Merck &17: Otolaryngology

Chapter 204: Clinical Evaluation of Complaints Referable to the Ears

Hearing loss, tinnitus, vertigo, earache, and otorrhoea are the principal symptoms attributed to the ears. A thorough history should be taken and a physical examination performed with emphasis on the ears, nose, nasopharynx, and paranasal sinuses to evaluate complaints referable to the ears. In addition, the teeth, tongue, tonsils, hypopharynx, larynx, salivary glands, and temporomandibular joints should be examined, since pain and discomfort may be referred from them to the ears. Radiography or CT of the temporal bones is usually indicated in trauma to the ear, possible basal skull fracture, perforation of the tympanic membrane, hearing loss, vertigo, facial paralysis, and otalgia of obscure origin. Measurements of auditory and vestibular function are of great diagnostic importance in patients with complaints referable to the ears.

Hearing Loss

Hearing loss caused by a lesion in the external auditory canal or the middle ear is called **conductive,** while hearing loss due to a lesion in the inner ear or the 8th nerve is called **sensorineural.** Conductive and sensorineural hearing loss can be differentiated by comparing the threshold of hearing by air conduction with that by bone conduction.

Clinical Measurement of Hearing

Hearing by air conduction is tested by presenting an acoustic stimulus, in air, to the ear. A hearing loss or elevation of the threshold demonstrated in this way can be caused by a defect in any part of the hearing apparatus - external auditory canal, middle ear, inner ear, 8th nerve, or central auditory pathways.

Hearing by bone conduction is tested by placing a sounding source (i.e. the oscillator of an audiometer or the stem of a tuning fork) in contact with the head. This causes vibration throughout the skull, including the walls of the bony cochlea, and stimulates the inner ear directly. Hearing by bone conduction bypasses the external and middle ear and tests the integrity of the inner ear, 8th nerve, and central pathways.

If the air conduction threshold is elevated and the bone conduction threshold is normal, the hearing loss is *conductive.* If both air and bone conduction thresholds are elevated equally, the hearing loss is *sensorineural.* Occasionally, a **composite** or **mixed** loss of hearing occurs, with both conductive and sensorineural components. Under these circumstances, both bone and air conduction thresholds are elevated, the air conduction more than the bone.

The Weber and Rinne tuning fork tests are used to differentiate a conductive from a sensorineural hearing loss. For these tests, tuning forks with frequencies of 256, 512, 1024, and 2048 Hz are used. The **Weber tuning fork test** is performed by placing the stem of a vibrating tuning fork on the midline of the head and having the patient indicate in which ear the tone is heard. The patient with a unilateral *conductive* hearing loss hears the tone louder in the affected ear, for reasons that are unclear. By contrast, the patient with a unilateral *sensorineural* loss hears the tone in the unaffected ear, because the tuning fork stimulates both

inner ear equally and the patient perceives the stimulus with the more sensitive, unaffected end organ and nerve.

The **Rinne tuning fork test** compares hearing ability by air conduction with that by bone conduction. The tines of a vibrating tuning fork are held near the pinna (air conduction) and then the stem of the vibrating tuning fork is placed in contact with the mastoid process (bone conduction) and the patient is asked to indicate which stimulus is louder. Normally the stimulus is heard longer and louder by air conduction than by bone conduction; i.e. 40 sec by air conduction and 20 sec by bone conduction (AC>BC). With a conductive hearing loss, this ratio is reversed; the bone conduction stimulus will be perceived longer and louder than the air conduction stimulus (BC>AC). With a sensorineural hearing loss, both air and bone conduction perception are reduced, but the ratio remains the same (AC>BC).

The **audiometer** is used to quantitate hearing loss. With this electronic device, acoustic stimuli of specific frequencies (pure tones) are delivered at specific intensities in order to determine the patient's hearing threshold for each frequency. The hearing for each ear is measured from 125 or 250 to 8000 Hz by air conduction (using earphones) and by bone conduction (using an oscillator in contact with the head). Hearing loss is measured in decibels **(dB),** which equal 10 times the logarithm of the ratio of the acoustic power required to achieve threshold in a normal individual. Test results are plotted on graphs called audiograms. If intense tones are presented to one ear, they may be heard in the other ear. The Rinne tuning fork test and audiometry require the use of masking for accurate results. **Masking** is presentation of sound (usually noise) to the ear not being tested so that responses are based on hearing in the ear being tested.

Speech audiometry: The **spondee threshold (ST),** the intensity at which speech is recognized as a meaningful symbol, is determined by presenting a list of spondee words (2 syllables equally accented, such as *railroad, staircase, baseball*) at specific intensities, noting the intensity at which the patient repeats 50% of the words correctly. The ST usually approximates the average hearing levels at speech frequencies of 500, 1000, and 2000 Hz.

Ability to discriminate the various speech sounds or phonemes is determined by presenting 50 phonetically balanced one-syllable words, containing the phonemes in the same relative frequency as in conversational English, at an intensity of 25 to 40 dB above the ST. The percentage of words correctly repeated by the patient is the **discrimination score,** normally 90 to 100%. The discrimination score remains in the normal range in conductive hearing losses, but is reduced in sensorineural hearing losses because analysis of the speech sounds by the inner ear and 8th nerve is impaired. Discrimination tends to be poorer in neural than in sensory hearing losses.

Tympanometry measures the impedance of the middle ear to acoustical energy without voluntary participation by the patient and requires only that the patient remain quiet during the test; a sounding source and microphone sealed in the external auditory canal can measure the acoustical energy absorbed (passing through) or reflected by the middle ear. In conductive hearing loss, the middle ear absorbs relatively less sound and reflects relatively more sound. Normally the greatest compliance of the middle ear occurs with a pressure in the external auditory canal equal to atmospheric pressure. Increasing or decreasing the pressure in the external auditory canal demonstrates various patterns of compliance. With a relatively

negative pressure in the middle ear, as in eustachian tube obstruction and middle ear effusion, maximal compliance occurs with a negative pressure in the external auditory canal. With discontinuity of the ossicular chain, as in necrosis or dislocation of the long process of the incus, no point of maximal compliance can be obtained. With fixation of the ossicular chain, as in stapedial footplate ankylosis in otosclerosis, compliance may remain normal or may be reduced. Tympanometry has been used to screen children for middle ear effusions (serous or secretory otitis media), to provide diagnostic clues, and to confirm the type of lesion in patients with conductive hearing losses.

This technic can detect changes in compliance produced by reflex contraction of the stapedius muscle; the acoustic reflex is initiated by presenting to the same or opposite ear a tone approximately 80 dB above the hearing threshold. The presence or absence of this reflex is important in the topographical diagnosis of facial nerve paralysis. The reflex adapts or decays in neural hearing losses, and presence or absence of acoustic reflex adaptation or decay, especially below 2000 Hz, aids in differential diagnosis of sensory and neural hearing losses. The acoustic reflex can also confirm voluntary threshold responses.

The minimum comprehensive audiologic assessment requires measuring pure-tone air and bone conduction thresholds, STs, and discrimination, performance intensity function for phonetically balanced words, tympanometry, and acoustic reflex testing, including reflex decay testing. Information gained from these procedures helps one decide if more definitive information of a sensory from a neural hearing loss as described below is indicated.

When the patient cannot or will not respond voluntarily to acoustic stimuli, measuring the cochlear microphonic response and action potentials of the 8th nerve **(electrocochleography)** and evoked responses from the brainstem and auditory cortex **(brainstem response audiometry)** to acoustic stimuli has been useful in evaluating infants and children suspected of having profound hearing loss, individuals suspected of feigning or exaggerating a hearing loss (psychogenic hypacusis), and patients with sensorineural hearing loss of obscure etiology. Seven sequential wave forms have been identified that occur in the 8th nerve and central auditory pathways in response to acoustic stimuli. Lesions of the 8th nerve and brainstem auditory pathways result in changes in the amplitude and latency of the wave forms; these changes in latency of the wave forms are often of diagnostic value. Brainstem-response audiometry is used in coma to determine the functional integrity of the brainstem.

Clinical Measurement of Hearing in Children

(same under Screening Procedures for Infants and CHildren in Ch. 182)

About 1:1000 neonates has a significant hearing loss. Detecting this problem in infancy depends on understanding high-risk conditions as well as behaviors and responses that suggest a hearing loss. **High-risk factors** include birth weight < 1500 gm; Apgar score ≤ 5 at 5 min; serum bilirubin > 22 mg/dL in an infant whose birth weight is > 2000 gm, 17 mg/dL in a baby < 2000 gm; anoxia; neonatal sepsis or meningitis; neonatal hyperbilirubinemia; seizures or apneic spells; congenital intrauterine infection, such as rubella, cytomegalovirus, toxoplasmosis; drugs such as streptomycin; or a history of early hearing loss in a parent or close relative.

In about 1/3 of infants deaf from birth, a hereditary recessive etiology that is not present in either parent is assumed. These children must be identified by **observations that the parents can learn to make.** By age 3 mo an infant can be expected to startle to a nearby loud sound, stir or awaken from sleep when someone talks or makes a noise, and be soothed by the mother's voice. By age 6 mo an infant should look toward an interesting sound, turn when his name is called, make sounds such as "moo", "ma", "da", and "di" to toys or objects, and coo when listening to music. By age 10 mo the infant should make sounds on his own, imitate some sounds made by others, and understand "no" and "bye-bye". By age 18 mo the appropriate use of a few single words, the understanding of many single words or commands, and babbling in sentence-like patterns is expected. Infants who do not pass these minimal performance standards should be referred for hearing testing.

Ear infection, serous fluid middle ear accumulation, or frequent respiratory infections may cause enough hearing loss in infants and children to seriously affect development of language skills. Prompt audiologic referral may be indicated.

Early identification and correction of hearing loss is essential for normal development of communication skills. If history-taking identifies risk factors, infant audiometry should be done by age 3 mo. Profound hearing loss may be suspected by parents if their infant does not seem to respond to a spoken voice or ordinary household sounds. Parents' observations are very important, and questions they raise about a child's hearing should be investigated.

Special audiometric technics, usually performed by an audiologist, can assess hearing ability starting at birth. These tests use reflexive, behavioral, and physiologic auditory responses to stimuli of controlled intensity.

In the infant from birth to age 6 mo, evaluation involves eliciting reflexive responses **(auropalpebral, Moro, startle)** to relatively intense levels of sound. These tests may be administered manually with a hand-held audiometric device or with automated devices designed to present auditory stimuli and record body movements in response to the stimulation.

In the child from age 6 mo to 2 yr, localization responses to tones and speech are evaluated. In Conditioned Orientation Response **(COR)** audiometry, sometimes called Visual Response audiometry **(VRA),** a lighted toy that is mounted on a loudspeaker is flashed following the presentation of the test tone. After a brief conditioning period, the child will localize toward the tone, if audible, in anticipation of the flashing toy. A threshold recorded in this manner is called a minimal response level **(MRL),** since true thresholds may be slightly less than the levels required to elicit these behavioral responses.

In the child ≥ 12 mo of age, the speech reception threshold **(SRT)** is determined by having the child point to body parts or identify common objects in response to speech of controlled intensity. Although this technic will quantify the child's hearing level for speech, it may fail to identify a low frequency conductive loss or a high frequency sensorineural loss that does not affect the speech frequencies. When depressed SRTs are noted by air conduction, comparison with bone conduction SRTs may determine if the loss is conductive or sensorineural in nature.

In the child above age 36 mo, play audiometry is used. This technic involves conditioning the child to perform a task (place a block in a box, etc) in response to a tone. Play audiometry is usually used until age 4 or 5, when the child can respond by raising his hand.

Tympanometry and acoustic reflex measurements can be used with any age child and do not depend on voluntary responses from the patient. The **SPAR** test (Sensitivity Prediction from the Acoustic Reflex) is sometimes used to estimate hearing ability. In the normal ear, the difference between acoustic reflex levels for broad band noise and pure tones is about 20 dB, but decreases in sensorineural losses. The determination of this difference can be used to predict hearing level in an uncooperative or unresponsive child. In addition to the SPAR, electrocohleography and brainstem-evoked response audiometry may be used.

Differentiation of Sensory (Cochlear) and Neural (8th Nerve) Hearing Losses

The term *sensorineural* indicates that it is not certain whether the loss of hearing is due to a lesion in the inner ear or in the 8th nerve. The differentiation between sensory (cochlear) and neural (8th nerve) hearing loss is clinically important. **Sensory hearing losses** result from end-organ lesions (acoustic trauma, viral endolymphatic labyrinthitis, ototoxic drugs, Ménière's disease) that usually represent no threat to life. On the other hand, **neural hearing losses** are frequently due to potentially fatal cerebellopontine angle tumors and a wide variety of other neurologic disorders.

Sensory and neural hearing losses may be differentiated on the basis of tests for discrimination, performance intensity function for phonetically balanced words **(PIPB),** recruitment, acoustic reflex decay, sensitivity to small increments in intensity, pathologic adaptation, and auditory brainstem responses (see also Table 204-1).

Sensory hearing losses due to cochlear lesions are characterized by mild to moderate loss of discrimination for speech, improved discrimination with increasing intensity, presence of recruitment, absence of acoustic reflex decay, high sensitivity for small increments in intensity, mild tone decay, and well-formed waves with normal latencies in brainstem response audiometry.

Neural hearing losses are characterized by severe loss of discrimination for speech, deteriorating discrimination with increasing intensity, absence of recruitment, presence of acoustic reflex decay, poor sensitivity for small increments in intensity, marked tone decay, and abnormally long latencies of wave forms or absence of wave forms in brainstem response audiometry.

The following diagnostic studies are used to differentiate sensory from neural hearing losses:

Discrimination for phonetically balanced words is described above.

Performance intensity function for phonetically balanced words (PI-PB), as mentioned above, is tested at 25 to 40 dB above the ST. Once the discrimination score is determined, discrimination may be determined at higher intensities. With sensory hearing

losses, discrimination usually improves at higher intensities. With neural hearing losses, discrimination characteristically deteriorates at higher intensities. If an articulation function (discrimination as a function of intensity) is plotted, a "rollover" or decrement in discrimination is seen with increasing intensity in patients with an 8th nerve lesion.

Recruitment (abnormal increase in the perception of loudness or the ability to hear loud sounds normally despite a hearing loss) can be demonstrated by having the patient compare the loudness of sounds in the affected ear with the loudness of sounds in the normal ear. In sensory hearing losses, the sensation of loudness in the affected ear increases more with each increment in intensity than it does in the normal ear. In neural hearing losses, the sensation of loudness in the affected ear increases no more with each increment in intensity than it does in the normal ear (no recruitment) or increases less with each increment in intensity than it does in the normal ear (decruitment).

Acoustic reflex decay: As mentioned above, this reflex response adapts or decays with continuous presentation of a tone, particularly below 2000 Hz, over time mildly in sensory hearing losses and severely in neural hearing losses.

Sensitivity to small increments in intensity can be demonstrated by presenting a continuous tone at a hearing level (dB level above audiometric zero) of 75 dB and increasing the intensity by 1 dB briefly at irregular intervals. The percentage of small increments that the patient can detect yields the **short increment sensitivity index (SISI).** A high SISI (60 to 100%) is characteristic of sensory hearing losses, while a patient with an 8th nerve lesion can detect < 30% of the small changes in intensity.

Pathologic adaptation is demonstrated when a patient cannot continue to perceive a constant tone above the threshold of hearing **(tone decay).** The tone decay is mild in sensory lesions and severe in neural lesions.

Several of these phenomena may be demonstrated with the **Békésy automatic audiometer,** in which the intensity of the stimulus can be controlled by the patient. The patients is instructed to depress a button when he hear the stimulus, which causes the intensity of the stimulus to decrease. When the stimulus is no longer audible, the patient releases the button, and the intensity begins to increase. In this way the patient traces back and forth across his threshold of hearing. Over the course of a 6.5 min period, the frequency of the test tone may be gradually increased from 100 to 10,000 Hz. If pathologic adaptation is present, it can be demonstrated by decay of the response to a continued presentation of the test stimulus. Decay of the response can be reduced or eliminated by interrupting the tone for 0.5 sec every second. Testing with continuous and interrupted tone presentations yields 5 patterns of tracings. In the Type I pattern, the continuous and interrupted tracings are superimposed. This pattern is found in normal hearing and in conductive hearing losses. In the Type II pattern, the 2 tracings are superimposed up to 1000 Hz. Above this frequency, the threshold for the continuous tones increases by about 20 dB from that of the interrupted tones, and in the higher frequencies the excursions of the continuous tracings become smaller. This pattern is characteristic of sensory hearing losses, as in Ménière's disease, and indicates mild pathologic adaptation. In the type III pattern, the continuous tracing separates sharply from the interrupted tracing at a lower frequency, and excursions of the continuous tracing do not become smaller. This pattern is characteristic of neural lesions, such as acoustic neuromas,

and indicates severe physiologic adaptation. In the Type IV pattern, the continuous tracing separates from the interrupted tracing at all frequencies, and excursions of the continuous tracing may or may not become smaller. This pattern indicates active cochlear lesions (such as a recent attack of Ménière's disease) or early neural lesions. In the Type V pattern, the apparent thresholds of the continuous and interrupted tones are separated but the apparent thresholds of the interrupted tones are greater than those of the continuous tones. This pattern occurs in psychogenic or feigned hearing loss.

Brainstem response audiometry (BRA) is the most powerful technic available to differentiate sensory from neural hearing losses. Five distinct electric waves, generated in the 8th nerve and brainstem in response to acoustic stimulation and categorized by Jewett as I, II, III, IV, and V, can be recorded from the head by computer averaging the responses to many stimuli. Each wave probably emanates from a distinct structure in the auditory pathway such as the 8th nerve, cochlear nuclei, superior olivary complex, lateral lemniscus, and inferior colliculus. With lesions of the 8th nerve, one or more wave forms may be lost, the latency of the wave forms from the onset of the acoustic stimuli may be increased, and the interwave latencies may be prolonged. With cochlear lesions, the wave forms are easily recognized and the latency relationship remain normal.

Patients with complaints referable to one cranial nerve, such as the 8th cranial nerve, deserve thorough neurologic evaluation. Emphasis has been placed in this discussion on thorough evaluation of the auditory division of the 8th nerve and its end-organ. Further evaluation of the patient should include vestibular testing (see below) and may require CT of the head with enhancement (paying special attention to the internal auditory canals) and air-contrast, computed cysternography.

Central Auditory Imperception

Lesions of the central auditory pathways (cochlear nuclei, brainstem pathway crossing the midline (trapezoid body, dorsal stria of Held, and stria of von Monakow), superior olivary complex, lateral lemniscus, inferior colliculus, medial geniculate body, auditory radiation, and auditory cortex) characteristically do not result in elevation of pure-tone and STs and decreased discrimination for single words. Special tests are required to bring out the deficit in auditory function with lesions of the central auditory pathways. These tests (1) measure discrimination of degraded or distorted connected speech, (2) measure discrimination in the presence of a competing message in the other ear, (3) evaluate the ability to fuse into a meaningful message incomplete or partial messages to each ear, and (4) localize sound in space (median plane localization) when the acoustic stimuli are delivered simultaneously to each ear.

Speech may be degraded or distorted with low frequency or high frequency filters, periodic interruptions, or time compression. There is a loss of discrimination of degraded or distorted connected speech in the ear contralateral to a cortical lesion. Likewise, presenting a competing message in the ipsilateral ear results in a loss of discrimination in the ear contralateral to a cortical lesion. Brainstem lesions produce a loss of ability to fuse incomplete messages presented to each ear into a meaningful message and impair the ability to make accurate localizations of sound in space.

Tinnitus

Perception of sound in the absence of an acoustic stimulus. Tinnitus, a subjective experience of the patient, is distinguished from **bruit,** noise that may be hear by the examiner and often by the patient as well.

Tinnitus may be of a buzzing, ringing, roaring, whistling, or hissing quality or may involve more complex sounds that vary over time. Tinnitus may be intermittent, continuous, or pulsatile (synchronous with the heart beat). An associated hearing loss is usually present.

The mechanism involved in tinnitus remains obscure. Tinnitus may occur as a symptom of nearly all ear disorders, including obstruction of the external auditory canal due to cerumen and foreign bodies, infectious processes (external otitis, myringitis, otitis media, labyrinthitis, petrositis, syphilis, meningitis), eustachian tube obstruction, otosclerosis, middle ear neoplasms such as the glomus tympanicum and glomus jugulare tumors, Ménière's disease, arachnoiditis, cerebellopontine angle tumors, ototoxicity (due to salicylates, quinine and its synthetic analogs, aminoglycoside antibiotics, certain diuretics, carbon monoxide, heavy metals, alcohol, etc), cardiovascular diseases (hypertension, arteriosclerosis, aneurysms, etc), anemia, hypothyroidism, hereditary sensorineural hearing loss, noise-induced hearing loss, acoustic trauma (blast injury), and head trauma.

Evaluation of the patient with tinnitus requires the minimum comprehensive audiologic assessment described above as well as CT of the temporal bone. Finding a sensorineural hearing loss indicates the testing described above for differentiating sensory and neural hearing losses. Pulsatile tinnitus requires investigation of the vascular system with carotid and vertebral arteriograms to exclude arterial obstruction, aneurysms, and vascular neoplasms.

Treatment

The patient's ability to tolerate the tinnitus varies. Treatment should be directed toward the underlying disease, since its amelioration may produce improvement in the tinnitus. Correction of the associated hearing loss usually results in relief of the tinnitus. Although there is no specific medical or surgical therapy for tinnitus, many patients find relief by playing background music to mask the tinnitus and even go to sleep with the radio playing. A hearing aid for the associated hearing loss often results in suppression of the tinnitus. Some patients benefit from use of a tinnitus masker, a device that is worn like a hearing aid and that presents a noise more pleasant than the tinnitus. Electrical stimulation of the inner ear, as with a cochlear implant, often reduces the tinnitus but is appropriate only for the profoundly deaf.

Clinical Evaluation of the Vestibular Apparatus

Patients with vertigo, difficulty with balance, or a sensorineural hearing loss of unknown etiology should have vestibular function tested. Evaluation of vestibular function includes rapidly alternating movement, finger-to-nose, heel-to-shin, and Romberg tests; gait testing; and electronystagmography **(ENG)** with caloric testing. Since the results in each ear can be compared, caloric tests are more useful clinically than stimulation with acceleration or deceleration in rotational, torsion swing, and lateral swing tests.

Artificial stimulation of the vestibular apparatus produces nystagmus, past-pointing, falling, and autonomic responses such as sweating, vomiting, hypotension, and bradycardia. **Nystagmus** (see also in Ch. 119), the most useful response, can be monitored visually or, more reliably, by recording changes in the corneoretinal potential (electronystagmography). Vestibular nystagmus is a rhythmic movement of the eyes. It has a quick and a slow component and may be rotary, vertical, or horizontal. The direction of the nystagmus is determined by the direction of the quick component because it is easier to see. However, the slow component is the more fundamental response to vestibular stimulation, while the quick component is compensatory. The slow component moves in the direction of the movement of the endolymph. Past-pointing and falling are also in the direction of movement of the endolymph. The hallucination of the movement of the environment is in the direction of endolymphatic flow, and the hallucination of the movement of the subject is in the direction opposite to that of endolymphatic flow.

ENG electronically detects spontaneous gaze, or positional nystagmus that might not be visually detectable. Eye tracking of a moving target and the response to optokinetic stimulation with a rotating striped drum are conveniently recorded electronically at the time of caloric testing.

Caloric stimulation produces convection currents within the endolymph. These currents cause movement of the cupula in the ampulla of the horizontal semicircular canal; the movement is in one direction during cooling and in the opposite direction during warming.

The **Hallpike caloric test,** an accurate and reproducible measure of vestibular sensitivity, is performed with the patient supine and the head elevated 30 degrees to bring the horizontal semicircular canal into a vertical position. Each ear is irrigated with 240 mL of water delivered in 40 sec, first at 30 °C (86 °F) and then at 44 °C (110 °F). The resulting nystagmus is monitored with the patient gazing straight ahead. Irrigation of the ear with cool water produces nystagmus to the opposite side, warm water produces nystagmus to the same side. A mnemonic device is COWS (cold to the opposite side and warm to the same).

The duration of the nystagmus, the velocity of the slow component, or the frequency of the nystagmus may be measured. **Canal paresis,** a unilateral reduction or absence of sensitivity, and **directional preponderance,** a relative exaggeration of the nystagmic response in one direction, can be demonstrated. Various combinations of canal paresis and directional preponderance may coexist. The presence of canal paresis, directional preponderance, or combination of the two signals an organic lesion - end organ, 8th nerve, brainstem, or cerebellar - but does not necessarily indicate on which side the lesion is. Occasionally, an important differential point rests on the caloric examination. Acoustic neurinomas frequently show canal paresis or complete lack of response on the side of the neoplasm.

Patients with vertigo should have a minimum comprehensive audiologic assessment and CT of the head with enhancement as well as the vestibular evaluation described above.

Earache

Pain occurs with infections and neoplasms in the external ear and middle ear, or is referred to the ear from remote disease processes. Even mild inflammation in the external

auditory canal produces severe pain. Perichondritis of the pinna produces severe pain and tenderness. With eustachian tube obstruction, abrupt changes in middle-ear pressure relative to atmospheric pressure may result in painful retraction of the tympanic membrane. Infection in the middle ear results in painful inflammation of the middle-ear mucous membrane and pain due to increased pressure in the middle ear with bulging of the tympanic membrane. The commonest cause of earache in children, acute otitis media, requires prompt examination by a physician and antibiotic therapy to prevent serious sequelae. In the absence of disease in the ear, the source of referred otalgia should be sought in those areas receiving sensory supply from the cranial nerves that subserve sensation in the external ear and middle ear - i.e. the trigeminal, glossopharyngeal, and vagus nerves. Specifically, the cause of obscure otalgia should be sought in the nose, paranasal sinuses, nasopharynx, teeth, gingiva, temporomandibular joints, mandible, tongue, palatine tonsils, pharynx, hypopharynx, larynx, trachea, and esophagus. Occult neoplasms in these locations often first make their presence known by pain referred to the ear.

Treatment depends on identifying the cause of the pain and providing the therapy appropriate for that disease.

Vertigo

An abnormal sensation of rotary movement associated with difficulty with balance, gait, and navigation in the environment. The sensation may be subjective: the patient feels he is moving relative to his environment; or it may be objective: he feels the environment is moving relative to him. Vertigo results from lesions or disturbance in the inner ear, 8th nerve, or vestibular nuclei and their pathways in the brainstem and cerebellum.

Chapter 205: External Ear

Obstructions

Cerumen may obstruct the ear canal and cause itching, pain, and a temporary conductive hearing loss. It may be removed by irrigation, but rolling the cerumen out of the ear canal with a blunt curet or loop is quicker, less messy, and more comfortable for the patient. Irrigation is contraindicated if there is a history of otorrhea, perforation of the tympanic membrane, or recurrent external otitis. Allowing water into the middle ear through a perforation may exacerbate chronic otitis media. Cerumen solvents are not recommended because they often do not solve the problem and frequently cause maceration of the skin of the canal and allergic reactions.

Children insert all types of objects into their ear canals, such as beads, erasers, or beans. A **foreign body** in the ear canal is best removed by raking it out with a blunt hook. Forceps tend to push smooth objects deeper. A foreign body lying medial to the isthmus is difficult to remove without injuring the tympanic membrane and ossicular chain. Metal and glass beads may sometimes be removed by irrigation, but with a hygroscopic foreign body (i.e. a bean) adding water causes it to swell and complicates its removal. A general anesthetic is needed for an uncooperative child or when there is a difficult mechanical problem.

Insects in the ear canal are most annoying while alive. Filling the ear canal with mineral oil will kill the insect and give immediate relief, and will facilitate its removal with forceps.

External Otitis

Infection in the ear canal may be localized **(furuncle)** or diffuse, involving the entire canal **(generalized or diffuse external otitis).** External otitis is more common during the summer swimming season. It is often called **swimmer's ear.**

Etiology

Generalized external otitis may be caused by a gram-negative rod such as *Escherichia coli, Pseudomonas aeruginosa,* or *Proteus vulgaris;* by *Staphylococcus aureus;* or, rarely, by a fungus. Furuncles are usually due to *S. aureus.* Certain persons (i.e. individuals with allergies, psoriasis, eczema, and seborrhea dermatitis) are particularly prone to develop external otitis. Predisposing factors include getting water or various irritants such as hair spray or hair dye in the ear canal, and trauma from cleaning the canal. The ear canal is self-cleansing by the movement of desquamated epithelium, like a conveyor belt, from the tympanic membrane outward. The patient's attempts to clean the canal with cotton applicators interrupt the self-cleansing mechanism and promote accumulation of debris by pushing it in the direction opposite to the movement of the desquamated epithelium. Debris and cerumen tend to trap water allowed into the canal; the resulting skin maceration sets the stage for invasion of pathogenic bacteria.

Symptoms and Signs

Patients with diffuse external otitis complain of itching, pain, a foul-smelling discharge, and loss of hearing if the canal becomes swollen or filled with purulent debris. Tenderness on traction of the pinna and on pressure over the tragus tends to distinguish it from otitis media. The skin of the external auditory canal appears red, swollen, and littered with moist, purulent debris.

Furuncles cause severe pain and, when a furuncle drains, brief sanguineous purulent otorrhea.

Treatment

Systemic antibiotics are seldom necessary unless there is a spreading cellulitis. In **diffuse external otitis,** topical antibiotics and corticosteroids are effective. The infected debris is first gently removed from the canal with suction or dry wipes of cotton. A solution containing neomycin sulfate 0.5% and polymyxin B sulfate 10,000 u./mL is effective against the usual gram-negative rods, while adding a topical corticosteroid such as 1% hydrocortisone reduces the swelling and allows antibiotic penetration into the depth of the canal; 5 drops are instilled tid for 7 days. External otitis also responds to alteration of the pH of the canal with topical 2% acetic acid 5 drops tid for 7 days. An analgesic such as codeine 30 mg orally q 4 h is usually necessary for the first 24 to 48 hours. If there is cellulitis extending beyond the ear canal, penicillin G 250 mg orally q 6 h for 7 days is indicated.

Furuncles should be allowed to drain spontaneously, since incision may lead to a spreading perichondritis of the pinna. Topical antibiotics are *ineffective.* Analgesics such as codeine 30 mg orally q 4 h are necessary to relieve the pain. Dry heat is also helpful in relieving the pain and hastens resolution.

Perichondritis

Trauma, insect bites, and incision of superficial infections of the pinna may initiate perichondritis, which causes an accumulation of pus between the cartilage and the perichondrium. The blood supply to the cartilage is provided by the perichondrium. If the perichondrium is separated from both sides of the cartilage, the resulting avascular necrosis leads to a deformed pinna. Septic necrosis also plays a role. The infection tends to be indolent, long-lasting, and destructive. Perichondritis is usually caused by a gram-negative rod. **Treatment:** Wide incision and suction drainage is used to reapproximate the blood supply to the cartilage. Systemic antibiotics therapy is indicated and should be guided by culture and sensitivity studies; often IV therapy with an aminoglycoside antibiotic and a synthetic penicillin is required.

Aural Eczematoid Dermatitis

Eczema, characterized by itching, redness, discharge, desquamation, and even fissuring leading to secondary infection, frequently involves the pinna and ear canal. Recurrences are common. **Treatment:** Dilute aluminum acetate solution (Burow's solution) is applied as often as required. Itching and inflammation can be reduced with topical corticosteroids. Topical antibiotic therapy as described above for diffuse external otitis may be needed occasionally.

Malignant External Otitis

Pseudomonas osteomyelitis *of the temporal bone.*

Malignant external otitis occurs mainly in elderly diabetics, beginning as an external otitis caused by *Pseudomonas aeruginosa* and becoming a pseudomonas osteomyelitis of the temporal bone. It is characterized by persistent and severe earache, foul-smelling purulent otorrhea, and granulation tissue in the external auditory canal. There may be varying degrees of conductive hearing loss. Frequently, facial nerve paralysis occurs. Increased radiodensity throughout the air-cell system in the temporal bone and middle ear and radiolucency of the temporal bone develop. Biopsy of the tissue in the ear canal is necessary to differentiate the condition from a malignant neoplasm. The osteomyelitis spreads along the base of the skull and may cross the midline. Surgical therapy is usually not helpful or necessary. Careful control of the diabetes and prolonged (6 wk) IV therapy with an aminoglycoside antibiotic and a synthetic penicillin result in complete resolution in most cases.

Trauma

Hematoma

A subperichondral hematoma may result from blunt trauma to the pinna. The external ear becomes a shapeless, reddish-purple mass when blood collects between the perichondrium

and the cartilage. Since the perichondrium carries the blood supply to the cartilage, avascular necrosis of the cartilage may occur. The "cauliflower ear" characteristic of wrestlers and boxers is the consequence of an organized and calcified hematoma. **Treatment:** The clot must be evacuated through an incision, and the skin and perichondrium are reapproximated to the cartilage with suction drainage to keep the cartilage and its blood supply in close approximation.

Lacerations

For lacerations of the external ear that penetrate the cartilage and the skin on both sides, the skin margins are sutured, the cartilage is splinted externally with benzoin-impregnated cotton, and a protective dressing is applied. Sutures should not extend into the cartilage.

Fractures

Forceful blows to the mandible may be transmitted to the anterior wall of the ear canal (posterior wall of the glenoid fossa). Displaced fragments from fractures of the anterior wall of the canal may cause stenosis of the canal and must be reduced or removed under general anesthesia.

Tumours

Sebaceous cysts, osteomas, and **keloids** may arise in and occlude the ear canal and cause retention of cerumen and a conductive hearing loss. Excision is the treatment of choice.

Ceruminomas arise in the outer third of the external auditory canal. Although these neoplasms appear benign histologically, *they behave in a malignant manner and should be excised widely.*

Basal cell and squamous cell carcinomas frequently develop on the external ear following regular exposure to the sun. Early lesions can be successfully treated with cautery and curettage or irradiation. More advanced lesions involving the cartilage require surgical excision of V-shaped wedges or larger amounts of the external ear. Invasion of cartilage makes irradiation therapy less effective and surgery the preferred treatment. Basal cell and squamous cell carcinomas may also arise in or secondarily invade the external auditory canal. Persistent inflammation in chronic otitis media may predispose to development of squamous cell carcinoma. Extensive resection is indicated, followed by radiation therapy. En bloc resection of the external auditory canal with sparing of the facial nerve is performed when lesions are limited to the canal and have not invaded the middle ear.

Chapter 206: Tympanic Membrane and Middle Ear

The patient with a middle ear disorder may present with one or more of the following complaints; a feeling of fullness or pressure in the ear; constant or intermittent, mild to excruciating pain; otorrhea; diminished hearing; tinnitus; and vertigo. In acute otitis media, systemic symptoms (i.e. fever) are commonly present in addition. The symptoms may begin with a feeling of fullness and progress serially in additive fashion. Infants and children,

especially, may be febrile and present with other prominent systemic manifestations (anorexia, vomiting, diarrhea, lethargy, etc).

The symptoms may result from infection, trauma, and disturbed pressure relationships secondary to eustachian tube obstruction. In determining the cause, the physician should elicit information about antecedent and associated symptoms (i.e. rhinorrhea, nasal obstruction, sore throat, upper respiratory infection, allergic manifestations; headache or other evidence of meningeal involvement; systemic symptoms). The appearance of the external auditory canal and tympanic membrane often yields diagnostic clues; the nose, nasopharynx, and oropharynx should also be examined for signs of infection and allergy and for evidence of an underlying disorder - i.e. a neoplasm of the nasopharynx.

The function of the middle ear should be evaluated with pneumatic otoscopy, the Weber and Rinne tuning fork tests, tympanometry, and audiometry.

Trauma

The tympanic membrane may be punctured and the tympanum penetrated by objects placed in the ear canal (i.e. cotton applicators) or entering the canal accidentally (i.e. twigs on a tree or missiles such as pencils or hot slag). A sudden overpressure, as in an explosion (acoustic trauma), a slap, or swimming and diving accidents, or a sudden negative pressure, as in a kiss over the ear, also can perforate the tympanic membrane. Penetration of the eardrum may cause dislocations of the ossicular chain, fracture of the footplate of the stapes, displacement of fragments of the ossicles of missile into the inner ear, a perilymph fistula from the oval or round window, or facial nerve paralysis.

Symptoms and Signs

Traumatic perforation of the tympanic membrane results in sudden severe pain followed by bleeding from the ear. A loss of hearing and tinnitus occur. The loss of hearing is more severe if there has been a disruption of the ossicular chain or trauma to the inner ear. Vertigo suggests an associated injury to the inner ear, as occasionally a portion of the stapes or a missile is driven into the inner ear. Purulent otorrhea may begin in 24 to 48 h, particularly if water gets into the middle ear.

Treatment

Following perforation, oral penicillin G or V 250 mg q 6 h should be given for 7 days to prevent infection. Aseptic technic is used in examining the ear. If necessary, under local anesthesia and microscopic control, the displaced flaps of tympanic membrane may be laid in their original position to facilitate healing. The ear is kept dry, and topical medication with 2% acetic acid, 5 drops tid, though not used prophylactically, may be employed if the ear becomes infected. Spontaneous closure of the perforation is usual; a tympanoplasty is indicated if the perforation does not heal spontaneously within 2 mo. A persistent conductive hearing loss suggests discontinuity of the ossicular chain, and the middle ear should be explored surgically and repaired. A sensorineural hearing loss or vertigo that persists for hours or longer following the injury indicates penetration of the inner ear and requires an exploratory tympanoplasty to repair the damage as soon as possible.

Barotitis Media

(Aerotitis)

Damage to the middle ear due to ambient pressure changes. During a sudden increase in ambient pressure, as in descent of an airplane or in deep sea diving (see appropriate chapters in &21), gas must move from the nasopharynx into the middle ear to maintain equal pressure on both sides of the tympanic membrane. If the eustachian tube is not functioning properly, as in URI or allergy, the pressure in the middle ear will result in retraction of the tympanic membrane, and a transudate of blood from the vessels in the lamina propria of the mucous membrane will form in the middle ear. If the difference in pressure becomes great, ecchymosis and subepithelial hematoma may develop in the mucous membrane of the middle ear and in the tympanic membrane. Very severe pressure differentials cause bleeding into the middle ear and rupture of the tympanic membrane. A perilymph fistula through the oval or round windows may occur. Pressure differentials between the middle ear and the ambient pressures usually produce severe pain and a conductive hearing loss. A sensorineural hearing loss or vertigo during descent suggests the possibility of a perilymph fistula while the same symptoms during ascent from an aquatic dive suggest bubble formation in the inner ear.

An individual with an acute URI or allergic reaction should be advised not to fly or dive, but if these activities are undertaken, a nasal vasoconstrictor such as phenylephrine 0.25% applied topically 30 min before descent is of prophylactic value.

Infectious Myringitis

(Bullous Myringitis)

Inflammation of the tympanic membrane secondary to viral or bacterial infections. Vesicles develop in the tympanic membrane in viral infections and in acute bacterial (particularly *Streptococcus (Diplococcus) pneumoniae)* and mycoplasmal otitis media. Pain is sudden in onset and persists for 24 to 48 h. Hearing loss and fever, when they occur, suggest bacterial otitis media. **Treatment:** Since it is difficult to differentiate a viral from a bacterial or mycoplasmal otitis, antibiotic therapy as for acute otitis media is indicated. Pain may be relieved by rupture of the vesicles with a myringotomy knife or by analgesia with a narcotic such as codeine orally q 4 h as necessary.

Acute Otitis Media

A bacterial or viral infection in the middle ear, usually secondary to a URI. While it can occur at any age, it is most common in young children, particularly from age 3 mo to 3 yr. Microorganisms may migrate from the nasopharynx to the middle ear over the surface of the eustachian tube mucous membrane or by propagating in the lamina propria of the mucous membrane as a spreading cellulitis or thrombophlebitis.

Etiology

In newborn infants, gram-negative enteric bacilli, particularly *Escherichia coli,* and *Staphylococcus aureus* cause suppurative otitis media. In older infants and young children (<

8 yr of age), *Streptococcus (Diplococcus) pneumoniae, Hemophilus influenzae,* Group A beta-hemolytic streptococci, and *S. aureus* are the causative microorganisms in suppurative acute otitis media. Viral otitis media usually becomes secondarily invaded by one of these microorganisms. In those > 8 yr of age, *H. influenzae* is a less frequent causative microorganism and *S. pneumoniae,* group A beta-hemolytic streptococci, and *S. aureus* are the causative organisms. The relative frequency of the microorganisms causing acute otitis media varies according to which microorganisms are epidemic in the community at any given time. After the neonatal period, *E. coli* rarely causes acute otitis media. Likewise, *Klebsiella pneumoniae* and *Bacteroides* species rarely cause acute otitis media.

Symptoms and Signs

The first complaint usually is of persistent, severe earache. Hearing loss may occur. Fever (up to 40.5 °C (105 °F)), nausea, vomiting, and diarrhea may occur in young children. The tympanic membrane is erythematous and may bulge; landmarks become indistinct, and the light reflex is displaced. Bloody, then serosanguinous and finally purulent, otorrhea may follow spontaneous perforation of the tympanic membrane.

Complications

Serious complications include acute mastoiditis, petrositis, labyrinthitis, facial paralysis, conductive and sensorineural hearing loss, epidural abscess, meningitis (the most common intracranial complication), brain abscess, lateral sinus thrombosis, subdural empyema, and otic hydrocephalus. Symptoms of an impending complication include headache, sudden profound hearing loss, vertigo, and chills and fever.

Diagnosis

Diagnosis is usually made on clinical grounds. The exudate obtained at myringotomy should be cultured, as should spontaneous otorrhea. Nasopharyngeal cultures may be helpful but do not correlate well with the causative agent.

Treatment

Antibiotic therapy is indicated for acute otitis media to relieve the symptoms, hasten resolution of the infection, and reduce the chance of labyrinthine and intracranial infectious complications and of residual damage to the hearing mechanism of the middle ear.

Penicillin G or V 250 mg orally q 6 h for 10 days is the drug of choice in patients over age 8 yr. Amoxicillin 35 to 70 mg/kg/day orally in 3 equal doses q 8 h for 10 days is preferred for those < 8 yr because of the frequency of *H. influenzae* infections. Treatment is continued for 10 to 14 days to ensure resolution and to prevent sequelae. Subsequent therapy depends on cultures, sensitivities, and the clinical course. In penicillin allergy, erythromycin 250 mg orally q 6 h for older children and adults, and a combination of erythromycin 30 to 50 mg/kg/day orally in equally divided doses q 6 h and sulfisoxazole 150 mg/kg/day orally in equally divided doses q 6 h for children < 8 yr may be given for 10 to 14 days.

To improve eustachian tube function, topical vasoconstrictors such as phenylephrine 0.25% 3 drops q 3 h may be instilled into each nasal cavity while the patient is supine and his neck is extended. Such therapy should not exceed 5 to 7 days. Systemic sympathomimetic amines such as ephedrine sulphate, pseudoephedrine, or phenylpropanolamine, 30 mg orally (for adults) q 4 h for 7 to 10 days, may also be helpful. Antihistamines, such chlorpheniramine 4 mg (for adults) orally q 4 to 6 h for 7 to 10 days, may improve eustachian tube function in allergic patients but are not indicated for nonallergic individuals.

Myringotomy should be considered if the tympanic membrane is bulging or if pain, fever, vomiting, and diarrhea are severe or persistent. The patient's hearing, tympanometry, and the appearance and movement of the tympanic membrane should be followed until there is complete resolution.

Secretory Otitis Media

(Serous Otitis Media)

An effusion in the middle ear resulting from incomplete resolution of acute otitis media or obstruction of the eustachian tube. The effusion is often sterile but may contain pathogenic bacteria. Secretory otitis media is common in children. The eustachian tube obstruction may be due to inflammatory processes in the nasopharynx, allergic manifestations, hypertrophic adenoids, or benign or malignant neoplasms. The middle ear is normally ventilated 3 to 4 times/min as the eustachian tube opens during swallowing. O_2 is absorbed by the blood in the vessels of the middle ear mucous membrane, and if the patency of the eustachian tube is impaired, a relative negative pressure develops within the middle ear.

Symptoms and Signs

At first there is mild retraction of the tympanic membrane, with displacement of the light reflex and accentuation of the landmarks; then in the middle ear a transudate from the blood vessels in the mucous membrane develops, recognizable by the amber or gray color that it gives the eardrum and the immobility of the tympanic membrane. An air-fluid level or bubbles of air may be seen through the tympanic membrane; conductive hearing loss occurs. Tympanometry demonstrates maximal compliance with negative pressures in the external auditory canal.

Treatment

In view of the role of pathogenic bacteria in middle ear effusions, a trial of antibiotic therapy as described under acute otitis media is often beneficial and is the first step to be considered in therapy. It is effective in relieving eustachian tube obstruction due to bacterial infection and in sterilizing the middle ear.

Systemic sympathomimetic amines such as ephedrine sulfate, pseudoephedrine, or phenylpropanolamine, 30 mg orally tid (for adults), may improve eustachian tube function by their vasoconstrictive effect. Antihistamines such as chlorpheniramine 4 mg (for adults) orally q 4 to 6 h may relieve eustachian tube obstruction in allergic individuals. Myringotomy may be necessary for aspiration of the fluid and for insertion of a tympanostomy tube, which

allows ventilation of the middle ear and ameliorates the eustachian tube obstruction regardless of the cause. The middle ear may be ventilated on a temporary basis with Valsalva's maneuver or politzerization.

Correction of any underlying condition in the nasopharynx is required. Children may require adenoidectomy, removing lymphoid aggregations on the torus of the eustachian tube and in Rosenmüller's fossa as well as the central adenoid tissue mass, to eradicate persistent and recurrent serous otitis media. Antibiotic therapy should be given to resolve bacterial rhinitis, sinusitis, and nasopharyngitis. Immunologic investigation is occasionally helpful. Any demonstrated allergen should be eliminated from the patient's environment, or immunotherapy should be tried.

Acute Mastoiditis

Infection in the mastoid process resulting in coalescence of the mastoid air cells due to bacterial infection. In acute purulent otitis media, the infection always extends into the mastoid antrum and cells, but progression and destruction of the bony portions of the mastoid process are aborted by suitable antibiotic therapy. The responsible bacteria are the same as those causing acute otitis media. Characteristically, **streptococcal mastoiditis** is preceded by early perforation of the tympanic membrane and profuse otorrhea; **pneumococcal mastoiditis** is likely to be less symptomatic but just as destructive, and advanced coalescence of the mastoid air cells may precede perforation of the tympanic membrane.

Symptoms and Signs

Acute mastoiditis becomes clinically apparent 2 wk or more after the onset of untreated acute otitis media, as one of the cortices of the mastoid process is destroyed. A postauricular subperiosteal abscess may develop as the lateral mastoid cortex is destroyed. Redness, swelling, tenderness, and fluctuation develop over the mastoid process and the pinna is displaced laterally and inferiorly. An exacerbation of the aural pain, fever, and otorrhea usually occurs. The pain tends to be persistent and throbbing, and a creamy, profuse discharge is common. Increasing hearing loss is characteristic.

In acute otitis media, radiographic density of mastoid air cells is increased due to swollen mucous membrane and purulent fluid in the air cells. In coalescent mastoiditis, cell partitions become indistinct and radio-opacity decreases. The individual septa can no longer be seen as the air cells coalesce.

Treatment

The initial antibiotic of choice is penicillin. After a sample of the otorrhea is taken for culture and determination of antibiotic sensitivities, penicillin G 1 million u. IV q 6 h is given. Subsequent IV therapy depends on cultures, sensitivities, and the clinical course. Antibiotic therapy should be continued for at least 2 wk.

A subperiosteal abscess calls for complete exenteration of mastoid air cells (mastoidectomy).

Chronic Otitis Media

A permanent perforation of the tympanic membrane.

Chronic otitis media can result from acute otitis media, eustachian tube obstruction, mechanical trauma, thermal or chemical burns, or blast injuries. Chronic otitis media can be divided into 2 major categories, depending on the type of perforation: (1) the benign central perforation of the pars tensa and (2) the dangerous attic perforations of the pars flaccida and marginal perforations of the pars tensa.

Some substance of the tympanic membrane remains between the rim of the perforation and the bony sulcus tympanicus in **central perforations.** These perforations result in a conductive hearing loss. Exacerbations of chronic otitis media may follows URIs or occur when water gains access to the middle ear in bathing and swimming. They are often caused by gram-negative rods and *Staphylococcus aureus,* and result in painless, purulent otorrhea, which may be foul-smelling. Persistent exacerbations may produce **aural polyps** (granulation tissue that prolapses from the middle ear through the perforation into the external auditory canal) and destructive changes in the middle ear such as necrosis of the long process of the incus.

Pars flaccida (attic) perforations lead into the epitympanum. **Marginal perforations** usually occur in the posterior-superior portion of the pars tensa and there is no substance of tympanic membrane between the edge of the perforation and the bony sulcus tympanicus. Marginal perforations result from an acute necrotizing otitis media that destroys large areas of the tympanic membrane, including the annulus tympanicus and the mucous membrane of the middle ear. These perforations may be associated with a conductive hearing loss, and exacerbations of otorrhea occur as with the central perforations. Complications such as labyrinthitis, facial paralysis, and intracranial suppuration are more likely to occur than with central perforations. Pars flaccida and marginal perforations are frequently associated with **cholesteatomas.**

During the healing of acute necrotizing otitis media, the remaining epithelium of the mucous membrane and the stratified squamous epithelium of the ear canal migrate to cover the denuded areas. Once the stratified squamous epithelium is established in the middle ear, it begins to desquamate and accumulate and a cholesteatoma results. Cholesteatomas may also develop from hyperplasia of the basal layer of the stratified squamous epithelium of the pars flaccida, from progressive retraction of the pars flaccida or the pars tensa, and from squamous metaplasia in the middle ear due to long-standing infection. The desquamated epithelium accumulates in ever-enlarging concentric layers, and collagenases in the epithelium destroy adjacent bone.

Cholesteatomas may be recognized on otoscopic examination by the white debris in the middle ear and the destruction of the external auditory canal bone adjacent to the perforation. Bone destruction due to an otherwise unsuspected cholesteatoma may be demonstrated radiographically. Aural polyps are usually associated with cholesteatomas. The presence of a cholesteatoma, particularly with a pars flaccid perforation, greatly increases the probability of a serious complication (i.e. purulent labyrinthitis, facial paralysis, or intracranial suppurations).

Treatment

In exacerbations of both types of chronic otitis media, the ear canal and middle ear are thoroughly cleaned with suction and dry wipes of cotton; then a solution of 2% acetic acid with hydrocortisone 1.0% is instilled into the ear, 5 to 10 drops tid for 7 to 10 days. Severe exacerbations require systemic therapy with a broad-spectrum antibiotics such as ampicillin 250 to 500 mg orally q 6 h for 10 days or tetracycline 250 mg orally q 6 h for 10 days. Subsequent treatment should be guided by culture and sensitivities of the isolated microorganisms as well as the clinical response of the patient.

In chronic otitis media, the middle ear can generally be repaired. A tympanoplasty restores the two major functions of the tympanic membrane: sound protection for the round window and sound pressure transformation to the oval window. If the ossicular chain has been disrupted, it may also be repaired at the tympanoplasty. Patients with marginal or attic perforations with cholesteatomas requires surgical treatment to exteriorize or to remove the cholesteatoma. Preservation and reconstruction of the middle ear mechanism is less likely in the presence of cholesteatoma.

Otosclerosis

A disease of the bone of the otic capsule and the most common cause of progressive conductive hearing loss in the adult with a normal tympanic membrane. Histologically, foci of otosclerosis show irregularly arranged, new, immature bone interspersed with numerous vascular channels. These foci enlarge and cause ankylosis of the footplate of the stapes and a consequent conductive hearing loss. Otosclerosis also may produce a sensory hearing loss, particularly when the foci of otosclerotic bone are adjacent to the scala media.

The tendency to otosclerosis is familial. About 10% of adult white populations have foci of otosclerosis, but only about 10% of affected persons develop conductive hearing loss. It becomes clinically evident in the late teenage and early adult years. The fixation of the stapes may progress rapidly during pregnancy.

Treatment is with microsurgical technics: The stapes is removed and replaced by a prosthesis; the hearing loss is corrected in most cases. A hearing aid may also improve the hearing of the patients with otosclerosis.

Tumours

Rarely, the middle ear is the site of origin of squamous cell carcinoma. The persistent otorrhea of chronic otitis media may be a predisposing factor. Radiation therapy and resection of the temporal bone is necessary.

Nonchromaffin paraganglioma (chemodectoma), known as glomus jugulare or glomus tympanicum tumors, arise in the temporal bone from glomus bodies in the jugular bulb or the medial wall of the middle ear. They produce a pulsatile red mass in the middle ear. The first symptom is often a tinnitus that is synchronous with the pulse. Hearing loss and, later, vertigo develop. Excision is the treatment of choice. Palliation is achieved with radiation therapy for tumors too large to resect.

Chapter 207: Inner Ear

Vertigo ("Dizziness")

A disturbance in which the individual has a subjective impression of movement in space (subjective vertigo) or of objects moving around him (objective vertigo), usually with a loss of equilibrium.

Etiology

True vertigo, as distinguished from faintness, lightheadedness, or other forms of "dizziness", results from a disturbance somewhere in the equilibratory apparatus: vestibule, semicircular canals; 8th nerve; vestibular nuclei in the brainstem and their temporal lobe connections; and eyes. These structures may be affected by any of a large variety of disorders: (1) **otogenic:** Ménière's syndrome, myringitis, otitis media, acute vestibular neuronitis, herpes zoster oticus, labyrinthitis, middle ear or labyrinthine tumors, petrositis, otosclerosis, obstructed external auditory canal or eustachian tube; (2) **toxic:** alcohol, streptomycin, opiates; (3) **psychogenic:** hysteria; (4) **environmental:** motion sickness; (5) **ocular:** diplopia; (6) **circulatory:** transient vertebrobasilar ischemic attacks; (7) **neurologic:** multiple sclerosis, skull fracture, temporal lobe seizures, encephalitis; (8) **neoplastic:** tumors of the pons, cerebellopontine angle, or 8th nerve; (9) **hematogenic:** leukemia involving the labyrinth.

Stimulation of proprioception in muscles, joints, and tendons amy induce a sense of disequilibrium, but this is not true vertigo.

Diagnosis

Nystagmus, past-pointing, inability to walk a straight line, and persistent deviation to one side when walking all indicate a disturbance of the labyrinthine vestibular apparatus or its CNS connections. Determining whether the vertigo is **peripheral** *(arising from the labyrinth or vestibular nerve)* or **central** *(arising from the vestibular nuclei or their higher connections)* is the first step in establishing the cause.

Peripheral nystagmus is conjugate, horizontal, or horizontal-rotatory, is maximal towards the affected labyrinth, and has its fast component away from the side of the lesion. **Central nystagmus** can be horizontal or vertical, characteristically has its fast component in the direction of gaze to either side, and may be pendular or unequal in the 2 eyes. Pronounced rotary, unidirectional upgaze or downgaze nystagmus always arises from central abnormalities.

Paroxysmal, episodic, or severe attacks of vertigo separated by normal interludes indicate a peripheral etiology. Persistent vertigo or disequilibrium accompanied by nystagmus and gait disturbances usually indicates CNS disease. Unilateral deafness and tinnitus indicate cochlear nerve involvement and are reliable indicators of a peripheral nerve lesion. Labyrinthine disease produces more intense symptoms than does involvement of vestibular nuclei. Headache is more common with central lesions; and other findings such as double vision, slurred speech, incoordination of an extremity, or unilateral weakness are not seen with peripheral lesions.

Vestibular function tests are important - absence of the caloric response indicates a dead labyrinth. Audiogram may differentiate between cochlear and neural hearing loss. Special studies of the vestibular apparatus and CNS often are necessary. Skull x-rays with special views and tomography of the petrous pyramids, CSF examination, and EEG help to exclude pathologic CNS changes. CT scan, MRI, or cerebral angiography may be indicated.

Sudden, episodic attacks of vertigo, tinnitus, and progressive deafness, accompanied by nausea and vomiting and persisting for minutes to hours, are characteristic of **Ménière's disease.** Vertigo persisting for days and weeks may be due to **vestibular neuronitis.** This diagnosis is made from the nonrecurrent nature of the attack, preservation of hearing, and absence of any neurologic signs except nystagmus and equilibratory disturbance. In patients with **postural hypotension** who become vertiginous on changing from recumbent to upright position, examination before and after this shift in posture demonstrates the exaggerated fall in BP. However, vertigo with sudden change in body or head position - i.e. on rising from a recumbent posture, or on rolling over in bed **(postural** or **positional vertigo)** - is more often due to labyrinthine disturbance, as occurs after skull fracture or frequently in older persons for unknown reasons. Vertigo that occurs on sudden turning or, more often, strong extending of the head rarely may be due to **vertebral artery insufficiency** or **tumors of the floor of the 4th ventricle.**

True vertigo is not a symptom of psychoneurosis, but giddiness and fear of losing one's balance while walking may be symptoms of an anxiety neurosis or depression. Diagnosis is established by absence of objective findings, by negative laboratory tests, and by psychologic evaluation.

Treatment

Treatment depends on determining and eliminating the cause. CNS disorders causing vertigo are posterior fossa tumors, cerebellar disorders, and multiple sclerosis.

Symptomatic relief may be obtained by bed rest and dimenhydrinate 50 to 100 mg orally q 4 to 6 h, perphenazine 4 to 8 mg orally or 5 mg IM tid, or meclizine 25 mg orally tid. All these drugs are moderately effective against both intermittent and continuous vertigo in ambulatory patients.

Meniere's Disease

A disorder characterized by recurrent prostrating vertigo, sensory hearing loss, and tinnitus, associated with generalized dilation of the membranous labyrinth (endolymphatic hydrops).

The etiology of Ménière's disease is unknown, and the pathophysiology is poorly understood. The attacks of vertigo appear suddenly, last from a few to 24 h, and subside gradually. The attacks are associated with nausea and vomiting. The patient may have a recurrent feeling of fullness or pressure in the affected ear. The hearing in the affected ear tends to fluctuate, but over the years the hearing progressively worsens. The tinnitus may be constant or intermittent, and may be worse before, after, or during an attack of vertigo. Although only one ear is usually affected, both ears are involved in 10 to 15% of patients.

In **Lermoyez's variant** of Ménière's disease, hearing loss and tinnitus precede the first attack of vertigo by months or years and the hearing may improve with the onset of the vertigo.

Treatment

Treatment is empirical. A number of operations, including sacculotomy, placement of a stainless steel tack through the footplate of the stapes, ultrasonic irradiation, endolymphatic-subarachnoid shunt, and cryosurgery have been advocated for patients who are disabled by the frequency of vertiginous attacks. Vestibular neurectomy relieves the vertigo and usually the hearing is preserved. A labyrinthectomy can be performed if the vertigo is sufficiently disabling and the hearing has degenerated to a useless level.

Symptomatic relief of the vertigo may be obtained with anticholinergic agents (i.e. atropine 1 to 2 mg orally or scopolamine 0.6 mg orally or IM q 4 to 6 h or by transdermal patch) to minimize vagal-mediated GI symptoms, antihistamines (i.e. diphenhydramine, meclizine, or cyclizine 50 mg orally or IM q 6 h) to sedate the vestibular system, or barbiturates (i.e. phenobarbital 100 mg orally or IM q 8 h) for general sedation. Diazepam 2 to 5 mg orally q 6 to 8 h is particularly effective in relieving the distress of severe vertigo by sedating the vestibular system.

Vestibular Neuronitis

A benign disorder characterized by sudden onset of severe vertigo that is persistent at first and then becomes paroxysmal. The disease is thought to be a neuronitis involving the vestibular division of the 8th nerve, and to be viral in origin because of its frequent epidemic occurrence, particularly among adolescents and young adults.

The first attack of vertigo is severe, is associated with nausea and vomiting, and lasts for 7 to 10 days. There is persistent nystagmus toward the affected side. The condition is self-limited and may occur as only a single episode, or several subsequent attacks may occur over the next 12 to 18 mo; each subsequent attack is less severe and of shorter duration. There is no associated hearing loss or tinnitus.

The diagnostic evaluation should include an audiologic assessment, electronystagmography with caloric testing, and CT of the head with enhancement with particular attention to the internal auditory canals to exclude other diagnostic possibilities, such as cerebellopontine angle tumor and brainstem hemorrhage or infarction.

Treatment: Acute attacks of vertigo may be suppressed symptomatically as in Ménière's disease. With prolonged vomiting, IV fluids and electrolytes may be required for replacement and maintenance.

Benign Paroxysmal Positional Vertigo

(Postural or Positional Vertigo; Cupulolithiasis)

Violent vertigo, lasting < 30 sec and induced by certain head positions. The vertigo occurs when the patient lies on one ear or the other. It also occurs when the patient tips his head backward to look up over his head. The vertigo is accompanied by nystagmus. There is no associated hearing loss or tinnitus. Benign paroxysmal positional vertigo usually subsides in several weeks or months but may recur after months or years.

Etiology

Granular basophilic masses in the cupula of the posterior semicircular canal have been demonstrated. It has been suggested that the cupular deposits represent calcium carbonate derived from the otoliths. Etiologic factors appear to be spontaneous degeneration of the utricular otolithic membranes, labyrinthine concussion, otitis media, ear surgery, and occlusion of the anterior vestibular artery.

Diagnosis

A **provocative test for positional nystagmus** may be performed. The patient is first seated on an examining table, then assumes the supine position with his head dependent over the end of the table and turned so that one ear is undermost. After the position has been assumed, a latent period of several seconds will be followed by vertigo, which is severe, is likely to last for 15 to 20 sec, and is accompanied by rotary nystagmus. If the left ear is affected, when it is put undermost the nystagmus will be clockwise; if the right ear, the nystagmus is counterclockwise. When the patient sits up, the response recurs, but the nystagmus is rotary in the reverse direction and is milder. The response fatigues, so that with immediate repetition of the test the response will be less strong.

Positional nystagmus may occur with end-organ or CNS lesion. The latency of the response, the severe subjective sensation, the fatigability of the response, the limited duration, and the direction of the rotary nystagmus distinguish benign paroxysmal positional vertigo from a CNS lesion. The positional nystagmus of CNS lesions lacks latency, fatigability, and the severe subjective sensation. The nystagmus may continue as long as the position is maintained. The positional nystagmus of CNS lesions may be vertical or changing in direction and, if rotary, is likely to be perverted (i.e. not in the anticipated direction).

The diagnostic evaluation should include an audiologic assessment, electronystagmography with caloric testing, and CT of the head with enhancement with particular attention to the internal auditory canals, to exclude other diagnostic possibilities.

Treatment

The patient is instructed to avoid the provocative positions. If benign positional paroxysmal vertigo lasts for as long as a year it can be relieved in most cases by dividing the nerve to the posterior semicircular canal of the affected ear at tympanostomy.

Herpes Zoster Oticus

(Ramsay Hunt Syndrome; Viral Neuronitis and Ganglionitis; Geniculate Herpes)

Invasion of the 8th nerve ganglia and the geniculate ganglion of the facial nerve by the herpes zoster virus, producing severe ear pain, hearing loss, vertigo, and paralysis of the facial nerve.

Vesicles can be seen on the pinna and in the external auditory canal in the distribution of the sensory branch of the facial nerve. Other cranial nerves are often involved, and some degree of meningeal inflammation is common. Lymphocytes may be present in the CSF, and the protein content is often increased. Evidence of a mild generalized encephalitis can be found in many patients. The hearing loss may be permanent or there may be partial or complete recovery. The vertigo lasts for days to several weeks. The facial paralysis may be transient or permanent.

Treatment

Corticosteroid therapy is the treatment of choice; i.e. prednisone 40 mg/day orally for 2 days, then 30 mg/day orally for 7 to 10 days, followed by gradual tapering of the dose. Pain is relieved with codeine 30 to 60 mg orally q 3 to 4 h as necessary, while the vertigo is effectively suppressed with diazepam 2 to 5 mg orally q 4 to 6 h. Decompression of the fallopian canal, indicated when the nerve excitability declines or electroneurography demonstrates a 90% decrement, occasionally relieves the facial paralysis.

Purulent Labyrinthitis

(Suppurative Labyrinthitis)

Invasion of the inner ear by a bacterium. Purulent labyrinthitis may occur secondary to acute otitis media or purulent meningitis. In acute otitis media, the microorganisms may gain access to the inner ear through the oval and round windows; in purulent meningitis, through the cochlear aqueduct. Purulent labyrinthitis is also frequently followed by meningitis as the microorganisms gain access to the subarachnoid space through the cochlear aqueduct.

Purulent labyrinthitis is characterized by severe vertigo and nystagmus. It invariably results in complete hearing loss and, in chronic otitis media and cholesteatoma, is often followed by facial paralysis. **Treatment** includes labyrinthectomy for drainage of the inner ear, radical mastoidectomy, and IV antibiotic therapy appropriate for meningitis.

Sudden Deafness

Severe sensorineural hearing loss that usually occurs in only one ear and develops over a period of few hours or less.

Sudden deafness occurs in about 1:5000 persons every year. Although the sudden onset suggests a vascular etiology (embolism, thrombosis, or hemorrhage) by analogy with vascular accidents in the CNS, the evidence supports a viral etiology in most cases. Sudden deafness

tends to occur in children and young and middle-aged adults who have no evidence of vascular disease. The histopathologic findings in the temporal bone in sudden deafness are unlike those seen in the inner ear of animals with experimental vascular occlusion or embolization, but are similar to those seen in human viral infections of the inner ear that result in sudden deafness - i.e. mumps and measles **(viral endolymphatic labyrinthitis).** The viruses of influenza, chickenpox, mononucleosis, the adenoviruses, and others also produce sudden deafness.

The pathologic findings in individuals with persistent hearing loss due to viral endolymphatic labyrinthitis are similar regardless of the causative virus. The organ of Corti is missing in the basal turn. Individual hair cells tend to be missing. Ganglion cell population are reduced in the basal turn. The stria vascularis becomes atrophic. The tectorial membrane is often rolled up and ensheathed in a syncytium. Reissner's membrane may be collapsed and adherent to the basilar membrane.

Perilymph fistulas between the inner and middle ears occasionally occur with severe ambient pressure changes and with strenuous activities like weight lifting. Fistulas in the oval or round windows result in a sudden or fluctuating sensory hearing loss and vertigo. The patient may experience an explosive sound in the affected ear when the fistula occurs.

Symptoms and Signs

The hearing loss is usually profound, but hearing returns to normal in most patients and partial recovery occurs in others. Tinnitus and vertigo may be present initially. The vertigo usually subsides in several days. If hearing is going to return, it is likely to do so in 10 to 14 days.

Treatment

Although vasodilators, anticoagulants, low mol wt dextran, corticosteroids, and vitamins have all been advocated, no form of treatment is of proven value. In view of the frequent micropetechiae and extravasation of blood that are characteristic of virus-induced inflammatory reactions, vasodilation and anticoagulation may not be indicated. Furthermore, in an inflammatory reaction the cochlear blood flow is already increased as much as is beneficial. Although corticosteroids are not of proven value, their use appears rational - i.e. prednisone 40 mg/day orally for 2 days, then 30 mg/day orally for 5 to 7 days followed by a tapering of dosage. Bed rest also seems advisable.

Surgical exploration of the middle ear should be carried out for a suspected perilymph fistula, and the fistula should be repaired with an autogenous soft tissue graft of fat or fascia.

Congenital Sensorineural Hearing Loss

In the past, epidemics of rubella resulted in the birth of large numbers of children with congenital deafness. Particularly during the first trimester of pregnancy, the rubella virus may invade the developing inner ear **(viral endolymphatic labyrinthitis)** and produce much destruction and a profound sensorineural hearing loss. Other causes of profound congenital sensorineural hearing loss are anoxia during birth, bleeding into the inner ear from trauma to

the base of the skull during delivery (particularly in premature infants), ototoxic drugs given to the mother, erythroblastosis fetalis, and numerous hereditary conditions including Waardenburg's syndrome, albinism, and Hurler's syndrome.

Diagnosis and Treatment

If a child does not develop speech normally, a differential diagnosis of deafness, mental retardation, aphasia, and autism must be considered.

Since children must hear language to learn it, deaf children do not develop language without special training. They require special education, beginning as soon as the hearing loss is identified. Because there is an optimum time for acquisition of language, early diagnosis of deafness in infants is essential.

Amplification with a **hearing aid** should be started as early as possible after diagnosis (even as early as 6 mo of age). In bilateral sensorineural hearing loss, binaural amplification using postauricular or in-the-ear aids is indicated to maximize hearing and permit development of auditory localization.

Noise-Induced Hearing Loss

Any source of intense noise, such as woodworking equipment, chain saws, internal combustion engines, heavy machinery, gunfire, or aircraft, may damage the inner ear. Exposure to intense noise results in loss of hair cells in the organ of Corti. Individuals vary greatly in susceptibility to noise-induced hearing loss, but nearly everyone will lose some hearing if exposed to sufficiently intense noise for a sufficient time. Any noise > 85 dB is damaging. Usually a high-frequency tinnitus accompanies the hearing loss. Loss occurs first at 4 kHz and gradually moves into the lower frequencies with further exposure. In contrast to most sensorineural hearing losses, damage is less at 8 kHz than at 4 kHz. Blast injury, termed acoustic trauma, produces the same kind of sensory hearing loss.

Prevention depends on limiting the length of exposure, reducing the noise at its source, and isolating the person from the sound source. As the intensity of the noise increases, the duration of exposure must be reduced to prevent damage to the inner ear. Noise may be attenuated by wearing ear protectors, i.e. plastic plugs in the ear canals or glycerine-filled cups over the ears. With severe noise-induced hearing loss, a hearing aid is usually helpful.

Presbyacusis

The sensorineural hearing loss that occurs as a part of normal aging. It begins after age 20, first affecting the highest frequencies (18 to 20 kHz) and gradually moving into the lower frequencies; it usually begins to affect the 4- to 8-kHz range by age 55 to 65, although there is considerable variation. Some individuals are severely handicapped by age 60 and some are essentially untouched at 90. Men are affected more often and more severely than women. Stiffening of the basilar membrane and deterioration of the hair cells, stria vascularis, ganglion cells, and cochlear nuclei may play a role in pathogenesis, and presbyacusis appears to be related in part to noise exposure.

Speech reading (lip reading); auditory training, making maximum use of nonauditory clues; and amplification with a hearing aid are helpful.

Ototoxic Drugs

The aminoglycoside antibiotics, salicylates, quinine and its synthetic substitutes, and the diuretics ethacrynic acid and furosemide can be ototoxic. Though affecting both the auditory and vestibular portions of the inner ear, these drugs are particularly toxic to the organ of Corti (cortitoxic). Nearly all ototoxic drugs are eliminated through the kidneys, and renal impairment predisposes to the accumulation of toxic levels. Ototoxic drugs should be avoided in topical medication for the ear in the presence of a perforated tympanic membrane, since they can be absorbed into the inner ear fluids through the secondary tympanic membrane at the oval window.

Streptomycin damages the vestibular portion of the inner ear more readily than the auditory portion. Although vertigo and difficulty with maintaining balance tend to be temporary and eventually completely compensated, severe and permanent loss of vestibular sensitivity may persist, causing difficulty when walking in the dark and Dandy's syndrome (bouncing of the environment with each step). From 4 to 15% of patients receiving 1 gm/day for > 1 wk develop a measurable hearing loss, which usually appears after a short latent period (7 to 10 days) and slowly becomes worse if treatment is continued. Complete, permanent deafness may follow.

Neomycin has the greatest cortitoxic effect of any antibiotic. With large doses given orally or by colonic irrigation for intestinal sterilization, enough may be absorbed to affect hearing, particularly if GI ulceration or other mucosal lesions are present. Neomycin should *not* be used for irrigating wounds or for intrapleural or intraperitoneal irrigation because massive amounts of neomycin may be retained and absorbed and cause deafness. **Kanamycin** and **amikacin** are close to neomycin in cortitoxic potential.

Viomycin shows both cochlear and vestibular toxicity. **Vancomycin** causes hearing loss, especially in the presence of renal insufficiency. **Gentamycin** and **tobramycin** have vestibular toxic properties in man and both vestibular and cochlear toxicity in laboratory animals.

Ethacrynic acid IV has caused profound and permanent hearing loss in gravely ill patients with renal failure who are given concomitant aminoglycoside antibiotic therapy. Transient hearing loss from **furosemide** has been reported.

Salicylates produces hearing loss and tinnitus that is usually reversible. **Quinine** and its synthetic substitutes produce a permanent loss of hearing.

Precautions

Ototoxic antibiotics should be **avoided** in pregnancy. Elderly persons and those with a preexisting hearing loss should not be treated with ototoxic drugs if other effective drugs are available. If possible, before treatment is begun with an ototoxic drug (especially an ototoxic antibiotic), the hearing should be measured in order to document a preexisting

hearing loss. Hearing should be monitored audiometrically as often as daily while treatment is continued. The highest frequencies are usually affected first, and high-pitched tinnitus or vertigo may develop - though they are not reliable warning symptoms. If renal function is impaired, the dosage of renally eliminated ototoxic drugs should be adjusted so that the blood levels do not exceed those required therapeutically. Serum levels of the agent should be monitored to insure that adequate therapeutic levels have been achieved but not exceeded. Although there is some individual variation in susceptibility, not exceeding the recommended blood level will usually conserve the hearing.

Fractures of the Temporal Bone

Ecchymosis in the postauricular skin (Battle's sign) suggests a fracture of the temporal bone. Bleeding from the ear following a skull injury is pathognomonic of a temporal bone fracture. The bleeding may be medial to an intact tympanic membrane, may come from the middle ear through a ruptured tympanic membrane, or may come from a fracture line in the ear canal. A hemotympanum gives the eardrum a blue-black color. CSF otorrhea signifies a communication between the middle ear and the subarachnoid space. Fractures longitudinal to the petrous pyramid (80%) extend through the middle ear and rupture the tympanic membrane; they produce facial paralysis in 15% of cases and a profound sensorineural hearing loss in 35%. The middle ear damage may include disruption of the ossicular chain. Transverse fractures (20%) cross the fallopian canal and the cochlea and nearly always produce facial paralysis and a permanent hearing loss. The hearing can be assessed initially with the Weber and Rinne tuning fork tests and subsequently with audiometry. With CT of the head with special attention to the temporal bone, the fracture can usually be demonstrated.

Treatment

IV penicillin G 1 gm q 6 h should be given for 7 to 10 days to prevent meningitis. Persistent facial paralysis requires decompression of the nerve. Tympanoplasty with repair of the ossicular chain is carried out weeks or months later.

Acoustic Neurinoma

(Vestibular Schwannoma; Acoustic Neuroma; Eight Nerve Tumor)

Acoustic neurinomas are derived from Schwann cells. They arise twice as often from the vestibular division of the 8th nerve as from the auditory division and account for approximately 7% of all intracranial tumors.

As the tumor increases in size, it projects from the internal auditory meatus into the cerebellopontine angle and begins to compress the cerebellum and brainstem. The 5th and later the 7th cranial nerves become involved.

Symptoms, Signs, and Diagnosis

A hearing loss and tinnitus are early symptoms. Although the patient complains of dizziness and unsteadiness, true vertigo is not usually present. The sensorineural hearing loss (see Ch. 204) is characterized by greater impairment of speech discrimination than would be

expected with a cochlear lesion. Recruitment is absent, and the short increment sensitivity index **(SISI)** is low. Tone decay is marked. Usually Békésy audiometry shows a Type III or IV pattern. Acoustic reflex decay and absence of wave forms and increase in latency of the 5th wave in the brainstem response audiometry provide further evidence of a neural lesion. As a rule, caloric testing demonstrates marked vestibular hypoactivity (canal paresis). Early diagnosis is based on the audiologic assessment, particularly brainstem response audiometry, and air contrast computed cisternography.

Treatment

Small tumors may be removed with microsurgical techniques that allow preservation of the facial nerve, using a middle cranial fossa route to preserve the remaining hearing or a translabyrinthine route if no useful hearing remains. Large tumors are removed by a combined translabyrinthine and suboccipital approach.

Hearing Aids

Amplification of sound with hearing aids is helpful for patients with conductive or sensorineural hearing losses that are > 30 dB in the speech frequencies. Hearing aids are also helpful to individuals with predominantly high-frequency sensorineural hearing losses and to individuals with unilateral hearing losses.

Air conduction hearing aids, usually coupled to the ear canal with an airtight seal or open tube, are generally superior to bone conduction hearing aids and are used except when there is some contraindication to using an ear mold or tube, as in atresia of the external auditory canal or persistent otorrhea. The **body aid** type, appropriate for profound hearing loss, is the most powerful; it is worn in the shirt pocket or in a body harness and connected by a wire to the earpiece, or receiver, which is coupled to the ear canal by a plastic insert, or earmold. In infants and young children with profound hearing loss in which it is not possible to determine which ear hears better, the amplification from the body aid is delivered to both ears by using a Y-cord. For moderate to severe hearing losses, a **postauricular** or **ear-level aid,** which fits behind the pinna and is coupled to the ear mold with flexible tubing, is appropriate. **Eyeglass aids,** which are built into the temple bar of eyeglasses, with tubing leading to the earmolds, are usually restricted to individuals who wear eyeglasses continually. The less powerful **in-the-ear aid** is contained entirely within the earmold and fits less conspicuously into the concha and the ear canal; it is appropriate for mild to moderate hearing losses. **Canal aids** are contained entirely within the ear canal and are cosmetically acceptable to many users who would otherwise refuse amplification. The **CROS aid** (for Contralateral Routing of Signals) is used for individuals with monaural hearing; a hearing-aid microphone is placed on the nonfunctioning ear and sound is routed to the functioning ear (either through a wire or miniature radio transmitter), allowing the patient to hear sound from the nonfunctioning side and to develop a limited ability to localize sound. If the better ear also has a loss of hearing, the sound from the poorer side can be amplified and the sound coming to the better side can be amplified, the so-called **BiCROS aid.**

A **bone conduction aid** is sometimes used in hearing losses in which an earmold or tube cannot be used, as in atresia of the ear canal or persistent otorrhea. The oscillator is placed in contact with the head, usually over the mastoid, with a spring band over the head,

and sound is conducted through the bone of the skull to the cochlea. Bone conduction hearing aids require more power and introduce more distortion than air conduction hearing aids and are less comfortable to wear.

In evaluating a patient for a hearing aid, professional advice is required. Selecting the proper hearing aid requires matching the electroacoustic characteristics of the hearing aid with the type of hearing loss on the basis of gain, saturation level, and frequency response. **Gain,** or amplification, refers to the difference between the input and the output of the hearing aid. The more severe the hearing loss, the more gain is generally required. **Saturation level,** the maximum output of the hearing aid regardless of input, is an important consideration for patients with reduced tolerance to sound (as in recruitment). In severe tolerance problems, special circuitry (automatic gain control, or AGC) is available that keeps the output of the aid at tolerable levels. **Frequency response** refers to the gain of the aid as a function of frequency. As a general rule, the frequency response should be selected to provide gain consistent with the patient's audiometric configuration. High frequency accentuation can also be achieved by venting the earmold, which benefits many individuals with a sensorineural hearing loss who have a greater loss in the high frequencies than in the low frequencies.

Cochlear Implants

Profoundly deaf individuals to whom hearing aids are of no help in speech reading (lip reading) or in hearing environmental sounds (doorbells, ringing of the telephone, alarms, etc) may benefit from a cochlear implant. This electronic device consists of a battery-powered processor that converts sound into modulation of an electric current, an internal and external induction coil system that transmits the electrical impulses through the skin, and an electrode (some implants have more than one electrode) connected to the internal induction coil that stimulates the remaining fibers of the auditory division of the 8th cranial nerve. At mastoid surgery, the electrode is inserted into the scala tympani of the basal turn of the inner ear. The internal induction coil is implanted into the bone of the skull posterior and superior to the ear; the external conduction coil is held in place on the skin over the induction coil by magnets in the two coils.

Cochlear implants help with speech reading by allowing the profoundly deaf to distinguish when a word begins and ends, the intonation of the word, the rhythm of the speech, and some speech percepts. Some cochlear implants even allow minimal discrimination of words *without* visual clues. Cochlear implants allow deaf person to hear and distinguish environmental sounds and warning signals. They also help deaf persons modulate their voices to make their speech more intelligible to hearing persons.

Chapter 208: Nose and Paranasal Sinuses

Fractures of the Nose

The nasal bones are fractured more frequently than are other facial bones. The fracture usually includes the ascending processes of the maxilla and often the septum. The torn mucous membrane results in nosebleed. Soft tissue swelling develops promptly and may obscure the break. Septal hematomas may occur between the perichondrium and the quadrilateral cartilage and may become infected; abscess formation leads to avascular and

septic necrosis of the cartilage, with a saddle deformity of the nose.

Diagnosis

A fracture should be suspected if blunt injury causes bleeding from the nose. Diagnosis can ordinarily be established by gently palpating the dorsum (bridge) of the nose for deformity, instability, crepitus, and point tenderness, and is confirmed by x-ray. The most common deformity is deviation of the dorsum of the nose in one direction and depression of the nasal bone and ascending process of the maxilla on the other side.

Treatment

Nasal fractures in adults may be reduced under local anesthesia; children require general anesthesia. The fracture is manipulated into a good position by internal and external traction. A blunt elevator is placed under the depressed nasal bone and the depressed bone is lifted anteriorly and laterally while pressure is applied to the other side of the nose, in order to bring the nasal dorsum to the midline. The position of the nose may be stabilized by internal packing and external splinting. Septal hematomas must be immediately incised and drained. Septal fractures are difficult to hold or position and often require nasal septal surgery later.

Foreign Bodies

(Same as in Ch. 199)

Common in young children, foreign bodies in the nose result in foul-smelling, bloody, unilateral discharge. Mineral salts are deposited on a long-retained foreign body, producing a rhinolith. **Treatment:** Removal usually requires general anaesthesia in a child. Vasoconstriction with a topically applied sympathomimetic amine (i.e. 10 drops of phenylephrine 0.25%) may facilitate removal. A blunt hook is placed behind the foreign body and then drawn forward. Attempts at grasping smooth, firm foreign bodies with forceps tend to push them farther posteriorly. Rhinoliths are difficult to remove because their shape tends to conform to the contour of the nasal passage.

Septal Deviation and Perforation

Deviations of the nasal septum from developmental abnormalities or trauma are common but often are asymptomatic and require no treatment. Septal deviation may cause varying degrees of nasal obstruction and predispose the patient to sinusitis (particularly if the deviation obstructs an ostium of a paranasal sinus) and to epistaxis as a result of drying air currents. **Treatment** of symptomatic deviation of the nasal septum is by septoplasty or submucous resection of the septum.

Septal **ulcers** and **perforations** may follow nasal surgery, repeated trauma such as picking the nose, and granulomatous infections such as TB and syphilis. Crusting about the margins and repeated epistaxis may result. Small perforations may whistle. Topically applied bacitracin 500 u./gm in a petrolatum base reduces the crusting. Although perforations of the nasal septum may be repaired by using buccal or septal mucous membrane flaps, the problem

can be more reliably controlled by closing the perforation with a silastic septal button.

Epistaxis

Bleeding from the nose occurs secondary to local infections such as vestibulitis, rhinitis, and sinusitis; systemic infections such as scarlet fever, malaria, and typhoid fever; drying of the nasal mucous membrane; trauma (digital, as in picking the nose, and blunt, as in nasal fractures); arteriosclerosis; hypertension; and bleeding tendencies associated with aplastic anaemia, leukaemia, thrombocytopenia, liver disease, the hereditary coagulopathies, and Osler-Weber-Rendu syndrome (hereditary hemorrhagic telangiectasia - Ch. 99).

Treatment

Most nasal bleeding occurs from a plexus of vessels in the anteroinferior septum (Kiesselbach's area). **Bleeding may be controlled by pinching the nasal alae together for 5 to 10 min.** If this fail, the bleeding site must be found. The bleeding point may be cauterized. Bleeding may be controlled temporarily by applying pressure over a cotton pledget impregnated with a vasoconstrictor such as phenylephrine 0.25% and a topical anaesthetic such as tetracaine 1% until the site is anaesthetized. Although electrocautery may be used, silver nitrate in a 75% applicator bead may be used to control bleeding without producing too deep a burn of the mucous membrane.

In epistaxis due to a haemorrhagic disorder, petrolatum gauze is used to apply pressure as atraumatically as possible to the bleeding point; cautery is not used, since the periphery of a cauterized area may begin to bleed. Attention is directed to identifying and correcting the bleeding disorder.

In arteriosclerosis and hypertension, bleeding is likely to be far posterior in the inferior meatus and may be difficult to control. Control requires ligating the internal maxillary artery and its branches or packing the posterior part of the nasal cavity. The arteries may be ligated under microscopic control with a surgical approach through the maxillary sinus. In order to pack the posterior part of the nasal cavity, the choana is obstructed with a postnasal pack made by folding and rolling 4-in. gauze squares into a tight bundle and tying the bundle with 2 strands of heavy silk suture. The ends of one suture are tied to a catheter that has been introduced through the nasal cavity on the side of the bleeding and brought out through the mouth. The catheter is withdrawn from the nose as the pack is placed behind the soft palate into the nasopharynx. The 2nd suture is trimmed below the level of the soft palate so that it can be used to removed the pack. Alternatively, the balloon of a Foley catheter may be inflated in the nasopharynx to obstruct the choana. The nasal cavity, particularly the posterior part of the inferior meatus, is firmly packed with petrolatum gauze and the first suture is tied over a roll of gauze at the anterior nares to secure the postnasal pack. The packing remains in place for 4 days. An antibiotic such as ampicillin 250 mg orally q 6 h is given to prevent sinusitic and otitis media. Postnasal packing lowers the arterial P_{O2}, and supplementary O_2 should be given while the packing is in place.

in Osler-Weber-Rendu syndrome, multiple severe nosebleeds may occur from arteriovenous aneurysms in the mucous membrane and result in profound and persistent anaemia that is not easily corrected with administration of iron. A split-thickness skin graft

(septal dermoplasty) reduces the episodes of epistaxis and allows the anaemia to be corrected.

Severe epistaxis is often associated with liver disease. Blood may have been swallowed in large amounts. It should be eliminated as promptly as possible with enemas and catharactics, and the GI tract sterilized with nonabsorbable antibiotics (i.e. neomycin 1 gm orally gid) to prevent the breakdown of blood and absorption of ammonia.

Need for blood replacement is determined by the Hb level, vital signs, and the central venous pressure.

Nasal Vestibulitis

Infection of the nasal vestibule. **Low-grade infections** and **folliculitis** produce annoying crusts, and bleeding occurs as the crusts come away. Bacitracin ointment 500 u./gm applied topically bid for 14 days is effective.

Furuncles of the nasal vestibule are usually staphylococcal; they may develop into a spreading cellulitis of the tip of the nose. Systemic antibiotics should be employed along with hot soaks; penicillin G or V is the drug of choice. Furuncles of the central portion of the face should be allowed to drain spontaneously. Incision and drainage increase the risk of retrograde thrombophlebitis and subsequent cavernous sinus thrombosis, and are **contraindicated.**

Rhinitis

The most frequent of the acute upper respiratory infections, characterized by edema and vasodilatation of the nasal mucous membrane, nasal discharge, and obstruction.

Acute rhinitis is the usual manifestation of a common cold (see under Respiratory Viral Diseases in Ch. 12); it may also be caused by streptococcal, pneumococcal, or staphylococcal infections. **Chronic rhinitis** may occur in syphilis, TB, rhinosclerosis, rhinosporidiosis, leishmaniasis, blastomycosis, histoplasmosis, and leprosy, all conditions characterized by granuloma formation and destruction of soft tissue, cartilage, and bone. Rhinosclerosis also causes progressive nasal obstruction from indurated inflammatory tissue in the lamina propria. These conditions produce nasal obstruction, purulent rhinorrhea, and frequent bleeding. Rhinosporidiosis is characterized by bleeding polyps.

Diagnosis and Treatment

Diagnosis and treatment of **acute bacterial rhinitis** are based on pathogen identification and antibiotic sensitivities. Topical vasoconstriction with a sympathomimetic amine (i.e. phenylephrine 0.25%), given q 3 to 4 h for not more than 7 days, provides symptomatic relief. Systemic sympathomimetic amines, such as pseudoephedrine 30 mg orally q 4 to 6 h, may be given for vasoconstriction of the nasal mucous membrane.

Diagnosis in **chronic rhinitis** is based on demonstrating the causative microorganism by culture or biopsy. Treatment consists of chemotherapy appropriate to the causative agent.

Atrophic Rhinitis

A chronic rhinitis characterized by an atrophic and sclerotic mucous membrane, abnormal patency of the nasal cavities, crust formation, and foul odor. The mucous membrane changes from ciliated pseudostratified columnar epithelium to stratified squamous epithelium, and the lamina propria is reduced in amount and vascularity. Anosmia results, and epistaxis may be recurrent and severe. The etiology is unknown, although bacterial infection plays a role.

Treatment is directed toward reducing the crusting and eliminating the odor. Topical antibiotics, such as bacitracin 500 u./gm in a petrolatum base and estrogens and vitamins A and D topically or systemically, may be effective. Occluding or reducing the patency of the nasal cavities, surgically or with a pledget of lamb's wool, decreases the crusting caused by the drying effect of air flowing over the atrophic mucous membrane.

Vasomotor Rhinitis

A chronic rhinitis characterized by intermittent vascular engorgement of the nasal mucous membrane, sneezing, and watery rhinorrhea. The turgescent mucous membrane varies from bright red to a purplish hue. The condition is marked by periods of remission and exacerbation. It appears to be aggravated by a dry atmosphere. The etiology is uncertain, and no allergy can be identified.

Treatment is empirical and not always satisfactory. Patients benefit from humidified air; i.e. from a humidified central heating system or a vaporizer in the workroom and bedroom. Systemic sympathomimetic amines - i.e. pseudoephedrine 30 mg orally (adult) q 4 to 6 h as necessary - give symptomatic relief but are not recommended for regular long-term use. Topical vasoconstrictors should be **avoided** because the vasculature of the nasal mucous membrane loses its sensitivity to stimuli - i.e. the humidity and temperature of the inspired air - that result in vasoconstriction. Vasodilatation results, except after application of a strong stimulus, such as a topical sympathomimetic amine.

Allergic Rhinitis

(Same as under Atopic Diseases in Ch. 21)

A symptom complex including hay fever and perennial allergic rhinitis, characterized by seasonal or perennial sneezing, rhinorrhea, nasal congestion, pruritus, and often conjunctivitis and pharyngitis.

Hay Fever

(Pollinosis)

Hay fever, the acute seasonal form of allergic rhinitis, is generally induced by wind-borne pollens. The **spring type** is due to tree pollens (i.e. oak, elm, maple, alder, birch, cottonwood); the **summer type,** to grass pollens (i.e. Bermuda, timothy, sweet vernal, orchard, Johnson) and to weed pollens (i.e. sheep sorrel, English plantain); the **fall type,** to

weed pollens (i.e. ragweed). Occasionally, seasonal hay fever is due primarily to airborne fungus spores rather than to pollens. Important geographic regional differences occur.

Symptoms and Signs

The nose, roof of the mouth, pharynx, and eyes begin to itch gradually or abruptly after onset of the pollen season. Lacrimation, sneezing, and clear, watery nasal discharge accompany or soon follow the pruritus. Frontal headaches, irritability, anorexia, depression, and insomnia may appear. The conjunctiva is injected, and the nasal mucous membranes are swollen and bluish red. Coughing and asthmatic wheezing may develop as the season progresses. Many eosinophils are present in the nasal mucus during the season.

Diagnosis

The nature of the allergic process and even the responsible allergen are often suspected from the history. Diagnosis is confirmed by the above physical findings, skin tests, and the accompanying eosinophilia in secretions.

Treatment

Symptoms may be diminished by avoidance of the allergen (see above). Most patients obtain adequate relief with oral antihistamines (i.e. chlorpheniramine, in sustained-release form , 12 mg q 8 h; tripirolidine 2.5 mg q 8 h). If these drugs are too sedating, a nonsedating but more expensive antihistamine may be used (terfenadine 60 mr orally q 12 h). Topical treatment is another alternative (see below). Sympathomimetics are often used in combination with antihistamines. Phenylpropanolamine, phenylephrine, or pseudoephedrine are available in many antihistamine-decongestant preparations. Ephedrine 25 mg orally q 4 h is more effective, but its central-stimulating effects limit its use. Sympathomimetic drugs taken by mouth can raise the BP, and patients with a tendency to hypertension should not use them without periodic monitoring.

If antihistaminic drugs are not satisfactory, then a nasal spray containing 4% cromolyn sodium may be used. It is delivered by means of a finger-activated pump. The dosage is one spray (5.2 mg) 2 to 6 times/day. Because cromolyn acts by blocking the reaction of allergen with tissue mast cells, it is most effective in preventing symptoms rather than in relieving acute symptoms. Because of higher cost and because its effect is limited to the nose, it usually is not the first drug to be tried for hay fever treatment.

When nasal symptoms are not relieved adequately by antihistaminic treatment, intranasal glucocorticoid spray usually is effective. Two doses bid to qid are used initially; beclomethasone dipropionate is freon-propelled from a container that delivers 0.042 mg (42 microg)/dose, and flunisolide 0.025% is delivered by a finger-activated pump, 0.025 mg/dose. When symptoms have been relieved, dosage is reduced to 1 dose bid for the remainder of the season. Severe intractable extranasal symptoms may require a short course of systemic corticosteroid treatment (prednisone 10 mg orally bid, with gradual reduction in dose; an alternate-day regimen may also be used).

Desensitization treatment (see above) is advised if drug treatment is poorly tolerated, if glucocorticoids are needed during the season, or if asthma develops. If the patient is allergic to pollens, treatment should begin soon after the pollen season has ended, in preparation for the following season.

Perennial Allergic Rhinitis

In contrast to hay fever, symptoms of perennial rhinitis vary in severity (often unpredictably) throughout the year. Extranasal symptoms such as conjunctivitis are uncommon, but chronic nasal obstruction is often prominent and may extend to eustachian tube obstruction. The resultant hearing difficulty is particularly common in children. The **diagnosis** of allergic rhinitis is supported by a positive history of atopic disease, the characteristic bluish-red mucosa, numerous eosinophils in the nasal secretions, and positive skin tests (particularly to house dust, dust mites, feathers, animal danders, or fungi). Some patients have complicating sinus infections and nasal polyps.

Certain patients suffer from chronic rhinitis, sinusitis, and polyps and have negative skin tests. These patients are not atopic but often have sensitivity to aspirin and other nonsteroidal anti-inflammatory drugs, and should be evaluated also for sensitivity to tartrazine (a yellow food coloring) by a trial elimination of this food additive from the diet. Despite the negative skin tests, these patients have numerous eosinophils in their tissues and nasal secretions. A subset suffers only from chronic rhinitis. Although they are not atopic, their nasal secretions have many eosinophils. This group of patients has been given the diagnosis of **eosinophilic nonallergic rhinitis.** Some patients with mild but annoying chronic continuous nasal obstruction or rhinorrhea have no demonstrable allergy, and no polyps, infection, or drug sensitivity. Their condition is called **vasomotor rhinitis.**

Treatment

Management is similar to that for hay fever if specific allergens are identified, except that systemic glucocorticoids, even though effective, should be avoided because of the need for prolonged use. Surgery (antrotomy and irrigation of sinuses, polypectomy, submucous resection) may be necessary after allergic factors have been controlled or ruled out. The subset of patients with eosinophilic nonallergic rhinitis mentioned above usually respond best to a topical glucocorticoid. For many patients the only treatment is reassurance, antihistamine and vasoconstrictor drugs, and advice to avoid topical decongestants, which produce after-congestion and, when used continuously for a week or more, may aggravate or perpetuate chronic rhinitis **(rhinitis medicamentosa).** Some patients with rhinitis may benefit from the frequent use of saline irrigation or nasal sprays.

Polyps

Allergic rhinitis predisposes to polyp formation; polyps may also occur in acute and chronic infections. Nasal polyps form at the site of massive dependent edema in the lamina propria of the mucous membrane, usually around the ostia of the maxillary sinuses. As a polyp develops, it becomes teardrop-shaped; when mature, it resembles a peeled seedless grape. In acute infections, polyps may regress after the infection resolves. Bleeding polyps occur in rhinosporidiosis. Unilateral polyps occasionally occur in association with benign or

malignant neoplasms of the nose or paranasal sinuses.

Treatment: Corticosteroids, such as beclomethasone dipropionate (42 microg/spray) or flunisolide (25 microg/spray) aerosols, 1 or 2 sprays in each nasal cavity bid, have reduced or eliminated polyps, although surgical removal is often still required. Polyps should be removed if they obstruct the airway or promote sinusitis, or if they are unilateral polyps that may be obscuring benign or malignant neoplasms. Polyps tend to recur unless the underlying allergy or infection is controlled. Following removal of nasal polyps, topical beclomethasone or flunisolide therapy tends to retard their recurrence. In severe and recurrent cases, maxillary sinusotomy or ethmoidectomy may be indicated.

Wegener's Granulomatosis

Wegener's granulomatosis, a vasculitis of unknown etiology characterized by granulomas of the nose and lung and glomerulitis of the kidney, is discussed fully in Ch. 110. Most destructive lesions of bone, cartilage, and soft tissue of the nose and paranasal sinuses are ultimately found on thorough biopsy to be malignant neoplasms such as a lymphoma or carcinoma.

Anosmia

Loss of the sense of smell. Anosmia requires careful evaluation for intranasal and intracranial diseases. Anosmia occurs when (1) intranasal swelling of other obstruction prevents odors from gaining access to the olfactory area; (2) the olfactory neuroepithelium is destroyed, as in viral infections, atrophic rhinitis, or the chronic rhinitis of granulomatous diseases and neoplasms; or (3) the olfactory nerve fila, olfactory bulbs and tracts, or their central connections are destroyed, as by head trauma, intracranial surgery, infections, or neoplasms. Head trauma is a major cause of anosmia in young adults; viral infections are a major cause in older adults. Anosmia occurs congenitally in male hypogonadism (Kallmann's syndrome). Most patients with anosmia have normal perception of salty, sweety, sour, and bitter substances, but they lack flavor discrimination, which is largely dependent on olfaction, and therefore often complain of loss of the sense of taste.

Diagnostic evaluation requires examination of the cranial nerves and upper respiratory tract (particularly the nose and nasopharynx), psychophysical measuring of odor and taste identification and threshold detection, and enhanced CT of the head to role out neoplasms and unsuspected fractures of the floor of the anterior cranial fossa.

Treatment for allergic rhinitis and removal of nasal polyps and benign neoplasms often result in recovery of the sense of smell. Conditions causing destruction of the olfactory neuroepithelium or its central pathways do not lend themselves to effective therapeutic intervention, although spontaneous recovery frequently follows regeneration of the olfactory neuroepithelium and its central pathways.

Sinusitis

An inflammatory process in the paranasal sinuses due to viral, bacterial, and fungal infections or allergic reactions.

Etiology and Pathogenesis

Acute sinusitis is caused by streptococci, pneumococci, *Hemophilus influenzae,* and staphylococci, and is usually precipitated by an acute viral respiratory tract infection. Exacerbations of chronic sinusitis may be caused by a gram-negative rod or anaerobic microorganisms. In about 25% of cases, chronic maxillary sinusitis is secondary to dental infection.

With a URI, the swollen mucous membrane obstructs the ostium of the paranasal sinus, and the O_2 in the sinus is absorbed into the blood vessels in the mucous membrane. The resulting relative negative pressure in the sinus **(vacuum sinusitis)** is painful. If the vacuum is maintained, a transudate from the mucous membrane develops and fills the sinus, where it serves as a medium for bacteria that enter through the ostium or through a spreading cellulitis or thrombophlebitis in the lamina propria of the mucous membrane. An outpouring of serum and leukocytes to combat the infection results, and painful positive pressure develops in the obstructed sinus. The mucous membrane becomes hyperemic and edematous.

Symptoms, Signs, and Diagnosis

The symptoms and signs of acute and chronic sinusitis are similar. The area over the involved sinus may be tender and swollen. Maxillary sinusitis causes pain in the maxillary area, toothache, and frontal headache. Frontal sinusitis produces pain in the frontal area and frontal headache. Ethmoid sinusitis causes pain behind and between the eyes, and a frontal headache that is often described as "splitting". Pain from sphenoid sinusitis is less well localized and is referred to the frontal or occipital area. There may be malaise. Fever and chills suggest an extension of the infection beyond the sinuses.

The nasal mucous membrane is red and turgescent; yellow or green purulent rhinorrhea may be present. The seropurulent or mucopurulent exudate may be seen in the middle meatus in maxillary, anterior ethmoid, and frontal sinusitis, and in the area medial to the middle turbinate in posterior ethmoid and sphenoid sinusitis.

The frontal and maxillary sinuses may be opaque to transillumination, but radiography of the paranasal sinuses more reliably defines the sites and the degree of involvement. Radio-opacity in acute sinusitis may be due to the swollen mucous membrane or a retained exudate. X-rays of the apices of the teeth are required in chronic maxillary sinusitis to exclude a periapical abscess.

Treatment

Improved drainage and control of infection are the aims of therapy in acute sinusitis. Steam inhalation effectively produces nasal vasoconstriction and promotes drainage. Topical vasoconstrictors such as phenylephrine 0.25% spray q 3 h are effective but should be used

for a maximum of 7 days; systemic vasoconstrictors such pseudoephedrine 30 mg orally (adult) q 4 to 6 h are less reliably effective.

In both acute and chronic sinusitis, antibiotics should be given for at least 10 to 12 days. In acute sinusitis, penicillin G or V 250 mg orally q 6 h is the initial antibiotic of choice, and erythromycin 250 mg orally q 6 h is the second choice. In exacerbations of chronic sinusitis, a broad-spectrum antibiotic such as ampicillin 250 or 500 mg or tetracycline 250 mg orally q 6 h is better. In chronic sinusitis, prolonged antibiotic therapy for 4 to 6 wk often results in complete resolution. The sensitiveness of pathogens isolated from the sinus exudate and the patient's response guide subsequent therapy. Sinusitis not responsive to antibiotic therapy may require operative intervention (Caldwell-Luc operation for the maxillary sinuses, ethmoidectomy for the ethmoid sinuses, and sphenoid sinusotomy for the sphenoid sinuses) to improve ventilation and drainage and to remove inspiccated mucopurulent material, epithelial debris, and hypertrophic mucous membrane. Chronic frontal sinusitis is managed with an osteoplastic obliteration of the frontal sinuses.

Neoplasms

Unilateral bloody nasal discharge and obstruction, and facial swelling and numbness indicate cancer of the nose or paranasal sinuses until proven otherwise. **Exophytic papillomas** are squamous cell papillomas with a branching, vascular connective tissue stalk with finger-like projections on the surface. In the nasal cavity they often require repeated excision, but have a benign course. **Inverted papillomas** are squamous cell papillomas in which the epithelium is invaginated into the vascular connective tissue stroma. They are invasive and behave in a locally malignant manner; excision must include a large margin of normal tissue, including the bone of the lateral wall of the nasal cavity in a procedure called a lateral rhinotomy.

Often benign tumors that occur in the nasal cavity are fibromas, hemangiomas, and neurofibromas. Fibromas, neurolemmomas, and ossifying fibromas occur in the paranasal sinuses.

Squamous cell carcinoma is the most common malignant tumor in the nose and paranasal sinuses; others are adenoid cystic and mucoepidermoid carcinomas, malignant mixed tumors, adenocarcinomas, lymphomas, fibrosarcomas, osteosarcomas, chondrosarcomas, and melanomas. Hypernephroma is the most common metastatic tumor in the paranasal sinuses. Combined irradiation and radical resection give the best survival rates for the primary neoplasm.

Chapter 209: Nasopharynx

Adenoid Hypertrophy

(Same as in Adenoid Hypertrophy under Bacterial Infections in Ch. 191 and Juvenile Angiofibroma in Ch. 199)

Enlargement of adenoidal tissue due to lymphoid hyperplasia.

Adenoidal lymphoid hyperplasia occurs in children and may be physiologic or secondary to infection or allergy. Consequent obstruction of the eustachian tubes may result in recurrent acute, chronic, or secretory (serous) otitis media; obstruction of the choanae may cause chronic sinusitis, mouth breathing, a hyponasal voice, and purulent rhinorrhea. Chronic adenoiditis is common.

Treatment

Adenoidectomy is frequently indicated in persistent serous and chronic otitis media to reduce exacerbations of chronic otitis media and improve the results of tympanoplasty. Adenoidectomy for recurrent acute otitis media depends on the duration of the earache after antibiotic is initiated, the presence of spontaneous perforation, the frequency with which myringotomy is required, and the severity of systemic symptoms. Adenoidectomy for nasal obstruction depends on the severity of the obstruction and the patient's age, since lymphoid hyperplasia reaches its maximum at puberty. Purulent rhinorrhea or sinusitis that recurs or persists despite adequate antibiotic treatment in otherwise normal children may be corrected by adenoidectomy.

Juvenile Angiofibroma

A benign tumor arising from the connective tissue in the nasopharyngeal vault and occurring almost exclusively in males at puberty. The angiofibroma is red and firm and is composed of fibrous tissue and numerous thin-walled vessels without contractile elements. Epistaxis is the major symptom. The angiofibroma may obstruct the nasal cavity, encroach upon the paranasal sinuses, and invade the orbit and the cranial cavity. The pterygomaxillary fissure is frequently widened by extension of the tumor into the infratemporal fossa. The widening of the fissure may be determined radiographically; the extent of the tumor may be determined with CT. The source of the blood supply and the presence of intracranial extension are determined with bilateral selective internal and external carotid angiography.

Although angiofibromas usually involute with maturity, **treatment** is usually necessary. To control recurrent massive bleeding, estrogen therapy with diethylstilbestrol (5 mg orally tid for 6 wk prior to excision) reduces the size and vascularity of the tumor. Angiographic embolization followed by excision is the more definitive method of treatment, but radiation therapy is the treatment of choice for patients with intracranial or orbital extension.

Tornwaldt's Cyst

(Nasopharyngeal Cyst)

A frequently infected cyst found in the midline of the nasopharynx. The cyst lies superficial to the superior constrictor muscle of the pharynx and is covered by the mucous membrane of the nasopharynx. If infected, it may cause persistent purulent drainage with a foul taste and odor, eustachian obstruction, and sore throat. Purulent exudate may be seen coming from the opening of the cyst. **Treatment** consists of marsupialization or excision.

Nasopharyngeal Carcinoma

Squamous cell carcinoma of the nasopharynx occurs in children and young adults. Rare in North America, it is one of the most common cancers in the Orient. The first symptom is often nasal or eustachian tube obstruction; the latter may result in middle ear effusion. Purulent bloody rhinorrhea, frank epistaxis, cranial nerve paralysis due to invasion of the parapharyngeal space and cranial cavity by the tumor, and cervical lymphadenopathy due to metastasis are common presenting complaints. **Diagnosis** is by biopsy of the primary nasopharyngeal tumor. Biopsy of the neck metastasis should be avoided until the nasopharynx has been inspected and palpated and any suspicious lesion there has been biopsied. The **treatment** of choice is supervoltage irradiation. The overall 5-yr survival rate is 35%.

Chapter 210: Oropharynx

Pharyngitis

Acute inflammation of the pharynx. Usually viral in origin, it may be due to a Group A beta-hemolytic streptococcus or occasionally to a pneumococcus or coagulase-positive staphylococcus. It is characterized by sore throat and pain on swallowing. Differentiating viral form from bacterial pharyngitis on the basis of physical examination alone is difficult. In both, the pharyngeal mucous membrane may be mildly injected or severely inflamed and may be covered by a membrane and a purulent exudate. Fever, cervical adenopathy, and leukocytosis are present in both viral and streptococcal pharyngitis but may be more marked in the latter. (For pharyngitis in gonorrhea and in other sexually transmitted diseases, see in Ch. 14.)

Treatment includes aspirin, to relive discomfort, and rest. Antibiotic therapy should usually be withheld pending positive cultures for bacteria. Penicillin G or V 250 mg orally q 6 h for 10 days is indicated for streptococcal pharyngitis and may be given for pneumococcal and staphylococcal pharyngitis if the symptoms and course warrant therapeutic intervention.

Tonsillitis

Acute inflammation of the palatine tonsils, usually due to streptococcal or, less commonly, viral infection. Epidemics of viral tonsillitis occur among military recruits. Tonsillitis is characterized by sore throat and pain, most marked on swallowing and often referred to the ears. Very young children may not complain of sore throat but will refuse to eat. High fever, malaise, headache, and vomiting are common.

Diagnosis

The tonsils are edematous and hyperemic. There may be a purulent exudate from the crypts and a membrane - white, thin, nonconfluent, and confined to the tonsil - that peels away without bleeding. The **differential diagnosis** includes diphtheria, Vincent's angina (trench mouth), and infectious mononucleosis. In diphtheria, the membrane is dirty, gray, thick, and tough, and bleeds if peeled away; it shows *Corynebacterium diphtheriae* on smear and culture. Vincent's angina, characterized by superficial, painful ulcers with erythematous

borders, is caused by a fusiform bacillus and a spirochete that are demonstrable on smear. Infectious mononucleosis tonsillitis characteristically is associated with micropetechiae of the soft palate, atypical lymphocytes on smear and a positive heterophil agglutination test confirm the diagnosis of mononucleosis.

Treatment

In viral tonsillitis, symptomatic therapy is as for pharyngitis. Penicillin G or V 250 mg orally q 6 h is the treatment of choice in streptococcal tonsillitis and should be continued for 10 days. When possible, the throat should be recultured 5 to 6 days later. Family members' throats should also be cultured initially so that carriers may be treated at the same time. Tonsillectomy should be considered if, despite these precautions, acute tonsillitis repeatedly develops after adequate treatment, or if chronic tonsillitis and sore throat persist or are relieved only briefly by antibiotic therapy.

Peritonsillar Cellulitis and Abscess

(Quinsy)

An acute infection located between the tonsil and the superior pharyngeal constrictor muscle. Peritonsillar abscesses are rare in children but common in young adults. Although usually due to a Group A beta-hemolytic streptococcus, anaerobic microorganisms such as bacteroides also cause peritonsillar infection. There is severe pain on swallowing; the patient is febrile and toxic, holds his head tilted toward the side of the abscess, and shows marked trismus. The tonsil is displaced medially by the peritonsillar cellulitis and abscess, the soft palate is erythematous and swollen, and the uvula is edematous and displaced to the opposite side.

Treatment

Cellulitis with pus formation will respond to penicillin in 24 to 48 h. Initially, penicillin G 1 million u. IV q 4 h is given. If pus is present and does not drain spontaneously, incision and drainage are required. Antibiotic therapy should be continued orally with penicillin G or V 250 mg q 6 h for 12 days. Peritonsillar abscesses tend to recur and tonsillectomy is indicated (usually performed 6 wk after the acute infection has subsided). With antibiotic therapy, the tonsillectomy can be performed at the time of the acute peritonsillar infection.

Parapharyngeal Abscess

Suppuration of a parapharyngeal lymph node with consequent abscess formation is usually secondary to pharyngitis or tonsillitis and may occur at any age. The abscess is lateral to the superior pharyngeal constrictor muscle and close to the carotid sheath. Pharyngeal inflammation may not be apparent. The anterior cervical triangle is markedly swollen. Penicillin G 150,000 u./kg/day IV in 4 equal doses (for a child) should be given initially and the abscess drained through a cervical, not pharyngeal, incision. Subsequently, penicillin G or V 250 mg orally q 6 h is given to complete 12 days of therapy.

Retropharyngeal Abscess

(Same under Bacterial Infections in Ch. 191)

Retropharyngeal abscesses usually occur in infants or young children as complications of suppurative retropharyngeal lymph nodes to which infection has spread from the pharynx, sinuses, adenoids, nose, or middle ear. These abscesses are unusual in adults because the retropharyngeal lymph nodes diminish or disappear after childhood. Occasional causes in adults or children are TB or perforation of the posterior pharyngeal wall by foreign bodies or instrumentation.

The major manifestations are painful swallowing, fever, cervical lymphadenopathy, and, if airway obstruction occurs, stridor, dyspnea, and hyperextension of the neck. The cervical vertebrae cannot be palpated through the posterior pharyngeal wall, which is boggy and fluctuant, with a definite, usually unilateral, bulging. Widening of the prevertebral space can be demonstrated on lateral radiographs of the neck. **Complications** include hemorrhage; rupture of the abscess into the airway, causing asphyxia or pulmonary aspiration; laryngeal spasm; mediastinitis; and suppurative thrombophlebitis of the internal jugular veins.

Treatment includes draining the abscess through an incision in the posterior pharyngeal wall and giving penicillin G 150,000 u./kg/day IV in 4 equal doses for 3 to 4 days; then orally for a total of 14 days, unless culture and sensitivity studies of the drained abscess material indicate use of an alternate antimicrobial agent.

Velopharyngeal Insufficiency

Incomplete closure of the velopharyngeal sphincter between the oropharynx and the nasopharynx, resulting in impaired speech and deglutition. The speech is characterized by nasal emission of air and weak oral plosive and fricative articulation.

Normal closure, achieved by the sphincteric action of the soft palate and the superior constrictor muscle, is impaired in patients with cleft palates, repaired cleft palates, congenitally short palates, submucous cleft palates, unusually large nasopharynges, and palatal paralysis.

Diagnosis and Treatment

Regurgitation of solid foods and fluids through the nose denotes gross velopharyngeal insufficiency, but normal speech is a more exacting criterion of competency. Inspection of the palate during phonation may reveal palatal paralysis. Palpation of the midline of the soft palate and transillumination with a nasopharyngoscope may demonstrate a submucous cleft. A lateral x-ray may demonstrate the congenitally short palate or an unusually large nasopharynx and, if taken during phonation, will indicate the degree of insufficiency; cinefluoroscopy during connected speech verifies an inability to maintain velopharyngeal valve closure.

Treatment requires speech therapy and surgical correction by a palatal push-back procedure, pharyngeal flap, pharyngoplasty, or Teflon paste injection of the posterior

pharyngeal wall.

Carcinoma of the Tonsil

Squamous cell carcinoma of the tonsil, second in frequency only to carcinoma of the larynx among malignancies of the upper respiratory tract, occurs predominantly in males and is associated with tobacco smoking and ethanol ingestion. Sore throat is the most common presenting complaint, and pain often radiates to the ear on the same side. A metastatic mass in the neck may be the first symptom. **Diagnosis** is made by biopsy. Direct laryngoscopy, bronchoscopy, and esophagoscopy are carried out to exclude a synchronous 2nd primary neoplasm. **Treatment** combines irradiation and surgery, consisting of radical resection of the tonsillar fossa, hemimandibulectomy, and radical neck dissection. The 5-yr survival rate approximates 50%.

Zenker's (Pharyngoesophageal) Diverticulum

(Same in Ch. 51 - Esophageal Diverticula)

There are several types of esophageal diverticula, each of different etiology. A **pharyngeal diverticulum (Zenker's)** is an outpouching of the mucosa and submucosa posteriorly through the cricopharyngeal muscle. It probably results from incoordination between pharyngeal propulsion and cricopharyngeal relaxation. **Mid-esophageal (traditionally called traction) diverticula** are either due to traction from mediastinal inflammatory lesions or secondary to motor disorders. An **epiphrenic diverticulum,** also probably of propulsive origin, occurs just above the diaphragm and usually accompanies an esophageal motor disturbance (achalasia, diffuse esophageal spasm).

Symptoms, Signs, and Diagnosis

A Zenker's diverticulum fills with food that may be regurgitated when the patient bends or lies down. Aspiration pneumonitis may result if regurgitation is nocturnal. Rarely, the pouch becomes large and causes dysphagia. Traction and epiphrenic diverticula are rarely symptomatic in themselves. All diverticular are diagnosed by barium-swallow x-ray.

Treatment

Specific treatment is usually not required, though surgical resection of the diverticulum is occasionally necessary.

Chapter 211: Larynx

Vocal Cord Polyps

Vocal cord polyps develop from voice abuse, chronic laryngeal allergic manifestations, and chronic inhalation of irritants such as industrial fumes and cigarette smoke. They consist of chronic edema in the lamina propria of the true vocal cord and result in hoarseness and a breathy voice quality. Biopsy of discrete lesions should be done to exclude carcinoma. Treatment involves surgical removal of the polyp to restore the voice, and attention to the

underlying cause to prevent recurrence, including voice therapy if voice abuse is the cause.

Vocal Cord Nodules

(Singer's, Teacher's, or Screamer's Nodules)

Vocal cord nodules are caused by chronic voice abuse, such as screaming or shouting, or using an unnaturally low fundamental frequency. The nodules are condensation of hyaline connective tissue in the lamina propria at the junction of the anterior 1/3 and the posterior 2/3 of the free edges of the true vocal cords. Hoarseness and a breathy voice quality result. Carcinoma should be excluded by biopsy. **Treatment** involves surgical removal of the nodules and correction of the underlying voice abuse. Vocal nodules in children usually regress with voice therapy alone.

Contact Ulcers

Unilateral or bilateral ulcers of the mucous membrane over the vocal process of the arytenoid cartilage resulting from voice abuse. Mild pain on phonation and swallowing and varying degrees of hoarseness result. Biopsy to exclude carcinoma is important. Prolonged ulceration leads to formation of nonspecific granulomas, which produce varying degrees of hoarseness.

Treatment consists of prolonged voice rest (6 wk minimum) for healing of the ulcers. Patients must recognize the limitations of their voices and learn to adjust their vocal activities to avoid recurrent ulcers. Granulomas tend to recur after surgical removal but respond to extensive voice therapy.

Laryngitis

Inflammation of the larynx.

Etiology

Viral and bacterial URIs are the most frequent causes of acute laryngitis. Although viral laryngitis is commoner, beta-hemolytic streptococcus and Streptococcus pneumoniae are causative microorganisms. It may also occur in the course of bronchitis, pneumonia, influenza, pertussis, measles, and diphtheria. Excessive use of the voice, allergic reactions, and inhalation of irritating substances can cause acute or chronic laryngitis.

Symptoms and Signs

Unnatural change of voice is usually the most prominent symptom. Hoarseness and even aphonia, together with a sensation of tickling, rawness, and a constant urge to clear the throat, may occur. Symptoms vary with the severity of the inflammation. Fever, malaise, dysphagia, and throat pain may occur in the more severe infections; dyspnea may be apparent if laryngeal edema is present. Laryngoscopic examination discloses a mild to marked erythema of the mucous membrane that may also be edematous. If a membrane is present, diphtheria must be suspected.

Treatment

There is no specific treatment for viral laryngitis. Penicillin G 250 mg orally q 6 h for 10 to 12 days is the drug of choice for streptococcal or pneumococcal laryngitis. Treatment of acute of chronic bronchitis may improve the laryngitis. Treatment of chronic bronchitis may require a broader spectrum antibiotic, such as ampicillin 250 or 500 mg or tetracycline 250 mg orally q 6 h for 10 to 14 days. Voice rest and steam inhalations give symptomatic relief and promote resolution of acute laryngitis.

Acute Epiglottitis

(Same in Acute Epiglottitis under Bacterial infections and Croup

under Viral Infection in Ch. 191)

A severe rapidly progressive infection of the epiglottis and surrounding tissues that may be quickly fatal due to sudden respiratory obstruction by the inflamed structures.

Etiology and Incidence

Hemophilus influenzae type B is almost exclusively the pathogen; very rarely, streptococci may be responsible. The incidence of *H. influenzae* type B epiglottitis is highest in children aged 2 to 5 yr; the disease is uncommon in children under 2 yr, but it may occur at any age.

Pathophysiology

Infection, acquired through the respiratory tract, may produce initial nasopharyngitis. Subsequent downward extension produces inflammation of the epiglottis and often of the lower tracheobronchial tree. Bacteremia is common. The inflamed epiglottis mechanically obstructs the airway, increasing the work of breathing, which may result in CO_2 retention and hypoxia. Clearance of inflammatory secretions is also impaired. These combined factors may result in fatal asphyxia within a few hours.

Symptoms and Signs

Onset frequently is acute and fulminating. Sore throat, hoarseness, and, usually, high fever develop abruptly in a previously well child. Dysphagia and respiratory distress characterized by drooling, dyspnea, tachypnea, and inspiratory stridor develop rapidly. On physical examination the child may appear moribund and in severe respiratory distress. There are deep suprasternal, supraclavicular, intercostal, and subcostal inspiratory retractions. Breath sounds may be diminished bilaterally and rhonchi may be heard. The pharynx is usually inflamed. The epiglottis is "beefy" red, stiff, and swollen, resembling a red cherry. Thick, superficial secretions may coat the epiglottis and glottis.

H. influenzae pneumonia, occasionally with empyema, may occur concurrently with epiglottitis. Metastatic infection to the joints, meninges, pericardium, or subcutaneous tissues

occurs infrequently, resulting in an abscess or cellulitis.

Diagnosis

The patient should be hospitalized immediately whenever the diagnosis is suspected clinically. Direct visualization of the epiglottis is diagnostic, but manipulation may initiate sudden, fatal airway obstruction. Visualization should only be attempted by trained personnel with equipment to establish an airway if necessary. H. influenzae Type B may be cultured from the upper respiratory tract and usually the blood.

The major differential diagnostic concern is acute viral croup (laryngo-tracheobronchitis). Croup is usually less fulminant in onset, and its characteristic barking cough is uncommon in epiglottitis. The epiglottis in croup may be erythematous, but is not markedly edematous and crimson as in epiglottitis. Lateral and anteroposterior neck X-rays will differentiate the two, showing subepiglottic narrowing and a normal-sized epiglottis in croup and, in epiglottitis, an enlarged epiglottis and distention of the hypopharynx. Diphtheria should be considered in an unimmunized patient.

Treatment

Speed is vital. A continually adequate airway must be assured, and specific parenteral antibiotics given. Sudden complete airway obstruction occurs so unpredictably that an airway must be secured immediately, preferably by nasotracheal intubation. Alternatively, tracheostomy may be performed. Careful and skilled nursing care is required, since secretions can cause obstruction even after intubation or tracheostomy. The nasotracheal tube is usually required only until the patient has been stable for 24 to 48 h (usually a total intubation time < 60 h). The inflammation is effectively controlled with parenteral antibiotics. Chloramphenicol 75 to 100 mg/kg/day IV in 6 divided doses should be used initially since ampicillin-resistant H. influenzae are currently common. Rarely, H. influenzae strains have been isolated that are resistant to chloramphenicol. Where this has occurred, both chloramphenicol and ampicillin therapy should be started. If the organism is isolated and proved to be ampicillin-sensitive, ampicillin 200 mg/kg/day IV in 6 divided doses can be given. Sedatives should be *avoided.*

Croup

(Acute Laryngotracheobronchitis)

An acute viral inflammation of the upper and lower respiratory tracts, characterized by inspiratory stridor, subglottic swelling, and respiratory distress that is most pronounced on inspiration.

Etiology, Epidemiology, and Pathophysiology

The parainfluenza viruses, especially Type I, are the major pathogens. Less common causes are respiratory syncytial virus (RSV) and influenza A and B viruses, followed by adeno-, entero-, rhino-, and measles viruses and Mycoplasma pneumoniae. Seasonal outbreaks are common; cases due to parainfluenza viruses tend to occur in the fall, and those due to

RSV and influenza viruses are likely to occur in the winter and spring. Spread is most likely to be by the airborne route or by contact with infected secretions. Croup is primarily a disease of children aged 6 mo to 3 yr, though it may occasionally occur earlier or later.

The infection produces inflammation of the larynx, trachea, bronchi, bronchioles, and lung parenchyma. However, obstruction, caused by swelling and inflammatory exudate, is most pronounced in the subglottic region. Obstruction increases the work of breathing and, as the child tires, results in hypercapnia. Hypoxemia without hypercapnia commonly occurs due to parenchymal pulmonary infection. Atelectasis may occur concurrently if the bronchioles become obstructed.

Symptoms and Signs

Croup is usually preceded by a URI. A "barking" often spasmodic cough, and hoarseness may mark the acute onset of inspiratory stridor, commonly at night. The child may awaken during the night with respiratory distress, tachypnea, and supraclavicular, suprasternal, substernal, and intercostal inspiratory retractions. In severe cases, cyanosis with increasingly shallow respirations may develop as the child tires. The obvious respiratory distress and the harsh inspiratory stridor are the most dramatic physical findings. Auscultation reveals prolonged inspiration and stridor, often with some expiratory rhonchi and wheezes. Rales also may be present. Breath sounds may be diminished with atelectasis. Fever is present in about 1/2 the children. Leukocytosis with increased polymorphonuclear cells may be present initially, with a subsequent shift to leukopenia and lymphocytosis. With involvement of the lung parenchyma, arterial blood gas analysis reveals hypoxemia with or without hypercapnia. Subepiglottic narrowing may be seen on anteroposterior neck X-ray. The child's condition may appear improved in the morning, but worsens again at night. The illness usually lasts 3 to 4 days.

Differential Diagnosis

Croup must be differentiated from the epiglottitis. Distinguishing features are given in the discussion of epiglottitis, above. A foreign body may cause respiratory distress and a typical croupy cough, but fever and a preceding URI are absent. X-rays of the neck may show a foreign body, but indirect and direct laryngoscopy may be required to confirm the diagnosis. Diphtheria is excluded by a history of adequate immunization, or confirmed by identification of the organism in special cultures of scrapings from the typical grayish diphtheric pharyngeal or laryngeal membrane. Rarely, retropharyngeal abscess may present with stridor. It may be diagnosed by finger palpation of the mass or by lateral x-ray of the neck.

Treatment

Home therapy: The mildly ill child may be cared for at home with supportive measures. The child should be made comfortable and should be kept well hydrated. Rest is important, as fatigue and crying can aggravate the condition. Home humidification devices (e.g., "cold-steam" vaporizers or humidifiers, or steam from a hot shower in an enclosed bathroom) may ameliorate upper airway drying, but the water droplets produced are too large to help in mobilizing secretions in the lower respiratory tract. Increasing or persistent dyspnea, tachycardia, fatigue, or dehydration indicates the need for hospitalization.

Hospital therapy: Since moderate hypoxemia may exist without cyanosis, arterial blood gas analysis is indicated in all hospitalized croup patients. If the Pa_{o2} is < 60 mmHg, O_2 should be administered. A 40% inspired O_2 concentration is usually adequate, and may be best achieved by face masks. Tents may also be used, but are less efficient. CO_2 retention (Pa_{co2} > 45 mmhg) generally indicates fatigue and necessitates close surveillance of the patient. The need for intubation should be anticipated, and equipment and personnel should be ready. The need for airway intervention is indicated by (1) increasing CO_2 retention despite adequate oxygenation, nebulized mist therapy, and hydration; (2) hypoxemia that is unresponsive to O_2 administration; and (3) secretions that cannot be mobilized by coughing. Nasotracheal intubation causes fewer complications than does tracheostomy if performed early by skilled personnel.

The viscosity of tracheobronchial secretions may be reduced and their clearance enhanced by mist therapy. Standard jet-type nebulizers may be used to enhance laryngeal humidification; but bronchiolar humidification requires use of an ultrasonic nebulizer fitted to a mask or an oxygen tent.

The viruses that most commonly cause croup do not predispose to secondary bacterial infection, and antibiotics are rarely indicated. Corticosteroids are not recommended. Nebulized racemic epinephrine has been used successfully to produce symptomatic improvement and relieve fatigue. However, it should only be used with the understanding that the effects are transient, and that the course of the illness, the Pa_{o2}, and the prognosis are not altered, and tachycardia and other side effects may occur.

Vocal Cord Paralysis

Etiology

Vocal cord paralysis may result from lesions at the nucleus ambiguous, its supranuclear tracts, the main trunk of the vagus, or the recurrent laryngeal nerves. Intracranial neoplasms, vascular accidents, and demyelinating diseases cause nucleus ambiguous paralysis. Tumors at the base of the skull and trauma of the neck cause vagus paralysis. Recurrent laryngeal paralysis is caused by neck or thoracic lesions, e.g., aortic aneurysm, mitral stenosis, neoplasms of the thyroid gland, esophagus, lung, and mediastinal structures, or trauma, thyroidectomy, neurotoxins (lead), neurotoxic infections (diphtheria), and viral illness. Vocal cord paralysis is often idiopathic.

Symptoms and Signs

Vocal cord paralysis results in loss of vocal cord abduction, or adduction and abduction; and may affect phonation, respiration, and deglutition; may result in aspiration of food and fluids into the trachea. The paralyzed cord generally lies 2 to 3 mm lateral to the midline, and in recurrent laryngeal nerve paralysis may move with phonation but not on inspiration. In unilateral vocal cord paralysis, the voice is hoarse and breathy. There is usually no airway obstruction because the normal cord abducts sufficiently. In bilateral vocal cord paralysis, the cords are within 2 to 3 mm of the midline and the voice is of limited intensity but good quality. The airway, however, is inadequate, resulting in stridor and dyspnea on moderate exertion.

Diagnosis

The cause must always be sought. The evaluation may include laryngoscopy, bronchoscopy, and esophagoscopy. Neurologic examination, x-rays of the base of the skull, thyroid gland scan, upper GI series, posteroanterior and lateral chest x-rays, and laminagraphy of the chest are also indicated. Cricoarytenoid arthritis may cause fixation of the cricoarytenoid joint and must be differentiated.

Treatment

In unilateral paralysis, augmenting the paralyzed cord by injection of a Teflon suspension may allow approximation of the cords for voice improvement and prevention of aspiration. Maintenance of an adequate airway is the problem in bilateral paralysis. Tracheotomy may be needed permanently or during URIs. An arytenoidectomy with lateralization of the true vocal cord will open the glottis and improve the airway but may adversely affect the voice quality.

Laryngoceles

Evaginations of the mucous membrane of the laryngeal ventricle. Internal laryngoceles displace and enlarge the false vocal cord and result in hoarseness and airway obstruction. External laryngoceles extend through the thyroid membrane, producing a mass in the neck. Laryngoceles are filled with air and can be expanded by Valsalva's maneuver. They tend to occur in musicians who play wind instruments. They appear on x-ray as smooth, ovoid, radiolucent masses. Laryngoceles may become infected or filled with mucoid fluid. **Treatment** is excision.

Neoplasms

Benign

Juvenile papillomas may grow so exuberantly at multiple sites in the larynx that tracheotomy is required to maintain an adequate airway. They are thought to be of viral etiology, and they may appear as early as 1 yr of age and occur in epidemics. Treatment is by periodic excision. Recurrence is common. Regression occurs spontaneously at puberty.

Other benign laryngeal tumors include hemangiomas, fibromas, chondromas, myxomas, and neurofibromas. They may involve any part of the larynx. Removal restores the voice, the functional integrity of the laryngeal sphincter, and the airway.

Malignant

Squamous cell carcinoma, the most common malignant neoplasm of the larynx, is also the most common malignancy of the head and neck. The incidence is higher in males, heavy drinkers, and heavy smokers. The true vocal cord (particularly the anterior portion), epiglottis, pyriform sinus, and postcricoid are common sites of origin. Hoarseness is an early symptom and all patients with hoarseness lasting 2 wk should have indirect laryngoscopy. A discrete lesion of the laryngeal mucous membrane should be biopsied at direct laryngoscopy.

Early treatment by irradiation or cordectomy results in a 5-yr survival rate of 85 to 95%. Since irradiation usually returns the voice to normal, it is the treatment of choice in early carcinoma. Surgery is necessary in advanced carcinoma with anterior commissure involvement, thyroid cartilage invasion, or impaired vocal cord mobility. Partial laryngectomy, preserving laryngeal phonatory and sphincteric functions, may be possible, but total laryngectomy with radical neck dissection on the side of the lesion is more frequently required. A combination of irradiation and surgery is more successful than surgery alone in advanced supraglottic and hypopharyngeal lesions.

Rehabilitation after total laryngectomy requires developing a new voice by using esophageal speech or creating a tracheoesophageal fistula. **Esophageal speech** involves taking air into the esophagus during the negative intrathoracic pressure of inspiration and gradually eructating the air through the pharyngoesophageal junction to produce a sound that is articulated into speech by the pharynx, palate, tongue, teeth, and lips.

A **tracheoesophageal fistula,** created by inserting a one-way valve between the trachea and the esophagus, forces air into the esophagus during expiration to produce a sound that is converted into speech. With this technic, fluids and food may be aspirated into the tracheobronchial tree if the tracheoesophageal valve misfunctions.

An alternative method, using an **electrolarynx** as a sounding source, requires holding the device in place while it produces the sound that is articulated into speech.

Chapter 212: Neoplasms of the Head and Neck

Neoplasms of specific organs are discussed elsewhere in The Manual. This discussion deals with important general principles of head and neck neoplasms and with the situation in which cervical metastasis is present and the primary neoplasm cannot be determined.

Etiology and Pathogenesis

A history of alcohol and tobacco consumption is present in 85% of patients with cancer of the head or neck. The most common cancer of the upper respiratory and alimentary tracts is squamous cell carcinoma of the larynx, followed by squamous cell carcinoma of the palatine tonsil and hypopharynx. People who practice poor oral hygiene, have ill-fitting dentures, dip snuff, or chew tobacco tend to develop oral cavity cancers.

The Epstein-Barr virus plays a role in pathogenesis of nasopharyngeal cancer. Patients who received small doses of radiation therapy 20 or more years ago (for acne, facial hair, enlarged thymus, hypertrophic tonsils, and adenoids) are predisposed to developing thyroid and salivary gland cancer.

Head and neck cancer usually spreads in an orderly fashion and remains localized to the head and neck for long periods. Local tissue invasion is followed by regional metastasis and then by distant metastasis. Hematogenous metastases are usually associated with large or recurrent tumors and with concurrent disease in immunosuppressed patients.

Epidemiology

In the head and neck, 90% of cancers are squamous cell (epidermoid) carcinoma; melanomas, lymphomas, and sarcomas make up another 5%. Patients with cancers of the salivary glands, thyroid, paranasal sinuses, and sarcomas are usually under age 59 yr, whereas those with squamous cell carcinoma of the oral cavity, pharynx, and larynx are generally over age 59 yr, the average age for all patients with head and neck cancers. Cancer of the nasopharynx is extremely prevalent in native-born and first-generation Chinese. Caucasians are more frequently afflicted with head and neck cancer than blacks.

Staging and Prognosis

Head and neck cancers are classified according to size and site of involvement of the primary neoplasm **(T),** number and size of metastases to the cervical lymph nodes **(N),** and evidence of distant metastases **(M). Stage I:** The primary neoplasm is < 2 cm in diameter or localized to one anatomic site without regional or distant metastasis ($T_1N_0M_0$). **Stage II:** The primary neoplasm measures 2 to 4 cm in diameter or involves 2 areas within a specific site (i.e. larynx) without regional or distant metastasis ($T_2N_0M_0$). **Stage III:** The primary neoplasm is > 4 cm or involves 3 adjacent areas in a specific head and neck site and/or has an isolated neck metastasis < 3 cm in diameter (T_3N_0 or any $T_{1-3}N_1M_0$). **Stage IV:** The cancer is massive, invades bone and cartilage and/or extends outside of its site of origin into another site (i.e. oral cavity into oropharynx); neck metastasis measures > 3 cm, involves multiple nodes or is fixed to surrounding tissue, and/or evidence exists of distant metastases ($T_4N_{1-3}M_{0-1}$).

With appropriate treatment, Stage I survival generally approaches 90%; Stage II, 75%; Stage III, 45-75%; and Stage IV, < 35%. Overall 5-yr survival is 65% for all patients with local Stage I and II squamous cell cancer of the head and neck. The rate drops to 30% for patients with metastasis to lymph nodes (Stage III). Patients over age 70 yr have longer disease-free intervals and better survival rates than younger patients.

Exophytic or verrucous-appearing tumors respond to treatment better than infiltrative, ulcerative, or indurated lesions. Cervical or distant metastasis is associated with limited survival. The more poorly differentiated the cancer, the greater the chance of regional and distant metastasis. Invasion of muscle, bone, or cartilage reduces cure rates. Perineural spread as evidence by pain, paralysis, or numbness indicates a highly aggressive neoplasm with a high propensity for recurrence.

Treatment

Small soft-tissue lesions < 2 cm in diameter, regardless of location within the upper respiratory or alimentary tracts, respond equally to surgery or radiation. A 5-yr cure rate of 90% can be expected. With radiation therapy, some surgical procedures may be needed in order to achieve the 90% cure rate. Lesions > 2 cm or with bone or cartilage invasion (with or without regional neck metastasis) require surgery. If lymph node metastases are found, postoperative radiation is necessary. Alternatively, fair survival rates can be attained with radiation with or without chemotherapy. If the cancer recurs, the patient has recourse to surgery. In advanced (most stage II and all stages III and IV) squamous cell carcinoma, combining surgery and radiation offers a better chance of cure than treatment with either

modality alone. Radiation therapy may be given preoperatively or postoperatively. Postoperative radiation is usually preferred.

Surgery is more effective than radiation and/or chemotherapy in controlling large primary cancers, while radiation is effective in controlling microscopic or nonpalpable metastases. Chemotherapy kills tumor cells at the local site, regional lymph nodes, and distant metastases. It is not clear whether adjuvant chemotherapy (in combination with surgery or radiation therapy) increases the cure rate; however, combined therapy does prolong the interval between cancer disappearance and recurrence. Several agents - cisplatin, fluorouracil, bleomycin, and methotrexate - provide useful palliation for pain and reduce neoplasm size in patients in whom surgery or radiation therapy cannot be used. A disadvantage of surgery is the need for rehabilitation for swallowing and speaking. Radiation produces skin changes, fibrosis, ageusia, xerostomia, and, rarely, osteoradionecrosis. Toxicities of chemotherapy include severe nausea and vomiting, transient hair loss, gastroenteritis, and hematopoietic and immune depression. In cancer excision after chemotherapy or irradiation, the surgeon must remove what was originally involved with the neoplasm before the anticancer therapy was started.

Recurring or persisting cancer: *A palpable mass or ulcerated lesion with edema or pain at the primary site after therapy.* Detecting recurrence after chemotherapy or radiation therapy is more difficult than after surgery alone; however, recurrence after surgery alone is usually more difficult to eradicate than recurrence after radiation and/or chemotherapy.

For adequate local control following surgical failure, all scar planes as well as reconstructive flaps must be excised in addition to the cancer. Radiation and/or chemotherapy following surgical failure is much less effective than when used before or with surgery. Gallium scan can in some cases detect recurrences or tumors that are ≥ 2 cm.

Unknown Primary and Cervical Metastases

After detection of cancerous cervical adenopathy, 80% of primary carcinoma will be found in the upper respiratory or alimentary tracts. If adenopathy occurs in the supraclavicular area, only 40% of patients will have the primary site of origin in the head and neck. Evaluation of a neck mass may require endoscopy and biopsy of all suspicious areas. Random biopsies of the tonsils, base of tongue, and nasopharynx are indicated if no lesions are found. Open biopsy of a neck mass suspicious for carcinoma is contraindicated unless definitive radical neck surgery is simultaneously undertaken; otherwise, open biopsy severely affects patient prognosis. In contradistinction, fine needle (20 to 22 gauge) aspiration biopsy of a cervical mass can be complementary to the workup of an unknown primary, without spreading cancer or affecting patient prognosis.

Treatment: Palpable cancerous cervical adenopathy requires surgical removal and radiation therapy to the nasopharynx, palatine tonsils, base of the tongue, and both sides of the neck. Nonpalpable cervical metastases with a squamous cell cancer primary in the head and neck respond equally to surgery or radiation.

Premalignant Lesions

(Same as in Ch. 251)

Chapter 251: Preneoplastic and Neoplastic Lesions

Oral Preneoplastic and Early Lesions

Approximately 20,000 new cases of oral cancer (mainly squamous) are reported each year in the USA and account for about 5% of cancers in men and 2% in women. More than any other factor, the stage of these cancers determines the prognosis (see Staging and Prognosis in Ch. 212). While oral cancers < 1 cm in diameter are easily cured, most lesions are *not* diagnosed before stage III or IV and ≥ 50% have metastasized to lymph nodes. Therefore, 5-yr survival rates remain at 30 to 40%. This unfortunate situation appears to result from inadequate knowledge of appropriate screening procedures and strongly entrenched misconceptions.

Persons at risk: While screening is easy enough to include all patients, careful attention to patients with clearly defined **risk factors** is mandatory; i.e. individuals ≥ 40 yr old, those who smoke ≥
1 pack of cigarettes/day, those who use smokeless ("chewing") tobacco, and those who drink alcoholic beverages regularly. Screening these individuals has been reported to increase discovery rates of early cancer in these high risk populations to 1/200 to 1/250.

Sites at risk: Next, it should be recognized that 90% of oral cancers are detected in only a few "high risk" sites: the floor of the mouth, the ventrolateral aspect of the tongue, and the soft palate complex (uvula, soft palate proper, anterior pillar, and lingual aspects of the retromolar trigone). Buccal carcinoma should be considered in people who smoke cigars or piper or use smokeless tobacco.

Misconceptions: Most physicians and dentists have been taught that **leukoplakia** (white lesions) are the most common precancerous lesions in the mouth and that early cancers are white lesions. In fact, < 5% of such lesions ultimately prove to be cancerous. Early, asymptomatic oral cancer appears most often as a **red (erythroplastic) lesion.** These lesions are not *pre*cancerous; they are early carcinomas. These areas look like an inflammation, probably as the result of a submucosal round-cell infiltrate that has arisen below the malignant squamous cells in response to the developing neoplasm. When dry, these red lesions appear more granular or slightly abraded. Therefore, they should be dried gently with a piece of gauze and examined carefully under good light. Two distinct types of erythroplastic lesions have been identified: (1) a red, granular lesion (appearing like worn velvet) speckled with islands of keratin (white) or normal mucosa within or peripheral to the red component, and (2) a smooth, nongranular, red lesion with minimal associated keratin, similat to a nonspecific inflammation. Both types may have irregular, ill-defined borders; generally, palpation is not helpful diagnostically, as few are indurated or raised. *Any erythroplastic lesion that does not repond to treatment and persists > 14 days should be considered carcinoma in situ or invasive carcinoma and requires biopsy.* Unfortunately, **squamous cell carcinoma** is not usually diagnosed in its earliest stages and later appears as a deep ulcer, with smooth,

indurated, rolled margins, fixed to deeper tissues.

Other malignancies of the oral cavity are epidermoid carcinoma of the lip, cheek, and tongue; lymphoepithelioma; melanoma; and myelocytic and lymphocytic leukemias. **Benign lesions that may be confused with oral cancer include** irritation, fibroma, papilloma, granuloma (including pregnancy tumor), the glossitis of avitaminosis, geographic tongue, median rhomboid glossitis, hemangioma and lymphangioma, fibrous hypertrophy of the gingiva, melanosis, myoblastoma, retention cysts (including ranula), xanthomatosis, torus palatinus, submandibular duct calculus, hypertrophy of the foliate papillae, radiculodental cysts, and ameloblastoma. Syphilis, erosive lichen planus, benign ulcer, TB, leukoplakia, and dental abscess should also be considered. Exfoliative cytology is useful in screening, but biopsy is essential to establish the diagnosis.

Multiple carcinomas: *Patients with an oral cancer are at high risk (up to 33%) of developing a second primary neoplasm in the mouth, pharynx, larynx, esophagus, or lung.* Therefore, patients identified as having an oral cancer should be screened for cancer in all these sites and reexamined at yearly intervals (i.e. examination of mouth and throat, indirect laryngoscopy, chest x-ray).

Treatment

Any recognizable irritation (i.e. faulty restorations and prosthetic devices) should be corrected or removed. Tobacco in any form should be eliminated. Mucosal drying agents such as alcoholic beverages and mouth rinses with alcoholic vehicles should be discontinued.

Treatment of oral neoplasms generally consists of wide local excision for small lesions and en bloc excision of larger lesions in continuity with radical neck dissection if lymph nodes are involved. Radiotherapy alone may be appropriate for certain small or large lesions or may be combined with surgery. Chemotherapy may be used as palliation or as an adjunct to surgery and radiotherapy.

Neoplasms of Specific Tissues

Lips, gingiva, and tongue: Smoker's patch is a firm, brownish, keratotic plaque on the vermilion border of the lower lip and is most common in smokers who hold a cigarette or pipe in one location. *Only a biopsy can rule out squamous cell carcinoma.* Cessation of smoking and careful observation are recommended even if the lesion is not malignant. **Actinic cheilosis** occurs in adults, especially redheads with fair skin, who spend much time out of doors. The lips are dry with many erosive areas. A person with this *precancerous* lesion should be seen every 3 to 6 mo, avoid prolonged exposure to sunlight, wear a broad-brimmed hat or cap, and use an antiactinic crea. **Squamous cell carcinoma** usually appear on the vermilion border of the lower lip as a nonhealing ulcer with a convex, indurated margin, or less commonly, as a keratotic patch. It may be fixed to the underlying tissues. If treated early, prognosis is excellent. In **leukemia,** the gingival tissue may be infiltrated and prone to bleed. **Rhabdomyoma** of the tongue causes a palpable interior mass. It is much rarer than squamous cell carcinoma, which arises in the mucosa.

Cheek: Irritation of the mucosa is commonly seen in the mucobuccal fold, when chewing tobacco or snuff may be habitually retained. The irritation may progress to erythroplakia or leukoplakia and squamous cell carcinoma. A firm, nodular, nonpainful swelling in the cheek covered by normal-appearing mucosa is rather common. It is usually the result of a cheek bite and is an **"irritation fibroma".** This is considered benign.

Palate: Accessory salivary gland tumor is usually a mixed tumor with both epithelial and mesenchymal components, an adenoic cystic carcinoma, or a mucoepidermoid carcinoma. Typically, it appears as a firm, smooth, painless mass lateral to the midline. Any such swelling that is not bony hard should be considered a salivary gland tumor until a biopsy proves otherwise. A **fullness of the palate** can represent extension of a malignant tumor of the lining of the nose or the antrum rather than a primary lesion of the palate. The soft palate may become immobile if a cancer is in the nasopharynx.

Jaws: If not initially detected on x-ray, jaw tumors are diagnosed clinically because their growth causes **swelling** of the face, palate, or alveolar process (the area of the jaw surrounding the teeth). They cause bone tenderness and severe pain originating in the involved bone. **Ameloblastoma,** the most common odontogenic neoplasm, most frequently arises in the posterior mandible and is slowly invasive, but rarely metastatic. On x-ray, it typically appears as a multiloculated or soap-bubble radiolucency. **Odontomas** are tumors of the dental follicle or the dental tissues that usually appear in the mandibles of young people; several types include fibrous odontomas and cementinomas. An absent molar tooth suggests a composite odontoma. **Other neoplasms** include osteogenic sarcoma, giant cell tumor, Ewing's tumor, multiple myeloma, and metastatic tumors.

Salivary glands: The two main types of tumors are the **mixed tumor (pleomorphic adenoma),** 60% of which occur in the parotid glands, and **mucoepidermoid carcinoma.** These tumors occur not only in major salivary glands but about 20% are found in accessory salivary glands located mainly in the palate and the buccal mucosa (see above). One of 6 tumors in the parotid gland, 1/3 of those in the submandibular gland, almost 1/2 of all palatal tumors, and nearly all sublingual gland tumors are malignant.

Slowly developing parotid swellings may be painless and the patient complains because of change in appearance; but acute swelling of the parotid gland is painful because of dense fascia surrounding it. A parotid tumor causes facial paralysis if it compresses or infiltrates the facial nerve, which may also be damaged inadvertently during surgery to remove the tumor. Tumors of the submandibular salivary glands are often painful because of close association with the lingual branch of the trigeminal nerve.