely on the degree of histologic dysplasia. The frequency of progression of a leukoplakia to carcinoma increases with the degree of the dysplasia (Figs. 3.26, 3.27).

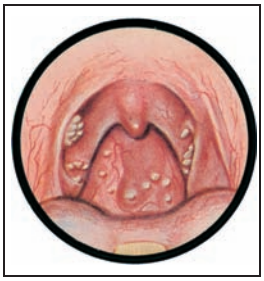


Fig. 3.26 Hyperkeratosis. Whitish-yellow, thorn-shaped or button-shaped structures are located singly or in groups on the tonsils, palatal arch, or posterior wall of the pharynx. Confusion with lacunar pharyngitis is unlikely, as the plaques are firmly attached to the underlying tissue.

Fig. 3.27 Leukoplakia of the buccal mucosa.

Fig. 3.28 Pemphigus. Two large, isolated serous bullae on the soft palate. The bullous stage is very short, so that this stage is rarely observed. Usually, the ruptured shreds of the blister membrane are all that can be seen.

Note: Leukoplakia should be regarded as being potentially premalignant and should therefore be investigated and followed up carefully.

Diagnosis. This is made by histology. The lesion should be entirely removed, if possible with a clear margin.

Differential diagnosis. Ulcerative stomatitis, mycoses, lichen ruber planus, lupus erythematosus, and pemphigus must be considered.

Treatment. This is by generous surgical removal, preferably at the time of biopsy and avoidance of possible causative agents.

Bowen Disease (Erythroplasia, Erythroplakia)

This is a premalignant lesion or carcinoma in situ. It is an intraepithelial squamous cell carcinoma with an intact basal membrane; the tumor has not yet invaded the subepithelial layer. It occurs on the skin or mucosa.

Clinical Features. Sharply demarcated full red foci of varying sizes, with a smooth surface, that occur on the mucosa of the cheek or tongue. It may also take the form of white patches of leukoplakia or verrucous vegetative papillomatous lesions the size of a hazelnut. The disease occurs mainly in men in the fourth to seventh decades of life. Progression to true squamous cell carcinoma is possible at any time and is very common.

Diagnosis. Biopsy distinguishes the lesion from other precancerous lesions of the mucosa such as leukoplakia and pure hyperplasia.

Treatment. The lesion must be excised with a healthy margin.

Inflammations of the Oral Mucosa in Dermatoses

Pemphigus

Clinical Features. The first symptom is often in the mouth and takes the form of flat soft or tense vesicles (Fig. 3.28). These give way to superficial epithelial erosions with a fibrin layer and epithelial tags at the edge. The course is episodic, and several stages may be present at any one time. There is oral fetor, often regional lymphadenopathy, and a bullous eruption on the skin. The disease usually begins between age 40 and 60.

Pathogenesis. Pemphigus vulgaris is a well-established autoimmune disease with autoantibodies directed against desmosomal proteins, primarily desmogleins. The main desmoglein of the mucosa is desmoglein 3. Almost all patients with oral involvement have antibodies against this desmoglein. Other components of the desmosome may also be targets.

Diagnosis. Routine histology may suggest the diagnosis by demonstrating acantholysis, but it has to be confirmed using direct and indirect immunofluorescence studies or molecular biological identification of autoantibodies.

Differential diagnosis. This includes stomatitis, hereditary epidermolysis bullosa, erythema multiforme, lichen planus, and mucosal pemphigoid.

Treatment. Systemic corticosteroids and immunosuppressive agents are administered under the supervision of a dermatologist.

Erythema Multiforme

Clinical Features. Fibrinous exudate, crust, and vesicles form on the lips and oral mucosa. There are simultaneous skin and joint lesions, and also fever. The skin lesions may be target-shaped and occur preferentially on the hands and feet. The symptoms are those of severe acute infection, with oral fetor, sialorrhea, pain, and regional lymphadenopathy. Far more common is recurrent disease, which is milder and often confined to the mucosa. Drug-induced or atypical erythema multiforme is more likely to be on the trunk and not to have target morphology, and it can progress to Stevens–Johnson syndrome or toxic epidermal necrolysis.

Pathogenesis. Acute disease is usually caused by *Mycoplasma* infection. Chronic disease is almost invariably caused by recurrent herpes simplex virus infection. A wide variety of drugs have been implicated in atypical erythema multiforme, including allopurinol, carbamazepine, co-trimoxazole, lamotrigine, nevirapine, nonsteroidal anti-inflammatory drugs (of the oxicam type) phenobarbital, phenytoin and sulfonamides.

Diagnosis. This is made on the basis of the generalized disease picture, and possibly by biopsy.

Differential diagnosis. Pemphigus, lichen planus, and enanthema due to drug reactions must be excluded.

Treatment. In severe primary disease, the underlying infection should be sought and treated properly. Most recurrent disease can be suppressed with either long-term low-dose or episodic promptly introduced antiviral drugs (usually acyclovir). Topical corticosteroids may be helpful, as are a bland liquid diet or even parenteral feeding for severe primary disease. A possible drug trigger should be investigated.

Prognosis. Primary disease may be life-threatening. Recurrent disease is annoying and reduces quality of life but rarely causes severe problems. Drug-induced disease may be life-threatening, advancing to Stevens-Johnson syndrome or toxic epidermal necrolysis. Ophthalmologic consultation should be obtained.

Fig. 3.29 Lichen planus (lichen ruber). Bluish-white papules with a mother-of-pearl shimmer, small and sharply bordered, posteriorly connected by fine lines forming a network. Delicate shapes are seen, usually with a symmetrical distribution. Leukoplakia and mucous plaques need to be included in the differential diagnosis.

Lichen Planus

Clinical Features. The mucosal appearance of lichen planus can be specific, with a lacy network of white lines or streaks (Wickham striae). More often, there are nonspecific eroded lesions on the buccal mucosa, gingiva, and tongue. Blue-gray, smooth, tiny papules are sometimes also found on the dorsum of the tongue. The lesions cannot be wiped off and are firm. The ulcerations can be very painful. Pruritic red-brown to pink papules, often polygonal, 2–3 mm in size, may be seen on the skin, preferentially on the volar surface of the wrist (Fig. 3.29).

Pathogenesis. The etiology is unknown.

Diagnosis. This is made on the basis of the full clinical picture, at times supported by biopsy. In contrast to the skin, lichen planus in the mouth is a potentially premalignant disease. It may be accompanied by *Candida* infection, which can complicate the clinical picture and diagnosis.

Differential diagnosis. This includes leukoplakia, hyperkeratosis, Bowen disease, mycosis, enanthema of drug reactions, and lupus erythematosus.

Treatment. Possible toxic agents such as sunlight, tobacco, and chemical agents should be excluded. The disease is not curable, but is controllable when significant or symptomatic. Topical corticosteroids or retinoids are the best approach. A cyclosporine oral rinse is expensive but helpful in refractory

Table 3.5 Oral fetor

Site of origin	Cause
Teeth, gingiva, and mouth	Dental caries, parodontosis, gingivitis, stomatitis, erythema multiforme, pemphigus, neglected false teeth, abscesses of the floor of the mouth, ulcerating tumors
Pharynx	Acute tonsillitis, Vincent angina, mononucleosis, peripharyngeal and retropharyngeal abscess, pharyngeal diphtheria, chronic tonsillitis and pharyngitis, foreign body in the nasopharynx, tertiary syphilis
Airway	Atrophic rhinitis, ozena, purulent rhinitis, sinusitis, bronchitis, bronchiectasis, bronchial foreign body, lung abscess, and pneumonia
Digestive tract	Hypopharyngeal or esophageal diverticulum, hiatus hernia, esophagitis, diseases of the stomach and the intestines with or without hiccup and vomiting
Generalized disorders	Diabetes mellitus with ketosis (acetone), renal failure (urine), and hepatic coma, which produces a sweet aromatic smell

cases. Careful follow-up is needed, as this is a premalignant lesion.

Other rare inflammatory lesions of the oral cavity include rhinoscleroma, leprosy, and sarcoidosis (see p. 147), as well as AIDS. See also page 251.

An overview of the common causes of oral fetor is given in Table 3.5.

Tongue

The inflammatory diseases described above usually also produce symptoms on the tongue. The following inflammatory disorders affect the tongue primarily.

Glossitis (Glossopyrosis and Glossodynia)

Clinical Features. These include burning of the tongue, especially at its tip and edges, and frequent parageusia or hypergeusia. On the tongue itself, only minimal mucosal lesions can be demonstrated, such as circumscribed inflammation or loss of papillae.

Pathogenesis. The cause may lie in mechanical irritation by sharp teeth, dental calculi, pressure from dentures, intolerance to dental materials—e. g., denture material or the use of metals that are electrically noninert, mouthwashes, drug sensitivity, vitamin B_{12} deficiency (Hunter glossitis), megaloblastic anemia, iron-deficiency anemia (Plummer–Vinson syndrome), diabetes, and gastrointestinal diseases, including cirrhosis and mycoses.

Diagnosis. The diagnosis is based on the detection or exclusion of mechanical irritation, sensitivity reactions, diabetes, gastrointestinal or hematologic diseases, and also on the mycologic findings. Finally, the diagnosis can be made by exclusion of all other causes. Often it is difficult to classify the symptoms. Fifty percent of cases have a psychosomatic etiology. Neurologic forms based on diseases of the glossopharyngeal nerve, lingual nerve, hypoglossal nerve or chorda tympani produce only localized symptoms. Episodes of pain may be induced by tumors of the brain or skull base. Myofascial pain syndrome (Costen syndrome) may be accompanied by glossodynia.

Differential diagnosis. This should include allergic glossitis or a depressed immune response.

Treatment. The cause should be eliminated if possible. A bland diet (the patient should avoid very hot, cold, or spicy foods and heavy alcohol consumption) is essential. Symptomatic topical treatment with anesthetics or corticosteroids is rarely helpful. Often, systemic psychotropic medications are required for relief of discomfort.

Median Rhomboid Glossitis

This is an anomaly that occurs when the tuberculum impar is displaced into the tongue during embryonic development. The anomalous structure persists, and the lingual papillae are not differentiated in that area.

Diagnosis. Visual inspection reveals a circumscribed, spindle-shaped area around the foramen cecum.

Treatment. Nystatin. Laser resection may be appropriate in patients with carcinophobia.

Allergic Glossitis

Clinical Features. The signs are similar to those of nonspecific glossitis, except that the disease begins suddenly with swelling and redness of the tongue,

with swelling and pain progressing to itching. There is a danger of respiratory obstruction if the reaction progresses to edema.

Pathogenesis. This is an allergic reaction localized to the tongue. Many substances are possible allergens, including serum injections, antibiotics, drugs such as phenothiazine, barbiturates, sulfonamide, aspirin, local anesthetics, and foods such as fruits, fish, protein, or nuts.

Diagnosis. This is based on the clinical picture of a sudden onset with marked symptoms, and on demonstration of the allergen.

Differential diagnosis. Acute infectious enanthema, mycosis, intoxication, local chemical damage, and nonspecific glossitis must be considered.

Treatment. This is both symptomatic and antiallergic. Allergen tests can be done and an allergen-free diet prescribed.

The tongue is also affected by the following specific or chronic inflammations: tuberculosis, syphilis, mycoses, actinomycosis, dermatomyositis, Sjögren disease, and progressive scleroderma.

Surface lesions of the tongue that encourage infection include:

1. *Geographic tongue:* The dorsum of the tongue is covered with smooth red patches, which resemble a map and may gradually change position. The cause is unknown, but this is a harmless condition that does not require treatment.
2. *Fissured tongue:* In this disease, there are clefts and folds of varying depth forming islands on the mucosa of the dorsal surface of the tongue. This appears to be a simple autosomal-dominant hereditary disease. It is one of the aspects of the Melkersson–Rosenthal syndrome of facial paralysis, edema of the face and lips, and fissured tongue. Fissured tongue occurs frequently in trisomy 21 (Down syndrome). Harmless inflammation may occur due to penetration of foreign material into the clefts. The treatment is symptomatic.

The common causes of a coated, red, or fissured tongue are summarized in Table 3.6, and the causes of burning of the tongue in Table 3.7.

Abscess of the Floor of the Mouth

Clinical Features. These include swelling and limitation of movement of the tongue, increasing pain, difficulty in articulation progressing to complete loss of speech, protrusion and induration of the floor of the mouth with marked sensitivity to pressure, severe difficulty in swallowing, and finally complete inability to eat, limitation of movement of the temporomandibular joint with trismus, fever, severe generalized symptoms, and occasionally stridor.

Pathogenesis. Infected material enters via rhagades of the tongue or the oral mucosa. The infection advances into the loose musculature of the tongue and into the numerous connective-tissue spaces (Fig. 3.30b). The base of the tongue, the lingual tonsil, or a carious tooth are also possible portals of infection (see Fig. 3.30a). The causal organism is usually common pathogenic bacteria. Penetration of small foreign bodies such as fish bones, bone splinters, kernels of corn, etc. are also possible causes. The infection may arise primarily in the sublingual or submandibular glands lying in the floor of the mouth. An abscess on the floor of the mouth is known as a *Ludwig angina.*

Diagnosis. This is made on the basis of the clinical picture of an inflammatory swelling of the floor of the mouth, severe pain, and a progressive course.

Differential diagnosis. Hematoma, gumma, tuberculosis, and malignancy.

Treatment. Therapy should be started at the onset of symptoms, with high doses of intravenous broad-spectrum antibiotics. If this does not induce remission, the antibiotic must be changed depending on the resistogram. If there is evidence of liquefaction, the abscess is opened widely and drained externally. Adequate diet and fluid intake must be ensured, if necessary with a nasogastric tube or parenteral feeding. A tracheotomy is indicated for respiratory obstruction. If *Ludwig angina* develops, the infected area is opened widely to prevent infection spreading to the larynx or mediastinum.

Course and prognosis. These are favorable provided that suppuration occurs quickly so that the abscess can be drained. Extension of the abscess to the deeper soft tissues of the neck (see Fig. 3.45a, b) and to the mediastinum is a life-threatening situation.

Actinomycosis. This is also a possible differential diagnosis, but it is now becoming very rare. This

Table 3.6 Surface lesions of the tongue

Type of lesion	Basic disease	Clinical symptoms	Details
Red tongue	Pernicious anemia	Initially dark red, later caramel-colored spots and striae on the dorsum of the tongue. The surface of the tongue is red, smooth, and shiny	Dysgeusia, paresthesia, xerostomia; the oral mucosa is also affected
	Scarlet fever	Strawberry tongue	Protrusion of papillae
	Hepatic cirrhosis	Glazed, shiny smooth red dry tongue with blue spots, “liver tongue”	Generalized symptoms prominent, glazed lips, yellow staining of the oral mucosa, brownish pallid face
	Sjögren syndrome	Dry, smooth, red glazed tongue	Swelling of the salivary glands, and salivary stones
	Median rhomboid glossitis	A raised or slightly sunken red area free of papillae in the center of the middle third of the tongue in the midline	A harmless lesion confined to the tongue causing no symptoms
	Vascular congestion	Red-violet swollen tongue	Right heart failure, hepatic cirrhosis and malignant tumors
	Hypertension	Pink to carmine red	Hypertension, myocardial insufficiency, left heart failure, valvular heart disease, local allergic reaction and shock
	Allergy	Strawberry or raspberry red, edema	Occurs in local allergic reaction and also in shock
Gray smooth tongue	Vitamin A deficiency	Bluish matt epithelial protrusions and bluish staining of the lips	Xerostomia, dysphagia
	Radiotherapy	Oral mucosal is sensitive to heat, circumscribed mucosal atrophy, mucosal induration	Ageusia, xerostomia
	Lichen planus	Milky bluish striae, spider web leukoplakia, papillae are retained and there is no membrane	Also affects the oral mucosa
	Progressive scleroderma	Dry tongue, limited mobility, initially edema of the tongue, later atrophy of the tongue and increasing rigidity	Dysphagia, interference with speech, mouth is too small, salivary deficiency
Black hairy tongue	Antibiotics, idiopathic, smoking, niacin shortage, liver disease	Hairy, greenish-black membrane, long black cornified papillae	Also occurs in mycosis
Fissured tongue	Lingua plicata	Surface of the tongue is furrowed and fissured	A benign normal hereditary variant
	Melkersson–Rosenthal syndrome	Folded tongue	Periodic swellings of the lips, tongue, and parotid glands, and intermittent facial paralysis
Coated tongue	Nonspecific oral infection	Whitish coat (horny scales)	Connected with reduced food intake in gastritis and enteritis and in feverish infections
	Oral thrush	Whitish, membranous adherent plaques with red edges	*Candida albicans* demonstrated on culture

Table 3.6 Surface lesions of the tongue (continued)

Type of lesion	Basic disease	Clinical symptoms	Details
Coated tongue	AIDS	Commonly associated with oral candidiasis and hairy leukoplakia of the tongue	Multifocal symptoms are typical. Diagnosis, see p. 251
	Scarlet fever	Dirty-white coating with reddened tip and edges to tongue	Pharyngitis, exanthemas; β-hemolytic streptococci found on culture
	Diphtheria	Grayish-white membranous coat, smells sweet and nasty	Adherent membrane, the underlying bed bleeds slightly, generalized symptoms
	Typhus	Grayish-white tongue with very red edges	Infection with *Salmonella typhi,* generalized symptoms
	Uremia	Brownish plaques	Renal insufficiency

AIDS, acquired immune deficiency syndrome.

Table 3.7 Burning of the tongue

Basic disease	Clinical symptoms	Details
Geographic tongue	Burning of the tongue, with red-spotted tongue, absence of filiform papillae	
Toxic stomatitis	Burning of the tongue and grayish-blue discoloration of the gingiva by bismuth and lead, reddened edematous mucosa in mercury poisoning	
Stomach and intestinal disorders of various causes	Manifest or latent symptoms, depending on the site of the cause	
Plummer–Vinson (Paterson–Kelly) syndrome (see p. 275)	Dry tongue, considerable dysphagia. Rhagades of the commissures, atrophic mucosa	Almost exclusively in women, dry pale skin, koilonychia, dry mucous membranes
Sjögren disease	Xerostomia, tough sticky saliva, papillary atrophy, smooth glazed tongue, dysphagia	Dryness affecting the mucosa of the oral cavity, the pharynx, larynx, and trachea; swelling of the major salivary glands
Glossitis in megaloblastic anemia	Burning of the tongue, dysgeusia, paresthesia, dryness, spotted tongue with purple-red areas in striae alternating with bluish areas, smooth surface but not papillary atrophy, partially swollen papillae	May involve the entire oral mucosa
Diabetes mellitus	Intermittent marked burning of the tongue with dry surface	Tendency to oral infections and mycoses
Food allergy and contact allergy	Begins suddenly; marked swelling and redness; burning of the tongue increasing to cause pain and feeling of tension	Typical history or evidence of allergic cause, involvement of the rest of the oral mucosa
Pellagra, niacin deficiency	Hypoesthesia of the tongue, salty taste, feeling of a "chapped" tongue, red swollen and occasionally coated tongue, later chessboard tongue with marked fissuring, and finally atrophy	Inflammation of the remaining oral mucosa, sialorrhea is more common than salivary deficiency
Mucoviscidosis	Dry, burning tongue, tough glutinous secretion	Increased sodium and chloride ions in mucus and saliva
Psychogenic glossodynia	Burning of the tongue without any demonstrable organic cause	Frequent in latent depression

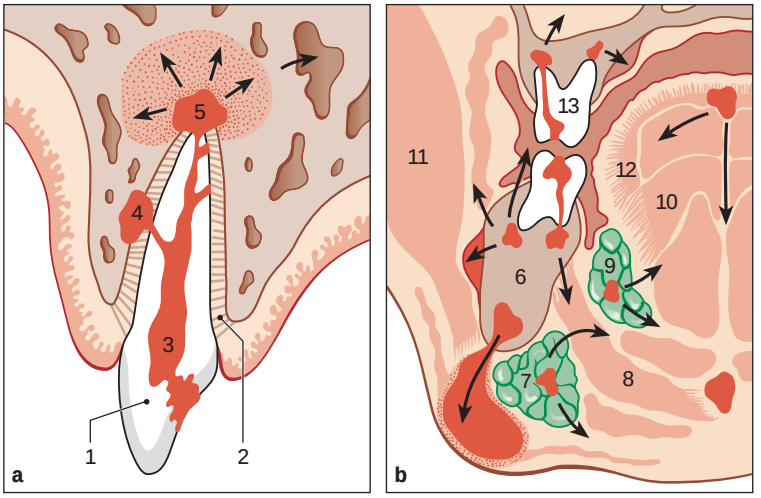


Fig. 3.30a, b Dental infections and infections in the floor of the mouth.
a Infections in and around the teeth. 1, Carious incisor tooth; 2, alveolar periosteum; 3, pulpitis; 4, periapical abscess; 5, apical granuloma.
b Sites of origin of inflammations of the floor of the mouth. 6, Mandibular, arising from the teeth or osteomyelitis; 7, submandibular gland, due to inflammation or a stone; 8, musculature of the floor of the mouth; 9, sublingual gland, inflammation or retention; 10, tongue musculature; 11, muscles of the cheek; 12, abscess of the body of the tongue; 13, apical granuloma of one of the upper teeth, extending to the antrum.

disease has an indolent course and causes relatively slight pain, but leads to the formation of multiple hard infiltrates with formation of abscesses and fistulas. It is mainly localized to the head and neck (98% of cases). The organisms are usually *Actinomyces israelii* and accompanying bacteria. *Actinomyces* agglomerations (sulfur granules) can be demonstrated in the pus from the abscess and in tissue specimens. A needle biopsy can be taken, with a specimen of pus being sent for bacteriologic identification. Serologic tests include agglutination and complement fixation. Precipitation tests and intracutaneous tests are not reliable. Treatment is with penicillin in the early phases, sulfonamides in the long term, and incision of any abscesses.

■ Pharyngeal Lymphatic Ring (Waldeyer Ring)

Acute Tonsillitis

Clinical Features. Acute tonsillitis usually begins with high temperature and possibly chills, especially in children. The patient complains of a burning sensation in the throat, persistent pain in the oropharynx, pain on swallowing, and pain radiating to the ear on swallowing. Opening the mouth is often difficult and painful, the tongue is coated, and there is oral fetor. The patient also complains of headaches, thick speech, a marked feeling of malaise, and swelling and tenderness of the regional lymph nodes. Both tonsils and the surrounding area, including the posterior pharyngeal wall, are deep red and swollen, but in catarrhal tonsillitis there is no exudate on the tonsil. Later, yellow spots corresponding to the lymphatic follicles form on the tonsils—hence the term *follicular tonsillitis.* Alternatively, yellow spots occur over the openings of the crypts, a condition known as *lacunar tonsillitis.* A membrane occurs in *pneumococcal tonsillitis,* but it is seldom confluent and rarely spreads beyond the tonsil. There is also swelling of neighboring organs such as the faucial pillars, uvula, and base of the tongue. The patient also reports sialorrhea and difficulty in eating (Figs. 3.31, 3.32).

Pathogenesis. The most common organism is β-hemolytic streptococci. Staphylococci, pneumococci, mixed flora, *Haemophilus influenzae,* and *E. coli* are much less common. If the symptoms worsen and multifocal symptoms occur, a generalized disorder expressing itself particularly in the lymphoepithelial organs should be suspected. On the other hand, there are also tonsillar infections in which the generalized symptoms are minimal and only the local changes can be recognized. Virus infections are particularly important in this respect—e. g., herpangina (see p. 263). The tonsillar parenchyma is infiltrated with leukocytes in tonsillitis, causing small abscesses in the parenchyma and in the crypts. In addition, a fibrinous exudate is formed and there are marked changes in the parenchyma and the epithelium.

Note: Bacteria are constantly present in the mouth and pharynx. These saprophytic organisms include: saprophytic *Streptococcus viridans,* pneumococci, fusiform bacteria, *Leptothrix, Neisseria, Lactobacillus,* staphylococci, *Sarcina,* and fungi. They may become pathogenic due to a change in the environment.
A virus infection may prepare the way for secondary bacterial infection.

Diagnosis. The diagnosis is based on the clinical presentation of an acute onset with high fever, a sore throat, painful swallowing, redness and exudate on the tonsils, general blood tests, the erythrocyte sedimentation rate (ESR), C-reactive protein (CRP), cardiovascular tests, and urinalysis. Rapid immunoassay testing can identify streptococcus bacteria in 10 min. Appropriate tests or even cultures are performed if diphtheria is suspected, and blood tests are done for mononucleosis (see below)

Differential diagnosis. This includes scarlet fever, diphtheria, infectious mononucleosis, agranulocytosis, leukemia, hyperkeratosis of the tonsils, stage 2 syphilis, and in unilateral disease, ulceromembranous tonsillitis, peritonsillar cellulitis or abscess, tuberculosis, and tonsillar tumors (see below).

Treatment. The standard treatment in patients with streptococcal tonsillitis is penicillin V for 10–14 days (Table 3.8). Oral cephalosporins or macrolides can be used in patients allergic to penicillin. As acute tonsillitis is a systemic rather than local disease, treatment should include bed rest, analgesics, a bland liquid diet, and ice packs. The patient should be observed for complications. Local care should include oral and dental hygiene. Local antibiotics should not be given, but antiseptic and analgesic mouthwashes can be used. Moist neck dressings and a sweat pack may be used in the early phases.

Course. Tonsillitis usually resolves within 1 week. However, complications such as respiratory obstruction due to laryngeal edema, otitis media, or rhinosinusitis may occur; sequelae may also develop.

Fig. 3.31 Lacunar tonsillitis. The palatine tonsils are bright red, swollen, and coated.

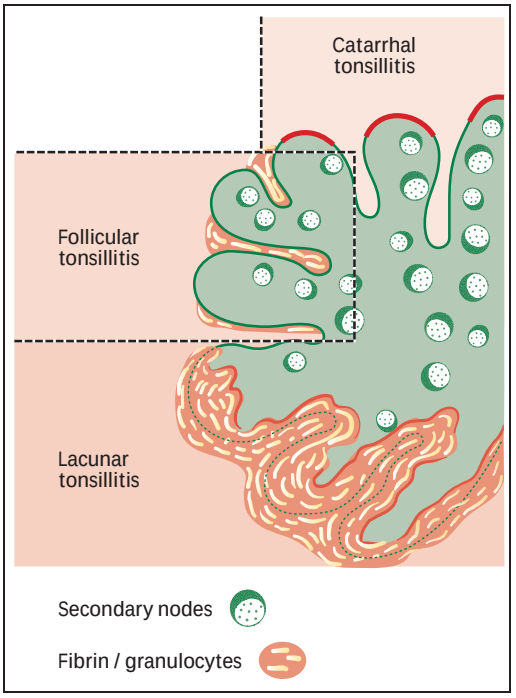


Fig. 3.32 The grades of severity and pathomorphology of catarrhal, follicular, and lacunar tonsillitis.

Note: The following laboratory investigations should be done in tonsillitis:
1. Smear and culture to exclude diphtheria and determine causative bacteria, ideally before commencing antibiotics.
2. Urinalysis to exclude nephritis.
3. Differential blood count to exclude mononucleosis and leukemia.

Table 3.8 Initial empirical treatment of acute tonsillitis

Most common pathogens	Initial treatment	Duration
Streptococcus pyogenes	Phenoxymethylpenicillin (penicillin V)	10 days
Haemophilus influenzae	Cefuroxime axetil, loracarbef	5 days
	Amoxicillin	
	Clarithromycin, azithromycin	
	Telithromycin	

Fig. 3.33a–c a Typical appearance of the mucosa and tonsils in a patient with infectious mononucleosis; the tonsils are swollen and red, with fibrin coatings.
b The nuchal and neck lymph nodes are visibly enlarged.
c A patient with a pseudoallergic rash following treatment with ampicillin.

Nasopharyngitis. The symptoms are as described above but are localized mainly or exclusively to the adenoid. Differential diagnosis includes viral nasopharyngitis.

Infection of the lingual tonsil. This is similar, with symptoms localized to the base of the tongue. Ipsilateral involvement of the larynx or abscess formation in the tongue are possible.

Other types of tonsillitis

1. Simple tonsillitis caused by nonspecific organisms.
2. Tonsillitis in infectious diseases.
3. Tonsillitis in hematologic diseases.
4. Ulceromembranous tonsillitis.

Infectious Mononucleosis (Glandular Fever, Benign Lymphadenosis)

Clinical Features. Salient features include fever in the range of 38–39°C and marked lymphadenopathy of the jugulodigastric group and the deep cervical chain, later becoming generalized (Fig. 3.33b). The lymph nodes are moderately tender. The tonsil is very swollen and covered with a fibrinous exudate or membrane (Fig. 3.33a). The patient has rhinopharyngitis, hepatosplenomegaly, pain in the neck on swallowing, and a marked feeling of being unwell. There is pain in the head and limbs. The blood picture initially shows leukopenia, and then leukocytosis with a white blood cell count to 20 000–30 000 or more, 80–90 % of which are mononuclear cells and atypical lymphocytes.

Two types of disease are distinguished:

1. The *pharyngeal type* is characterized by a very severe sore throat and painful swallowing. The tonsils have a heavy fibronectin coating, and the lymph nodes are enlarged. The virus is located mainly in the mucosa-associated lymphoid tissue (MALT).
2. The *hematogenic type* is characterized by a high fever, severe malaise, abdominal pain, and circulation of the causative virus in lymphocytes (liver, spleen).

Microbiology. The causative organism is the Epstein–Barr virus, which chiefly affects children and adolescents. The disease is probably spread by droplet infection. The incubation period is 7–9 days.

Diagnosis. This is made from the picture of generalized lymphadenopathy and tonsillitis, the characteristic blood picture, a monospot test, and the Paul–Bunnell test (demonstration of heterophile antibodies in the serum; a positive titer is > 1 : 28).

Differential diagnosis. This includes diphtheria (by culture), Vincent angina, scarlet fever, syphilis, rubella, acute leukemia, toxoplasma, listeriosis, and tularemia.

Treatment. Symptomatic treatment includes oral hygiene and measures to reduce fever. Antibiotics may be given against secondary bacterial infection if there is marked ulceration. Cephalosporin or clindamycin is advisable. An allergy-like rash may occur in response to ampicillin (Fig. 3.33c). Tonsillectomy may be indicated for severe local symptoms

such as respiratory obstruction and dysphagia, and persistent fever.

Complications. The course may be protracted, with paralyses affecting cranial nerves VII and X, serous meningitis and encephalitis, myocarditis (cardiologic supervision), hemolytic anemia, hemorrhagic complications in the gastrointestinal tract, the pharynx, and the skin, hematuria, obstruction of the airway, and a danger of asphyxia. An associated bleeding disorder may also occur in severe disease. Tracheotomy should only be performed in extreme emergencies.

Note: If penicillin does not induce a rapid fall in fever in patients thought to have tonsillitis, they are probably suffering from infectious mononucleosis.

Course. Resolution is marked by a long convalescence (months), during which liver values should be monitored. The patient should refrain from competitive sports during this time, especially if there is any evidence of splenomegaly.

Infection of the Lateral Bands

This is a specific form of infection of the lateral, tubopharyngeal bands, especially in patients who have had their tonsils removed. It may be a "substitute" infection in the absence of tonsils. There is swelling, redness, and yellow spots around the lateral bands, and also in the solitary follicles on the posterior pharyngeal wall (Fig. 3.34).

Treatment. Penicillin is administered as in tonsillitis, as complications may also occur in this disease. If the disease recurs frequently, the area should be cauterized with a 2–5 % silver nitrate solution; careful use of a cryoprobe is also possible.

Herpangina

This disease causes marked generalized symptoms such as high fever, headache, pains in the neck, and loss of appetite and mainly affects children up to the age of 15 years. Vesicles form initially, particularly on the anterior faucial pillar, but are very fleeting and are therefore not often seen. The tonsils are often only slightly red and swollen. Occasionally, they are covered in milky white vesicles up to the size of a lentil, arranged like a chain of pearls, or there are small flat ulcerations of the tonsil. Similar eruptions may occur on the palate or buccal mucosa (Fig. 3.35).

Fig. 3.34 Lateral pharyngitis. Inflammation of the lateral pharynx. Both salpingopharyngeal folds are markedly swollen and reddened, and their surface dotted with numerous small, yellowish-white spots. There is very severe pain, radiating to the ears, and the complaints and generalized symptoms are as in acute tonsillitis.

Fig. 3.35 Herpetic angina is usually caused by group A coxsackieviruses, less often by those of group B, retroviruses, or echoviruses.

Microbiology. The organism is coxsackievirus A, which has an incubation period of 4–6 days.

Diagnosis. This is based on the presence of vesicles, minimal lesions in the tonsils, and the benign rapid course over several days.

Treatment. Symptomatic, including oral hygiene and mouth rinses with chamomile and antiseptic solutions.

Scarlet Fever

The tonsils and pharyngeal mucosa are deep red; there is pain on swallowing, severe malaise, progression to lacunar tonsillitis, and regional lymphadenopathy. After ≈ 24 h, a typical exanthema appears, beginning on the upper part of the body. At the same time, a definite reddening of the tip and edges of the tongue appears, extending later to the entire tongue—strawberry tongue. Facial reddening does not include the perioral skin. It should be noted that the exanthema may not appear. Desquamation of the skin begins on about the eighth day (Fig. 3.36).

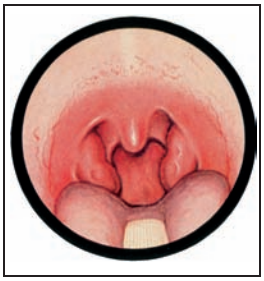


Fig. 3.36 Scarlet fever (scarlatina). Scarlet fever pharyngitis on the first day of the illness. The tonsils and palatal arches are swollen and dusky red. The erythema of the palatal arch is sharply demarcated. A vascular network is frequently visible on the soft palate. The tonsils may have a transient, whitish-gray coating. Necrosis may occur in severe cases.

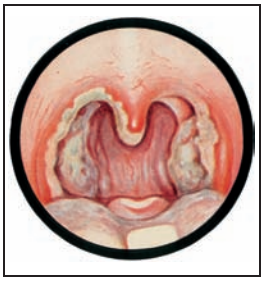


Fig. 3.37 Diphtheria. The tonsils, palatal arches, and uvula are coated with dirty gray–whitish membranes. The tissue is already ulcerated. On forceful depression of the tongue, the epiglottis is visualized. It is not coated.

Microbiology. The type A hemolytic streptococcus is responsible.

Diagnosis. This is made on the basis of the clinical appearances of redness and swelling of the tonsil, strawberry tongue, small erythematous spots on the soft palate, and the Rumpel–Leede phenomenon (petechiae distal to an inflated sphygmomanometer cuff). The blood picture shows leukocytosis and a left shift, and eosinophilia from the fifth day.

Differential diagnosis. This includes diphtheria, which is excluded by smear and/or culture.

Treatment. This is based on penicillin, oral hygiene, and analgesics.

Diphtheria

Two clinical forms are distinguished:

1. Local, benign pharyngeal diphtheria.
2. Primary toxic, malignant diphtheria.

Clinical Features. Patients experience a mild prodromal phase, usually with a temperature of ≈ 38°C and no more than 39°C. There is slight pain on swallowing and often a very high pulse rate. The tonsils are moderately reddened and swollen, with a white or gray velvety membrane that becomes confluent, extends beyond the boundaries of the tonsil to the faucial pillars and soft palate, and is fixed firmly to its base. The membrane can only be wiped off with difficulty and it then leaves a bleeding surface behind. The jugulodigastric lymph nodes are very swollen, tender, and often hard. There is a characteristic smell of acetone on the breath. Sixty percent of cases are localized to the pharynx including the tonsils, and in 8% the larynx is involved in addition. Albuminuria is common (Fig. 3.37).

Microbiology. The infection is due to the diphtheria bacillus, *Corynebacterium diphtheriae*. The disease is transmitted by contact, droplets, or contamination by oral or nasal secretions. The incubation period is 3–5 days. In localized forms, the disease is restricted to the tonsil, the nose, the larynx, or a wound. The generalized form is progressive and toxic.

Diagnosis. This is based on: (1) a bacteriologic smear from the tonsils and pharynx—Gram staining of a smear from the pseudomembrane provides the result within 1 h; (2) culture provides the answer at the earliest after 10 h; (3) isolation of the organism confirms the diagnosis in 2–8 days; (4) there is a membrane that is firmly adherent and extends beyond the tonsil. This is a notifiable disease.

Differential diagnosis. This includes nonspecific tonsillitis, infectious mononucleosis, Vincent angina, candidiasis, agranulocytosis, leukemia, and syphilis.

Treatment. On the earliest reasonable suspicion of diphtheria (possibly *before* bacteriologic confirmation), the patient should be isolated and treated with antiserum administered intramuscularly at a dosage of 200–500 IU/kg. A skin test should be given to exclude allergy to the antitoxin before it is administered. In severe cases, a high dose of 1000 IU/kg may be given, accompanied by antibiotic coverage. In addition, penicillin G should be given, initially 3 × 2 million IU i.v., or alternatively erythromycin for 14 days.

Treatment also includes bed rest, oral hygiene, neck dressings changed several times a day, and steam inhalation. Diphtheria immunization with diphtheria toxoid is protective but does not become effective for several weeks.

Complications. These include general toxicity, failure of the heart and circulation, hemorrhagic nephritis or nephrosis, palatal paralysis due to polyneuritis, airway obstruction, and danger of asphyxia. A proportion of the population are silent carriers of the disease.

Normally, excretion of virulent diphtheria bacteria ceases after several weeks. However, carriers may continue to be a source of infection for months or even years. Cultures are therefore necessary until three cultures at weekly intervals are negative.

Long-term carriers should be treated with local and parenteral antibiotics and local disinfection. If this does not eradicate the organism, it may be necessary to carry out tonsillectomy, accompanied by adenoidectomy in children, to remove the source.

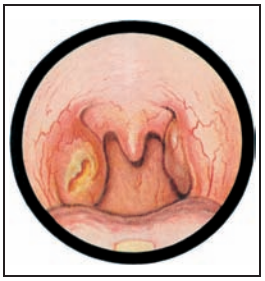


Fig. 3.38 Primary complex on the right tonsil. Among the rare extragenital sites of syphilitic infection, the tonsils are second only to the lips in frequency of involvement. Sharply demarcated ulcerations on the thickened, firm, reddened right tonsil. Painless, hard swelling of the regional lymph nodes, which are readily displaceable. Definitive diagnosis is possible only if spirochetes are found on dark-field microscopy.

Syphilitic Tonsillitis

Clinical Features. All stages of syphilis may be encountered in the oropharynx. After 3 weeks, the primary lesion appears on the lips, buccal mucosa, tonsils, and tongue. Typically, the primary chancre is painless, consisting initially of a papular lesion that develops into an ulcer. Palpable regional lymph nodes are present. Approximately 6 weeks after the primary infection, white, hazy mucosal enanthemas (*plaques opalines*) appear on the tonsils, faucial pillars, and soft palate. The hard palate is usually spared. Later, they progress to dark-red papules. Signs of stage 2 infection are usually present in other parts of the body. The tertiary stage appears after 20–30 years.

Diagnosis. Any ulcer in the oropharynx, especially involving the soft palate and uvula, is suspicious. The diagnosis is confirmed by dark-field examination of organisms from the primary lesion and by serologic testing (Fig. 3.38).

Serologic tests. *Treponema pallidum* hemagglutination assay (TPHA), fluorescent treponemal antibody absorption (FTA-ABS) test, 3 weeks after infection, follow-up test of the patient's response to therapy.

Treatment. Penicillin G (600 000 IU daily for 2 weeks). Erythromycin should be used in patients allergic to penicillin.

Tonsillar Tuberculosis

This disease causes a superficial erosive ulcer with a necrotic slough (see p. 389).

Agranulocytosis

Clinical Features. The generalized symptoms are prominent and include high fever and chills. The patient feels very sick and has a typical blood picture. The disease occurs mainly in older individuals. There is ulceration and necrosis of the tonsils and pharynx, with a blackish exudate, severe pain in the neck and on swallowing, sialorrhea, and oral fetor. There is no regional lymphadenopathy.

Pathogenesis. Severe injury to the leukopoietic system may be caused by drugs or by occupational or other toxins.

Differential diagnosis. This includes diphtheria, infectious mononucleosis, Vincent angina, and acute leukemia.

Treatment. This consists of elimination of all possible leukotoxic drugs, avoidance of other sources of injury, and prevention of secondary infection by high-dose penicillin, blood transfusion, and careful oral hygiene. The patient should be in the care of a hematologist.

Vincent Angina (Ulceromembranous Pharyngitis, Trench Mouth)

Clinical Features. The patient usually describes unilateral pain on swallowing, and there is ipsilateral swelling of the jugulodigastric nodes. There is an ulcer, which is often deep, on *one* tonsil, with a

Fig. 3.39 Vincent angina (ulceromembranous pharyngitis, trench mouth). The right tonsil is enlarged and reddened. On its upper end, there is an irregular, crater-like ulcer, the bottom of which is coated with a dirty grayish layer of mucus.

Fig. 3.40 Fungal (*Candida*) pharyngitis. On the soft palate, small, irregularly shaped, slightly prominent, white spots are seen that can be removed easily with a cotton swab. The diagnosis is easy and is based on microscopic findings.

Fig. 3.41 Chronic tonsillitis. When pressure is applied with a spatula to the swollen tonsil, crumbly, fetid cores of pus and liquid pus emerge from the crypts. Taken alone, demonstration of cores of pus is insufficient for diagnosis of chronic tonsillitis, as they may also occur in completely normal tonsils lacking any signs of irritation.

whitish exudate, and the site of predilection for this is the upper pole. The local findings are often impressive in contrast to the symptoms, which are often slight. There may only be a feeling of a foreign body in the throat, and the patient also has a characteristic oral fetor. There is usually no fever. The exudate, which can be easily wiped off, may extend to the palate, buccal mucosa, and gingiva.

Microbiology. Fusiform and spirochetal organisms are always both present.

Diagnosis. This is made on the basis of the clinical picture of a typical infection usually of *one* tonsil, with unilateral lymphadenopathy, and on the results of bacterial culture (Fig. 3.39).

Differential diagnosis. This includes diphtheria, tuberculosis, syphilis, tonsillar neoplasms, acute leukemia, agranulocytosis, and infectious mononucleosis.

Treatment. Penicillin is given for 3–6 days. Local treatment with 2–5% silver nitrate is applied topically.

Course. Usually short, with a good prognosis.

Fungal (*Candida*) Pharyngitis

Clinical Features. A white superficial punctate exudate forms, which can be wiped off and later becomes confluent. There is usually only slight redness in the surrounding mucosa. The tonsils, palate, posterior pharyngeal wall, and buccal mucosa may be affected. Subjective symptoms are few (Fig. 3.40).

Treatment. Antimycotic agents. Nystatin and imidazole are given in the form of a suspension or gel. Ketoconazole and fluconazole may be used for longer-term therapy. Systemic treatment with fluconazole is often required and essential in patients with HIV/AIDS.

Hyperkeratosis of the Tonsil (Differential Diagnosis of Interest)

This is a typical yellowish-brown or white, flat, or somewhat nodular prominent hyperkeratotic process on the tonsillar surface which *cannot* be wiped off.

Caution: This condition is frequently misdiagnosed as tonsillitis.

The cause is a benign circumscribed keratinization of the tonsil, especially of the epithelium of the crypt. No treatment is necessary.

Chronic Tonsillitis

Chronic tonsillitis requires particular attention on diagnostic grounds, as it is difficult to differentiate from a normal tonsil. It may also be a focus of infection, with effects on the entire body (Fig. 3.41).

Clinical Features. The history usually shows recurrent attacks of tonsillitis, but this is not always the case. There is often little or no pain in the neck and little or no difficulty in swallowing. There is halitosis and a bad taste in the mouth. The jugulodigastric lymph nodes are often enlarged. Chronic tonsillitis often remains more or less symptom-free. The sys-

temic effect may become evident through reduced resistance, fatigue, a tendency to catch colds, unexplained high temperature, and loss of appetite.

Pathogenesis. The organisms are usually a mixed flora of aerobic and anaerobic bacteria in which streptococci predominate. Group A β-hemolytic streptococci are especially likely to cause focal symptoms. Poor drainage of the branching crypts leads to retention of cell debris, which forms a good culture medium for bacteria (see p. 260). From crypt abscesses of this type, the infection extends via epithelial defects in the reticular epithelium into the tonsillar parenchyma to produce a cryptic parenchymatous tonsillitis. Alternatively, it can penetrate into the capillaries surrounding the crypts, allowing toxins and organisms to enter the general circulation intermittently or continuously. In the long term, the tonsillar parenchyma undergoes fibrosis and atrophy.

Diagnosis

- The history shows recurrent acute or subacute attacks of tonsillitis.
- *Local findings:*
  - The tonsils are more or less fixed to their base, as shown by the depressor test (see Fig. 3.13e).
  - The tonsillar surface is fissured or scarred.
  - *Watery* pus and grayish-yellow material can be pressed out of the opening of the crypts by a tongue depressor (see Fig. 3.13e).
  - Reddening of the anterior faucial pillar is present.
  - Peritonsillar tenderness is present.
  - Lymphadenopathy of the jugulodigastric group is found.
- *General findings:*
  - The history includes recurrent tonsillitis, unexplained high temperature, lowering of resistance, etc.
  - Blood picture: the ESR and antistreptolysin titer are raised (see below).

Note: Fixed yellow concrements observed when pressure is exerted on the crypts with a tongue depressor do *not* signify chronic tonsillitis, but are a physiologic phenomenon (tonsillar plugs).
The size of the tonsil is also *not a* criterion for the presence of chronic tonsillitis. This disease can occur in large hyperplastic tonsils, but is more common in small and medium-sized tonsils. It is not always possible to make the diagnosis of chronic tonsillitis on the basis of the local findings. The history and general findings also need to be assessed critically. The examiner's judgment and experience are often decisive. Immunology has so far not proved to be of any practical diagnostic value.

Focal infection. A "focus" is any local change in the body that is capable of producing remote pathologic effects beyond its immediate surroundings. Poststreptococcal infections after tonsillitis are of special interest.

In clinical experience, diseases that may be based on a focal process include the following:

- Rheumatic fever (acute, febrile joint, and muscle disease).
- Glomerulonephritis and focal nephritis.
- Localized pustular psoriasis.
- Eruptive psoriasis (in children).
- Chronic urticaria.
- Endocarditis, myocarditis, and pericarditis.
- Polyserositis.
- Inflammatory disorders of the nerves and eyes (iridocyclitis).
- Vascular diseases (e. g., recurrent thromboangiitis, nodular vasculitis).

Treatment. Chronic tonsillitis requires treatment whenever it fulfils the criteria for a local pathologic process outlined above (see p. 247). Conservative measures such as gargling, painting, and suctioning are of no benefit. Even antibiotics can at best favorably influence the local process and risk of dissemination during the course of treatment. When the antibiotic is withdrawn, the pathogenic mechanism becomes reactivated, as the anatomic situation has not changed.

Tonsillectomy thus appears to be the only definitive treatment that can be considered for chronic tonsillitis. This applies even to patients with a coexisting coagulation disorder that will require appropriate replacement measures before surgery, special surgical techniques (e. g., "sealing" of the wound surface with fibrin glue and collagen fleece), and meticulous local supervision for a period of several days.

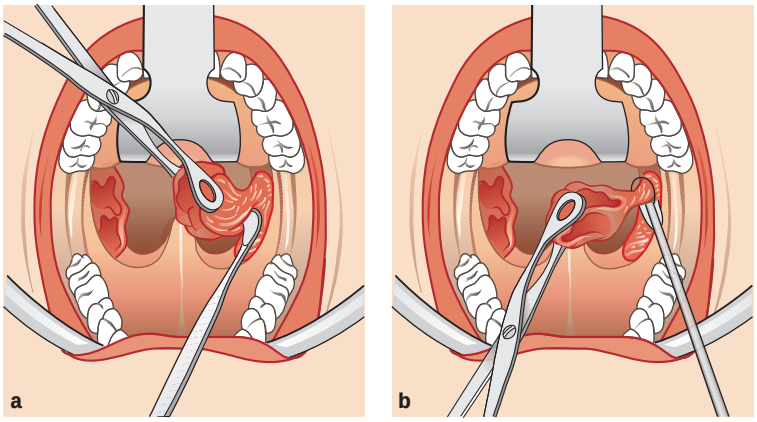


Fig. 3.42a, b Tonsillectomy with the head extended. The tongue is above, the upper incisors are below.
a Incision.
b Dissection and removal of the tonsil.

Tonsillectomy is indicated for the following diseases:

- Chronic tonsillitis.
- Recurrent tonsillitis.
- Peritonsillar abscess.
- Tonsillogenic septicemia.
- Tonsillogenic or posttonsillitis focal symptoms.
- Marked hypertrophy of the tonsil causing mechanical obstruction.
- If a tonsillar tumor is suspected.

Relative indications include:

- Resistant carriers of diphtheria bacilli.
- Stubborn oral fetor as a result of excess production of tonsillar plugs.
- Cervical lymph nodes that are tuberculous (bovine type), in which the tonsil is a possible portal of entry (see p. 389).

Contraindications include pharyngitis sicca, leukemia, agranulocytosis, serious generalized disorders such as tuberculosis or diabetes, and ulcerative or destructive processes extending beyond the tonsil if the diagnosis has not been confirmed. In some cases, cleft palate also represents a contraindication.

The patient's age is not a contraindication in doubtful cases.

The balance between immunobiologic considerations and the local pathologic findings needs to be weighed up carefully before tonsillectomy is recommended, particularly in children.

Principles of tonsillectomy. The operation can be performed under either local or general intubation anesthesia.

Tonsillectomy is usually performed with the patient under intubation anesthesia, with the head extended (Fig. 3.42a, b). An incision is made into the anterior faucial pillar, and the connective-tissue layer between the tonsillar parenchyma and the pharyngeal constrictor muscles is demonstrated. The tonsil is then freed by combined blunt and sharp dissection, proceeding from the upper pole to the base of the tongue, preserving the faucial pillars. The *entire* tonsillar tissue has to be removed. Hemostasis is secured by pressure, ligatures, or electrocautery. The same procedure is then performed on the opposite side.

Complications include hemorrhage, which may occur up to the 14th postoperative day. See also page 269.

As tonsillectomy is the most common surgical procedure in otolaryngology, there has been a desire to reduce the risk of hemorrhage through innovative surgical techniques and equipment. The National Prospective Tonsillectomy Audit, conducted in England and Northern Ireland in 2003–04, compared the overall risk of hemorrhage in relation to different surgical techniques (Table 3.9).

The following questions are often asked about tonsillectomy:

1. *The tonsils are apparently protective organs. Is it not bad for the body as a whole if they are removed?* The tonsils ought to be removed only if they are a source of irreversible inflammatory disease, if they are acting as a focus of infection, or if they are impeding breathing or swallowing due to their size. In these cases, it is the tonsils' pathologic characteristics that predominate, and their original protective function is reduced or has been lost. There is no doubt that this protective immunologic

function can be taken over satisfactorily by the remaining lymphoepithelial organs and structures in the pharynx. Removing irreversibly diseased or very enlarged tonsils, and those suspected of harboring a focus of infection, is therefore not a disadvantage, but a prerequisite for good health.

2. *Is there a tendency to catch upper respiratory infections more often after tonsillectomy?* A distinction needs to be made between pharyngitis and tonsillitis. Susceptibility to pharyngitis is not reduced by tonsillectomy itself, but it may be reduced if the tonsillectomy restores the airway to normal (e.g., in tonsillar hypertrophy) and if the flora in the mouth and pharynx are restored to normal (e.g., in chronic tonsillitis). Simple pharyngitis can therefore occur just as often after tonsillectomy as before, but tonsillitis can no longer occur, and intratonsillar abscess (a dangerous condition), focal symptoms, and tonsillogenic complications are also eliminated.
3. *What are the best age and the best time of year for tonsillectomy? The* operation can be performed at any age and at any time of year, but in patients under the age of 4 or over the age of 60 it should only be done with the strictest indications.
4. *Does tonsillectomy have any deleterious effects on the voice and speech?* If it is performed with a careful, conservative technique, tonsillectomy usually does not have any deleterious effects on the voice and speech. However, considerable caution is needed before recommending the operation in a patient who has an open, corrected, or occult cleft palate. Reinforcement of palatal insufficiency is described on page 281. In singers, the resonating space may be changed, although this is usually only temporary, and care is advised in these patients.

Tonsillotomy. There has been renewed interest in the traditional procedure of tonsillotomy, which was previously abandoned due to the risk of scarring of the crypts and follicles, with postoperative debris creating a nidus for abscess formation. A major reason for this renaissance is that there is a reduced risk of infection and abscess formation after tonsillotomy. The procedure can be performed using various types of laser (CO_2, diode), radiofrequency surgery, or the Coblation technique. It maintains open crypts (Fig. 3.43) and may preserve the immunological function of the tonsils in children younger than 4 years of age.

Indications: Tonsillotomy is only indicated in children or adults who have hyperplasia of the palatine tonsils. It is contraindicated if there is a history of recurrent infections or abscess.

Table 3.9 Results with five commonly used tonsillectomy techniques, based on the findings of the National Prospective Tonsillectomy Audit (Royal College of Surgeons of England, London, 2005)

Cold steel tonsillectomy using ties and /or packs	Technique with by far the lowest risk of postoperative hemorrhage (1.3%) and return to theater (1.0%)
Cold steel dissection with (bipolar or monopolar) diathermy hemostasis	Hemorrhage rate of 2.9%; 1.7% of the patients returned to theater
Bipolar (forceps or scissors) diathermy for dissection and hemostasis	Hemorrhage rate of 3.9%; 2.4% of the patients returned to theater
Monopolar diathermy for dissection and hemostasis	Hemorrhage rate of 6.1%; 4.0% of the patients returned to theater
Coblation for dissection and hemostasis	Hemorrhage rate of 4.4%; 3.1% of the patients returned to theater

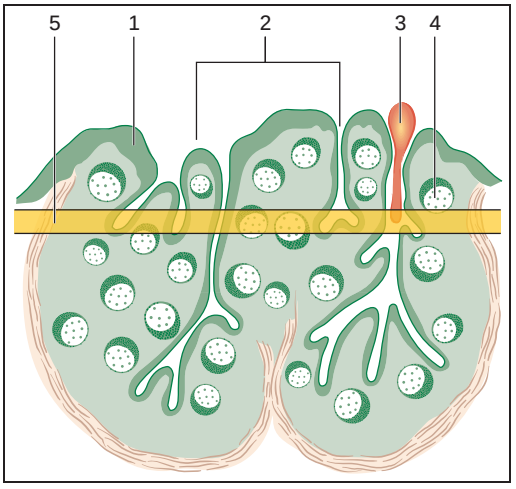


Fig. 3.43 The principle of laser tonsillotomy.1, Squamous epithelium; 2, tonsillar crypts; 3, cryptic content; 4, secondary nodes; 5, dissection level.

Local and General Complications During and After Tonsillitis

Post-tonsillitis complications include rheumatic fever, often with a symptom-free interval of 4–6 weeks, and endocarditis, myocarditis, or pericarditis. Acute glomerulonephritis and focal nephritis, which require urinalysis after the tonsillitis has