اکسیر

بهترین
نرم افزار مطب

+ قلم نوری
+ مالی و نسخ الکترونیک
+ اثـر انگشـت بیمـاران

The Ear

PART XXVIII

Chapter 676

General Considerations and Evaluation of the Ear

Joseph Haddad Jr.

CLINICAL MANIFESTATIONS

Diseases of the ear and temporal bone typically manifest with one or more of eight clinical signs and symptoms.

Otalgia usually is associated with inflammation of the middle ear (about 50% of cases) or external ear, but it can represent pain referred from involvement of the teeth, temporomandibular joint, or pharynx (Table 676.1). In young infants, pulling or rubbing the ear along with general irritability or poor sleep, especially when associated with fever, may be the only signs of ear pain. Ear pulling alone is not diagnostic of ear pathology.

Purulent otorrhea is a sign of otitis externa, otitis media (OM) with perforation of the tympanic membrane (TM), drainage from the middle ear through a patent tympanostomy tube, or, rarely, drainage from a first branchial cleft sinus. Bloody drainage may be associated with acute or chronic inflammation (often with granulation tissue and/or an ear tube), trauma, neoplasm, foreign body, or blood dyscrasia. Clear drainage suggests a perforation of the TM with a serous middle-ear effusion or, rarely, a cerebrospinal fluid leak draining through defects (congenital or traumatic) in the external auditory canal or from the middle ear.

Hearing loss results either from disease of the external or middle ear (conductive hearing loss) or from pathology in the inner ear, retrocochlear structures, or central auditory pathways (sensorineural hearing loss [SNHL]); the underlying etiology can be genetic or nongenetic, syndromic or nonsyndromic, or idiopathic. The most common cause of hearing loss in children is OM.

Swelling around the ear most commonly is a result of inflammation (e.g., external otitis, perichondritis, mastoiditis), trauma (e.g., hematoma), benign cystic masses, or neoplasm.

Vertigo is a specific type of dizziness that is defined as any illusion or sensation of motion. **Dizziness** is less specific than vertigo and refers to a sensation of altered orientation in space. Vertigo is an uncommon complaint in children; the child or parent might not volunteer information about balance unless asked specifically. The most common cause of dizziness in young children is eustachian tube–middle-ear disease, but true vertigo also may be caused by labyrinthitis, perilymphatic fistula between the inner and middle ear as a result of trauma or a congenital inner ear defect, cholesteatoma in the mastoid or middle ear, vestibular neuronitis, benign paroxysmal vertigo, Meniere disease, or disease of the central nervous system. Older children might describe a feeling of the room spinning or turning; younger children might express the dysequilibrium only by falling, stumbling, or clumsiness.

Nystagmus may be unidirectional, horizontal, or jerk nystagmus. It is vestibular in origin and usually is associated with vertigo.

Tinnitus rarely is described spontaneously by children, but it is common, especially in patients with eustachian tube–middle-ear disease or SNHL. Children can describe tinnitus if asked directly about it, including laterality and the quality of the sound.

FACIAL PARALYSIS

The facial nerve may be dehiscent in its course through the middle ear as a normal variant in as many as 50% of people. Infection with local inflammation, most commonly in acute OM, can lead to a temporary paralysis of the facial nerve. It also can result from Lyme disease, cholesteatoma, Bell palsy, Ramsay Hunt syndrome (herpes zoster oticus), fracture, neoplasm, or infection of the temporal bone. Congenital facial paralysis can result from birth trauma or congenital abnormality of the seventh nerve or from a syndrome such as Möbius or CHARGE (coloboma, heart defects, atresia choanae, retarded growth, genital hypoplasia, and ear anomalies), or it may be associated with other cranial nerve abnormalities and craniofacial anomalies.

PHYSICAL EXAMINATION

Complete examination with special attention to the head and neck can reveal a condition that can predispose to or be associated with ear disease in children. The facial appearance and the character of speech can give clues to an abnormality of the ear or hearing. Many craniofacial anomalies, such as cleft palate, mandibulofacial dysostosis (Treacher Collins syndrome), and trisomy 21 (Down syndrome), are associated with disorders of the ear and eustachian tube. Mouth breathing and hyponasality can indicate intranasal or postnasal obstruction. **Hypernasality** is a sign of velopharyngeal insufficiency. Examining the oropharyngeal cavity might uncover an overt cleft palate or a submucous cleft (usually associated with a bifid uvula), both of which predispose to OM with effusion. A nasopharyngeal tumor with nasal and eustachian tube blockage may be associated with OM.

The position of the patient for examination of the ear, nose, and throat depends on the patient's age and ability to cooperate, the clinical setting, and the examiner's preference. The child can be examined on an examination table or on the parent's lap. The presence of a parent or assistant usually is necessary to minimize movement and provide better examination results. An examining table may be desirable for uncooperative older infants or when a procedure, such as microscopic evaluation or tympanocentesis, is performed. Lap examination is adequate in most infants and young children; the parent may assist in restraining the child by folding the child's wrists and arms over the child's own abdomen with one hand and holding the child's head against the parent's chest with the other hand. If necessary, the child's legs can be held between the parent's knees. To avoid ear trauma with movement, the examiner should hold the otoscope with the hand placed firmly against the child's head or face, so that the otoscope moves with the head. Pulling up and out on the pinna straightens the ear canal and allows better exposure of the TM.

When examining the ear, inspecting the auricle and external auditory meatus for infection can aid in evaluating complications of OM. External otitis can result from acute OM with discharge, or inflammation of the posterior auricular area can indicate a periostitis or subperiosteal abscess extending from the mastoid air cells. The presence of preauricular pits or skin tags also should be noted because affected children have a slightly higher incidence of SNHL; ear pits can develop chronic infection.

Cerumen is a protective, waxy, water-repellent coating in the ear canal that can interfere with examination. Cerumen usually is removed using the surgical head of the otoscope, which allows passage of a wire loop or a blunt curette under direct visualization. Other methods include gentle irrigation of the ear canal with warm water, which should be performed only if the TM is intact, or instillation of a solution such as diluted hydrogen peroxide in the ear canal (with intact TM only) for a few minutes to soften the wax for suction removal or irrigation. Commercial preparations such as trolamine polypeptide oleate–condensate (Cerumenex) may cause dermatitis of the external canal with chronic use and should be used only under a physician's supervision.

Inflammation of the ear canal with associated pain often indicates external otitis. Abnormalities of the external auditory canal include

Table 676.1 Causes of Otalgia and Sources for Referred Pain

INTRINSIC
External Ear
External otitis
Cerumen impaction
Foreign body
Perichondritis
Preauricular cyst or sinus
Impacted insects
Myringitis
Trauma
Tumor
Middle Ear, Eustachian Tube, and Mastoid
Barotrauma
Middle ear effusion
Negative intratympanic pressure (eustachian tube dysfunction)
Acute otitis media
Mastoiditis
Aditus block
Complication of otitis media
Gradenigo syndrome (otorrhea, CN VI palsy, pain in CN V distribution, petrositis)
Tumor
Eosinophilic granuloma
Granulomatosis with polyangiitis
EXTRINSIC
Trigeminal Nerve
Dental
Jaw
Temporomandibular joint
Oral cavity (tongue)
Infratemporal fossa tumors
Facial Nerve
Bell palsy
Tumors
Herpes zoster
Glossopharyngeal Nerve
Tonsil
Oropharynx
Nasopharynx
Vagus Nerve
Laryngopharynx
Esophagus
Gastroesophageal reflux
Thyroid
Cervical Nerves
Lymph nodes
Cysts
Cervical spine
Neck infections
Miscellaneous
Migraine
Neuralgias
Paranasal sinuses
Central nervous system
Drug induced (mesalazine, sulfasalazine)
Factitious disorder by proxy

From Bluestone CD, Stool SE, Alper CM, et al. *Pediatric Otolaryngology*, 4th ed., vol. 1. Philadelphia: Saunders; 2003:288.

stenosis (common in children with trisomy 21), bony exostoses, otorrhea, and the presence of foreign bodies. Cholesteatoma of the middle ear can manifest in the canal as intermittent foul-smelling drainage, sometimes associated with white debris; cholesteatoma of the external canal can appear as a white, pearl-like mass in the canal skin. White or gray debris of the canal suggests fungal external otitis. Newborn ear canals are filled with vernix caseosa, which is soft and pale yellow and should disappear shortly after birth.

The TM and its mobility are best assessed with a pneumatic otoscope. The normal TM is in a neutral position; a bulging TM may be caused by increased middle-ear air pressure, with or without pus or effusion in the middle ear; a bulging drum can obscure visualization of the malleus and annulus. Retraction of the TM usually indicates negative middle-ear pressure, but it also can result from previous middle-ear disease with fixation of the ossicles, ossicular ligaments, or TM. When retraction is present, the bony malleus appears more prominent, and the incus may be more visible posterior to the malleus.

The normal TM has a silvery gray, "waxed paper" appearance. A white or yellow TM can indicate a middle-ear effusion. A red TM alone might not indicate pathology, because the blood vessels of the membrane may be engorged as a result of crying, sneezing, or nose blowing, though hemorrhagic redness is associated with acute OM. A normal TM is translucent, allowing the observer to visualize the middle-ear landmarks: incus, promontory, round window niche, and often the chorda tympani nerve. If a middle-ear effusion is present, an air-fluid level or bubbles may be visible (Fig. 676.1). Inability to visualize the middle-ear structures indicates opacification of the drum, usually caused by thickening of the TM or a middle-ear effusion or both. Assessment of the light reflex often is not helpful, because a middle ear with effusion reflects light as well as a normal ear. Bullae (blister of the TM) formation is associated with acute OM.

TM mobility is helpful in assessing middle-ear pressures and the presence or absence of fluid (see Fig. 676.1). To best perform pneumatic otoscopy, a speculum of adequate size is used to obtain a good seal and allow air movement in the canal. A rubber ring around the tip of the speculum can help obtain a better canal seal. Normal middle-ear pressure is characterized by a neutral TM position and brisk TM movement to both positive and negative pressures.

Eardrum retraction is most common when negative middle-ear pressure is present; with even moderate negative middle-ear pressure, there is no visible inward movement with applied positive pressure in the ear canal. However, negative canal pressure, which is produced by releasing the rubber bulb of the pneumatic otoscope, can cause the TM to bounce out toward the neutral position. The TM can retract in both the presence and absence of middle-ear fluid, and if the middle-ear fluid is mixed with air, the TM might still have some mobility. Outward eardrum movement is less likely in the presence of severe negative middle-ear pressure or middle-ear effusion.

The TM that exhibits fullness (bulging) moves to applied positive pressure but not to applied negative pressure if the pressure within the middle ear is positive. A full TM and positive middle-ear pressure without an effusion may be seen in young infants who are crying during the otoscopic examination, in older infants and children with nasal obstruction, and in the early stage of acute OM. When the middle-ear–mastoid air cell system is filled with an effusion and little or no air is present, the mobility of the TM is severely decreased or absent in response to both applied positive and negative pressures.

Tympanocentesis, or aspiration of the middle ear, is the definitive (but not usually needed) method of verifying the presence and type of a middle-ear effusion and is performed by inserting an 18-gauge spinal needle attached to a syringe or a collection trap through the inferior portion of the TM (Fig. 676.2). Culturing of the ear canal and alcohol cleansing should precede tympanocentesis and culture of the middle-ear aspirate; a canal culture is taken first to help determine whether organisms cultured from the middle ear are contaminants from the external canal or true middle-ear pathogens.

Further diagnostic studies of the ear and hearing include audiometric evaluation, impedance audiometry (tympanometry), acoustic reflectometry, and specialized eustachian tube function studies. Diagnostic imaging studies, including CT and MRI, often provide further information about anatomic abnormalities and the extent of inflammatory processes or neoplasms. Specialized assessment of labyrinthine function should be considered in the evaluation of a child with a suspected vestibular disorder (see Chapter 682).

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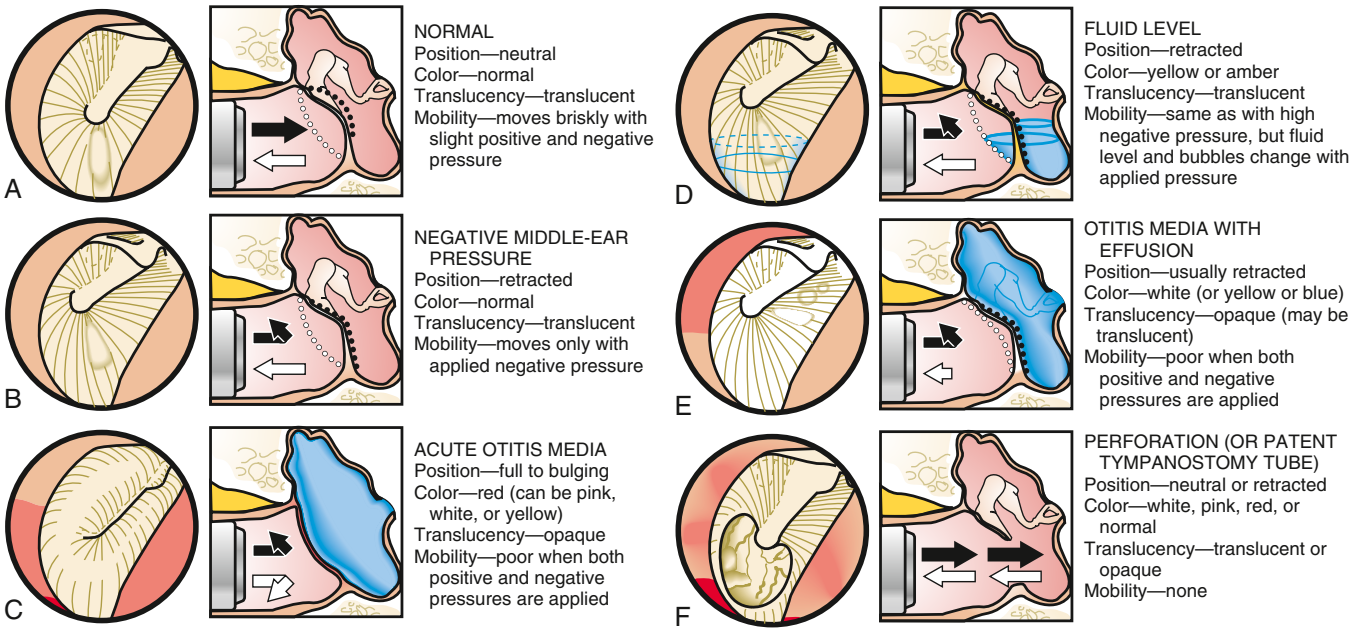

Fig. 676.1 A–F, Common conditions of the middle ear, as assessed with the otoscope. *(From Bluestone CD, Klein JO. Otitis Media in Infants and Children, 3rd ed. Philadelphia: Saunders; 2001:131.)*

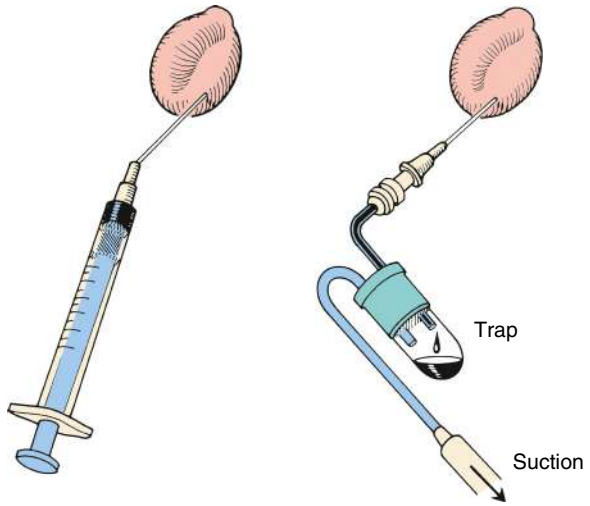

Fig. 676.2 Tympanocentesis can be performed with a needle attached to a tuberculin syringe *(left)* or by using an Alden-Senturia collection trap (Storz Instrument Co, St. Louis). *(From Bluestone CD, Klein JO. Otitis Media in Infants and Children, 2nd ed. Philadelphia: Saunders; 1995:127.)*

Chapter 677

Hearing Loss

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See also Chapter 55.

INCIDENCE AND PREVALENCE

Bilateral neural hearing loss is categorized as **mild** (20-30 dB hearing level [HL]), **moderate** (30-50 dB HL), **moderately severe** (50-70 dB HL), **severe** (75-85 dB HL), or **profound** (>85 dB). The World Health Organization estimates that approximately 360 million people (5% of the world's population, including 32 million children) have disabling hearing loss. An additional 364 million people have mild hearing loss. Half of these cases could have been prevented. In the United States, the average incidence of neonatal hearing loss is 1.6 per 1,000 infants; the rate by state varies from 0.22 to 3.61 per 1,000. Among children and adolescents, the prevalence of mild or greater hearing loss is 3.1% and is higher among persons from lower-income families.

Onset of hearing loss in children can occur at any time in childhood. When less severe hearing loss or the transient hearing loss that commonly accompanies middle-ear disease in young children is considered, the number of affected children increases substantially.

TYPES OF HEARING LOSS

Hearing loss can be peripheral or central in origin. Peripheral hearing loss can be conductive, sensorineural, or mixed. **Conductive hearing loss (CHL)** is the most common type of hearing loss in children and occurs when sound transmission is physically impeded in the external and/or middle ear, most commonly by otitis media (OM) with effusion. **Sensorineural hearing loss (SNHL)** is caused by damage to or maldevelopment of structures of the inner ear, including hair cell destruction, cochlear malformation; perilymphatic fistula of the round or oval window membrane, and failure in development or lesions of the acoustic division of the eighth nerve. Coexistent CHL and SNHL is considered a **mixed hearing loss**.

An auditory deficit originating along the central auditory nervous system pathways from the proximal eighth nerve to the cerebral cortex usually is considered **central** (or **retrocochlear**) **hearing loss**. Tumors or demyelinating disease of the eighth nerve and cerebellopontine angle can cause hearing deficits but spare the outer, middle, and inner ear. These causes of hearing loss are rare in children. Functional disorders of the eighth nerve and/or brainstem pathways may manifest in a variety of clinical defects known collectively as *auditory neuropathy spectrum disorder* (ANSD) or *auditory dyssynchrony,* without abnormalities demonstrable on imaging. Other forms of central auditory deficits, known as **central auditory processing disorders**, include those that make it difficult even for children with normal hearing sensitivity to listen selectively in the presence of noise, to combine information from the two ears properly, to process speech when it is slightly degraded, and to integrate auditory information when it is delivered faster, although they can process it when delivered at a slow rate. These deficits can manifest as specific language disorders or poor attention or as academic or behavior problems

in school. Strategies for coping with such disorders are available for older children, and identification and documentation of the central auditory processing disorder allow parents and teachers to make appropriate accommodations to enhance learning.

ETIOLOGY

Most CHL is acquired, with middle-ear fluid the most common cause. Congenital causes include anomalies of the pinna, external ear canal, tympanic membrane (TM), and ossicles. Rarely congenital cholesteatoma or other masses in the middle ear manifest as CHL. TM perforation (e.g., trauma, OM), ossicular discontinuity (e.g., infection, cholesteatoma, trauma), tympanosclerosis, acquired cholesteatoma, or masses in the ear canal or middle ear (Langerhans cell histiocytosis, salivary gland tumors, glomus tumors, rhabdomyosarcoma) also can manifest as CHL. Uncommon diseases that affect the middle ear and temporal bone and can manifest with CHL include otosclerosis, osteopetrosis, fibrous dysplasia, and osteogenesis imperfecta. Rare autoimmune or inflammatory syndromes may include hearing loss. **Susac syndrome** is manifest as a subacute encephalopathy, visual impairment, and hearing loss. **Cogan syndrome** presents with interstitial keratitis, sudden hearing loss, and vestibular impairment.

SNHL may be congenital or acquired. Acquired SNHL may be caused by genetic, infectious, autoimmune, anatomic, traumatic, ototoxic, and idiopathic factors (Tables 677.1-677.4). The recognized risk factors account for approximately 50% of cases of moderate to profound SNHL.

Infectious Causes

The most common infectious cause of congenital SNHL is **cytomegalovirus (CMV)**, which infects 1 in 100 newborns in the United States (see Chapters 149 and 302). Of these, 6,000-8,000 infants each year have clinical manifestations, including approximately 75% with SNHL. Congenital CMV warrants special attention because it is associated with hearing loss in its symptomatic and asymptomatic forms with bilateral and unilateral hearing loss, respectively; the hearing loss may be progressive. Some children with congenital CMV have suddenly lost residual hearing at 4-5 years of age. Much less common congenital infectious causes of SNHL include toxoplasmosis and syphilis. Congenital CMV, toxoplasmosis, and syphilis also can manifest with delayed onset of SNHL months to years after birth. Rubella, once the most common viral cause of congenital SNHL, is very uncommon because of effective vaccination programs. In utero infection with herpes simplex virus is rare, and hearing loss is not an isolated manifestation.

Other postnatal infectious causes of SNHL include neonatal group B streptococcal sepsis and bacterial meningitis at any age. *Streptococcus pneumoniae* is the most common cause of bacterial meningitis that results in SNHL after the neonatal period and has become less common with the routine administration of pneumococcal conjugate vaccine. *Haemophilus influenzae* type b, once the most common cause of meningitis resulting in SNHL, is rare owing to the *H. influenzae* type b conjugate vaccine. Uncommon infectious causes of SNHL include Lyme disease, parvovirus B19, and varicella. Mumps, rubella, and measles, all once common causes of SNHL in children, are rare owing to vaccination programs. When these infectious etiologies occur, the resulting hearing loss is frequently bilateral and severe.

Genetic Causes

Genetic causes of SNHL probably are responsible for as many as 50% of SNHL cases (see Tables 677.3 and 677.4). These disorders may be associated with other abnormalities, may be part of a named syndrome, or can exist in isolation. SNHL often occurs with abnormalities of the ear and eye and with disorders of the metabolic, musculoskeletal, integumentary, renal, and nervous systems.

Autosomal dominant hearing losses account for approximately 10% of all cases of childhood SNHL. Waardenburg (types I and II) and branchiootorenal syndromes represent two of the most common autosomal dominant syndromic types of SNHL. Types of SNHL are coded with a four-letter code and a number, as follows: **DFN** = deafness, A = dominant, B = recessive, and number = order of discovery (e.g., DFNA 13). Autosomal dominant conditions in addition to those just discussed include DFNA 1-18, 20-25, 30, 36, 38, and pathologic variants in the crystallin gene *(CRYM)*.

Autosomal recessive genetic SNHL, both syndromic and nonsyndromic, accounts for approximately 80% of all childhood cases of

Table 677.1 Indicators Associated with Hearing Loss

INDICATORS ASSOCIATED WITH SENSORINEURAL AND/OR CONDUCTIVE HEARING LOSS

Neonates (Birth to 28 Days) When Universal Screening Is Not Available

- Family history of hereditary childhood sensorineural hearing loss
- In utero infection, such as cytomegalovirus, rubella, syphilis, herpes simplex, or toxoplasmosis
- Craniofacial anomalies, including those with morphologic abnormalities of the pinna, ear canal, ear tags, ear pits, and temporal bone anomalies
- Birth weight <1,500 g (3.3 lb)
- Hyperbilirubinemia at a serum level requiring exchange transfusion
- Ototoxic medications, including but not limited to the aminoglycosides, used in multiple courses or in combination with loop diuretics
- Bacterial meningitis
- Apgar scores of 0-4 at 1 min or 0-6 at 5 min
- Mechanical ventilation lasting ≥5 days; extracorporeal membrane oxygenation
- Stigmata or other findings associated with a syndrome known to include a sensorineural and/or conductive hearing loss; white forelock

Infants and Toddlers (Age 29 Days to 2 Yr) When Certain Health Conditions Develop that Require Rescreening

- Parent or caregiver concern regarding hearing, speech, language, and/or developmental delay
- Bacterial meningitis and other infections associated with sensorineural hearing loss
- Head trauma associated with loss of consciousness or skull fracture
- Stigmata or other findings associated with a syndrome known to include a sensorineural and/or conductive hearing loss; neurofibromatosis, osteopetrosis, and Usher Hunter, Waardenburg, Alport, Pendred, or Jervell and Lange-Nielsen syndrome
- Ototoxic medications, including but not limited to chemotherapeutic agents or aminoglycosides used in multiple courses or in combination with loop diuretics
- Recurrent or persistent otitis media with effusion for 3 mo or longer
- Skeletal dysplasia

Infants and Toddlers (Age 29 Days to 3 Yr) Who Require Periodic Monitoring of Hearing

- Some newborns and infants pass initial hearing screening but require periodic monitoring of hearing to detect delayed-onset sensorineural and/or conductive hearing loss. Infants with these indicators require hearing evaluation at least every 6 mo until age 3 yr and at appropriate intervals thereafter

INDICATORS ASSOCIATED WITH DELAYED-ONSET SENSORINEURAL HEARING LOSS

- Family history of hereditary childhood hearing loss
- In utero infection, such as cytomegalovirus, rubella, syphilis, herpes simplex, or toxoplasmosis
- Neurofibromatosis type 2 and neurodegenerative disorders
- Cogan syndrome (vasculitis: keratitis, uveitis, vertigo, arthritis, dermatitis)

INDICATORS ASSOCIATED WITH CONDUCTIVE HEARING LOSS

- Recurrent or persistent otitis media with effusion
- Anatomic deformities and other disorders that affect eustachian tube function
- Neurodegenerative disorders

Note: At all ages, parents' concern about hearing loss must be taken seriously even in the absence of risk factors.

Adapted from American Academy of Pediatrics, Joint Committee on Infant Hearing. Joint Committee on Infant Hearing 1994 position statement. *Pediatrics.* 1995;95:152.

Table 677.2 Infectious Pathogens Implicated in Sensorineural Hearing Loss in Children

CONGENITAL INFECTIONS
Cytomegalovirus
Lymphocytic choriomeningitis virus
Rubella virus
Toxoplasma gondii
Treponema pallidum
ACQUIRED INFECTIONS
Borrelia burgdorferi (Lyme disease)
Cytomegalovirus
Epstein-Barr virus
Haemophilus influenzae
Herpes simplex
Lassa fever virus
Measles virus
Mumps virus
Neisseria meningitidis
Nonpolio enteroviruses
Plasmodium falciparum
Rubella
Streptococcus pneumoniae
Syphilis
Varicella-zoster virus

From Smith RJH, Bale JF Jr., White KR. Sensorineural hearing loss in children. *Lancet.* 2005;365:879–890.

SNHL. Usher syndrome (types 1, 2, and 3: all associated with blindness and retinitis pigmentosa), Pendred syndrome, and Jervell and Lange-Nielsen syndrome (one form of long QT syndrome) are three of the most common syndromic recessive types of SNHL. Other autosomal recessive conditions include Alström syndrome, type 4 Bartter syndrome, biotinidase deficiency, and DFNB 1-18, 20-23, 26-27, 29-33, 35-40, 42, 44, 46, 48, 49, 53, and 55.

Unlike children with an easily identified syndrome or with anomalies of the outer ear, who may be identified as being at risk for hearing loss and consequently monitored, children with nonsyndromic hearing loss present greater diagnostic difficulty. Pathogenic variants of the connexin-26 and -30 genes are identified in autosomal recessive (DNFB 1) and autosomal dominant (DNFA 3) SNHL and in sporadic patients with nonsyndromic SNHL; up to 50% of nonsyndromic SNHLs may be related to a pathogenic variation of connexin-26. Pathologic variants of the *GJB2* gene co-localize with DFNA 3 and DFNB 1 loci on chromosome 13, are associated with autosomal nonsyndromic susceptibility to deafness, and are associated with as many as 30% of cases of sporadic severe to profound congenital deafness and 50% of cases of autosomal recessive nonsyndromic deafness. In addition, pathogenic variants in *GJB6* are associated with approximately 5% of recessive nonsyndromic deafness. Sex-linked disorders associated with SNHL, thought to account for 1–2% of SNHLs, include Norrie disease, otopalatal digital syndrome, Nance deafness, and Alport syndrome. Chromosomal abnormalities such as trisomy 13-15, trisomy 18, and trisomy 21 also can be accompanied by hearing impairment. Patients with Turner syndrome have monosomy for all or part of one X chromosome and can have CHL, SNHL, or mixed hearing loss. The hearing loss may be progressive. Mitochondrial genetic abnormalities (MELAS, MERRF) also can result in SNHL (see Table 677.3).

Many genetically determined causes of hearing impairment, both syndromic and nonsyndromic, do not express themselves until sometime after birth. Alport, Alström, Down, and Hunter-Hurler syndromes and von Recklinghausen disease are genetic diseases that can have SNHL as a late manifestation.

Physical Causes

Agenesis or malformation of cochlear structures may be genetic; these include Scheibe, Mondini (Fig. 677.1), Alexander, and Michel anomalies, enlarged vestibular aqueducts (in isolation or associated with Pendred syndrome), and semicircular canal anomalies. These anomalies most likely develop before the eighth week of gestation and result from arrest in normal development, aberrant development, or both. Many of these anomalies also have been described in association with other congenital conditions such as intrauterine CMV and rubella infections. These abnormalities are quite common; in as many as 20% of children with SNHL, obvious or subtle temporal bone abnormalities are seen on high-resolution CT scanning or MRI.

Conditions, diseases, or syndromes that include craniofacial abnormalities may be associated with CHL and possibly with SNHL. Pierre Robin sequence, Stickler syndrome, and Treacher Collins, Klippel-Feil, Crouzon, and branchiootorenal syndromes and osteogenesis imperfecta often are associated with hearing loss. Congenital anomalies causing CHL include malformations of the ossicles and middle-ear structures and atresia of the external auditory canal.

SNHL also can occur secondary to exposure to toxins, chemicals, antimicrobials, and noise exposure. Early in pregnancy, the embryo is particularly vulnerable to the effects of toxic substances. Ototoxic drugs, including aminoglycosides, loop diuretics, and chemotherapeutic agents (cisplatin) also can cause SNHL. Congenital SNHL can occur secondary to exposure to these drugs as well as to thalidomide and retinoids. Certain chemicals, such as quinine, lead, and arsenic, can cause hearing loss both prenatally and postnatally. Among adolescents, the use of personal listening devices at high volume settings has been found to be correlated with hearing loss.

Trauma, including temporal bone fractures, inner ear concussion, head trauma, iatrogenic trauma (e.g., surgery, extracorporeal membrane oxygenation), radiation exposure, and noise, also can cause SNHL. Other uncommon causes of SNHL in children include autoimmune disease (systemic or limited to the inner ear), metabolic abnormalities, and neoplasms of the temporal bone.

EFFECTS OF HEARING IMPAIRMENT

The effects of hearing impairment depend on the nature and degree of the hearing loss and on the individual characteristics of the child. Hearing loss may be unilateral or bilateral, conductive, sensorineural, or mixed; mild, moderate, severe, or profound; of sudden or gradual onset; stable, progressive, or fluctuating; and affecting a part or all of the audible spectrum. Other factors, such as intelligence, medical or physical condition (including accompanying syndromes), family support, age at onset, age at time of identification, and promptness of intervention, also affect the impact of hearing loss on a child (see Chapter 55).

Most hearing-impaired children have some useable hearing. Only 6% of those in the hearing-impaired population have bilateral profound hearing loss. Hearing loss very early in life can affect the development of speech and language, social and emotional development, behavior, attention, and academic achievement. Some cases of hearing impairment are misdiagnosed because affected children have sufficient hearing to respond to environmental sounds and can learn some speech and language but when challenged in the classroom cannot perform to full potential.

Even mild or unilateral hearing loss can have a detrimental effect on the development of a young child and on school performance. Children with such hearing impairments have greater difficulty when listening conditions are unfavorable (e.g., background noise and poor acoustics), as can occur in a classroom. The fact that schools are auditory-verbal environments is unappreciated by those who minimize the impact of hearing impairment on learning. Hearing loss should be considered in any child with speech and language difficulties or below-par performance, poor behavior, or inattention in school (Table 677.5).

Children with moderate, severe, or profound hearing impairment and those with other impairing conditions often are educated in classes or schools for children with special needs. There is a strong trend toward integrating a child with hearing loss into the least restrictive learning environment; this approach can only be successful if there are sufficient supportive services available for auditory and other learning needs. The auditory management and choices regarding modes of communication

Table 677.3 Common Types of Early-Onset Hereditary Nonsyndromic Sensorineural Hearing Loss

LOCUS	GENE	AUDIO PHENOTYPE
DFN3*	*POU3F4*	Conductive hearing loss as a result of stapes fixation mimicking otosclerosis; superimposed progressive SNHL.
DFNA1	*DIAPH1*	Low-frequency loss beginning in the first decade and progressing to all frequencies to produce a flat audio profile with profound losses throughout the auditory range.
DFNA2	*KCNQ4* *GJB3*	Symmetric high-frequency sensorineural loss beginning in the first decade and progressing over all frequencies. Symmetric high-frequency sensorineural loss beginning in the third decade.
DFNA3	*GJB2* *GJB6*	Childhood-onset, progressive, moderate to severe high-frequency sensorineural hearing impairment. Childhood-onset, progressive, moderate to severe high-frequency sensorineural hearing impairment.
DFNA6, 14, and 38	*WFS1*	Early-onset low-frequency sensorineural loss; approximately 75% of families dominantly segregating this audio profile carry missense mutations in the C-terminal domain of wolframin.
DFNA8, and 12	*TECTA*	Early-onset stable bilateral hearing loss affecting mainly mid to high frequencies.
DFNA10	*EYA4*	Progressive loss beginning in the second decade as a flat to gently sloping audio profile that becomes steeply sloping with age.
DFNA11	*MYO7A*	Ascending audiogram affecting low and middle frequencies at young ages and then affecting all frequencies with increasing age.
DFNA13	*COL11A2*	Congenital midfrequency sensorineural loss that shows age-related progression across the auditory range.
DFNA15	*POU4F3*	Bilateral progressive sensorineural loss beginning in the second decade.
DFNA20, and 26	*ACTG1*	Bilateral progressive sensorineural loss beginning in the second decade; with age, the loss increases with threshold shifts in all frequencies, although a sloping configuration is maintained in most cases.
DFNA22	*MYO6*	Postlingual, slowly progressive, moderate to severe hearing loss.
DFNB1	*GJB2, GJB6*	Hearing loss varies from mild to profound. The most common genotype, 35delG/35delG, is associated with severe to profound SNHL in about 90% of affected children; severe to profound deafness is observed in only 60% of children who are compound heterozygotes carrying 1 35delG allele and any other *GJB2* SNHL-causing allele variant; in children carrying 2 *GJB2* SNHL-causing missense mutations, severe to profound deafness is not observed.
DFNB3	*MYO7A*	Severe to profound sensorineural hearing loss.
DFNB4	*SLC26A4*	DFNB4 and Pendred syndrome (see Table 677.5) are allelic. DFNB4 hearing loss is associated with dilation of the vestibular aqueduct and can be unilateral or bilateral. In the high frequencies, the loss is severe to profound; in the low frequencies, the degree of loss varies widely. Onset can be congenital (prelingual), but progressive postlingual loss also is common.
DFNB7, and 11	*TMC1*	Severe to profound prelingual hearing impairment.
DFNB9	*OTOF*	*OTOF*-related deafness is characterized by two phenotypes: prelingual nonsyndromic hearing loss and, less frequently, temperature-sensitive nonsyndromic auditory neuropathy spectrum disorder. The nonsyndromic hearing loss is bilateral severe to profound congenital deafness.
DFNB12	*CDH23*	Depending on the type of mutation, recessive mutations of *CDH23* can cause nonsyndromic deafness or type 1 Usher syndrome (USH1), which is characterized by deafness, vestibular areflexia, and vision loss as a result of retinitis pigmentosa.
DFNB16	*STRC*	Early-onset, nonsyndromic, autosomal recessive sensorineural hearing loss.
mtDNA	*MTRNR1* *MTTS1*	Degree of hearing loss varies from mild to profound but usually is symmetric; high frequencies are preferentially affected; precipitous loss in hearing can occur after aminoglycoside therapy; variable penetrance.

*Approximately 45 DFNA genes are inherited as autosomal recessive or dominant and, less often, X-linked.
SNHL, Sensorineural hearing loss.
Adapted from Smith RJH, Bale JF Jr,. White KR. Sensorineural hearing loss in children. *Lancet*. 2005;365:879–890.

and education for children with hearing impairments must be individualized, because these children are not a homogeneous group. A team approach to individual case management is essential because each child and family unit have unique needs and abilities (see Chapter 55).

HEARING SCREENING

Hearing impairment can have a major impact on a child's development, and because early identification improves prognosis, screening programs have been widely and strongly advocated. The National Center for Hearing Assessment and Management estimates that the detection and treatment at birth of hearing loss saves $400,000 per child in special education costs; screening costs approximately $8-$50/child. Data from the Colorado newborn screening program suggest that if hearing-impaired infants are identified and treated by age 6 months, these children (with the exception of those with bilateral profound impairment) should develop the same level of language as their age-matched peers who are not hearing impaired. These data provide compelling support for establishing mandated newborn hearing screening programs for all children. The American Academy of Pediatrics endorses the goal of universal detection of hearing loss in infants before 3 months of age, with appropriate intervention no later than 6 months of age. The Centers for Disease Control and Prevention estimates that of the approximately 4 million infants born in the United States in 2014, 97.9% were screened for hearing loss.

Table 677.4 Common Types of Syndromic Sensorineural Hearing Loss

SYNDROME	GENE	PHENOTYPE
DOMINANT		
Waardenburg (WS1) (may also be recessive)	*PAX3*	Major diagnostic criteria include dystopia canthorum, congenital hearing loss, heterochromic irises, white forelock, and an affected first-degree relative. Approximately 60% of affected children have congenital hearing loss; in 90%, the loss is bilateral.
Waardenburg (WS2) (WS2D) (WS2E) (WS4A) (WS4B)	*MITF*, others *SNAI12* *SOX10* *EDNRB* *EDN3*	Major diagnostic criteria are as for WS1 but without dystopia canthorum. Approximately 80% of affected children have congenital hearing loss; in 90%, the loss is bilateral.
Branchiootorenal	*EYA1* *SIX1* *SIX5*	Diagnostic criteria include hearing loss (98%); preauricular pits (85%); and branchial (70%), renal (40%), and external ear (30%) abnormalities. The hearing loss can be conductive, sensorineural, or mixed and mild to profound in degree.
CHARGE syndrome	*CHD7*	Choanal atresia, colobomas, heart defect, retardation, genital hypoplasia, ear anomalies, deafness. Can lead to sensorineural or mixed hearing loss. Can be autosomal dominant or isolated cases.
Goldenhar syndrome	Unknown	Part of the hemifacial microsomia spectrum. Facial hypoplasia, ear anomalies, hemivertebrae, and parotid gland dysfunction. Can cause conductive or mixed hearing loss. Can be autosomal dominant or sporadic.
Stickler	*COL2A1* *COL11A1* *COL11A2* *COL9A1*	Myopia, cleft palate, hearing loss, joint hypermobility, micrognathia.
RECESSIVE		
Pendred syndrome	*SLC26A4*	Diagnostic criteria include sensorineural hearing loss that is congenital, nonprogressive, and severe to profound in many cases, but can be late-onset and progressive; bilateral dilation of the vestibular aqueduct with or without cochlear hypoplasia; and an abnormal perchlorate discharge test or goiter.
Alport syndrome	*COL4A3, COL4A4,* and *COL4A5* (X-linked)	Nephritis, deafness, lens defects, retinitis. Can lead to bilateral sensorineural hearing loss in the 2,000-8,000 Hz range. The hearing loss develops gradually and is not generally present in early infancy.
Usher syndrome type 1 (USH1)	*USH1A, MYO7A, USH1C, CDH23, USH1E, PCDH15, USH1G*	Diagnostic criteria include congenital, bilateral, and profound hearing loss; vestibular areflexia; and retinitis pigmentosa (commonly not diagnosed until tunnel vision and nyctalopia become severe enough to be noticeable).
Usher syndrome type 2 (USH2)	*USH2A, USH2B, USH2C, WHRN, ADGRV1*	Diagnostic criteria include mild to severe, congenital, bilateral hearing loss and retinitis pigmentosa; hearing loss may be perceived as progressing over time because speech perception decreases as diminishing vision interferes with subconscious lip reading.
Usher syndrome type 3 (USH3)	*USH3* *CLRN1*	Diagnostic criteria include postlingual, progressive sensorineural hearing loss; late-onset retinitis pigmentosa; and variable impairment of vestibular function.
Jervell and Lange-Nielsen syndrome	*KCNQ1* *KCNE1*	Severe hearing loss, prolonged QT interval (ECG), and sudden death.

Adapted from Smith RJH, Bale JF Jr., White KR. Sensorineural hearing loss in children. *Lancet.* 2005;365:879–890.

Hearing screening is mandated in at least 45 states, but until screening programs are universally mandated, some hospitals will continue to use other criteria to screen for hearing loss. Some use the high-risk criteria (see Table 677.1) to decide which infants to screen, some screen all infants who require intensive care, and some do both. The problem with using high-risk criteria to screen is that 50% of cases of hearing impairment will be missed, either because the infants are hearing impaired but do not meet any of the high-risk criteria or because they develop hearing loss after the neonatal period.

The recommended hearing screening techniques are either otoacoustic emissions (OAE) testing or auditory brainstem evoked responses (ABRs). The ABR test, an auditory evoked electrophysiologic response that correlates highly with hearing, has been used successfully and cost-effectively to screen newborns and to identify further the degree and type of hearing loss. OAE tests, used successfully in most universal newborn screening programs, are quick, easy to administer, and inexpensive, and they provide a sensitive indication of the presence of hearing loss. Results are relatively easy to interpret. OAE tests elicit no response if hearing is worse than 30-40 dB, no matter what the cause; children who fail OAE tests undergo an ABR for a more definitive evaluation, as ABR has a higher sensitivity and specificity. It is recommended that both OAE measurement and ABR screening be used in the intensive care unit setting. Screening methods such as observing behavioral responses to uncalibrated noisemakers or using automated systems such as the Crib-o-gram (Canon) or the auditory response cradle (in which movement of the infant in response to sound is recorded by motion sensors) *are not recommended.*

Many children become hearing impaired after the neonatal period and therefore are not identified by newborn screening programs. Often it is not until children are in preschool or kindergarten that further hearing screening takes place; an evidence-based systematic review has identified pure-tone and OAE screening to be effective, with pure-tone screening having higher sensitivity. Among adolescents, high-frequency hearing loss is associated with exposure to loud noises, so

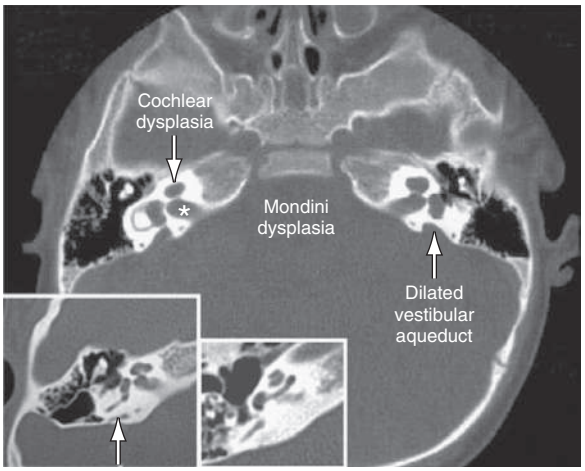

Fig. 677.1 Mondini dysplasia shown by CT of the temporal bone in a child with Pendred syndrome. Both dilation of the vestibular aqueduct and cochlear dysplasia are present in this section. In the larger of the two inset images of a normal temporal bone, the vestibular aqueduct is visible but much smaller *(arrow)*. The cochlea appears normal, and in the smaller inset image of a more inferior axial section, the expected number of cochlear turns can be clearly counted. Internal auditory canal *(asterisk)*. *(From Smith RJH, Bale JF Jr., White KR. Sensorineural hearing loss in children. Lancet. 2005;365:879–890.)*

attention should be paid to those frequencies on a hearing screen; most noise-induced hearing loss is around 4 kHz. Figure 677.2 provides recommendations for postneonatal screening.

IDENTIFICATION OF HEARING IMPAIRMENT

The impact of hearing impairment is greatest on an infant who has yet to develop language; consequently, identification, diagnosis, description, and treatment should begin as soon as possible. Infants with a prenatal or perinatal history that puts them at risk (see Table 677.3) or those who have failed a formal hearing screening should be evaluated by an experienced clinical audiologist until a reliable assessment of auditory sensitivity has been obtained. Primary care clinicians should encourage families to cooperate with the follow-up plan. Infants who are born at risk but who were not screened as neonates (e.g., because of transfer from one hospital to another) should have a hearing screening by age 3 months.

Hearing-impaired infants who are born at risk or are screened for hearing loss in a neonatal hearing screening program account for only a portion of hearing-impaired children. Children who are congenitally deaf because of autosomal recessive inheritance or subclinical congenital infection often are not identified until 1-3 years of age. Usually those with more severe hearing loss are identified at an earlier age, but identification often occurs later than the age at which intervention can provide an optimal outcome, especially in countries lacking technologic resources. Children who hear normally develop extensive receptive and expressive language by 3 years of age (Table 677.6) and exhibit behavior reflecting normal auditory

Table 677.5 Hearing Impairment as a Function of Average Hearing Threshold Level of the Better Ear

AVERAGE THRESHOLD LEVEL (dB) AT 500-2,000 Hz (ANSI)	DESCRIPTION	COMMON CAUSES	WHAT CAN BE HEARD WITHOUT AMPLIFICATION	DEGREE OF IMPAIRMENT (IF NOT TREATED IN FIRST YEAR OF LIFE)	PROBABLE NEEDS
0-15	Normal range	Conductive hearing loss	All speech sounds	None	None
16-25	Slight hearing loss	Otitis media, TM perforation, tympanosclerosis; eustachian tube dysfunction; some SNHL	Vowel sounds heard clearly, may miss unvoiced consonant sounds	Mild auditory dysfunction in language learning Difficulty in perceiving some speech sounds	Consideration of need for hearing aid, speech reading, auditory training, speech therapy, appropriate surgery, preferential seating
26-30	Mild	Otitis media, TM perforation, tympanosclerosis, severe eustachian dysfunction, SNHL	Hears only some speech sounds, the louder voiced sounds	Auditory learning dysfunction Mild language retardation Mild speech problems Inattention	Hearing aid Lip reading Auditory training Speech therapy Appropriate surgery
31-50	Moderate hearing loss	Chronic otitis, ear canal/middle ear anomaly, SNHL	Misses most speech sounds at normal conversational level	Speech problems Language retardation Learning dysfunction Inattention	All of the above, plus consideration of special classroom situation
51-70	Severe hearing loss	SNHL or mixed loss due to a combination of middle-ear disease and sensorineural involvement	Hears no speech sound of normal conversations	Severe speech problems Language retardation Learning dysfunction Inattention	All of the above; probable assignment to special classes
71+	Profound hearing loss	SNHL or mixed	Hears no speech or other sounds	Severe speech problems Language retardation Learning dysfunction Inattention	All of the above; probable assignment to special classes or schools

ANSI, American National Standards Institute; *SNHL*, sensorineural hearing loss; *TM*, tympanic membrane.
Modified from Northern JL, Downs MP. *Hearing in Children*, 4th ed. Baltimore: Williams & Wilkins; 1991.

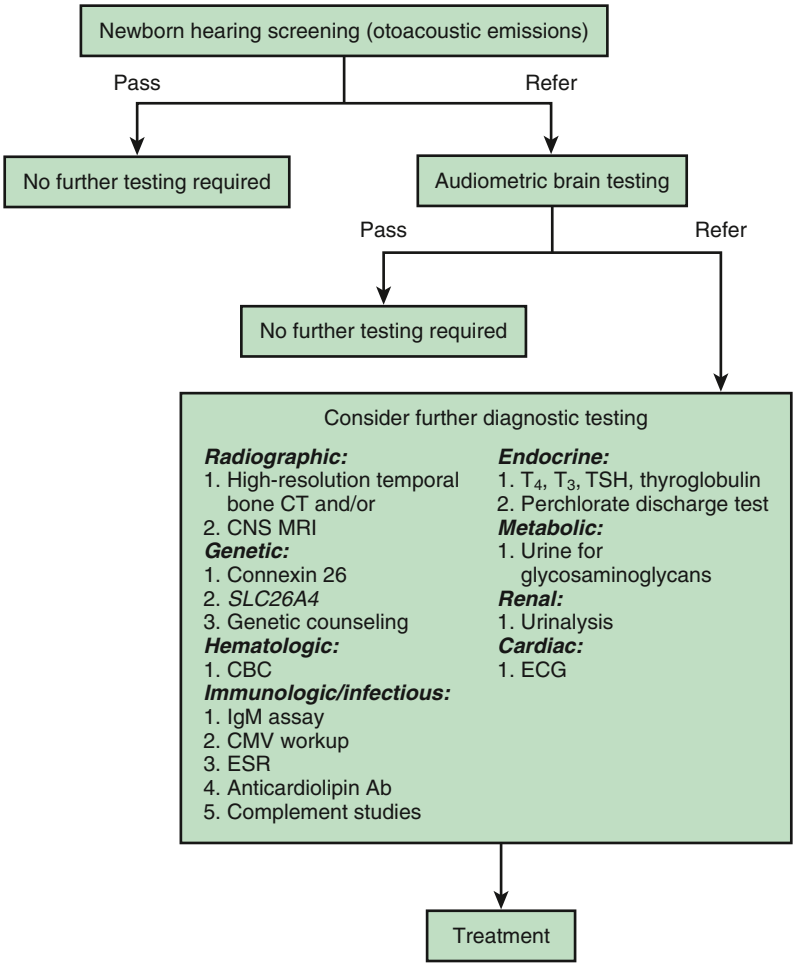

Fig. 677.2 Algorithm for newborn hearing screening. Ab, Antibody; CBC, complete blood count; CMV, cytomegalovirus; CNS, central nervous system; CT, computed tomography; ECG, electrocardiogram; ESR, erythrocyte sedimentation rate; IgM, immunoglobulin M; MRI, magnetic resonance imaging; T3, triiodothyronine; T4, thyroxine; TSH, thyroid-stimulating hormone. *(From Norton SJ, Bhama PK, Perkins JA. Early detection and diagnosis of infant hearing impairment. In: Flint PW, Haughey BH, Lund VJ, et al., eds. Cummings Otolaryngology–Head and Neck Surgery, 5th ed. Philadelphia: Mosby; 2010: Fig. 190.1.)*

Table 677.6 Criteria for Referral for Audiologic and Speech and Language Evaluation

SHOULD ABSOLUTELY REFER FOR A SPEECH-LANGUAGE EVALUATION IF:		
AT AGE (mo)	**RECEPTIVE**	**EXPRESSIVE**
15	Does not look/point at 5-10 objects/people named by parent	Not using three words
18	Does not follow simple directions ("get your shoes.")	Not using Mama, Dada, or other names
24	Does not point to pictures or body parts when they are named	Not using 25 words
30	Does not verbally respond or nod/shake head to questions	Not using unique two-word phrases, including noun-verb combinations
36	Does not understand prepositions or action words; does not follow two-step directions	Vocabulary <200 words; does not ask for things by name; echolalia to questions; regression of language after acquiring two-word phrases

From Schum RL. Language screening in the pediatric office setting. *Pediatr Clin North Am.* 2007;54(3):425–436.

Table 677.7 Guidelines for Referral of Infants/Toddlers with Suspected Hearing Loss

AGE (mo)	NORMAL DEVELOPMENT
0-4	Should startle to loud sounds, quiet to mother's voice, momentarily cease activity when sound is presented at a conversational level
5-6	Should correctly localize to sound presented in a horizontal plane, begin to imitate sounds in own speech repertoire or at least reciprocally vocalize with an adult
7-12	Should correctly localize to sound presented in any plane Should respond to name, even when spoken quietly
13-15	Should point toward an unexpected sound or to familiar objects or persons when asked
16-18	Should follow simple directions without gestural or other visual cues; can be trained to reach toward an interesting toy at midline when a sound is presented
19-24	Should point to body parts when asked; by 21-24 mo, can be trained to perform play audiometry

From Matkin ND. Early recognition and referral of hearing-impaired children. *Pediatr Rev.* 1984;6:151–156.

function (Table 677.7). Failure to fulfill these criteria should be the reason for an audiologic evaluation. Parents' concern about hearing and any delayed development of speech and language should alert the pediatrician, because parents' concern usually precedes formal identification and diagnosis of hearing impairment by 6-12 months.

CLINICAL AUDIOLOGIC EVALUATION

When hearing impairment is suspected in a young child, reliable and valid estimates of auditory function can be obtained using electrophysiologic and age-appropriate behavioral measurement. Successful treatment strategies for hearing-impaired children rely on prompt identification and ongoing assessment to define the dimensions of auditory function. Cooperation among primary care providers and specialists in areas such as audiology, speech and language pathology, education, and child development is necessary to optimize auditory-verbal development. Therapy for hearing-impaired children may include an amplification device, a **frequency modulation** (FM) system in the classroom, close monitoring of hearing and auditory skills, speech and language therapy, counseling of parents and families, advising teachers, and dealing with public agencies.

Audiometry

Audiologic evaluation technique varies as a function of the age and developmental level of the child, the reason for the evaluation, and the child's otologic condition or history. An audiogram provides the fundamental description of hearing sensitivity (Fig. 677.3). Hearing thresholds are assessed as a function of frequency using pure tones (single-frequency stimuli) at octave intervals from 250 to 8,000 Hz. When the child is old enough to accept their placement, earphones typically are used to assess each ear independently. Before this stage, testing may be performed in a sound-treated environment with stimuli delivered via speakers; this approach permits description only of the better-hearing ear.

Air-conducted signals are presented through earphones (or loudspeakers) and are used to provide information about the sensitivity of the entire auditory system. These same test sounds can be delivered to the ear through an oscillator that is placed on the head, usually on the mastoid. Such signals are considered bone-conducted because the bones of the skull transmit vibrations as sound energy directly to the inner ear, essentially bypassing the outer and middle ears. In a normal ear, and also in children with SNHL, the air- and bone-conduction thresholds are equivalent. In those with CHL, bone-conduction thresholds are more sensitive than air- conducted responses; this is called the **air-bone gap**, which indicates the amount of hearing loss attributable to dysfunction in the outer and/or middle ear. In mixed hearing loss, both the bone- and air-conduction thresholds are abnormal, and there is additionally an air-bone gap.

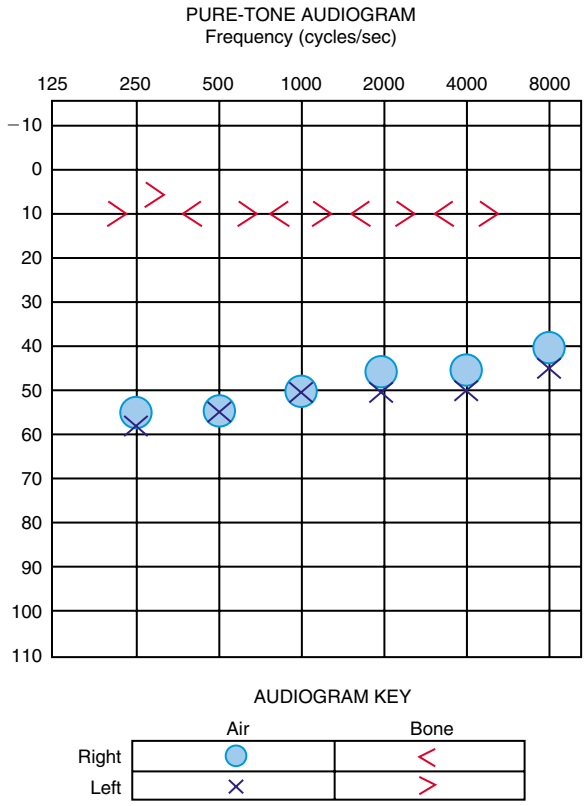

Fig. 677.3 Audiogram showing bilateral conductive hearing loss.

Speech-Recognition Threshold

Another measure useful for describing auditory function is the **speech-recognition threshold (SRT)**, which is the lowest intensity level at which a score of approximately 50% correct is obtained on a task of recognizing spondee words. Spondee words are 2-syllable words or phrases that have equal stress on each syllable, such as *baseball, hotdog,* and *pancake*. Listeners must be familiar with all the words for a valid test result to be obtained. The SRT should correspond to the average of pure-tone thresholds at 500, 1,000, and 2,000 Hz, the pure-tone average. The SRT is relevant as an indicator of a child's potential for development and use of speech and language; it also serves as a check of the validity of a test because children with nonorganic hearing loss (malingerers) might show a discrepancy between the pure-tone average and SRT. An SRT may be obtained in a child with expressive speech or language limitations using modified techniques, such as picture-pointing responses.

The basic battery of hearing tests concludes with an assessment of a child's ability to understand monosyllabic words when presented at a comfortable listening level. Performance on such word recognition tests assists in the differential diagnosis of hearing impairment and provides a measure of how well a child performs when speech is presented at loudness levels similar to those encountered in conversation. For speech recognition as well, a picture-pointing response may be obtained with standardized tests.

Play Audiometry

Hearing testing technique is age dependent. For children at or above the developmental level of a 5- to 6-year-old, conventional test methods can be used. For children 30 months to 5 years of age, play audiometry can be used. Responses in play audiometry usually are conditioned motor activities associated with a game, such as dropping blocks in a bucket, placing rings on a peg, or completing a puzzle. The technique can be used to obtain a reliable audiogram for a preschool child.

Visual Reinforcement Audiometry

For children between the ages of about 6 and 30 months, **visual reinforcement audiometry (VRA)** is commonly used. In this technique, the child is conditioned to turn the head in response to a tonal signal from a speaker in the same location as an animated (mechanical) toy or video reinforcer. If infants are properly conditioned, by presenting sounds associated with the reinforcer, VRA can provide reliable estimates of hearing sensitivity for tonal signals and speech sounds. In most applications of VRA, sounds are presented by loudspeakers in a sound field, so *ear-specific* information is not obtained. Assessment of an infant often is designed to rule out hearing loss that would be sufficient to affect the development of speech and language. Normal sound-field response levels of infants indicate sufficient hearing for this purpose despite the possibility of different HLs in the two ears. When ear-specific information is needed in this age-group, the ABR is conducted under sleep-deprived or sedated conditions.

Behavioral Observation Audiometry

Used as a screening device for infants <5 months of age, **behavioral observation audiometry** is limited to unconditioned, reflexive

responses to complex (not frequency-specific) test sounds such as warble tones, narrow-band noise, speech, or music presented using calibrated signals from a loudspeaker. Response levels can vary widely within and among infants and usually do not provide a reliable estimate of hearing sensitivity. The types of responses observed during this testing may include alterations in sucking behavior, initiation or cessation in crying, pupillary dilatation, and alterations in respiration.

Assessment of a child with suspected hearing loss is not complete until pure-tone hearing thresholds and SRTs (a reliable audiogram) have been obtained in each ear. Behavioral observation audiometry and VRA in sound-field testing give estimates of hearing responsivity in the *better-hearing ear*. When significant hearing loss is suspected in infants, electrophysiologic assessments must be conducted to permit early intervention.

Acoustic Immittance Testing

Acoustic immittance testing is a standard part of the clinical audiologic test battery and includes tympanometry, acoustic reflex threshold measurement, and acoustic reflex decay testing. It is a useful objective assessment technique that provides information about the status of the TM, middle ear, and acoustic reflex arc. Tympanometry can be performed in a physician's office and is helpful in the diagnosis and management of OM with effusion, a common cause of mild to moderate hearing loss in young children.

Tympanometry

Tympanometry provides a graph (tympanogram) of the middle ear's ability to transmit sound energy (admittance or compliance) or impede sound energy (impedance) as a function of air pressure in the external ear canal. Because most immittance test instruments measure acoustic admittance, the term *admittance* is used here. The principles apply to whatever units of measurement are used.

A probe is inserted into the entrance of the external ear canal so that an airtight seal is obtained. A manometer in the probe varies air pressure, while a sound generator presents a tone, and a microphone measures the sound pressure level reflected back. The sound pressure measured in the ear canal relative to the known intensity of the probe signal is used to estimate the acoustic admittance of the ear canal and middle-ear system. Admittance can be expressed in a unit called a millimho (mmho) or as a volume of air (mL) with equivalent acoustic admittance. Additionally, an estimate can be made of the volume of air enclosed between the probe tip and TM. The acoustic admittance of this volume of air is deducted from the overall admittance measure to obtain a measure of the admittance of the middle-ear system alone. Estimating ear canal volume also has a diagnostic benefit, because an abnormally large value is consistent with the presence of an opening in the TM (perforation, pressure equalization tube, or surgical defect).

Once the admittance of the air mass in the external auditory canal has been eliminated, it is assumed that the remaining admittance measure accurately reflects the admittance of the entire middle-ear system. Its value is controlled largely by the dynamics of the TM. Abnormalities of the TM can dictate the shape of tympanograms, thus obscuring abnormalities medial to the TM. In addition, the frequency of the probe tone, the speed and direction of the air pressure change, and the air pressure at which the tympanogram is initiated can all influence the outcome. The effect of the probe tone frequency is well documented, and in young children (<4-6 months) with small ear canals, use of a high-frequency probe tone, either 678 or 1,000 Hz, is recommended.

When air pressure in the ear canal is equal to that in the middle ear, the middle-ear system is functioning optimally. That is, the pressure equalization function of the eustachian tube permits the middle ear to rest at atmospheric pressure, equivalent to the condition in the ear canal. Therefore the ear canal pressure at which there is the greatest flow of energy (admittance) should be a reasonable estimate of the air pressure in the middle-ear space. This pressure is determined by finding the maximum or **peak admittance** on the tympanogram and obtaining its value on the *x* axis. The value on the *y* axis at the tympanogram peak is an estimate of peak admittance based on admittance tympanometry (Table 677.8). This peak measure sometimes is referred to as **static acoustic admittance**, even though it is estimated from a dynamic measure. Normative values for peak admittance as a function of air pressure are well established.

Table 677.8 Norms for Peak (Static) Admittance Using a 226-Hz Probe Tone for Children and Adults

AGE GROUP	ADMITTANCE (mL)	SPEED OF AIR PRESSURE SWEEP	
		≤50 daPa/sec*	200 daPa/sec†
Children (3-5 yr)	Lower limit	0.30	0.36
	Median	0.55	0.61
	Upper limit	0.90	1.06
Adults	Lower limit	0.56	0.27
	Median	0.85	0.72
	Upper limit	1.36	1.38

*Ear canal volume measurement based on admittance at lowest tail of tympanogram.
†Ear canal measurement based on admittance at lowest tail of tympanogram for children and at +200 daPa for adults.
daPa, decaPascals.
Adapted from Margolis RH, Shanks JE. Tympanometry: basic principles of clinical application. In: Rintelman WS, ed. *Hearing Assessment*, 2nd ed. Austin: PRODED; 1991: pp. 179–245.

Tympanometry in Otitis Media with Effusion

Children who have OM with effusion often have reduced peak admittance or high negative tympanometric peak pressures (see Fig. 680.5C). However, in the diagnosis of effusion, the tympanometric measure with the greatest sensitivity and specificity is the shape of the tympanogram rather than its peak pressure or admittance. The tympanogram is classified based on shape and peak admittance location. The greater the stiffening of the TM and ME, the lower the peak. As negative pressure within the middle ear increases, the peak becomes more negatively displaced. The more rounded the peak (or, in an absent peak, a flat tympanogram), the higher is the probability that an effusion is present (see Fig. 680.5B). The stage of OM may affect the tympanometric findings. An immobile TM/ME system based on significant effusion, as reflected in flat tympanogram, may evolve into findings of negative ME pressure and later positive pressure as the OM resolves, returning to a normal tympanogram.

Acoustic Reflex Threshold Test

The acoustic reflex threshold test also is part of the immittance test battery. With a properly functioning middle-ear system, admittance at the TM decreases due to the stiffening action of the middle ear muscles (stapedius and, to a lesser extent, tensor tympani). In healthy ears, the stapedial reflex occurs after exposure to loud sounds as a protective mechanism. Admittance instruments are designed to present reflex-activating signals (pure tones of various frequencies or noise), either to the same ear or the contralateral ear, while measuring the concomitant changes in admittance. Very small admittance changes that are time locked to presentations of the signal are considered to be a result of middle-ear muscle reflexes. Admittance changes may be absent when the hearing loss is sufficient to prevent the signal from reaching the loudness level necessary to elicit the reflex or when a middle-ear condition affects HLs or introduces sufficient stiffening to obscure reading the reflex activity. The acoustic reflex test also is used in the assessment of SNHL and the integrity of the neurologic components of the reflex arc, including crossed and uncrossed activity of cranial nerves VII and VIII.

Auditory Brainstem Response

The auditory brainstem response (ABR) test is used to screen newborn hearing, confirm hearing loss in young children, obtain

ear-specific information in young children, and test children who cannot, for whatever reason, cooperate with behavioral test methods. It also is important in the diagnosis of auditory dysfunction (i.e., estimation of hearing thresholds) and of disorders of the auditory nervous system. The ABR test is a far-field recording of minute electrical discharges from numerous neurons. The stimulus therefore must be able to cause synchronous discharge of the large numbers of neurons involved. Stimuli with very rapid onset, such as clicks or tone bursts, must be used. Unfortunately, the rapid onset required to create a measurable ABR also causes energy to be spread in the frequency domain, reducing the frequency specificity of the response.

The ABR result is not affected by sedation or general anesthesia. Infants and children from about 4 months to 4 years of age routinely are sedated to minimize electrical interference caused by muscle activity during testing. The ABR also can be performed in the operating room when a child is anesthetized for another procedure. Children younger than 4 months of age might sleep for a long enough period after feeding to allow an ABR to be done.

The ABR is recorded as 5-7 waves. Waves I, III, and V can be obtained consistently in all age-groups; waves II and IV appear less consistently. The latency of each wave (time of occurrence of the wave peak after stimulus onset) increases and the amplitude decreases with reductions in stimulus intensity; latency also decreases with increasing age, with the earliest waves reaching mature latency values earlier in life than the later waves. Age-specific normative data have been obtained in several studies.

The ABR test has two major uses in a pediatric setting. As an audiometric test, it provides information on the ability of the peripheral auditory system to transmit information to the auditory nerve and beyond. It also is used in the differential diagnosis or monitoring of central nervous system pathology. For hearing threshold estimation, the goal is to find the minimum stimulus intensity that yields an observable ABR, generally relying on wave V, the most robust aspect of morphology. Plotting latency versus intensity for various waves also aids in the differential diagnosis of hearing impairment. A major advantage of auditory assessment using the ABR test is that ear-specific threshold estimates can be obtained on infants or patients who are difficult to test. ABR thresholds using click stimuli correlate best with behavioral hearing thresholds in the higher frequencies (1,000-4,000 Hz); responsivity in the low frequencies requires different stimuli (tone bursts/pips or filtered clicks) or the use of masking, neither of which isolates the low-frequency region of the cochlea in all cases, and this can affect interpretation.

The ABR test does not assess "hearing." It reflects auditory neuronal electrical responses that can be correlated to behavioral hearing thresholds, but a normal ABR result only suggests that the auditory system, up to the level of the midbrain, is responsive to the stimulus used. Conversely, a failure to elicit an ABR indicates an impairment of the system's synchronous response but does not necessarily mean that there is no "hearing." The behavioral response to sound sometimes is normal when no ABR can be elicited, such as in neurologic demyelinating disease.

Hearing losses that are sudden, progressive, or unilateral are indications for ABR testing. Although it is believed that the different waves of the ABR reflect activity in increasingly rostral levels of the auditory system, the neural generators of the response have not been precisely determined. Each ABR wave beyond the earliest waves probably is the result of neural firing at many levels of the system, and each level of the system probably contributes to several ABR waves. High-intensity click stimuli are used for the neurologic application. The morphology of the response and wave, interwave latencies, and interaural latency differences are examined with respect to age-appropriate forms. Delayed or missing waves in the ABR result often have diagnostic significance.

The ABR and other electrical responses are extremely complex and difficult to interpret. A number of factors, including instrumentation design and settings, environment, degree and configuration of hearing loss, and patients' characteristics, can influence the quality of the recording. Therefore testing and interpretation of electrophysiologic activity as it possibly relates to hearing should be carried out by trained audiologists to avoid the risk that unreliable or erroneous conclusions will affect a patient's care.

Otoacoustic Emissions

During normal hearing, OAEs originate from the outer hair cells in the cochlea and are detected by sensitive amplifying processes. They travel from the cochlea through the middle ear to the external auditory canal, where they can be detected using miniature microphones. Transient evoked OAEs (TEOAEs) may be used to check the integrity of the cochlea. In the neonatal period, detection of OAEs can be accomplished during natural sleep, and TEOAEs can be used as screening tests in infants and children for hearing down to the 30 dB level of hearing loss. They are less time consuming and elaborate than ABRs and may be used when behavioral tests cannot be accomplished. TEOAEs are reduced or absent owing to various dysfunctions in the middle and inner ears. They are absent in patients with >30 dB of hearing loss and are not used to determine the hearing threshold; rather, they provide a screen for whether hearing is present at >30-40 dB. CHL, such as OM or congenitally abnormal middle-ear structures, reduces the transfer of TEOAEs and may be incorrectly interpreted as a cochlear hearing disorder. If a hearing loss is suspected based on the absence of OAEs, the ears should be examined for the evidence of pathology, tympanometry should be conducted, and then ABR testing should be used for confirmation and identification of the type, degree, and laterality of hearing loss.

TREATMENT

With widespread hearing screening within the United States, early diagnosis and treatment of children with hearing loss are common. Testing for hearing loss is possible even in very young children, and it should be done if parents suspect a problem. Any child with a known risk factor for hearing loss should be evaluated in the first 6 months of life.

Once a hearing loss is identified, a full developmental and speech and language evaluation is needed. Counseling and involvement of parents are required in all stages of the evaluation and treatment or rehabilitation. A CHL often can be corrected through treatment of a middle-ear effusion (i.e., ear tube placement) or surgical correction of the abnormal sound-conducting mechanism. Dependent on the level of hearing loss, children with SNHL should be evaluated for possible hearing aid use by a pediatric audiologist. Current guidelines indicate that within 1 month of diagnosis of SNHL, children should be fitted with **hearing aids**, and hearing aids may be fitted for children as young as 1 month of age. Compelling evidence from the hearing screening program in Colorado shows that identification and amplification before age 6 months make a very significant difference in the speech and language abilities of affected children compared with cases identified and amplified after the age of 6 months. In these children, repeat audiologic testing is needed to reliably identify the degree of hearing loss and to fine-tune the use of hearing aids. Hearing aids remain the rehabilitative device of choice, in the context of an individually designed treatment plan, for children with mild, moderate, or moderately severe CHL, mixed HL, or SNHL. For children with severe or profound SNHL, a trial with hearing aids is needed to determine if this approach is sufficient for the development of language; other options may need to be explored if there are indications that speech and language are delayed with a hearing aid in this HL group. Importantly, efficacy of hearing aids depends on their consistent use. There is great variability in how often children wear their hearing aids. Though there is no specific recommendation regarding the minimal number of hours per day that the hearing aids should be worn, parents should be encouraged to have their child use hearing aids full-time in order to facilitate speech and language development.

When it is clear that hearing aids are not providing the auditory stimulation needed to support language development, the parents require counseling to consider alternative treatments. A **cochlear implant** may be necessary to facilitate intelligible oral communication (i.e., oralism). This approach requires years of intensive speech and language training and is dependent on providing the best possible auditory stimulation. This option is very attractive to parents with hearing because it is the most familiar form of communication to them. Although there is a heavy emphasis in the medical world valuing the development of oral language (speech production), parents should also be provided with information about alternatives such as sign language, total communication, and cued speech (see Chapter 55). Each of these communication modalities has advantages and disadvantages. **Sign language** allows the child to develop a language system early and can support academic training. The consequence of this option is that the dominant hearing world does not interact easily with users of sign language, and the child may face significant challenges integrating into hearing society. Such possibilities as academic success and college/graduate school training are not excluded by the use of sign language, but a narrower set of venues may be available to accommodate the child's learning needs. Whereas this option may be acceptable to deaf parents already in the deaf community, many hearing parents are uncomfortable with this path for their child. This option also requires that the parents become fluent in sign language.

Total communication is an educational philosophy in which both sign and oral language as well as other forms of communication are encouraged. In theory, the two systems support and clarify information transfer and enhance academic progress. Depending on the particular school and/or teachers, one system may be emphasized over the other. **Cued speech** is an approach in which the development of oral language is supported by a system of hand gestures near the mouth and throat to disambiguate confusions that result from lip reading alone. This system can be highly successful in supporting spoken language and requires that parents become fluent in the use of the cues. Other factors should be considered in making the choice of communication modality. Significant comorbidities, such as visual impairment or other developmental delays, may limit the ability of a child to derive benefit from some choices. Support for the parents in making this decision may require counseling from an audiologist, social worker, deaf educator, and/or psychologist. Organizations of parents of deaf children, such as the A.G. Bell Association and the John Tracy Clinic, can provide a wealth of support and information to parents in this process.

Infants and young children with profound congenital or prelingual onset of deafness have benefited from **multichannel cochlear implants** (Fig. 677.4). Cochlear implants are systems that combine internal (surgically implanted) and externally worn components. These implants consist of 4 main components: the externals—which include a microphone, a minicomputer sound (speech) processor, and a transmitter—and the internal—an electrode array. These implants bypass injury to the organ of Corti and provide neural stimulation through the digitization of auditory stimuli into digital radiofrequency impulses. Specifically, sound is initially detected by the microphone and then is processed by the speech processor. The speech processor is programmed by an audiologist to implement the manufacturer's proprietary speech processing strategies that are highly sophisticated manipulations of the input signal. Signals from the speech processor are transmitted across the skin by an FM signal to the internal receiver, which converts these signals into electrical impulses. Finally, these electrical impulses are sent to the electrode array located in the cochlea, where electrical fields are created that act on the cochlear nerve. This contrasts with the transmission of sound in a healthy ear, which involves the transmission of sound vibrations to the hair cells of the cochlea, the release of ions and neurotransmitters in the cochlea, and the transmission of neural impulses to the cochlear nerve and then the brain.

Surgical implantation is done under general anesthesia and involves mastoidectomy and widening of the facial recess because the approach to the cochlea is through the facial recess. After fastening the internal stimulator package in the mastoid process, the cochlea must be opened to insert the electrode array, which is most commonly done through an opening made in the round window. Care is taken to avoid contamination of the cochlear fluids by bone dust or blood. After the cochlea is closed, generally with fascia, the wound is closed. An audiologist performs testing in the operating room to verify the functional integrity of the implanted device. These electrophysiologic responses from cranial nerve VIII are critical to determining a starting point for programming the external device after the wound has healed. A plain x-ray is often performed in the operating room as well to document placement of the array in the scala tympani.

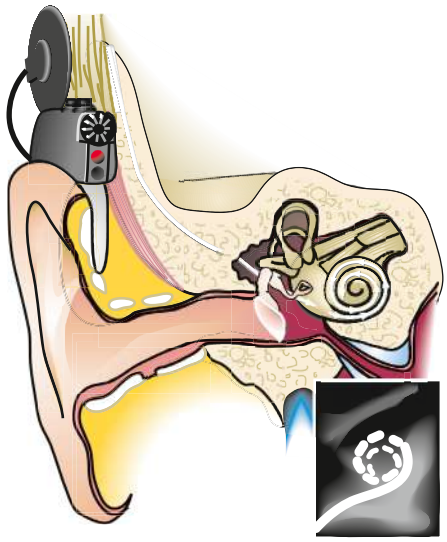

Fig. 677.4 All cochlear implants share key components, including a microphone, speech processor, and transmitter coil, shown in a behind-the-ear position in this diagram. The microphone and speech processor pick up environmental sounds and digitize them into coded signals. The signals are sent to the transmitter coil and relayed through the skin to the internal device embedded in the skull. The internal device converts the code to electronic signals, which are transmitted to the electrode array wrapping around the cochlea. The inset shows the radiographic appearance of the stimulating electrode array. *(From MED-EL Corporation, Innsbruck, Austria. From Smith RJH, Bale JF Jr., White KR. Sensorineural hearing loss in children. Lancet. 2005;365:879–890.)*

The healing process after surgery is approximately 3-4 weeks for a child. During this time, the child cannot hear. When the child is brought in for the first stimulation using the external equipment, programs are developed that provide the first access to sound. The methods to create the programs entail a combination of electrophysiologic measures and behavioral testing that is like the pediatric audiologic assessments described earlier. The initial programs are a starting point, followed by modifications and enhancements that are based on the parents' and audiologist's observations of changing auditory awareness and vocalization.

When parents elect to pursue cochlear implantation for their child, a long-term commitment is necessary to ongoing engagement with a team of rehabilitation specialists. Audiologic management entails consistent monitoring of the child's response to the implant and impact on emerging language skills. Speech and language therapy is necessary to stimulate language and to teach parents skills to support speech development. The child should be in a preschool setting in which speech, language, social, and academic precursor skills are fostered. For some parents, this engagement is very challenging, not only in terms of time required but also in terms of the emotional consequences of attempting to minimize the impact of hearing loss on their child's future; support for the parents is often needed in this process from the team.

Table 677.9 Recommended Pneumococcal Vaccination Schedule for Persons with Cochlear Implants

AGE AT FIRST PCV13 DOSE (mo)*	PCV12 PRIMARY SERIES	PCV13 ADDITIONAL DOSE	PPV23 DOSE
2-6	3 doses, 2 mo apart†	1 dose at 12-15 mo of age‡	Indicated at ≥24 mo of age§
7-11	2 doses, 2 mo apart†	1 dose at 12-15 mo of age‡	Indicated at ≥24 mo of age§
12-23	2 doses, 2 mo apart¶	Not indicated	Indicated at ≥24 mo of age§
24-59	2 doses, 2 mo apart¶	Not indicated	Indicated§
≥60	Not indicated	Not indicated**	Indicated

*A schedule with a reduced number of total 13-valent pneumococcal conjugate vaccine (PCV13) doses is indicated if children start late or are incompletely vaccinated. Children with a lapse in vaccination should be vaccinated according to the catch-up schedule (see Chapter 228).
†For children vaccinated at younger than age 1 yr, minimum interval between doses is 4 wk.
‡The additional dose should be administered 8 wk or more after the primary series has been completed.
§Children younger than age 5 yr should complete the PCV13 series first; 23-valent pneumococcal polysaccharide vaccine (PPV23) should be administered to children 24 mo of age or older 8 wk or more after the last dose of PCV13 (see Chapter 228) (Centers for Disease Control and Prevention Advisory Committee on Immunization Practices: Preventing pneumococcal disease among infants and young children: Recommendations of the Advisory Committee on Immunization Practices [ACIP]. *MMWR Recomm Rep*. 2000;49[RR-9]:1–35, and Licensure of a 13-valent pneumococcal conjugate vaccine [PCV13] and recommendations for use among children—Advisory Committee on Immunization Practices [ACIP]. *MMWR Morb Mortal Wkly Rep*. 2010;59[9]:258–261.)
¶Minimum interval between doses is 8 wk.
**PCV13 is not recommended generally for children age 5 yr or older.
PCV, Pneumococcal conjugate vaccine; PPV, pneumococcal polysaccharide vaccine.
From Centers for Disease Control and Prevention Advisory Committee on Immunization Practices. Pneumococcal vaccination for cochlear implant candidates and recipients: Updated recommendations of the Advisory Committee on Immunization Practices. *MMWR Morb Mortal Wkly Rep*. 2003;52(31):739–740.

A serious possible complication of cochlear implantation is pneumococcal meningitis. All children receiving a cochlear implant must be vaccinated with the pneumococcal polyvalent vaccine PCV13 (Table 677.9), and rates of pneumococcal meningitis have declined considerably since implementation of the vaccine.

The Food and Drug Administration (FDA) has approved cochlear implantation in patients over 9 months of age with severe to profound bilateral hearing loss not benefitting from hearing aids; off-label use of cochlear implants has demonstrated efficacy in younger children and those with residual hearing. Cochlear implantation before age 2 years improves hearing and speech, enabling more than 90% of children to be in mainstream education. Most develop age-appropriate auditory perception and oral language skills. There is increasing evidence to support expansion of the candidacy for cochlear implantation in children to be based on outcomes of advanced testing using speech stimuli, especially in noise. To date, implantation of children with devices that combine acoustic input (like a hearing aid) with electric stimulation from a cochlear implant has not been approved by the FDA. These devices, called *electroacoustic cochlear implants,* or *hybrids,* may offer hope for children using hearing aids but struggling with noise in the classroom or social contexts.

Valganciclovir has been effective in treating hearing deficits in children with congenital CMV and isolated SNHL (see Chapter 302).

GENETIC COUNSELING

Families of children with the diagnosis of SNHL or a syndrome associated with SNHL and/or CHL should be referred for genetic counseling. This will give the parents an idea of the likelihood of similar diagnoses in future pregnancies, and the geneticist can assist in the evaluation and testing of the patient to establish a diagnosis.

677.1 Idiopathic Sudden Sensorineural Hearing Loss

Joseph Haddad Jr.

Sudden SNHL in a previously healthy child is uncommon but may be from OM or other cochlear pathologies such as autoimmunity (Cogan syndrome, others). Usually, these causes are obvious from the history and physical examination. Sudden loss of hearing in the absence of obvious causes may also be the result of a vascular event affecting the cochlear apparatus or nerve, such as embolism or thrombosis (secondary to prothrombotic conditions) or hemorrhage. Additional causes include perilymph fistula, medications, trauma, and the first episode of Meniere syndrome. In adults, sudden SNHL is often idiopathic and unilateral; it may be associated with a sensation of ear fullness, tinnitus, and vertigo. Identifiable causes of sudden SNHL include infections (Epstein-Barr virus, varicella-zoster virus, herpes simplex virus) (see Table 677.2), vascular injury to the cochlea, enlarged vestibular aqueduct, endolymphatic hydrops, and autoimmune inflammatory diseases. In most (~75%) patients with sudden SNHL, no etiology is discovered, and it is termed **idiopathic sudden SNHL**. This entity is defined by a rapid (≤72 hours) onset that may be unilateral (bilateral in 25%) and associated with a hearing loss of ≥30 dB. Patients should be evaluated immediately to exclude other etiologies and obtain a focused MRI of the auditory vestibular region.

Management of **idiopathic sudden SNHL** has included oral prednisone, intratympanic (also called transtympanic) dexamethasone perfusion, or a combination of both; the latter combination may be the most useful. Recovery of hearing is more likely in patients with early treatment and with mild or moderate hearing loss and those with unilateral involvement.

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Chapter 678

Congenital Malformations of the Ear

Joseph Haddad Jr.

The external and middle ears, derived from the first and second branchial arches and grooves, grow throughout puberty, but the inner ear, which develops from the otocyst, reaches adult size and shape by midfetal development. The ossicles are derived from the first and second arches (malleus and incus), and the stapes arises from the second arch and the otic capsule. The malleus and incus achieve adult size and shape by the 15th week of gestation, and the stapes achieves adult size and shape by the 18th week of gestation. Although the pinna, ear canal, and tympanic membrane (TM) continue to grow after birth, congenital abnormalities of these structures develop during the first half of gestation. Malformed external and middle ears may be associated with serious renal anomalies, mandibulofacial dysostosis, hemifacial microsomia, and other craniofacial malformations (Table 678.1). Facial nerve abnormalities may be associated with any of the congenital abnormalities of the ear and temporal bone. Malformations of the external and middle ears also may be associated with abnormalities of the inner ear and both conductive hearing loss (CHL) and sensorineural hearing loss (SNHL).

Congenital ear problems may be either minor and mainly cosmetic or major, affecting both appearance and function. Any child born with an abnormality of the pinna, external auditory canal, or TM should have a complete audiologic evaluation in the neonatal period. Imaging studies are necessary for evaluation and treatment; in the patient with other craniofacial abnormalities, a team approach with other specialists can assist in guiding therapy.

PINNA MALFORMATIONS

Severe malformations of the external ear are rare, but minor deformities are common. Isolated abnormalities of the external ear occur in approximately 1% of children (Fig. 678.1). A **pitlike** depression just in front of the helix and above the tragus may represent a cyst or an epidermis-lined fistulous tract (Fig. 678.2). These are common, with an incidence of approximately 8 in 1,000 children, and may be unilateral or bilateral and familial. The pits require surgical removal only if there is recurrent infection. Accessory **skin tags**, with an incidence of 1-2/1,000 live births, can be removed for cosmetic reasons by simple ligation if they are attached by a narrow pedicle. If the pedicle is broad based or contains cartilage, the defect should be corrected surgically. An unusually prominent or "lop" ear results from lack of bending of the cartilage that creates the antihelix. It may be improved cosmetically in the neonatal period by applying a firm framework (sometimes soldering wire is used) attached by Steri-Strips to the pinna and worn continuously for weeks to months. Otoplasty for cosmetic correction can be considered in children older than 5 years of age, when the pinna has reached approximately 80% of its adult size.

The term **microtia** may indicate subtle abnormalities of the size, shape, and location of the pinna and ear canal or major abnormalities with only small nubbins of skin and cartilage and the absence of the ear canal opening; **anotia** indicates complete absence of the pinna and ear canal (Fig. 678.3). Microtia can have a genetic or environmental predisposition. Several hereditary forms of microtia have been identified that exhibit either autosomal dominant or recessive mendelian inheritance. In addition, some forms due to chromosomal aberrations have been reported. Most of the responsible genes that have been identified are homeobox genes, which are involved in the development of pharyngeal arches. Microtic ears often are more anterior and inferior in placement than normal auricles, and the location and function of the facial nerve may be abnormal. Surgery to correct microtia is considered for both cosmetic and functional reasons; children who have some pinna can wear regular glasses, a hearing aid, and earrings and feel more normal in appearance. If the microtia is severe, some patients may opt for creation and attachment of a prosthetic ear, which cosmetically closely

Table 678.1 Diseases with Anomalies of the External and Middle Ears Listed by Pathologic Defect and Traditional Name

PATHOLOGIC NAME	EPONYM
4p-syndrome	Wolf-Hirschhorn syndrome
Acrocephalosyndactyly type I	Apert syndrome
Acrocephalosyndactyly type III	Saethre-Chotzen syndrome
Acrocephalosyndactyly type V	Pfeiffer syndrome
Anus imperforate with hand, foot, and ear anomalies	Townes-Brocks syndrome
Arteriohepatic dysplasia	Alagille syndrome
Branchio-oto-renal syndrome	Melnick Fraser syndrome
Brevicollis	Klippel-Feil syndrome
Cervico-oculoacoustic syndrome	Wildervanck syndrome
Cleft palate, microcephaly, large ears, and short stature	Say syndrome
Cleft palate, micrognathia, and glossoptosis	Pierre Robin sequence
Congenital contractural arachnodactyly	Beals syndrome
Congenital facial diplegia	Möbius syndrome
Constitutional aplastic pancytopenia with multiple anomalies	Fanconi syndrome
Craniofacial dysostosis	Crouzon disease
Craniometaphyseal dysplasia	Pyle disease
Dyschondrosteosis	Léri-Weill syndrome
Exomphalos-macroglossia-gigantism syndrome	Beckwith-Wiedemann syndrome
Faciodigitogenital syndrome	Aarskog syndrome
Gargoylism	Hurler syndrome
Gonadal aplasia	Turner syndrome
Hemifacial microsomia (oculoauriculovertebral dysplasia)	Goldenhar syndrome
Lacrimoauriculodentodigital syndrome	Levy-Hollister syndrome
Mandibulofacial dysostosis	Treacher Collins syndrome
Orofaciodigital syndrome type II	Mohr syndrome
Osteodysplasty	Melnick-Needles syndrome
Osteopetrosis	Albers-Schönberg disease
Renal agenesis, bilateral	Potter syndrome
Third and fourth pharyngeal pouch syndrome	DiGeorge syndrome
Trisomy 13-15 syndrome	Patau syndrome
Trisomy 18 syndrome	Edwards syndrome
Trisomy 21 syndrome	Down syndrome

From Lesperance MM, Flint PW, eds. *Cummings Pediatric Otolaryngology.* Philadelphia: Elsevier; 2015, Table 15.1, p. 197.

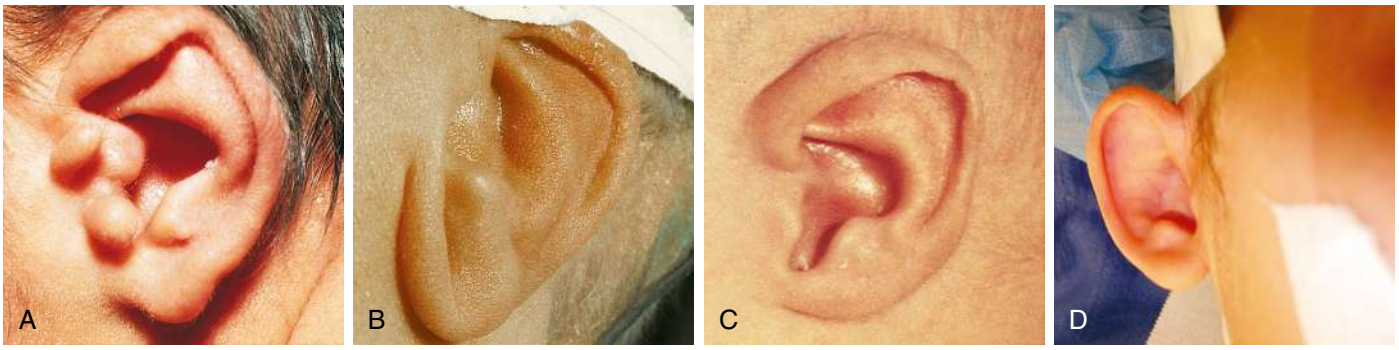

Fig. 678.1 Minor congenital auricular deformities. **A,** In this infant, the superior portion of the helix is folded over, obscuring the triangular fossa; the antihelix is sharply angulated; and there are three preauricular skin tags. **B,** This neonate with orofaciodigital and Turner syndromes has a simple helix and a redundant folded lobule. The ear is low set and posteriorly rotated, and the antitragus is anteriorly displaced. **C,** This infant with Rubinstein-Taybi syndrome has an exaggerated elongated intertragal notch. **D,** Prominent ear in an otherwise normal child. The auricular cartilage is abnormally contoured, making the ear protrude forward. *(**C** courtesy Dr. Michael E. Sherlock, Lutherville, Maryland; from Zitelli BJ, McIntire SC, Nowalk AJ, eds. Zitelli and Davis' Atlas of Pediatric Physical Diagnosis, 7th ed. Philadelphia: Elsevier; 2018: Fig. 24.17, p. 875.)*

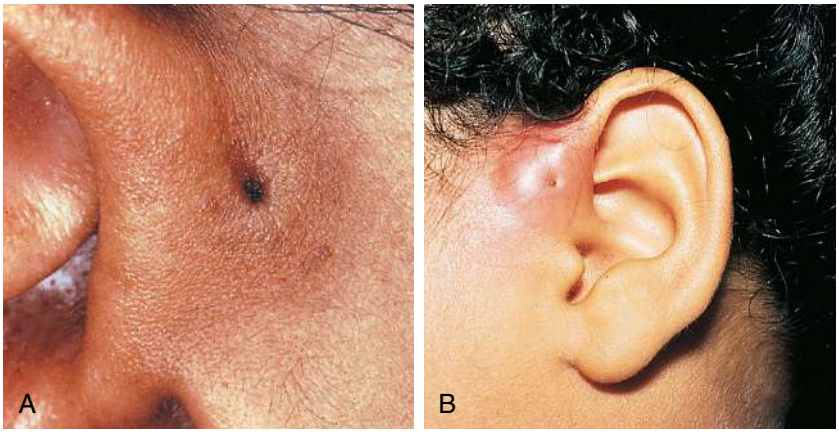

Fig. 678.2 Preauricular sinuses. **A,** These congenital remnants are located anterior to the pinna and have an overlying surface dimple. **B,** In this child, the sinus has become infected, forming an abscess. *(**A** courtesy Michael Hawke, MD; from Zitelli BJ, McIntire SC, Nowalk AJ, eds. Zitelli and Davis' Atlas of Pediatric Physical Diagnosis, 7th ed. Philadelphia: Elsevier; 2018: Fig. 24.18, p. 876.)*

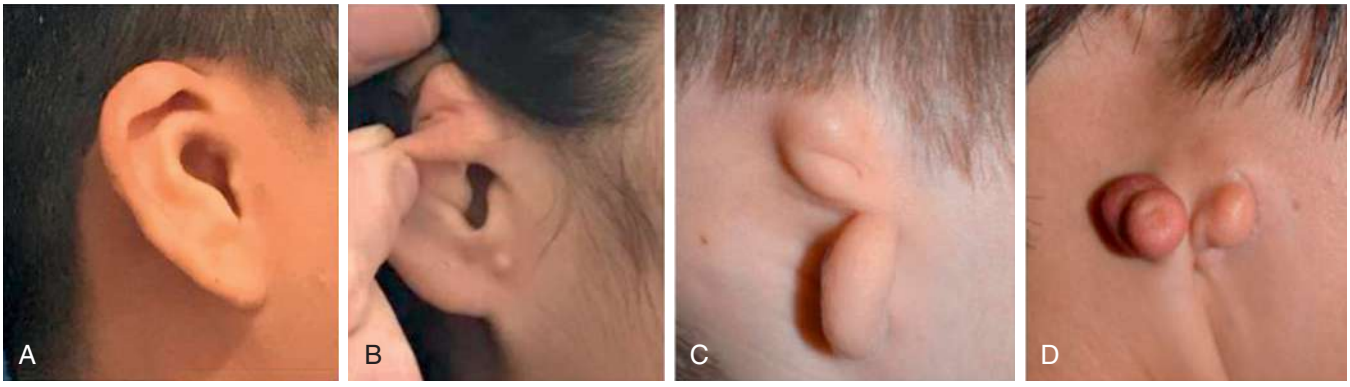

Fig. 678.3 A, Type I microtia with constricted ear and minimal tissue deficiency. **B,** Type II microtia, conchal type: absence of superior portions of the ear and preservation of inferior conchal anatomy. **C,** Type III microtia, lobular type: markedly deformed and no identifiable concha and preservation of the lobule. **D,** Type IV, anotia. *(From Lesperance MM, ed. Cummings Pediatric Otolaryngology, 2nd ed, Philadelphia: Elsevier; 2022: Fig. 18.2, p. 250.)*

resembles a real ear. Surgery to correct severe microtia may involve a multistage procedure, including carving and transplantation of autogenous cartilage rib grafts and local soft tissue flaps. Cosmetic reconstruction of the auricle usually is performed between 5 and 7 years of age and is performed before canal atresia repair in children deemed appropriate for this surgery.

CONGENITAL STENOSIS OR ATRESIA OF THE EXTERNAL AUDITORY CANAL

Stenosis or atresia of the ear canal often occurs in association with malformation of the auricle and middle ear. Malformations can occur in isolation or as part of a genetic syndrome. For example, the ear canal is narrow in trisomy 21, and external canal stenosis or atresia is common in branchiooculofacial syndrome, leading to CHL. Audiometric evaluation of these children should be undertaken as early in life as possible. Most children with significant CHL secondary to bilateral atresia wear bone conduction hearing aids for the first several years of life. Diagnosis, evaluation, and surgical planning often are aided by CT, and sometimes MRI, of the temporal bone. Mild cases of ear canal stenosis do not require surgical enlargement unless the patient develops chronic external otitis or severe cerumen impaction that affects hearing.

Reconstructive ear canal and middle-ear surgery for atresia usually is considered for children older than 5 years of age who have bilateral deformities resulting in significant CHL. The aim of reconstructive surgery is to improve hearing to a point where the child may not need a hearing aid or to provide an ear canal and pinna so that the child can derive improved benefit from an air-conduction hearing aid. Hearing results for atresiaplasty range from fair to excellent. CT evidence of an adequate middle-ear cleft, ossicles, and mastoid is required to perform the surgery; the position of the facial nerve, which often is in an abnormal location in these children, also must be considered (Fig. 678.4). The use of bone-anchored hearing aids is a safe, reliable, and low-risk alternative to atresiaplasty, and hearing results are generally excellent. Bone-anchored hearing aids may also be useful for rehabilitation of nonoptimal atresiaplasty hearing results. These devices are approved by the US Food and Drug Administration for surgical placement in children age 5 years and older; before age 5 years, they can be worn with a soft band around the head. Disadvantages include the fact that cosmesis is not very good (a bone-anchored hearing aid has a visible titanium abutment and snap-on hearing aid) and frequent wound care is required. Middle ear implants are effective alternatives for those who cannot tolerate foreign bodies in the ear for medical reasons or rely on good perception of high-frequency sounds.

CONGENITAL MIDDLE-EAR MALFORMATIONS

Children may have congenital abnormalities of the middle ear as an isolated defect or in association with other abnormalities of the temporal bone, especially the ear canal and pinna, or as part of a syndrome. Affected children usually have CHL but may have mixed CHL and SNHL. Most malformations involve the ossicles, with the incus most commonly affected. Other less common abnormalities of the middle ear include persistent stapedial artery, high-riding jugular bulb, and abnormalities of the shape and volume of the aerated portion of the middle ear and mastoid; all present problems for a surgeon. Depending on the type of abnormality and the presence of other anomalies, surgery may be considered to improve hearing.

CONGENITAL INNER EAR MALFORMATIONS

Congenital inner ear malformations are classified as a result of improvements in imaging modalities (Table 678.2). As many as 20% of children with SNHL may have anatomic abnormalities identified on CT or MRI. Congenital malformations of the inner ear usually are associated with SNHL of various degrees, from mild to profound. These malformations are most commonly found in infants and may occur as isolated anomalies or in association with other syndromes, genetic abnormalities, or structural abnormalities of the head and neck (Table 678.3). High-resolution temporal bone CT can identify enlarged vestibular aqueducts and cochlear nerve canal stenosis in association with SNHL. Although no therapy exists for this condition, it may be associated with progressive SNHL in some children; therefore diagnosis may have some prognostic value.

Congenital perilymphatic fistula of the oval or round window membrane may present as a rapid-onset, fluctuating, or progressive SNHL with or without vertigo and often is associated with congenital inner ear abnormalities. Middle-ear exploration may be required to confirm this

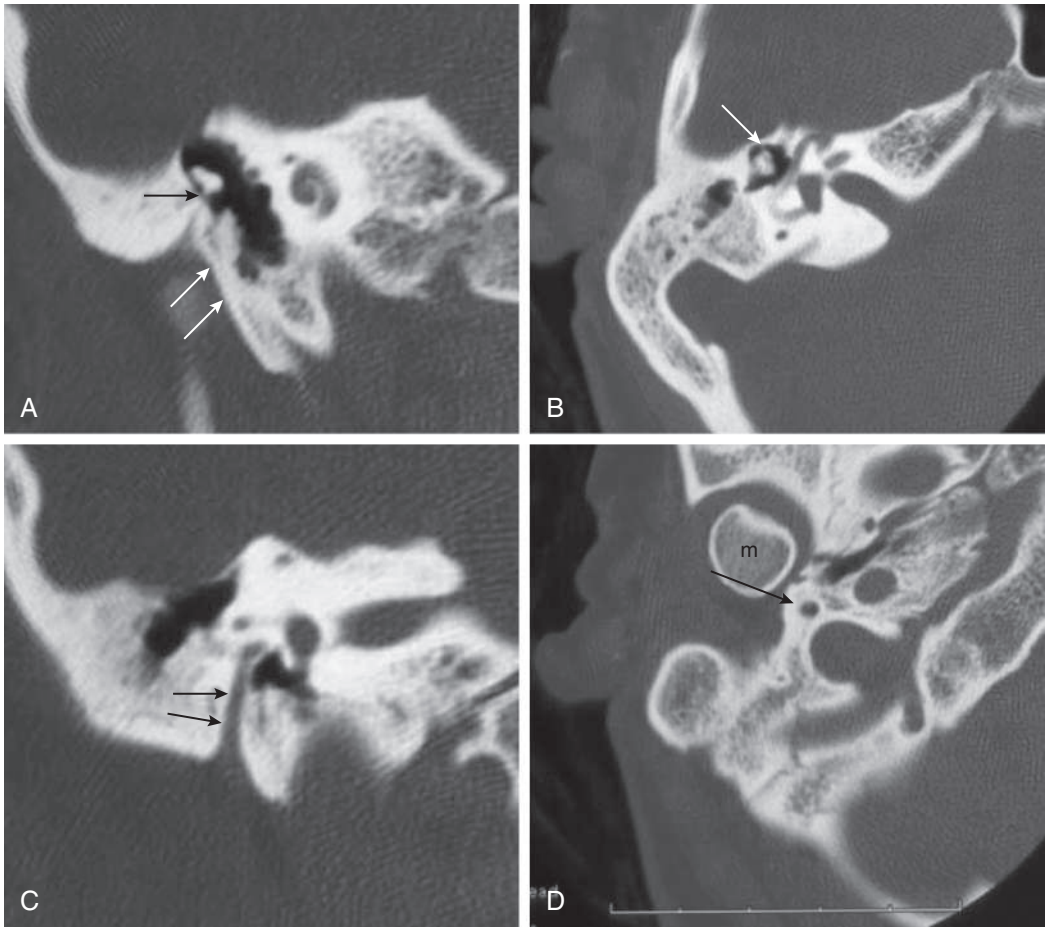

Fig. 678.4 External auditory canal atresia on CT scans. **A,** Coronal scan of right ear shows absent external auditory canal with thick bony atresia plate *(white arrows).* Malleus neck is rotated and fused to superior portion of atresia plate *(black arrow).* **B,** Axial scan through attic shows fused ossicular mass *(arrow).* **C,** Coronal scan more posterior to **(A)** shows mastoid segment of facial nerve canal positioned more anteriorly than normal *(arrows).* **D,** Axial scan more inferior to **(B)** shows anterior-posterior mastoid segment of the facial nerve en face *(arrow).* Note abnormally close relationship to mandibular condyle. *(From Faerber EN, Booth TN, Swartz JD. Temporal bone and ear. In Slovis TL, ed. Caffey's Pediatric Diagnostic Imaging, 11th ed. Philadelphia: Mosby; 2008: Fig. 44.7, p. 584.)*

diagnosis, because no reliable nonoperative diagnostic test exists. It may be necessary to repair a perilymphatic fistula to prevent possible spread of infection from the middle ear to the labyrinth or meninges, to stabilize hearing loss, and to improve vertigo when present.

Table 678.2 Classification of Congenital Inner Ear Malformations

MALFORMATIONS LIMITED TO THE MEMBRANOUS LABYRINTH
Complete membranous labyrinthine dysplasia
Limited membranous labyrinthine dysplasia
Cochleosaccular dysplasia (Scheibe)
Cochlear basal turn dysplasia
MALFORMATIONS OF THE OSSEOUS AND MEMBRANOUS LABYRINTH
Complete labyrinthine aplasia (Michel)
Cochlear anomalies
Cochlear aplasia
Cochlear hypoplasia
Incomplete partition (Mondini)
Common cavity
Labyrinthine anomalies
Semicircular canal dysplasia
Semicircular canal aplasia
Aqueductal anomalies
Enlargement of the vestibular aqueduct
Enlargement of the cochlear aqueduct
Internal auditory canal anomalies
Narrow internal auditory canal
Wide internal auditory canal
Eighth cranial nerve anomalies
Hypoplasia
Aplasia

From Lesperance MM, ed. *Cummings Pediatric Otolaryngology*, 2nd ed, Philadelphia: Elsevier; 2022: Box 13.1, p. 178.

CONGENITAL CHOLESTEATOMA

A congenital cholesteatoma (approximately 2–5% of all cholesteatomas) is a nonneoplastic, destructive, cystic lesion that usually appears as a white, round, cystlike structure medial to an intact TM. Cysts are seen most commonly in boys and in the anterior-superior portion of the middle ear, although they can present in other locations and within the TM or in the skin of the ear canal. They can be classified as "open," meaning in direct continuity with mucosa of the middle ear, or "closed." Affected children often have no prior history of otitis media. One theory for the pathogenesis is that the cyst derives from a congenital rest of epithelial tissue that persists beyond 33 weeks of gestation, when it ordinarily would disappear. Other theories include squamous metaplasia of the middle ear, entrance of squamous epithelium through a nonintact eardrum into the middle ear, ectodermal implants between the first and second branchial arch remnants, and residual amniotic fluid squamous debris. Congenital or acquired cholesteatoma should be suspected when deep retraction pockets, keratin debris, chronic drainage, aural granulation tissue, or a mass behind or involving the TM is present. Congenital cholesteatoma is often asymptomatic, whereas acquired cholesteatoma commonly presents with otorrhea. Besides acting as a benign tumor causing local bone destruction, the keratinaceous debris of a cholesteatoma serves as a culture medium, leading to chronic otitis media. Complications include ossicular erosion with hearing loss, bone erosion into the inner ear with dizziness, or exposure of the dura, with consequent meningitis or a brain abscess. Evaluation includes a CT scan (Fig. 678.5) to detect bone erosion and audiometry to assess air and bone conduction and speech reception and discrimination. Treatment includes cholesteatoma removal, repair of damaged small middle ear bones, and mastoidectomy in 50% of congenital and >90% of acquired cholesteatoma cases. A second-look procedure 6-9 months after primary surgery is usually recommended to detect and remove small amounts of residual disease before more extensive recurrence or development of complications. Higher initial stage of disease, erosion of ossicles, cholesteatoma abutting or enveloping the incus or stapes,

Table 678.3 Inner Ear Malformations in Syndromic Hearing Loss

SYNDROME	GENETIC ABNORMALITY	RADIOGRAPHIC ANOMALIES	ASSOCIATED ANOMALIES
Apert	*FGFR2*	Vestibular dysplasia, high-riding jugular bulb	Stapes footplate fixation
Branchio-oto-renal	*EYA1*	Hypoplastic cochlea, enlarged VA	Malleus or incus dysplasia, aberrant facial nerve
CHARGE	*CHD7*	Hypoplastic or absent SCC	Coloboma, heart defects, choanal atresia, growth retardation, genital/urinary abnormalities
Down	Trisomy 21	Hypoplastic cochlea, SCC dysplasia, cochlear nerve canal hypoplasia	Narrow external auditory canal, eustachian tube dysfunction
Edwards	Trisomy 18	Hypoplastic cochlea, absent SCC	Microtia, low-set ears
Pendred	*SLC26A4*	Hypoplastic cochlea, enlarged VA, modiolar deficiency	Goiter, hypothyroidism
Waardenburg type I	*PAX3*	Hypoplastic cochlea and/or SCC, enlarged VA	Telecanthus, white forelock, heterochromia iridum
Waardenburg type II	*MITF, SNAI2, SOX10*	Enlarged vestibule, hypoplastic SCC	White forelock, heterochromia iridum
Waardenburg type III	*PAX3*	Hypoplastic cochlea and/or SCC, enlarged VA	Upper limb abnormalities, white forelock, heterochromia iridum
Waardenburg type IV	*EDNRB, EDN3, SOX10*	Enlarged vestibule, hypoplastic SCC	Hirschsprung disease, white forelock, heterochromia iridum

SCC, Semicircular canal; VA, vestibular aqueduct.
Modified from Lesperance MM, ed. *Cummings Pediatric Otolaryngology,* 2nd ed. Philadelphia: Elsevier; 2022: Table 13.2, p. 189.

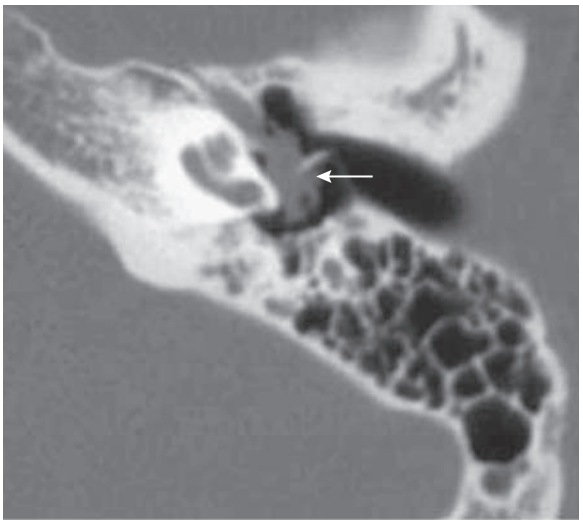

Fig. 678.5 Congenital cholesteatoma. Axial CT of left ear shows soft tissue mass *(arrow)* in the middle ear. This mass was noted otoscopically behind an intact membrane. *(From Faerber EN, Booth TN, Swartz JD. Temporal bone and ear. In Slovis TL, ed. Caffey's Pediatric Diagnostic Imaging, 11th ed. Philadelphia: Mosby; 2008: Fig. 44.31, p. 598.)*

and need for removal of the ossicles are associated with increased likelihood of residual cholesteatoma, which occurs in ~10% of congenital and ~25% of acquired cases. More extensive disease at initial surgery is associated with poorer hearing outcomes. Children with significant inflammation or extensive scarring may require a 2-stage procedure with initial removal of the cholesteatoma and subsequent repair of damaged middle ear structures.

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Chapter 679

External Otitis (Otitis Externa)

Joseph Haddad Jr.

In an infant, the outer two thirds of the ear canal is cartilaginous and the inner one third is bony. In an older child and adult, the outer one third is cartilaginous and the inner two thirds is bony. The epithelium is thinner in the bony portion than in the cartilaginous portion, there is no subcutaneous tissue, and epithelium is tightly applied to the underlying periosteum; hair follicles, sebaceous glands, and apocrine glands are scarce or absent. The skin in the cartilaginous area has well-developed dermis and subcutaneous tissue and contains hair follicles, sebaceous glands, and apocrine glands. The highly viscid secretions of the sebaceous glands and the watery, pigmented secretions of the apocrine glands in the outer portion of the canal combine with exfoliated surface cells of the skin to form **cerumen**, a protective, waxy, water-repellent coating.

The normal flora of the external canal consists mainly of aerobic bacteria and includes coagulase-negative staphylococci (see Chapter 227.3), *Corynebacterium* (diphtheroids; see Chapter 233), *Micrococcus,* and occasionally *Staphylococcus aureus* (see Chapter 227.1), viridans streptococci (see Chapter 231), and *Pseudomonas aeruginosa* (see Chapter 251.1). **Excessive wetness** (swimming, bathing, increased environmental humidity), **dryness** (dry canal skin and lack of cerumen), the presence of other skin pathologic conditions (previous infection, eczema, or other forms of dermatitis), and trauma (due to digital or foreign body, use of cotton-tipped swabs) make the skin of the canal vulnerable to infection by the normal flora or exogenous bacteria and predispose to colonization with gram-negative bacteria.

ETIOLOGY

External otitis (**swimmer's ear**, although it can occur without swimming) is caused most by *P. aeruginosa* (up to 60%), but *S. aureus*, *Enterobacter aerogenes, Proteus mirabilis, Klebsiella pneumoniae,* streptococci, coagulase-negative staphylococci and diphtheroids, and fungi such as *Candida* and *Aspergillus* also may rarely be isolated. External otitis results from chronic irritation and maceration from excessive moisture in the canal. The loss of protective cerumen may play a role, as may trauma, but cerumen impaction with trapping of water also can cause infection. Inflammation of the ear canal due to herpesvirus, varicella-zoster virus, other skin exanthems, and eczema also may predispose to external otitis.

CLINICAL MANIFESTATIONS

The predominant symptom is acute rapid onset (typically within 48 hours) of ear pain (otalgia), often severe, accentuated by manipulation of the pinna or by pressure on the tragus and by jaw motion. The severity of the pain and tenderness (tragus or pinna, or both) may be disproportionate to the degree of inflammation, because the skin of the external ear canal is tightly adhered to the underlying perichondrium and periosteum. Itching often is a precursor of pain and usually is characteristic of chronic inflammation of the canal or resolving acute otitis externa. Conductive hearing loss (CHL) may result from edema of the skin and tympanic membrane (TM), serous or purulent secretions, or the canal skin thickening associated with chronic external otitis.

Edema of the ear canal, erythema, and thick, clumpy otorrhea are prominent signs of acute disease. The cerumen usually is white and soft in consistency, as opposed to its usual yellow color and firmer consistency (Fig. 679.1). The canal often is so tender and swollen that the entire ear canal and TM cannot be adequately visualized, and complete otoscopic examination may be delayed until the acute swelling subsides. If the TM can be visualized, it may appear either normal or opaque. TM mobility may be normal, or, if the TM is thickened, mobility may be reduced in response to positive and negative pressure.

Other physical findings may include palpable and tender lymph nodes in the periauricular region and erythema and swelling of the pinna and periauricular skin. Rarely, facial paralysis, other cranial nerve abnormalities, vertigo, and/or sensorineural hearing loss are present. If these occur, **necrotizing (malignant) otitis externa**, an invasive infection of the temporal bone and skull base, is probable. Fortunately, this disease is rare in children and is seen only in association with immunocompromise or severe malnourishment. In adults, it is associated with diabetes mellitus.

DIAGNOSIS

Diffuse external otitis may be confused with **furunculosis**, **otitis media (OM)**, and **mastoiditis** (Table 679.1). Furuncles occur in the lateral hair-bearing part of the ear canal; furunculosis usually causes a localized swelling of the canal limited to one quadrant, whereas external otitis is associated with concentric swelling and involves the entire ear canal. In OM, the TM may be perforated, severely retracted, or bulging and immobile; hearing usually is impaired. If the middle ear is draining through a perforated TM or tympanostomy tube, secondary external otitis may occur; if the TM is not visible owing to drainage or ear canal swelling, it may be difficult to distinguish acute OM with drainage from an acute external otitis. Pain on manipulation of the

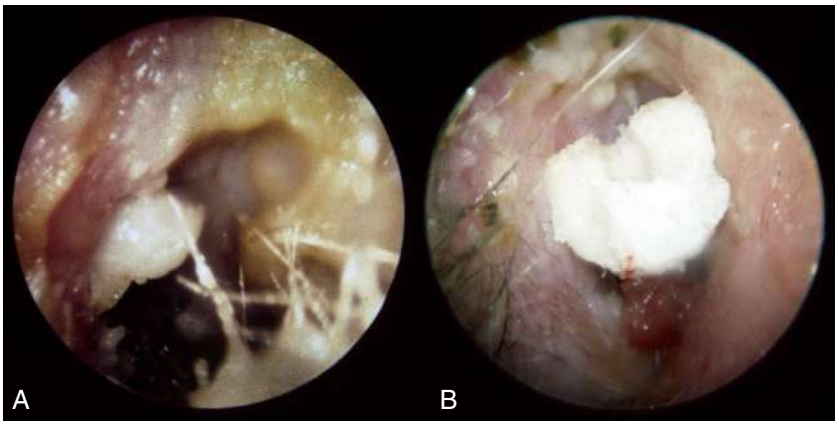

Fig. 679.1 Acute otitis externa. **A,** Erythema, edema, and copious purulent debris. **B,** In some cases an edematous canal with granulation tissue necessitates the placement of an ear wick to assist with topical drug delivery in the acute setting. *(Courtesy Dr. John W. House, Los Angeles.)*

Table 679.1 Differential Diagnosis of Painful External Ear and Auditory Canal Disorders

DISORDER	CLINICAL FEATURES
Acute otitis externa	Diffuse redness, swelling, and pain of the canal with greenish to whitish exudate; often very tender pinna
Necrotizing (malignant) otitis externa (± skull base osteomyelitis)	Rapidly progressive, severe swelling and redness of pinna; pinna may be laterally displaced; risk factors include diabetes mellitus, congenital or acquired immunodeficiency, severe neutropenia
Dermatitis	
Eczema	History of atopy, presence of lesions elsewhere; lesions are scaly, red, pruritic, and weeping
Contact	History of cosmetic use or irritant exposure; lesions are scaly, red, pruritic, and weeping
Seborrhea	Scaly, red, papular dermatitis; scalp may have thick, yellow scales
Psoriasis	History or presence of psoriasis elsewhere; erythematous papules that coalesce into thick, white plaques
Cellulitis	Diffuse redness, tenderness, and swelling of the pinna
Furuncles	Red, tender papules in areas with hair follicles (distal third of the ear canal)
Infected periauricular cyst	Discrete, palpable lesions; history of previous swelling at same site; cellulitis may develop, obscuring cystic structure
Insect bites	History of exposure; lesions are red, tender papules
Herpes zoster oticus	Painful, vesicular lesions in the ear canal and tympanic membrane, hearing loss, vestibulitis; with addition of seventh cranial nerve palsy—Ramsay Hunt syndrome
Perichondritis	Inflammation of the cartilage, usually secondary to cellulitis
Relapsing polychondritis	Recurrent episodes, involves other cartilage sites (nose)
Granulomatosis with polyangiitis	Fever, weight loss, respiratory and/or renal manifestations
Tumors including Langerhans cell histiocytosis	Palpable mass, destruction of surrounding structures
Foreign body	Foreign body may cause secondary trauma to the ear canal or become a nidus for an infection of the ear canal
Trauma	Bruising and swelling of external ear; there may be signs of basilar skull fracture (cerebrospinal fluid otorrhea, hemotympanum)
Red ear syndrome	Paroxysmal unilateral or bilateral burning and reddening. Associated with migraines or trigeminal cephalgias
Erythromelalgia	Burning, erythema from heat exposure relieved by cold; isolated ear involvement unusual

Modified from Kliegman R, Bordini B, Toth H, Basel D, eds. *Nelson Pediatric Symptom-Based Diagnosis,* 2nd ed. Philadelphia: Elsevier; 2022: Table 5.2.

auricle and significant lymphadenitis are not common features of OM, and these findings assist in the differential diagnosis. In some patients with external otitis, the periauricular edema is so extensive that the auricle is pushed forward, creating a condition that may be confused with acute mastoiditis and a subperiosteal abscess. In mastoiditis, the postauricular fold is obliterated, whereas in external otitis, the fold is usually better preserved. In acute mastoiditis, a history of OM and hearing loss is usual; tenderness is noted over the mastoid and not on movement of the auricle; and otoscopic examination may show sagging of the posterior canal wall.

Referred otalgia may come from disease in the paranasal sinuses, teeth, pharynx, parotid gland, neck and thyroid, and cranial nerves

(trigeminal neuralgia; herpes simplex virus, varicella-zoster virus; see Table 676.1).

TREATMENT

Topical otic preparations containing acetic acid, with or without hydrocortisone, or neomycin (active against gram-positive organisms and some gram-negative organisms, notably *Proteus* spp.), polymyxin (active against gram-negative bacilli, notably *Pseudomonas* spp.), or a quinolone (ciprofloxacin), with or without hydrocortisone, are all highly effective in treating most forms of acute external otitis. A nonototoxic (quinolone) antibiotic should be chosen in the setting of known TM perforation or tympanostomy tube. If canal edema is marked, the patient may need referral to a specialist for cleaning and possible wick placement. An otic antibiotic and corticosteroid eardrop is often recommended. A wick can be inserted into the ear canal and topical antibiotics applied to the wick 3 times a day for 24-48 hours. The wick can be removed after 2-3 days, at which time the edema of the ear canal usually is markedly improved and the ear canal and TM are better seen. Topical antibiotics are then continued by direct instillation. When the pain is severe, oral analgesics (e.g., ibuprofen, acetaminophen) may be necessary for a few days.

Someone other than the patient should place the drops in the ear canal while the patient is recumbent with the affected ear facing up. The drops should fill the canal, and the patient should remain in place for 3-5 minutes. Gently moving the ear to and fro may enhance the drops to fill the ear canal. Patients should respond to initial therapeutics in 48-72 hours. Failure to improve in this time frame should prompt assessment of drug delivery and adherence to therapy, consideration for change in therapy, and consideration of alternative diagnoses. Careful evaluation for underlying conditions should also be undertaken in patients with severe or recurrent otitis externa. Figure 679.2 outlines an approach to managing acute external otitis.

As the inflammatory process subsides, cleaning the canal with a suction or cotton-tipped applicator to remove the debris enhances the effectiveness of the topical medications. In subacute and chronic infections, periodic cleansing of the canal is essential. In severe acute external otitis associated with fever and lymphadenitis, oral or parenteral antibiotics may be indicated; an ear canal culture should be done, and empirical antibiotic treatment can then be modified if necessary, based on susceptibility of the organism cultured. A fungal infection of the external auditory canal, or **otomycosis**, is characterized by fluffy white debris, sometimes with black spores seen; treatment includes cleaning and application of antifungal solutions such as clotrimazole or nystatin; other antifungal agents include boric acid powder, m-cresyl acetate 25%, gentian violet 2%, and thimerosal 1:1,000.

Necrotizing otitis externa, commonly caused by *P. aeruginosa* (see Chapter 251.1), requires immediate culture, intravenous antibiotics,

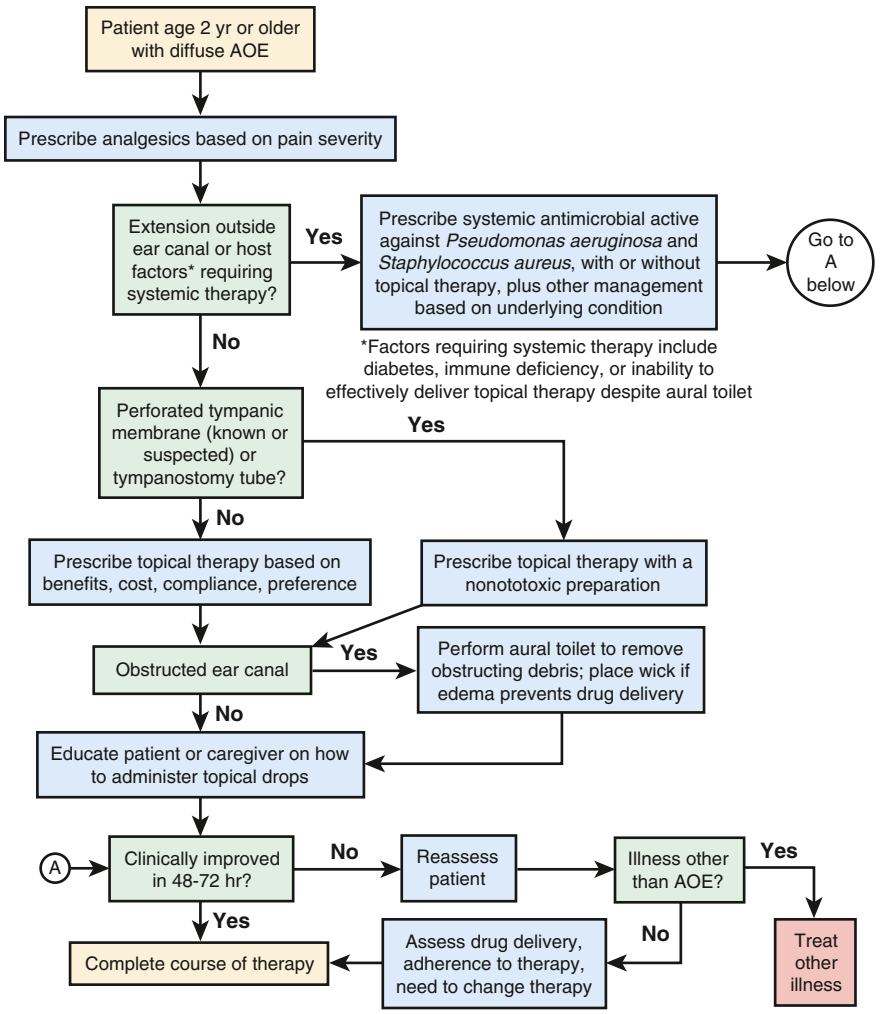

Fig. 679.2 Management algorithm for acute otitis externa (AOE). *(From Rosenfeld RM, Brown L, Cannon CR, et al. Clinical practice guideline: acute otitis externa. Otolaryngol Head Neck Surg. 2006;134:S4–S23. Copyright 2006 American Academy of Otolaryngology—Head and Neck Surgery Foundation, Inc.)*

and imaging studies to evaluate the extent of the disease. Surgical intervention to obtain cultures or debride devitalized tissue may be necessary.

PREVENTION

Preventing external otitis may be necessary for individuals susceptible to recurrences, especially children who swim. The most effective prophylaxis is instillation of dilute alcohol or acetic acid (2%) immediately after swimming or bathing. During an acute episode of otitis externa, patients should not swim, and the ears should be protected from excessive water during bathing. A hair dryer may be used to clear moisture from the ear after swimming as a method of prevention. Cotton-tipped swabs (or another material) may cause trauma to the ear canal, including tympanic membrane perforation, cerumen impaction, or retained foreign body and should be avoided.

OTHER DISEASES OF THE EXTERNAL EAR

Furunculosis

Furunculosis, caused by *S. aureus,* affects only the hair-containing outer third of the ear canal and typically occurs at the inferior entrance to the meatus. Mild forms are treated with oral antibiotics active against *S. aureus*. If an abscess develops, incision and drainage may be necessary.

Acute Cellulitis

Acute cellulitis of the auricle and external auditory canal usually is caused by group A streptococcus and occasionally by *S. aureus*. The skin is red, hot, and indurated, without a sharply defined border. Fever may be present with little or no exudate in the canal. Parenteral administration of penicillin G or a penicillinase-resistant penicillin is the therapy of choice.

Perichondritis and Chondritis

Perichondritis is an infection involving the skin and perichondrium of the auricular cartilage; extension of infection to the cartilage is termed **chondritis**. The ear canal, especially the lateral aspect, also may be involved. Early perichondritis may be difficult to differentiate from cellulitis because both are characterized by skin that is red, edematous, and tender. The main cause of perichondritis/chondritis and cellulitis is trauma (accidental or iatrogenic, laceration or contusion), including ear piercing, especially when done through the cartilage. The most commonly isolated organism in perichondritis and chondritis is *P. aeruginosa,* although other gram-negative and, occasionally, gram-positive organisms may be found. Treatment involves systemic, often parenteral, antibiotics; surgery to drain an abscess or remove nonviable skin or cartilage may also be needed. Removal of all ear jewelry is mandatory in the presence of infection.

Dermatoses

Various dermatoses (seborrheic, contact, infectious eczematoid, or neurodermatoid) are common causes of inflammation of the external canal; scratching and the introduction of infecting organisms cause acute external otitis in these conditions.

Seborrheic dermatitis is characterized by greasy scales that flake and crumble as they are detached from the epidermis; associated changes in the scalp, forehead, cheeks, brow, postauricular areas, and concha are usual.

Contact dermatitis of the auricle or canal may be caused by earrings or by topical otic medications such as neomycin, which may produce erythema, vesiculation, edema, and weeping. Poison ivy, oak, and sumac also may produce contact dermatitis. Hair care products have been implicated in sensitive individuals.

Infectious eczematoid dermatitis is caused by a purulent infection of the external canal, middle ear, or mastoid; the purulent drainage infects the skin of the canal or auricle, or both. The lesion is weeping, erythematous, or crusted.

Atopic dermatitis occurs in children with a familial or personal history of allergy; the auricle, particularly the postauricular fold, becomes thickened, scaly, and excoriated.

Neurodermatitis is recognized by intense itching and erythematous, thickened epidermis localized to the concha and orifice of the meatus.

Treatment of these dermatoses depends on the type but should include application of an appropriate topical medication, elimination of the source of infection or contact when identified, and management of any underlying dermatologic problem. In addition to topical antibiotics (or antifungals), topical steroids are helpful if contact dermatitis (see Chapter 696.1), atopic dermatitis (see Chapter 696), or eczematoid dermatitis is suspected.

Herpes Simplex Virus

See Chapter 299.

Herpes simplex virus may appear as vesicles on the auricle and lips. The lesions eventually become encrusted and dry and may be confused with impetigo. Topical application of a 10% solution of carbamide peroxide in anhydrous glycerol is symptomatically helpful. **Ramsay Hunt syndrome** (herpes zoster oticus with facial paralysis) may present initially with otalgia, with subsequent appearance of vesicles in the ear canal and on the pinna and with facial paralysis and pain. Other cranial nerves may be affected as well, especially the eighth nerve. Treatment of herpes zoster oticus includes systemic antiviral agents, such as acyclovir, and systemic corticosteroids. As many as 50% of patients with Ramsay Hunt syndrome do not completely recover their facial nerve function.

Bullous Myringitis

Commonly associated with an acute upper respiratory tract infection, bullous myringitis presents as an ear infection with more severe pain than usual. On examination, hemorrhagic or serous blisters (bullae) may be seen on the TM. The disease sometimes is difficult to differentiate from acute OM because a large bulla may be confused with a bulging TM. The organisms involved are the same as those that cause acute OM, including both bacteria and viruses. Treatment consists of empiric antibiotic therapy and pain medications. In addition to ibuprofen or codeine for severe pain, a topical anesthetic eardrop may also provide some relief. Incision of the bullae, although not necessary, promptly relieves the pain.

Exostoses and Osteomas

Exostoses represent benign hyperplasia of the perichondrium and underlying bone. Those involving the auditory canal tend to be found in people who swim often in cold water. Exostoses are broad based, often multiple, and bilateral. Osteomas are benign bony growths in the ear canal of uncertain cause (see Chapter 550.2). They usually are solitary and attached by a narrow pedicle to the tympanosquamous or tympanomastoid suture line. Both are more common in males; exostoses are more common than osteomas. Surgical treatment is recommended when large masses cause cerumen impaction, ear canal obstruction, or hearing loss.

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Chapter 680

Otitis Media

Brittany Player

The term **otitis media (OM)** has two main categories: acute infection, which is termed suppurative or **acute otitis media (AOM)**, and inflammation accompanied by **middle-ear effusion (MEE)**, termed nonsuppurative or **secretory OM**, or **otitis media with effusion (OME)**. These two main types are interrelated: acute infection usually is succeeded by residual inflammation and effusion that, in turn, predispose children to recurrent infection. MEE is a feature of both AOM and OME and is an expression of the underlying middle-ear mucosal inflammation. MEE results in the conductive hearing loss associated with OM, ranging from none to as much as 50 dB of hearing loss (moderate hearing loss).

The peak incidence and prevalence of OM are during the first 2 years of life. More than 80% of children experience at least one episode of OM by the age of 3 years. Conservative estimates for OM-related costs place the annual burden at $3-5 billion per year. Accounting for >10% of pediatric primary care encounters, OM is a leading reason for pediatric visits and use of antibiotics and figures importantly in the differential diagnosis of fever. Recurrent OM is defined as a minimum of three episodes of AOM in 6 months or four episodes of AOM in 12 months with at least one of those episodes occurring in the preceding 6 months. It often serves as the main justification for myringotomy with insertion of tympanostomy tubes and adenoidectomy, the most frequently performed operations in infants and young children. OM is also the most common cause of acquired hearing loss in children. OM has a propensity to become chronic and recur. The earlier in life a child experiences the first episode, the greater the frequency of recurrence, severity, and persistence of MEE.

Accurate diagnosis of AOM in infants and young children may be difficult (Figs. 680.1-680.3). Symptoms may not be apparent, especially in early infancy and in chronic stages of the disease. Accurate visualization of the tympanic membrane (TM) and middle-ear space may be difficult because of anatomy, patient cooperation, or blockage by cerumen, removal of which may be arduous and time consuming. Abnormalities of the eardrum may also be subtle or difficult to appreciate. In the face of these difficulties, both underdiagnosis and overdiagnosis occur.

EPIDEMIOLOGY

Several factors affect the occurrence of OM, including age, gender, genetic background, socioeconomic status, breast milk feeding, degree of exposure to tobacco smoke, degree of exposure to other children, presence or absence of respiratory allergy, season of the year, and pneumococcal vaccination status. Children with certain types of immune deficiencies and congenital craniofacial anomalies (i.e., cleft palate, Down syndrome) are particularly prone to OM.

Age

The age of onset of OM is an important predictor of the development of recurrent and chronic OM, with earlier age of onset having an increased risk for exhibiting these difficulties later in life. The development of at least one episode of OM is reported as 63–85% by 12 months and 66–99% by 24 months of age. The percentage of days with MEE is 5–27% during the first year of life and 6–18% during the second year of life. Across groups, rates are highest at 6-20 months of age. After the age of 2, the incidence and prevalence of OM decline progressively, although the disease remains relatively common into the early school-age years. The most likely reasons for the higher rates in infants and younger children include less well-developed immunologic defenses and less favorable eustachian tubal structure and function.

Gender

Epidemiologic data suggest an incidence of OM greater in males than in females, although some studies have found no gender-related differences in the occurrence of OM.

Genetic Background

That middle-ear disease tends to run in families is a commonplace observation, suggesting that OM may have a heritable component. The degree of concordance for the occurrence of OM is much greater among monozygotic than among dizygotic twins. OM is especially prevalent and severe among Native American, Inuit, and Indigenous Australian children.

Socioeconomic Status

Elements contributing to the association of poverty with OM include crowding, limited hygienic facilities, suboptimal nutritional status, limited access to medical care, and limited resources for complying with prescribed medical regimens.

Breast Milk Compared with Formula Feeding

Most studies have found a protective effect of breast milk feeding against OM. This protective effect may be greater in socioeconomically

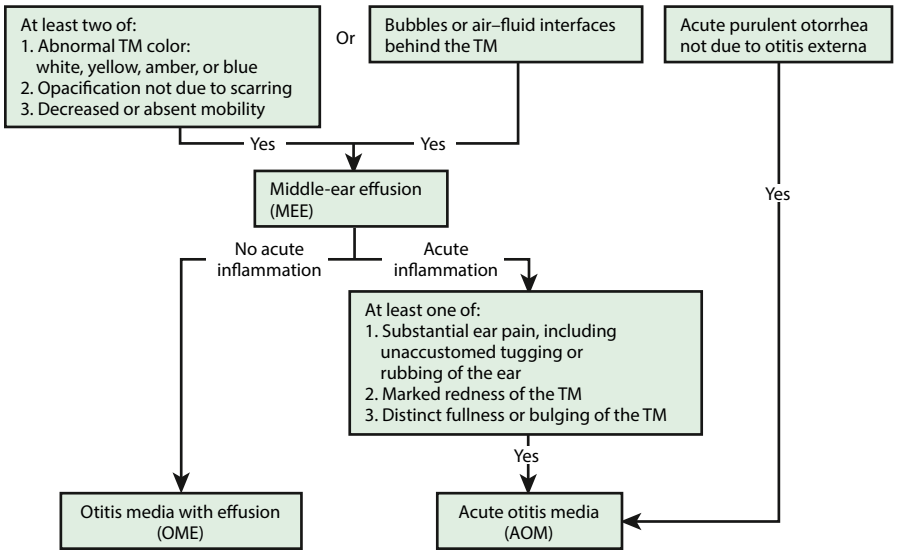

Fig. 680.1 Algorithm for distinguishing between acute otitis media and otitis media with effusion. TM, Tympanic membrane.

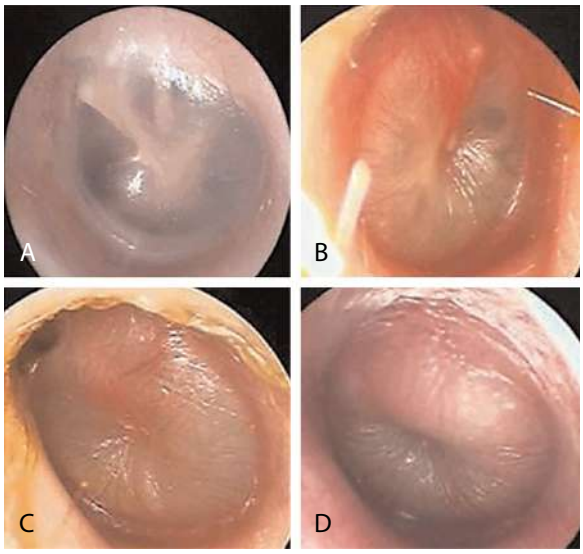

Fig. 680.2 Examples of normal tympanic membrane (A) and of mild bulging (B), moderate bulging (C), and severe bulging (D) of the tympanic membrane from middle-ear effusion. *(Courtesy Alejandro Hoberman, MD.)*

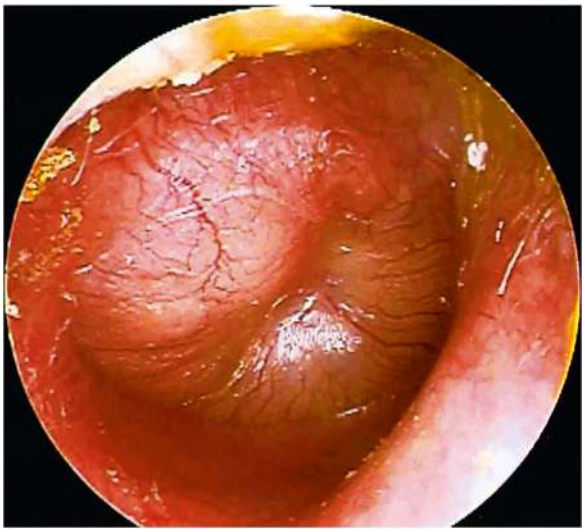

Fig. 680.3 Tympanic membrane in acute otitis media.

disadvantaged children than in more advantaged children. The protective effect is attributable to the milk itself rather than to the mechanics of breastfeeding.

Exposure to Tobacco Smoke

Tobacco smoke exposure is an important preventable risk factor in the development of OM. Studies that have used objective measures to determine infant exposure to secondhand tobacco smoke, such as cotinine levels, have consistently identified a significant linkage between tobacco smoke and OM.

Exposure to Other Children

OM is more common with repeated exposure to other children, whether at home or in out-of-home group daycare. Together, but independently, family socioeconomic status and the extent of exposure to other children appear to constitute two of the most important identifiable risk factors for developing OM.

Season

In keeping with the pattern of occurrence of upper respiratory tract infections in general, the highest rates of occurrence of OM are observed during cold weather months and the lowest rates during warm weather months. In OM, it is likely that these findings strongly depend on the significant association of OM with viral respiratory illnesses.

Congenital Anomalies

OM is universal among infants with unrepaired palatal clefts and is also highly prevalent among children with submucous cleft palate, other craniofacial anomalies, and Down syndrome (see Chapter 57). The common feature in these congenital anomalies is a deficiency in eustachian tube function, which predisposes these children to middle-ear disease.

Other Factors

Pacifier use is linked with an increased incidence of OM and recurrence of OM, although the effect is small. Neither maternal age nor birthweight nor season of birth appears to influence the occurrence of OM once other demographic factors are accounted for. Some suggest an association of OM with bottle feeding in the recumbent position (propped bottle). Children with HIV infection have a high risk for recurrent OM.

ETIOLOGY

Acute Otitis Media

Pathogenic bacteria can be isolated by standard culture techniques from middle-ear fluid in most documented AOM cases. Three pathogens predominate in AOM: *Streptococcus pneumoniae* (see Chapter 228), nontypeable *Haemophilus influenzae* (see Chapter 240), and *Moraxella catarrhalis* (see Chapter 242). The overall incidence of these organisms has changed with the use of the conjugate pneumococcal vaccine. Widespread use of the expanded serotype coverage 13-valent as compared with the 7-valent pneumococcal conjugate vaccine has further reduced the prevalence of *S. pneumoniae* as a cause of AOM, particularly the virulent 19A serotype. Less common pathogens include group A streptococcus (see Chapter 229), *Staphylococcus aureus* (see Chapter 227.1), and gram-negative organisms. Gram-negative organisms and *S. aureus* are found most commonly in neonates and very young infants who are hospitalized; in outpatient settings, the distribution of pathogens in these young infants is similar to that in older infants. Molecular techniques to identify nonculturable bacterial pathogens have suggested the importance of other bacterial species such as *Alloiococcus otitidis.*

Respiratory viruses may also be identified in MEEs of children with AOM, either alone or, more commonly, in association with pathogenic bacteria. Of these viruses, rhinovirus (see Chapter 310) and respiratory syncytial virus (RSV; see Chapter 307) are found most often. AOM is a known complication of bronchiolitis; middle-ear aspirates in children with bronchiolitis regularly contain bacterial pathogens, suggesting that RSV is rarely, if ever, the sole cause of their AOM. Viral pathogens have a negative impact on eustachian tube function, can impair local immune function and increase bacterial adherence, and can change the pharmacokinetic dynamics, reducing the efficacy of antimicrobial medications. However, it remains uncertain whether viruses alone can cause AOM or whether their role is limited to setting the stage for bacterial invasion, and perhaps also to amplify the inflammatory process, thereby interfering with resolution of the bacterial infection.

Otitis Media with Effusion

Using standard culture techniques, the pathogens typically found in AOM are recoverable in only 30% of children with OME. However, using polymerase chain reaction (PCR), MEEs contain evidence of bacterial DNA and viral RNA in much larger proportions of these children. Biofilms of pathogenic bacteria are present on the middle-ear mucosa and adenoid pad in most children with chronic OM. Biofilms consist of aggregated and adherent bacteria, embedded in an extracellular matrix and in neutrophil extracellular traps, allowing for

protection against antimicrobials; their presence may contribute to the persistence of pathogens and the recalcitrance of chronic OM to antibiotic treatment.

PATHOGENESIS

A multifactorial disease process, risk profile, and host-pathogen interactions play important roles in the pathogenesis of OM. Such events as alterations in mucociliary clearance through repeated viral exposure experienced in daycare settings or through exposure to tobacco smoke may tip the balance of pathogenesis in less virulent OM pathogens in their favor, especially in children with a unique host predisposition.

Anatomic Factors

Patients with significant craniofacial abnormalities affecting eustachian tube function have an increased incidence of OM. During the pathogenesis of OM, the eustachian tube demonstrates decreased effectiveness in ventilating the middle-ear space.

Under usual circumstances the eustachian tube is passively closed and is opened by contraction of the tensor veli palatini muscle. In relation to the middle ear, the tube has three main functions: ventilation, protection, and clearance. The middle-ear mucosa depends on a continuing supply of air from the nasopharynx delivered by way of the eustachian tube. Interruption of this ventilatory process by tubal obstruction initiates an inflammatory response that includes secretory metaplasia, compromise of the mucociliary transport system, and effusion of liquid into the tympanic cavity. Measurements of eustachian tube function have demonstrated that the tubal function is suboptimal during the events of OM with increased opening pressures.

Eustachian tube obstruction may result from extraluminal blockage via hypertrophied nasopharyngeal adenoid tissue or tumor or may result from intraluminal obstruction via inflammatory edema of the tubal mucosa, most commonly as a consequence of a viral upper respiratory tract infection. Progressive reduction in tubal wall compliance with increasing age may explain the progressive decline in the occurrence of OM as children grow older. The protection and clearance functions of the eustachian tube may also be involved in the pathogenesis of OM. Thus, if the eustachian tube is patulous or excessively compliant, it may fail to protect the middle ear from reflux of infective nasopharyngeal secretions, whereas impairment of the mucociliary clearance function of the tube might contribute to both the establishment and persistence of infection. The shorter and more horizontal orientation of the tube in infants and young children may increase the likelihood of reflux from the nasopharynx and impair passive gravitational drainage through the eustachian tube.

Children with **craniofacial abnormalities** experience an increased incidence of OM associated with the abnormal eustachian tube function. In children with cleft palate, where OM is a universal finding, a main factor underlying the chronic middle-ear inflammation appears to be impairment of the opening mechanism of the eustachian tube. Possible factors include muscular changes, tubal compliance factors, and defective velopharyngeal valving, which may result in disturbed aerodynamic and hydrodynamic relationships in the nasopharynx and proximal portions of the eustachian tubes. In children with other craniofacial anomalies and with Down syndrome, the high prevalence of OM has also been attributed to structural and/or functional eustachian tubal abnormalities.

Host Factors

The effectiveness of a child's immune system in response to the bacterial and viral infections of the upper airway and middle ear during early childhood probably is the most important factor in determining which children are otitis prone. The maturation of this immune system during early childhood is most likely the primary event leading to the decrease in incidence of OM with increased age. Immunoglobulin A (IgA) deficiency is found in some children with recurrent AOM, but the significance is questionable; many children with IgA deficiency do not experience recurrent episodes of AOM. Selective immunoglobulin G (IgG) subclass deficiencies (despite normal total serum IgG) may be found in children with recurrent AOM in association with recurrent sinopulmonary infection, and these deficiencies probably underlie the susceptibility to infection. Children with HIV infection have recurrent and difficult-to-treat episodes of AOM in the first and second year of life. Children with recurrent OM that is not associated with recurrent infection at other anatomic sites rarely have a readily identifiable immunologic deficiency. Evidence that subtle immune deficits play a role in the pathogenesis of recurrent AOM is provided by studies involving antibody responses to various types of infection and immunization; by the observation that breast milk feeding, as opposed to formula feeding, confers some protection against the occurrence of OM in infants with cleft palate; and by studies in which young children with recurrent AOM achieved a measure of protection from intramuscularly administered bacterial polysaccharide immunoglobulin or intravenously administered polyclonal immunoglobulin. This evidence, along with the documented decrease in incidence of upper respiratory tract infections and OM as children's immune systems develop and mature, is indicative of the importance of a child's innate immune system in the pathogenesis of OM (see Chapter 164).

Viral Pathogens

Although OM may develop and persist in the absence of apparent respiratory tract infection, many, if not most, episodes are initiated by viral or bacterial upper respiratory tract infection. Among children in group daycare, AOM was observed in approximately 30–40% of children with respiratory illness caused by RSV (see Chapter 307), influenza viruses (see Chapter 305), or adenoviruses (see Chapter 309) and in approximately 10–15% of children with respiratory illness caused by parainfluenza viruses (see Chapter 306), rhinoviruses (see Chapter 310), or enteroviruses (see Chapter 297). Viral infection of the upper respiratory tract results in release of cytokines and inflammatory mediators, some of which may cause eustachian tube dysfunction.

Respiratory viruses also may enhance nasopharyngeal bacterial colonization and adherence and impair host immune defenses against bacterial infection.

CLINICAL MANIFESTATIONS

Symptoms of AOM are variable, especially in infants and young children. In young children, evidence of ear pain may be manifested by irritability or a change in sleeping or eating habits and, occasionally, holding or tugging at the ear. *Pulling at the ear alone has a low sensitivity and specificity.* Fever may also be present and may occasionally be the only sign. Rupture of the TM with purulent otorrhea is uncommon. Systemic symptoms and symptoms associated with upper respiratory tract infections also occur; occasionally there may be no symptoms, with the disease having been discovered at a routine health examination. The Acute Otitis Media Severity of Symptom (AOM-SOS) scale is a five-item validated symptom score that has proven beneficial as a tool to monitor AOM symptoms in patients and studies of antimicrobial effectiveness in OM. OME often is not accompanied by overt complaints of the child but can be accompanied by hearing loss. This hearing loss may manifest as changes in speech patterns but often goes undetected if unilateral or mild in nature, especially in younger children. Balance difficulties or disequilibrium can rarely be associated with OME, and older children may complain of mild discomfort or a sense of fullness in the ear.

EXAMINATION OF THE TYMPANIC MEMBRANE

Otoscopy

Two types of otoscope heads are available: **surgical** or **operating** and **diagnostic** or **pneumatic**. The surgical head embodies a lens that can swivel over a wide arc and an unenclosed light source, thus providing ready access of the examiner's instruments to the external auditory canal and TM. Use of the surgical head is optimal for removing cerumen or debris from the canal under direct observation and is necessary for satisfactorily performing tympanocentesis or myringotomy. The

diagnostic head incorporates a larger lens, an enclosed light source, and a nipple for the attachment of a rubber bulb and tubing. When an attached speculum is fitted snugly into the external auditory canal, an airtight chamber is created comprising the vault of the otoscope head, the bulb and tubing, the speculum, and the proximal portion of the external canal. Although examination of the ear in young children is a relatively invasive procedure that is often met with lack of cooperation by the patient, this task can be enhanced if done with as little pain as possible. The outer portion of the ear canal contains hair-bearing skin and subcutaneous fat and cartilage that allow a speculum to be placed with relatively little discomfort. Closer to the TM the ear canal is made of bone and is lined only with skin and no adnexal structures or subcutaneous fat; a speculum pushed too far forward and placed in this area often causes skin abrasion and pain. Using a rubber-tipped speculum or adding a small sleeve of rubber tubing to the tip of the plastic speculum may serve to minimize patient discomfort and enhance the ability to achieve a proper fit and an airtight seal, facilitating pneumatic otoscopy.

Learning to perform **pneumatic otoscopy** is a critical skill in being able to assess a child's ear and in making an accurate diagnosis of AOM. The degree of TM mobility in response to both positive and negative pressure can be estimated by observing as the bulb is alternately squeezed gently and released, thus providing a critical assessment of middle-ear fluid, which is a hallmark sign of both AOM and OME (see Fig. 680.1). With both types of otoscope heads, bright illumination is also critical for adequate visualization of the TM.

Clearing the External Auditory Canal

Children's ears are "self-cleaning" because of squamous migration of ear canal skin. Cleaning of cerumen with cotton-tipped swabs should be avoided, as it often worsens impaction by pushing cerumen deeper into the canal, compacting it. If the TM is obscured by cerumen, the cerumen should be removed. This can be accomplished through direct visualization using a headlight or through the surgical head of the otoscope by using an ear curette or gentle suction with a No. 5 or 7 French ear suction tube. During this procedure, it may be most advantageous to restrain the infant or young child in the prone position, turning the child's head to the left or right as each ear is cleared. In children old enough to cooperate, clearing of the external canal may be achieved more easily and less traumatically by lavage than by mechanical removal, provided one can be certain that a TM perforation is not present.

Tympanic Membrane Findings

Important characteristics of the TM consist of contour, color, translucence, structural changes, if any, and mobility. The TM is anatomically divided into the pars tensa and pars flaccida. The pars tensa comprises the lower two thirds of the drum inferior to the lateral process of the malleus. Its contour is normally **slightly concave**; abnormalities consist of fullness or bulging or, conversely, extreme retraction. The normal color of the pars tensa is **pearly gray**, with the pars flaccida being slightly more vascular in nature. Erythema may be a sign of inflammation or infection, but unless intense, erythema alone may result from crying or vascular flushing. Abnormal whiteness of the membrane may result from either scarring or the presence of effusion in the middle-ear cavity; this effusion also may impart an amber, pale yellow, or, rarely, bluish color. Rarely a persistent focal white area may be indicative of a congenital cholesteatoma in the middle-ear space. Normally, the membrane is translucent, although some degree of opacity may be normal in the first few months of life; later, opacification denotes either scarring or, more commonly, underlying effusion. Structural changes include scars, perforations, and retraction pockets. Retractions or perforations, especially in the posterior-superior quadrant, or pars flaccida, of the TM may be a sign of cholesteatoma formation. Of all the visible characteristics of the TM, mobility is the most sensitive and specific in determining the presence or absence of MEE. Mobility is generally not an all-or-none phenomenon. A total absence of mobility does exist with a TM perforation that can develop after a substantial increase in middle-ear pressure associated with effusion. When a perforation is not present, substantial impairment of mobility is the more common finding with MEE. Bulging of the TM is the most specific finding of AOM (97%) but has lower sensitivity (51%) (see Figs. 680.2 and 680.3).

Diagnosis

A **diagnosis of AOM** should be made in children who present with:

- Moderate to severe bulging of the TM or new-onset otorrhea not caused by otitis externa
- Mild bulging of the TM and recent (<48 hours) onset of ear pain or intense TM erythema

A **diagnosis of AOM** should ***not*** be made in children without MEE.

AOM and OME may evolve into the other without any clearly differentiating physical findings; any schema for distinguishing between them is to some extent arbitrary. In an era of increasing bacterial resistance, distinguishing between AOM and OME is important in determining treatment, because OME in the absence of acute infection does not require antimicrobial therapy. Purulent otorrhea of recent onset is indicative of AOM. Difficulty in distinguishing clinically between AOM and OME is limited to circumstances in which purulent otorrhea is not present. Both AOM without otorrhea and OME are accompanied by physical signs of MEE, namely, the presence of at least two of three TM abnormalities: white, yellow, amber, or (rarely) blue discoloration; opacification other than that caused by scarring; and decreased or absent mobility. Alternatively, in OME, either air-fluid levels or air bubbles outlined by small amounts of fluid may be visible behind the TM, a condition often indicative of impending resolution (see Fig. 680.4).

To support a diagnosis of AOM instead of OME in a child with MEE, distinct fullness or bulging of the TM may be present, with or without accompanying erythema, or, at a minimum, MEE should be accompanied by ear pain that appears clinically important. Unless intense, erythema alone is insufficient because erythema, without other abnormalities, may result from crying or vascular flushing. In AOM, the malleus may be obscured, and the TM may resemble a bagel without a hole but with a central depression (see Fig. 680.3). Rarely, the TM may be obscured by surface bullae or may have a cobblestone appearance. Bullous myringitis is a physical manifestation of AOM and not an etiologically discrete entity. Within days after onset, fullness of the membrane may diminish, even though infection may still be present.

In OME, bulging of the TM is absent or slight or the membrane may be retracted (Fig. 680.4); erythema also is absent or slight but may increase with crying or with superficial trauma to the external auditory canal incurred in clearing the canal of cerumen.

Both before and after episodes of OM and also in the absence of OM, the TM may be retracted as a consequence of negative middle-ear air pressure. The presumed cause is diffusion of air from the middle-ear

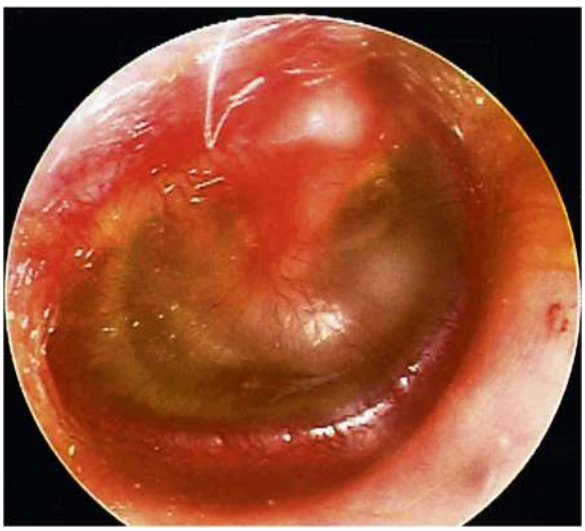

Fig. 680.4 Tympanic membrane in otitis media with effusion.

cavity more rapidly than it is replaced via the eustachian tube. Mild retraction is generally self-limited, although in some children it is accompanied by mild conductive hearing loss. More extreme retraction is of concern, as discussed later in the section on sequelae of OM.

Conjunctivitis-Associated Otitis Media

Simultaneous appearance of purulent and erythematous conjunctivitis with an ipsilateral OM is a well-recognized presentation, caused by nontypeable *H. influenzae* in most children. The disease often is present in multiple family members and affects young children and infants. Topical ocular antibiotics are ineffective. In an era of resistant organisms, this clinical association can be important in antibiotic selection, with oral antibiotics (see later) effective against resistant forms of nontypeable *H. influenzae.*

Asymptomatic Purulent Otitis Media

Rarely, a child will present during a routine exam without fever, irritability, or other overt signs of infection, but on exam, the patient will demonstrate an obvious purulent MEE and bulging TM. Although an uncommon presentation of "acute" OM, the bulging nature of the TM and the obvious purulence of the effusion do warrant antimicrobial therapy.

Tympanometry

Tympanometry, or **acoustic immittance testing**, is a simple, rapid, atraumatic test that, when performed correctly, offers objective evidence of the presence or absence of MEE. The tympanogram provides information about **TM compliance** in electroacoustic terms that can be thought of as approximately equivalent to TM mobility as perceived visually during pneumatic otoscopy. The absorption of sound by the TM varies inversely with its stiffness. The stiffness of the membrane is least, and its compliance is greatest, when the air pressures impinging on each of its surfaces—middle-ear air pressure and external canal air pressure—are equal. Anything tending to stiffen the TM, such as TM scarring or middle-ear fluid, reduces the TM compliance, which is recorded as a flattening of the curve of the tympanogram. An ear filled with middle-ear fluid generally has a very noncompliant TM and therefore a flattened tympanogram tracing.

Tympanograms may be grouped into one of three categories (Fig. 680.5). Tracings characterized by a relatively steep gradient, sharp-angled peak, and middle-ear air pressure (location of the peak in terms of air pressure) that approximates atmospheric pressure (see Fig. 680.5A) (type A curve) are assumed to indicate normal middle-ear status. Tracings characterized by a shallow peak or no peak are often termed "flat" or type B (see Fig. 680.5B) and usually are assumed to indicate the presence of a middle-ear abnormality that is causing decreased TM compliance. The most common such abnormality in infants and children is MEE. Tracings characterized by intermediate findings—somewhat shallow peak, often in association with a gradual gradient (obtuse-angled peak) or negative middle-ear air pressure peak (often termed type "C"), or combinations of these features (see Fig. 680.5C)—may or may not be associated with MEE and must be considered nondiagnostic or equivocal with respect to OM. However, type C tympanograms do suggest eustachian tube dysfunction and some ongoing pathology in the middle ear and warrant follow-up.

When reading a tympanogram, it is important to look at the volume measurement. The type B tympanometric response is analyzed within the context of the recorded volume. A flat, "low"-volume (≤1 mL) tracing typically reflects the volume of the ear canal only, representing MEE, which impedes the movement of an intact eardrum. A flat, high-volume (>1 mL) tracing typically reflects the volume of the ear canal and middle-ear space, representing a perforation (or patent tympanostomy tube) in the TM. In a child with a tympanostomy tube present, a flat tympanogram with a volume <1 mL would suggest a plugged or nonfunctioning tube and middle-ear fluid, whereas a flat tympanogram with a volume >1 mL would suggest a patent tympanostomy tube.

Although tympanometry is quite sensitive in detecting MEE, it can be limited by patient cooperation, the skill of the individual administering the test, and the age of the child, with less reliable results in very young children. Use of tympanometry may be helpful in office screening, may supplement the examination of difficult-to-examine patients, and may help to identify patients who require further attention because their tympanograms are abnormal. Tympanometry also may be used to help confirm, refine, or clarify questionable otoscopic findings; to objectify the follow-up evaluation of patients with known middle-ear disease; and to validate otoscopic diagnoses of MEE. Even though tympanometry can predict the probability of MEE, it cannot distinguish the effusion of OME from that of AOM.

PREVENTION

General measures to prevent OM that have been supported by numerous investigations include avoiding exposure to individuals with respiratory infection, appropriate vaccination strategies against pneumococci and influenzae, avoiding environmental tobacco smoke, and breast milk feeding.

IMMUNOPROPHYLAXIS AND VACCINATION STATUS

Heptavalent pneumococcal conjugate vaccine (PCV7) reduced the overall number of episodes of AOM by only 6–8% but with a 57% reduction in serotype-specific episodes. Reductions of 9–23% are seen in children with histories of frequent episodes, and a 20% reduction is seen in the number of children undergoing tympanostomy tube insertion. The 13-valent pneumococcal polysaccharide-protein conjugate vaccine (PCV13) contains the seven serotypes included in PCV7 (serotypes 4, 6B, 9V, 14, 18C, 19F, and 23F) and six additional serotypes (serotypes 1, 3, 5, 6A, 7F, and 19A). Early data indicate a significant reduction in the number of invasive pneumococcal mastoiditis cases since the introduction of PCV13. With the widespread use of PCV13,

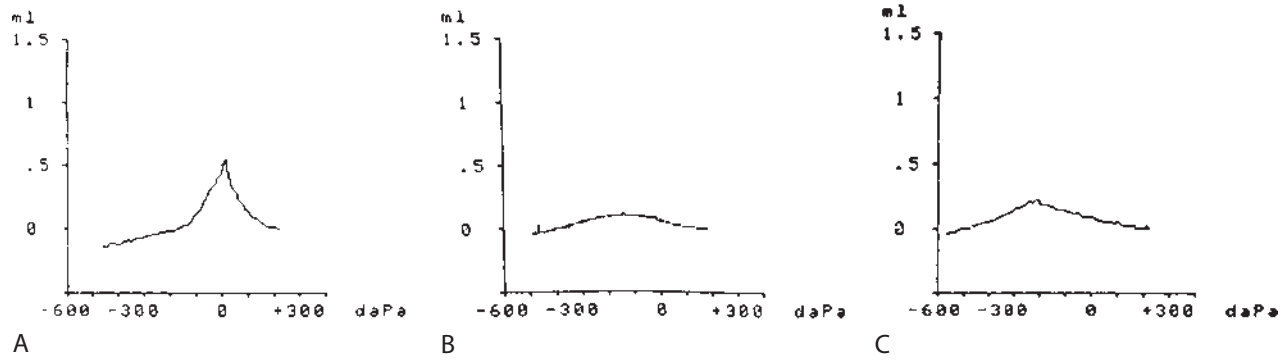

Fig. 680.5 Tympanograms obtained with a Grason-Stadler GSI 33 Middle Ear Analyzer, exhibiting (**A**) high admittance, steep gradient (i.e., sharp-angled peak), and middle-ear air pressure approximating atmospheric pressure (0 decaPascals [daPa]); (**B**) low admittance and indeterminate middle-ear air pressure; and (**C**) somewhat low admittance, gradual gradient, and markedly negative middle-ear air pressure.

continued surveillance will be necessary to detect other emerging serotypes, which are also demonstrating increasing resistance. Although the influenza vaccine also provides a measure of protection against OM, the relatively limited time during which individuals and even communities are exposed to influenza viruses limits the vaccine's effectiveness in broadly reducing the incidence of OM. Limitation of OM disease is only a portion of the benefit realized from the vaccinations for pneumococci and influenza viruses.

TREATMENT

Management of Acute Otitis Media

AOM can be very painful. Whether or not antibiotics are used for treatment, pain should be assessed and, if present, treated (Table 680.1). Individual episodes of AOM have traditionally been treated with antimicrobial drugs, and a sharp decline in complications from AOM during the last half-century seems, at least in part, attributable to the widespread routine use of antimicrobials. However, concerns about increasing bacterial resistance have prompted consideration for withholding antimicrobial treatment in certain, well-defined clinical situations (Table 680.2). Although viral and bacterial pathogens have been cultured from middle-ear fluid obtained by tympanocentesis, two factors may argue in favor of prescribing antimicrobial therapy for children who have AOM. First, symptomatic improvement and resolution of infection may occur more promptly and more consistently with antimicrobial treatment than without, even though most untreated cases eventually resolve. Second, prompt and adequate antimicrobial treatment may prevent the development of suppurative complications. Whereas data from the Netherlands in the 1990s suggested an increased incidence of acute mastoiditis with watchful waiting as the initial approach to treatment of AOM, other studies from several countries have contradicted these findings, demonstrating no increased incidence in acute mastoiditis with changes in recommendations for more conservative antimicrobial prescribing practices.

Given that most episodes of OM will spontaneously resolve, consensus guidelines have been published to assist clinicians who wish to consider a period of "watchful waiting" or observation before treating AOM with antibiotics (see Table 680.2 and Table 680.3; Fig. 680.6). The most important aspect of these guidelines is that close follow-up of the patient must be ensured to assess for lack of spontaneous resolution or worsening of symptoms and that patients should be provided with adequate analgesic medications (acetaminophen, ibuprofen) during the period of observation. When pursuing the practice of watchful waiting in patients with AOM, the certainty of the diagnosis, the patient's age, and the severity of the disease should be considered. For patients <2 years of age, it is recommended to treat all confirmed diagnoses of AOM. In patients, <6 months of age, even presumed episodes of AOM should be treated because of the increased potential of significant morbidity from infectious complications. In children between 6 and 24 months of age who have a questionable diagnosis of OM but severe disease, defined as temperature of >39°C (102°F), significant otalgia, or toxic appearance, antibiotic therapy is also recommended. Children in this age-group with a questionable diagnosis and nonsevere disease can be observed for a period of 2-3 days with close follow-up. In children older than 2 years of age, observation might be considered in all episodes of nonsevere OM or episodes of questionable diagnosis, whereas antibiotic therapy is reserved for confirmed, severe episodes of AOM. Information from Finland suggests that the "watchful waiting" or delayed treatment approach does not worsen the recovery from AOM or increase the complication rates.

Accurate diagnosis is the most crucial aspect of the treatment of OM. In studies using stringent criteria for diagnosis of AOM, the benefit of antimicrobial treatment is enhanced. In addition, subpopulations of patients clearly receive more benefit from oral antimicrobial therapy than others. *Younger children, children with otorrhea, and children with bilateral AOM have a significantly enhanced benefit from antimicrobial therapy in comparison with older children, children without otorrhea, or children with unilateral AOM.*

Table 680.1 Therapy for Otalgia in Acute Otitis Media

APPROACH	RECOMMENDATIONS
Acetaminophen, ibuprofen	Preferred therapy
Benzocaine, antipyrine (topical)	Brief, benefit over acetaminophen in patients older than 5 yr
Topical antibiotics (fluoroquinolones) with or without steroids for chronic suppurative otitis (perforated tympanic membrane)	Preferred treatment with ear canal cleaning; must culture
Homeopathic agents	Not recommended
Narcotic analgesia with codeine or analogs	Not recommended
Tympanostomy/myringotomy	Not recommended for initial approach; an option for otitis media unresponsive to antibiotic therapy

Bacterial Resistance

Persons at greatest risk of harboring resistant bacteria are those who are younger than 2 years of age; who are in regular contact with large groups of other children, especially in daycare settings; or who recently have received antimicrobial treatment. The development of resistant bacterial strains and their rapid spread have been fostered and facilitated by selective pressure resulting from extensive use of antimicrobial drugs, the most common target of which in children is OM. Many strains of each of the pathogenic bacteria that commonly cause AOM are resistant to commonly used antimicrobial drugs.

Although antimicrobial resistance rates vary between countries, in the United States approximately 40% of strains of nontypeable *H. influenzae*

Table 680.2 Recommendations for Initial Management for Uncomplicated Acute Otitis Media*

AGE	OTORRHEA WITH AOM*	UNILATERAL OR BILATERAL AOM* WITH SEVERE SYMPTOMS†	BILATERAL AOM* WITHOUT OTORRHEA	UNILATERAL AOM* WITHOUT OTORRHEA
6 mo to 2 yr	Antibiotic therapy	Antibiotic therapy	Antibiotic therapy	Antibiotic therapy or additional observation
≥2 yr	Antibiotic therapy	Antibiotic therapy	Antibiotic therapy or additional observation	Antibiotic therapy or additional observation‡

*Applies only to children with well-documented AOM with high certainty of diagnosis.
†A toxic-appearing child, persistent otalgia more than 48 hours, temperature ≥39°C (102.2°F) in the past 48 hours, or if there is uncertain access to follow-up after the visit.
‡This plan of initial management provides an opportunity for shared decision-making with the child's family for those categories appropriate for additional observation. If observation is offered, a mechanism must be in place to ensure follow-up and begin antibiotics if the child worsens or fails to improve within 48-72 hr of AOM onset.
NOTE: For infants younger than age 6 mo, a suspicion of AOM should result in antibiotic therapy.
AOM, Acute otitis media.
From Lieberthal AS, Carroll AE, Chonmaitree T, et al. The diagnosis and management of acute otitis media. *Pediatrics*. 2013;131:e964–e999, Table 4.

Table 680.3 Suggested Antibiotics for Treatment of Otitis Media and for Patients Who Have Failed First-Line Antibiotic Treatment

INITIAL IMMEDIATE OR DELAYED ANTIBIOTIC TREATMENT		ANTIBIOTIC TREATMENT AFTER 48-72 HR OF FAILURE OF INITIAL ANTIBIOTIC TREATMENT	
RECOMMENDED FIRST-LINE TREATMENT	**ALTERNATIVE TREATMENT (IF PENICILLIN ALLERGY OR SUSPICION OF β-LACTAMASE–PRODUCING ORGANISMS)**	**RECOMMENDED TREATMENT**	**ALTERNATIVE TREATMENT**
Amoxicillin (pathogens include *Pneumococcus, H. influenzae* non–type B, *Moraxella)*	Cefdinir	Amoxicillin-clavulanate	Ceftriaxone
or	*or*	*or*	
Amoxicillin-clavulanate Ceftriaxone	Cefpodoxime Ceftriaxone Levofloxacin (type I hypersensitivity to penicillin) Clindamycin + third generation cephalosporin (non–type I hypersensitivity to penicillin)	Ceftriaxone	Tympanocentesis*

ANTIBIOTIC DOSAGE

- Amoxicillin 80-90 mg/kg/day bid for 10 days**
- Amoxicillin-clavulanate (ratio 14:1) 80-90 mg/kg/day of amoxicillin component bid for 10 days**
- Ceftriaxone 50 mg/kg/day daily IM or IV for 1-3 days
- Cefdinir 14 mg/kg/day daily for 10 days**
- Cefpodoxime 10 mg/kg/day bid for 10 days**
- Levofloxacin 20 mg/kg/day bid if ≤5 yr for 10 days; 10 mg/kg/day bid if >5 yr for 10 days

*Tympanocentesis for those who fail second-line therapy.
**Durations of antibiotic therapy may vary based on symptom severity and patient age. For patients 2-5 yr old with mild or moderate disease severity, a 7-day course may be appropriate. For patients 6 yr and older, a 5- to 7-day course may be adequate for mild to moderate symptoms.
IM, Intramuscular; IV intravenous; bid, twice daily.

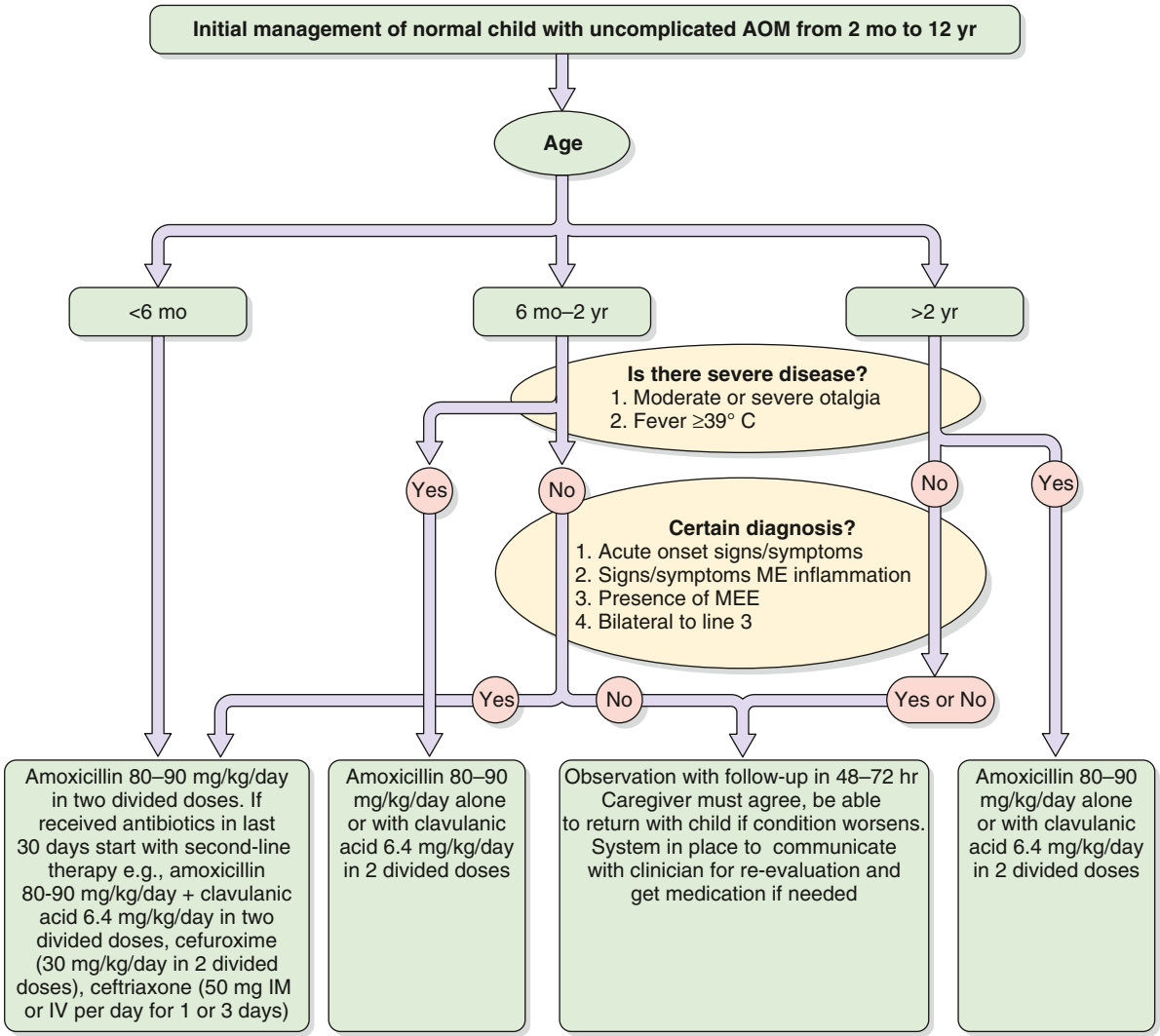

Fig. 680.6 Algorithm for management of acute otitis media. *(From Mazer BD: Otitis media. In: Leung DYM, Szefler SJ, Bonilla FA, et al., eds. Pediatric Allergy: Principles and Practices, 3 ed, Philadelphia: Elsevier; 2016: Fig. 25-3.)*

and almost all strains of *M. catarrhalis* are resistant to aminopenicillins (e.g., ampicillin and amoxicillin). In most cases the resistance is attributable to production of β-lactamase and can be overcome by combining amoxicillin with a β-lactamase inhibitor (clavulanate) or by using a β-lactamase–stable antibiotic. Occasional strains of nontypeable *H. influenzae* that do not produce β-lactamase are resistant to aminopenicillins and other β-lactam antibiotics by virtue of alterations in their penicillin-binding proteins. It is worth noting that bacterial resistance rates in Northern European countries where antibiotic use is not routine are lower (β-lactamase resistance in 6–10% of isolates) than in the United States.

In the United States, approximately 50% of strains of *S. pneumoniae* are penicillin-nonsusceptible, divided approximately equally between penicillin-intermediate and, even more difficult to treat, penicillin-resistant strains. A much higher incidence of resistance is seen in children attending daycare. Resistance by *S. pneumoniae* to the penicillins and other β-lactam antibiotics is mediated not by β-lactamase production but by alterations in penicillin-binding proteins. This mechanism of resistance can be overcome if higher concentrations of β-lactam antibiotics at the site of infection can be achieved for a sufficient time interval. Many penicillin-resistant strains of *S. pneumoniae* are also resistant to other antimicrobial drugs, including sulfonamides, macrolides, and cephalosporins. In general, as penicillin resistance increases, so also does resistance to other antimicrobial classes. Resistance to macrolides, including azithromycin and clarithromycin, by *S. pneumoniae* has increased, rendering these antimicrobials far less effective in treating AOM. One mechanism of resistance to macrolides also results in resistance to clindamycin, which otherwise is effective against resistant strains of *S. pneumoniae.* Unlike resistance to β-lactam antibiotics, macrolide resistance cannot be overcome by increasing the dose.

First-Line Antimicrobial Treatment

Amoxicillin remains the drug of choice for uncomplicated AOM under many circumstances because of its excellent safety record, relative efficacy, palatability, and low cost. Amoxicillin is the most efficacious of available oral antimicrobial drugs against both penicillin-susceptible and penicillin-nonsusceptible strains of *S. pneumoniae*. Increasing the dose from the traditional 40-45 mg/kg/day to 80-90 mg/kg/day will generally provide efficacy against penicillin-intermediate and some penicillin-resistant strains. This higher dose should be used particularly in children younger than 2 years old, in children who have recently received treatment with β-lactam drugs, and in children who are exposed to large numbers of other children because of their increased likelihood of an infection with a nonsusceptible strain of *S. pneumoniae*. A limitation of amoxicillin is that it may be inactivated by the β-lactamases produced by many strains of nontypeable *H. influenzae* and most strains of *M. catarrhalis*. However, episodes of AOM caused by these two pathogens often resolve spontaneously.

Allergies to penicillin antibiotics should be categorized into type I hypersensitivity, consisting of urticaria or anaphylaxis, and those that fall short of type I reactions, such as rash formation. For children with a non–type I reaction in which cross reactivity with cephalosporins is less of a concern, first-line therapy with cefdinir would be an appropriate choice. In children with a type I reaction or known sensitivity to cephalosporin antibiotics, there are far fewer choices. Resistance to trimethoprim-sulfamethoxazole by many strains of both nontypeable *H. influenzae* and *S. pneumoniae* and a reported high clinical failure rate in children with AOM treated initially with this antimicrobial argue against its use. Similarly, increasing rates of macrolide resistance argue against the efficacy of azithromycin. Because of concerns for arthropathy and damage to weight-bearing bones in animal models, fluoroquinolone use in children has been limited; fluoroquinolone use is recommended in certain clinical situations and/or when alternative therapies are deemed suboptimal. Early alternative management in allergic patients with tympanostomy tubes can allow for lessening of the severity of their disease and the use of topical antimicrobials.

Duration of Treatment

The duration of treatment of AOM has historically been set at 10 days, and most efficacy studies examining antimicrobial treatment in AOM have used this duration as a benchmark. Studies comparing shorter with longer durations of antimicrobial treatment have reported higher treatment failure rates in younger children, particularly those younger than 2 years old. Antibiotic courses shorter than 10 days, more severe disease, exposure to larger groups of other children, such as in daycare settings, and in children with a history of OME are also associated with higher treatment failure rates. The clinical practice guidelines published in 2013 by the American Academy of Pediatrics suggest shorter durations of antibiotic therapy may be adequate for treatment of AOM in children older than 2 years. However, compelling evidence substantiating the efficacy of shorter antibiotic courses to treat AOM in older children is limited.

Most patients improve within 72 hours; however, patients should not stop therapy. If there are persistent symptoms after 72 hours of therapy, the child should be reexamined for persistent OM or a complication such as mastoiditis (see Chapter 681).

Follow-Up

The principal goals of follow-up are to assess the outcome of treatment and to differentiate between inadequate response to treatment and early recurrence. The appropriate interval for follow-up should be individualized. Follow-up within days is advisable in the young infant with a severe episode or in a child of any age with continuing pain. Follow-up within 2 weeks is appropriate for the infant or young child who has been having frequent recurrences. At that point, the TM is not likely to have returned to normal, but substantial improvement in its appearance should be evident. In the child with only a sporadic episode of AOM and prompt symptomatic improvement, follow-up 1 month after initial examination is early enough, or in older children, no follow-up may be necessary. The continuing presence of MEE alone after an episode of AOM is not an indication for additional or second-line antimicrobial treatment. However, persisting MEE does warrant additional follow-up to ensure that the effusion resolves and does not lead to persisting hearing loss or other complications.

Unsatisfactory Response to First-Line Treatment

AOM is essentially a closed-space infection, and its resolution depends both on eradication of the offending organism and restoration of middle-ear ventilation. Factors contributing to unsatisfactory response to first-line treatment, in addition to inadequate antimicrobial efficacy, include poor compliance with treatment regimens, concurrent or intercurrent viral infection, persistent eustachian tube dysfunction and middle-ear under-aeration, reinfection from other sites or from incompletely eradicated middle-ear pathogens, and immature or impaired host defenses. The identification of biofilm formation in the middle ear of children with chronic OM also indicates that, in some children, eradication with standard antimicrobial therapy is likely to be unsuccessful. Despite these many potential factors, switching to an alternative or second-line drug is reasonable when there has been inadequate improvement in symptoms or in middle-ear status as reflected in the appearance of the TM or when the persistence of purulent nasal discharge suggests that the antimicrobial drug being used has less-than-optimal efficacy. Second-line drugs may also appropriately be used when AOM develops in a child already receiving antimicrobial therapy, or in an immunocompromised child, or in a child with severe symptoms whose previous experience with OM has been problematic.

Second-Line Treatment

When treatment of AOM with a first-line antimicrobial drug has proven inadequate, numerous second-line alternatives are available (see Table 680.3). Drugs chosen for second-line treatment should be effective against β-lactamase–producing strains of nontypeable *H. influenzae* and *M. catarrhalis* and against susceptible and most nonsusceptible strains of *S. pneumoniae.* Because high-dose amoxicillin (80-90 mg/kg/day) is effective against most strains of *S. pneumoniae* and because the addition of clavulanate extends the effective antibacterial spectrum of amoxicillin to include β-lactamase–producing bacteria, high-dose amoxicillin-clavulanate is particularly well-suited as a

second-line drug for treating AOM. The 14:1 amoxicillin-clavulanate formulation contains twice as much amoxicillin as the previously available 7:1 formulation. Diarrhea, especially in infants and young children, is a common adverse effect and usually is not severe enough to require cessation of treatment. Cefdinir has demonstrated efficacy in treatment, is generally well tolerated with respect to taste, and can be given as a once-daily regimen. The ability to also use cefdinir in most children with mild type I hypersensitivity reactions has further added to its favorable selection as a second-line agent. Intramuscular ceftriaxone has important limitations for use in young children; treatment entails both the pain of intramuscular injection and substantial cost, and the injection may need to be repeated once or twice at 2-day intervals to achieve the desired degree of effectiveness. Nonetheless, use of ceftriaxone is appropriate in severe cases of AOM when oral treatment is not feasible, in highly selected cases after treatment failure using orally administered second-line antimicrobials, or when highly resistant *S. pneumoniae* is found in aspirates obtained from diagnostic tympanocentesis.

Clarithromycin and azithromycin have only limited activity against nonsusceptible strains of *S. pneumoniae* and against β-lactamase–producing strains of nontypeable *H. influenzae*. Macrolide use also appears to be a major factor in causing increases in rates of resistance to macrolides by group A streptococcus and *S. pneumoniae*. Clindamycin is active against most strains of *S. pneumoniae,* including resistant strains, but is not active against nontypeable *H. influenzae* or *M. catarrhalis*.

Other antimicrobial agents that have been traditionally used in the management of AOM have such significant lack of effectiveness against resistant organisms that use seldom outweighs the potential side effects or complications. This includes cefaclor, loracarbef, cefixime, trimethoprim-sulfamethoxazole, and erythromycin-sulfisoxazole. Cefpodoxime has demonstrated reasonable effectiveness in some investigations but is generally poorly tolerated because of its taste.

ANTIMICROBIAL PROPHYLAXIS

In children who have developed frequent episodes of AOM, antimicrobial prophylaxis with subtherapeutic doses of an aminopenicillin or a sulfonamide has been used in the past to provide protection against recurrences of AOM (although not of OME). However, because of the increased incidence of resistant organisms and the contribution of antimicrobial usage to bacterial resistance, the risks of sustained antimicrobial prophylaxis clearly outweigh potential benefits.

Myringotomy and Tympanocentesis

Myringotomy is a long-standing treatment for AOM but is not commonly needed in children receiving antimicrobials. **Indications for myringotomy** in children with AOM include severe, refractory pain; hyperpyrexia; complications of AOM such as facial paralysis, mastoiditis, labyrinthitis, or central nervous system infection; and immunologic compromise from any source. Myringotomy should be considered as third-line therapy in patients who have failed two courses of antibiotics for an episode of AOM. In children with AOM in whom clinical response to vigorous, second-line treatment has been unsatisfactory, either diagnostic tympanocentesis or myringotomy is indicated to enable identification of the offending organism and its sensitivity profile. Either procedure may be helpful in effecting relief of pain. Tympanocentesis with culture of the middle-ear aspirate may also be indicated as part of the sepsis workup in very young infants with AOM who show systemic signs of illness such as fever, vomiting, or lethargy and whose illness accordingly cannot be presumed to be limited to infection of the middle ear. Performing tympanocentesis can be facilitated by use of a specially designed tympanocentesis aspirator. Studies reporting the use of strict, individualized criteria for the diagnosis of AOM that include office tympanocentesis with bacterial culture followed by culture-guided antimicrobial therapy demonstrate significant reduction in the frequency of recurrent AOM episodes and tympanostomy tube surgery. However, many primary care physicians do not feel comfortable performing this procedure, there is the potential for complications, and parents may view this procedure as traumatic. Often, children requiring this intervention have a strong enough history of recurrent OM to warrant the consideration of tympanostomy tube placement, so that the procedure can be performed under general anesthesia.

Early Recurrence After Treatment

Recurrence of AOM after apparent resolution may be caused by either incomplete eradication of infection in the middle ear or upper respiratory tract reinfection by the same or a different bacteria or bacterial strain. Recent antibiotic therapy predisposes patients to an increased incidence of resistant organisms, which should also be considered in choosing therapy, and, generally, initiating therapy with a second-line agent is advisable (see Table 680.3).

Myringotomy and Insertion of Tympanostomy Tubes

When AOM is recurrent, despite appropriate medical therapy, consideration of surgical management of AOM with tympanostomy tube insertion is warranted. Although this surgical procedure may improve the quality of life in patients with recurrent AOM, some studies have suggested the procedure is effective at reducing the rate of AOM, whereas others found postprocedure rates of AOM were no different than patients managed with antibiotics. Individual patient factors, including the risk profile, severity of AOM episodes, child's development and age, presence of a history of adverse drug reactions, concurrent medical problems, and parental wishes, will affect the timing of a decision to consider referral for this procedure. When a patient experiences three episodes of AOM in a 6-month period or four episodes in a 12-month period with one episode in the preceding 6 months, potential surgical management of the child's AOM should be discussed with the parents. Guidelines on tympanostomy tube placement indicate that if MEE is persistent in one or two ears and present at the time of evaluation by the otolaryngologist, then myringotomy is indicated. However, if MEE has cleared, the guidelines recommend holding off on myringotomy and offering observation unless there are additional considerations such as difficulty with tolerating antibiotic therapy (allergic concerns or other tolerance difficulties), severe episodes of acute OM, or other developmental considerations. Not infrequently, one or more of these additional considerations do affect a child's care.

AOM-Associated Tube Otorrhea

Although tympanostomy tubes may reduce the incidence of AOM in some children, patients with tympanostomy tubes may still develop AOM. One advantage of tympanostomy tubes in children with recurrent AOM is that if patients do develop an episode of AOM with a functioning tube in place, these patients will manifest purulent drainage from the tube. By definition, children with functioning tympanostomy tubes without otorrhea do not have bacterial AOM as a cause for a presentation of fever or behavioral changes and should not be treated with oral antibiotics. If tympanostomy tube otorrhea develops, ototopical treatment, and not oral antibiotics, should be considered as first-line therapy. With a functioning tube in place, the infection is able to drain, there is usually negligible pain associated with the infection, and the possibility of developing a serious complication from an episode of AOM is remote. Importantly, strict water precautions after tympanostomy tube placement do not appear to affect the occurrence of posttympanostomy otorrhea, and as such, water precautions are no longer recommended in children with myringotomy tubes. However, when otorrhea does occur, it is important to keep the ear canal dry while ototopical treatment is administered. The current quinolone otic drops approved by the U.S. Food and Drug Administration for use in the middle-ear space in children are formulated with ciprofloxacin/dexamethasone (Ciprodex) and ofloxacin (Floxin). The topical delivery of these otic drops allows them to use a higher antibiotic concentration than can be tolerated by administering oral antibiotics, and they have excellent coverage of even the most resistant strains of common middle-ear pathogens, as well as coverage of *S. aureus* and *Pseudomonas aeruginosa*. The high rate of success of these topical preparations, their broad coverage, the lower likelihood of their contributing to the

development of resistant organisms, the relative ease of administration, the lack of significant side effects, and the lack of ototoxicity make them the first choice for tube otorrhea. Oral antibiotic therapy should be reserved for cases of tube otorrhea that have other associated systemic symptoms, patients who have difficulty in tolerating the use of topical preparations, or, possibly, patients who have failed an attempt at topical otic drops. Despite these advantages of ototopical therapy, survey data have indicated that, compared with otolaryngologists, primary care practitioners are less likely to prescribe ototopicals as first-line therapy in tympanostomy tube otorrhea.

As a result of the relative ease in obtaining fluid for culture and the possibility of the development of fungal otitis, which has shown an increase with the use of broad-spectrum quinolone ototopicals, patients who fail topical therapy should also have culture performed. Other otic preparations are available; although these either have some risk of ototoxicity or have not received approval for use in the middle ear, many of these preparations were widely used before the development of the current quinolone drops and were generally considered reasonably safe and effective. In all cases of tube otorrhea, attention to aural toilet (e.g., cleansing the external auditory canal of secretions and avoidance of external ear water contamination) is important. In some cases, with very thick, tenacious discharge, topical therapy may be inhibited due to lack of delivery of the medication to the site of infection. Suctioning and removal of the secretions, often done through referral to an otolaryngologist, may be quite helpful. When children with tube otorrhea fail to improve satisfactorily with conventional outpatient management, they may require tube removal, hospitalization to receive parenteral antibiotic treatment, or both.

MANAGEMENT OF OTITIS MEDIA WITH EFFUSION

Management of OME depends on an understanding of its natural history and its possible complications and sequelae. Children with OME should be assessed for any baseline sensory, physical, cognitive, or behavioral factors that may portend risk of learning problems from MEE. Moreover, clinicians should evaluate developmentally at-risk children for OME at the time of diagnosis of an at-risk condition such as Down syndrome, autism, speech and language delay, permanent hearing loss, craniofacial syndromes, cleft palate, blindness, or global developmental delay and at 12-18 months of age (if diagnosed as being at risk before this time). However, children who do not have symptoms that could be attributed to OME, such as hearing difficulties, ear discomfort, balance (vestibular) problems, poor school performance, or behavioral problems, and are not at developmental risk should not be routinely screened for OME. When MEE persists for longer than 3 months, an age-appropriate hearing test and consideration of referral to an otolaryngologist are appropriate. In older children (generally older than 4 years) and depending on the expertise in the primary care office, hearing screening may be achieved in primary care. For any child who fails a hearing screening in the primary care office, referral to an otolaryngologist is warranted. In considering the decision to refer the patient for consultation, the clinician should attempt to determine the impact of the OME on the child and educate the family in this regard. Most cases of OME resolve without treatment within 3 months. For children with OME being managed expectantly, the guidelines for management of OME recommend examination should be performed at 3- to 6-month intervals, until the effusion is no longer present, significant hearing loss is identified, or structural abnormalities of the eardrum or middle ear are suspected. Although hearing loss may be of primary concern, OME causes several other difficulties in children that should also be considered. These include predisposition to recurring AOM, pain, disturbance of balance, and tinnitus. In addition, long-term sequelae that have been demonstrated to be associated with OME include pathologic middle-ear changes, atelectasis of the TM and retraction pocket formation, adhesive OM, cholesteatoma formation and ossicular discontinuity, and conductive and sensorineural hearing loss. Long-term adverse effects on speech, language, cognitive, and psychosocial development have also been demonstrated. This impact is related to the duration of effusion present, whether the effusion is unilateral or bilateral, the degree of underlying hearing loss, and other developmental and social factors affecting the child. In considering the impact of OME on development, it is especially important to take into consideration the overall presentation of the child. Although it is unlikely that OME causing unilateral hearing loss in the mild range will have long-term negative effects on an otherwise healthy and developmentally normal child, even a mild hearing loss in a child with other developmental or speech delays certainly has the potential to compound this child's difficulties (Table 680.4). At a minimum, children with OME persisting longer than 3 months deserve close monitoring of their hearing levels with skilled audiologic evaluation, frequent assessment of developmental milestones, including speech and language assessment, and attention paid to their rate of recurrent AOM.

Variables Influencing Otitis Media with Effusion Management Decisions

Patient-related variables that affect decisions on how to manage OME include the child's age, the frequency and severity of previous episodes of AOM and interval since the last episode, the child's current speech and language development, presence of a history of adverse drug reactions, concurrent medical problems, or risk factors such as daycare attendance, and parental wishes. In considering surgical management of OME with tympanostomy tubes, particular benefit is seen in patients with persisting OME punctuated by episodes of AOM, because the tubes may provide resolution of both conditions. Persistence of MEE after recurrent AOM (three episodes in 6 months or four in 12 months) may be treated with tympanostomy tube placement. Disease-related variables that most otolaryngologists consider in the treatment of OME include whether the effusion is unilateral or bilateral, the apparent quantity of effusion, the duration if known, the degree of hearing impairment, the presence or absence of other possibly related symptoms (such as tinnitus, vertigo, or disturbance of balance), and the presence or absence of mucopurulent or purulent rhinorrhea, which, if sustained for longer than 2 weeks, would suggest that concurrent nasopharyngeal or paranasal sinus infection is contributing to continuing compromise of middle-ear ventilation.

Medical Treatment

Antimicrobials have demonstrated limited efficacy in resolving OME, presumably because they help to eradicate nasopharyngeal infection, unapparent middle-ear infection, or both. The most significant effects of antibiotics for OME have been shown with treatment durations of 4 weeks and 3 months. However, with the risk of bacterial antimicrobial resistance, the small potential benefit of antimicrobial therapy is outweighed by the negative potential of treatment and is not recommended. Instead, antibiotic treatment should be limited to cases in which there is evidence of associated bacterial upper respiratory tract infection or untreated middle-ear infection. For this purpose, the most broadly effective drug available should be used as recommended for AOM.

Table 680.4 Sensory, Physical, Cognitive, or Behavioral Factors that Place Children Who Have Otitis Media with Effusion at an Increased Risk for Developmental Difficulties (Delay or Disorder)

Permanent hearing loss independent of otitis media with effusion
Suspected or diagnosed speech and language delay or disorder
Autism spectrum disorder and other pervasive developmental disorders
Syndromes (e.g., Down) or craniofacial disorders that include cognitive, speech, and language delays
Blindness or uncorrectable visual impairment
Cleft palate with or without associated syndrome
Developmental delay

From Rosenfeld RM, Shin JJ, Schwartz SJ, et al. American Academy of Otolaryngology-Head and Neck Surgery Foundation. Clinical practice guideline. Otitis media with effusion (update). *Otolaryngol Head Neck Surg.* 2016;154(2):201–214, Table 3.

The efficacy of corticosteroids in the treatment of OME has been demonstrated to be short term. Therefore the risk-to-benefit ratio for steroids is such that they are no longer recommended for treatment of OME. Antihistamine-decongestant combinations are not effective in treating children with OME and are not indicated. Antihistamines alone, decongestants alone, and mucolytic agents are also ineffective and not recommended. The risk profile for decongestants and antihistamines in children are such that, unless there is some other medical condition such as documented allergic disease for antihistamine therapy, these medications are contraindicated for OME treatment. Randomized controlled trials do not support the use of topical intranasal steroid sprays to treat the manifestations of eustachian tube dysfunction, and their use for OME resolution is also not recommended. Inflation of the eustachian tube by the Valsalva maneuver or other means has not demonstrated long-term efficacy. Other "alternative" therapies, including spinal manipulation, currently have no demonstrated efficacy or role in children with OME.

Myringotomy and Insertion of Tympanostomy Tubes

When OME persists despite an ample period of watchful waiting, generally 3-6 months or perhaps longer in children with unilateral effusion, consideration of surgical intervention with tympanostomy tubes is appropriate. Myringotomy alone, without tympanostomy tube insertion, permits evacuation of MEE and may sometimes be effective, but often the incision heals before the middle-ear mucosa returns to normal, and the effusion soon reaccumulates. Inserting a tympanostomy tube offers the likelihood that middle-ear ventilation will be sustained for at least as long as the tube remains in place and functional. Tympanostomy tubes have a variable duration of efficacy based on design. Tubes that are designed for a shorter duration, 6-12 months, have a lesser impact on disease-free middle-ear spaces in children. Some studies comparing the efficacy of tympanostomy tube types, including shorter-acting tubes, with watchful waiting provide a less helpful assessment of the differences between these approaches. Tubes that are somewhat longer acting, effective for 12-18 months, are generally more appropriate for most children undergoing tube placement. Regardless of type, tympanostomy tube placement nearly uniformly reverses the conductive hearing loss associated with OME. Occasional episodes of obstruction of the tube lumen and premature tube extrusion may limit the effectiveness of tympanostomy tubes, and tubes can also be associated with otorrhea. However, placement of tympanostomy tubes is generally quite effective in providing resolution of OME in children. Tympanostomy tubes generally extrude on their own but rarely require surgical removal after several years in place. Sequelae after tube extrusion include residual perforation of the eardrum, tympanosclerosis, localized or diffuse atrophic scarring of the eardrum (which may predispose to the development of a retraction pocket), residual conductive hearing loss, and cholesteatoma. The more serious of these sequelae are quite infrequent. Recurrence of MEE after the extrusion of tubes does develop, especially in younger children. However, most children without underlying craniofacial abnormalities require only one set of tympanostomy tubes. In developed countries, immunologic maturity and other developmental changes provide improved middle-ear health and resolution of chronic OME by the time of tube extrusion. However, in some populations, including Australian Aboriginal people, Indigenous Nations in the United States, and Alaskan Natives, even with an absence of craniofacial abnormalities, there is a preponderance of chronic OME; these patients should have increased follow-up after tube extrusion. Because persistent OME may clear spontaneously during the summer months, watchful waiting through the summer season may be advisable in children with OME who are otherwise well and without developmental or speech concerns. In considering surgical management of OME in children, primarily in those with bilateral disease and hearing loss, it has been demonstrated that placement of tympanostomy tubes results in a significant improvement in their quality of life.

Adenoidectomy

Adenoidectomy may reduce the risk of subsequent recurrences of both AOM and OME in older children who have undergone tube insertion and in whom, after extrusion of tubes, OM continues to be a problem. Efficacy appears to be independent of adenoid size and probably derives from removal of the focus of infection in the nasopharynx as a site of biofilm formation, chronic inflammation affecting eustachian tube function, and recurrent seeding of the middle ear via the eustachian tube. Current guidelines state that adenoidectomy should not be performed at the time of tympanostomy tube insertion in children younger than 4 years old unless a distinct indication exists (nasal obstruction, chronic adenoiditis). However, in children older than 4 years, one should recommend tympanostomy tubes, adenoidectomy, or both when surgery is performed for OME.

Complications of Acute Otitis Media

Most complications of AOM consist of the spread of infection to adjoining or nearby structures, the development of chronicity, or both. Suppurative complications are relatively uncommon in children in developed countries but occur not infrequently in disadvantaged children whose medical care is limited. The complications of AOM may be classified as either intratemporal or intracranial (Table 680.5).

Intratemporal Complications

Direct but limited extension of AOM leads to complications within the local region of the ear and temporal bone. These complications include dermatitis, TM perforation, chronic suppurative OM (CSOM), mastoiditis, petrositis, brain abscess, hearing loss, facial nerve paralysis, cholesteatoma formation, and labyrinthitis.

Infectious Dermatitis

This is an infection of the skin of the external auditory canal resulting from contamination by purulent discharge from the middle ear. The skin is often erythematous, edematous, and tender. Management consists of proper hygiene combined with systemic antimicrobials and ototopical drops as appropriate for treating AOM and tube otorrhea.

Tympanic Membrane Perforation

Rupture of the TM can occur with episodes of either AOM or OME. Although damage to the TM from these episodes generally heals spontaneously, chronic perforations can develop in a small number of cases and require further surgical intervention.

Chronic Suppurative Otitis Media

CSOM consists of persistent middle-ear infection with discharge through a TM perforation. The disease is initiated by an episode of AOM with rupture of the membrane. The mastoid air cells are invariably involved. The most common etiologic organisms are *P. aeruginosa* and *S. aureus;* however, the typical AOM bacterial pathogens may also be the cause, especially in younger children or in the winter months. Treatment is guided by the results of microbiologic investigation. If an associated cholesteatoma is not present, parenteral antimicrobial treatment combined with assiduous aural cleansing is likely to be successful in clearing the infection, but in refractory cases, tympanomastoidectomy can be required.

Mastoiditis

Mastoiditis is an important complication associated with OM (see Chapter 681).

Facial Paralysis

As it traverses the middle ear and mastoid bone, the facial nerve may be affected by adjacent infection. Facial paralysis occurring as a complication of AOM is uncommon and often resolves after myringotomy and parenteral antibiotic treatment. Facial paralysis in the presence of AOM requires urgent attention because prolonged infection can result in the development of permanent facial paralysis, which can have a devastating effect on a child. Facial paralysis in an infant or child requires complete and unequivocal examination of the TM and middle-ear space. Any difficulty in examination requires urgent consultation with an otolaryngologist. Any examination that demonstrates an ear abnormality also requires urgent referral to an otolaryngologist. If facial paralysis

Table 680.5 Manifestations of the Sequelae and Complications of Otitis Media

COMPLICATION	CLINICAL FEATURES
ACUTE	
Perforation with otorrhea	Immobile tympanic membrane secondary to visible perforation, exudate in ear canal
Acute mastoiditis with periostitis	Tenderness and erythema over mastoid process, no destruction of bony trabeculae
Acute mastoid osteitis	Destruction of bony trabeculae; tenderness and erythema over mastoid process coupled with outward displacement of pinna
Petrositis	Infection of perilabyrinthine cells; may present with otitis, paralysis of lateral rectus, and ipsilateral orbital or facial pain (Gradenigo syndrome)
Facial nerve palsy	Peripheral cranial nerve VII paralysis
Labyrinthitis	Vertigo, fever, ear pain, nystagmus, hearing loss, tinnitus, nausea and vomiting
Lateral (or transverse) sinus thrombosis	Headache, fever, seizures, altered states of consciousness, septic emboli
Meningitis	Fever, headache, nuchal rigidity, seizures, altered states of consciousness
Extradural empyema	Fever, headache, seizures, altered states of consciousness
Subdural empyema	Fever, headache, seizures, altered states of consciousness
Brain abscess	Fever, headache, seizures, altered states of consciousness, focal neurologic examination
Gradenigo syndrome	OM, sixth cranial nerve palsy, pain in first and second distribution of trigeminal nerve.
NONACUTE	
Chronic perforation	Immobile tympanic membrane secondary to perforation
Otitis media with effusion (OME)	Immobile, opaque tympanic membrane
Adhesive otitis	Irreversible conductive hearing loss secondary to chronic OME
Tympanosclerosis	Thickened white plaques may cause conductive hearing loss
Chronic suppurative otitis media	After acute otitis media with perforation, secondary infection with *Staphylococcus aureus, Pseudomonas aeruginosa*, or anaerobes develops, causing chronic otorrhea
Cholesteatoma	White, pearl-like destructive tumor with otorrhea arising near or within tympanic membrane; may be secondary to chronic negative middle ear pressure
Otitic hydrocephalus	Increased intracranial pressure secondary to AOM; signs and symptoms include severe headaches, blurred vision, nausea, vomiting, papilledema, diplopia (abducens paralysis)

AOM, Acute otitis media.
From Kliegman RM, Bordini BJ, Toth H, Basel D, eds. *Nelson Pediatric Symptom-Based Diagnosis*. Philadelphia: Elsevier; 2022: Table 5.6.

develops in a child with mastoid osteitis or with CSOM, mastoidectomy should be undertaken urgently.

Cholesteatoma

Cholesteatoma is a cystlike growth originating in the middle ear, lined by keratinized, stratified squamous epithelium and containing desquamated epithelium and/or keratin (see Chapter 678; Fig. 680.7).

Acquired cholesteatoma develops most often as a complication of long-standing chronic OM. The condition also may develop from a deep retraction pocket of the TM or as a consequence of epithelial implantation in the middle-ear cavity from traumatic perforation of the TM or insertion of a tympanostomy tube. Cholesteatomas tend to expand progressively, causing bony resorption, often extend into the mastoid cavity, and may extend intracranially with potentially life-threatening consequences. Acquired cholesteatoma commonly presents as a chronically draining ear in a patient with a history of previous ear disease. Cholesteatoma should be suspected if otoscopy demonstrates an area of TM retraction or perforation with white, caseous debris persistently overlying this area. Along with otorrhea from this area, granulation tissue or polyp formation identified in conjunction with this history and presentation should prompt suspicion of cholesteatoma. The most common location for cholesteatoma development is in the superior portion of the TM *(pars flaccida)*. Most patients also present with conductive hearing loss on audiologic evaluation. When cholesteatoma is suspected, otolaryngology consultation should be sought immediately. Delay in recognition and treatment can have significant long-term consequences, including the need for more extensive surgical treatment, permanent hearing loss, facial nerve injury, labyrinthine damage with loss of balance function, and intracranial extension. The required treatment for cholesteatoma is tympanomastoid surgery.

Congenital cholesteatoma is an uncommon condition generally identified in younger patients (Fig. 680.8). The etiology of congenital cholesteatoma is thought to be a result of epithelial implantation in the middle-ear space during otologic development in utero. Congenital cholesteatoma most commonly presents in the anterior-superior quadrant of the TM but can be found elsewhere. Congenital cholesteatoma appears as a discrete, white opacity in the middle-ear space on otoscopy. Unlike patients with acquired cholesteatoma, there is generally not a strong history of OM or chronic ear disease, history of otorrhea, or changes in the TM anatomy such as perforation or retraction. Similar to acquired cholesteatoma, many patients do have some degree of abnormal findings on audiologic evaluation, unless identified very early. Congenital cholesteatoma also requires surgical resection.

Labyrinthitis

Labyrinthitis occurs uncommonly as a result of the spread of infection from the middle ear and/or mastoid to the inner ear (see Chapter 682). Cholesteatoma or CSOM is the usual source. Symptoms and signs include vertigo, tinnitus, nausea, vomiting, hearing loss, nystagmus, and clumsiness. Treatment is directed at the underlying condition and must be undertaken promptly to preserve inner-ear function and prevent the spread of infection.

INTRACRANIAL COMPLICATIONS

Meningitis, epidural abscess, subdural abscess, focal encephalitis, brain abscess (see Chapters 643, 644, and 681), transverse (lateral) and sigmoid sinus thrombosis and otitic hydrocephalus each may develop as a complication of acute or chronic middle-ear or mastoid infection through direct extension, hematogenous spread, or thrombophlebitis. Bony destruction adjacent to the dura is often involved, and a cholesteatoma may be present. In a child with middle-ear or mastoid infection, the presence of any systemic symptom, such as high spiking fevers, headache, or lethargy of extreme degree or a finding of meningismus or of any abnormal central nervous system sign on physical examination should prompt suspicion of an intracranial complication.

When an intracranial complication is suspected, lumbar puncture should be performed only after imaging studies establish that there is no evidence of mass effect or hydrocephalus. In addition to examination

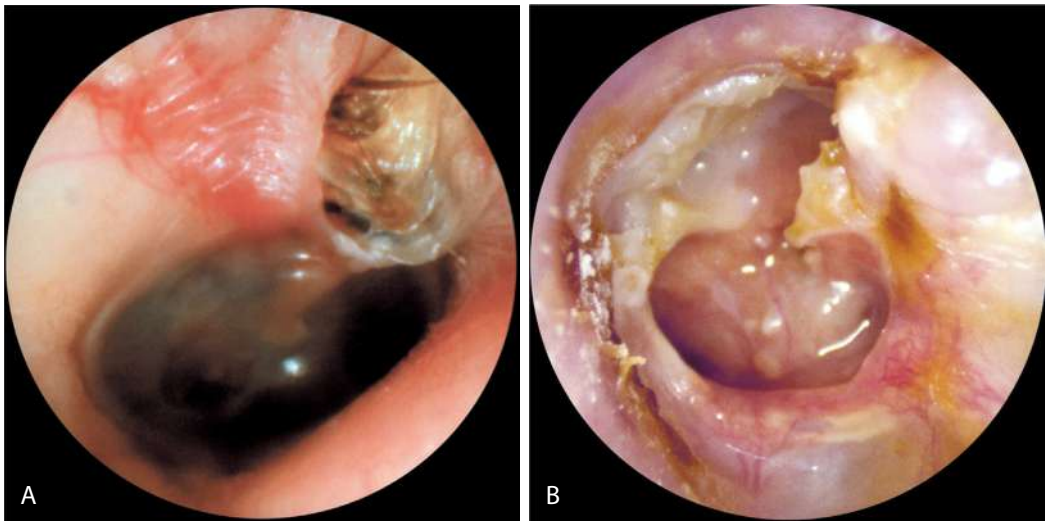

Fig. 680.7 A, Primary acquired cholesteatoma in the region of the pars flaccida with scutum erosion. B, Cholesteatoma developing at the margin of perforation (secondary acquired cholesteatoma) with secondary infection. *(From Chole RA, Sudhoff HH. Chronic otitis media, mastoiditis, and petrositis. In: Flint PW, Haughey BH, Lund VJ, et al, eds. Cummings Otolaryngology—Head and Neck Surgery, 5th ed. Philadelphia: Elsevier; 2010: Figs. 139-4, 139-5.)*

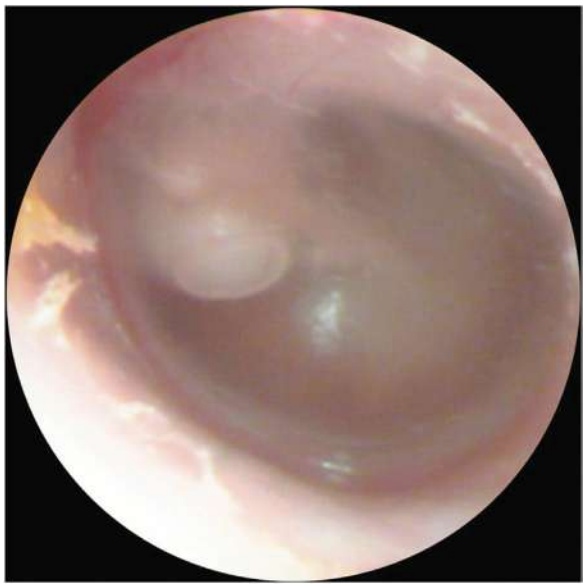

Fig. 680.8 Congenital chronic otitis media with cholesteatoma. *(From Chole RA, Sudhoff HH. Chronic otitis media, mastoiditis, and petrositis. In: Flint PW, Haughey BH, Lund VJ, et al., eds. Cummings Otolaryngology—Head and Neck Surgery, 5th ed. Philadelphia: Elsevier; 2010: Fig. 139-6.)*

of the cerebrospinal fluid, culture of middle-ear exudate obtained via tympanocentesis may identify the causative organism, thereby helping to guide the choice of antimicrobial medications. Myringotomy should be performed to permit middle-ear drainage. Concurrent tympanostomy tube placement is preferable to allow for continued decompression of the "infection under pressure" that is the causative event leading to intracranial spread of the infection.

Treatment of intracranial complications of OM requires urgent, otolaryngologic, and often, neurosurgical consultation, intravenous antibiotic therapy, drainage of mastoiditis, and tympanomastoidectomy in patients with coalescent mastoiditis. Many brain abscesses may be managed without drainage but require long-term broad-spectrum antibiotic therapy (see Chapter 644).

Lateral (transverse) or sigmoid sinus thrombosis may be complicated by dissemination of infected thrombi with resultant development of septic infarcts in various organs. Diagnosis is confirmed by MRI. Mastoidectomy may be required even in the absence of osteitis or coalescent mastoiditis, especially in the case of propagation or embolization of infected thrombi. In the absence of coalescent mastoiditis, sinus thrombosis can often be treated with tympanostomy tube placement, anticoagulation, and intravenous antibiotics. Otolaryngology consultation should be obtained before initiating this anticoagulation to coordinate the possible need for surgical intervention before anticoagulation.

Otitic hydrocephalus, a form of **idiopathic intracranial hypertension**, or **pseudotumor cerebri** (see Chapter 645), is an uncommon condition that consists of increased intracranial pressure without dilation of the cerebral ventricles occurring in association with acute or chronic OM or mastoiditis. The condition is commonly also associated with lateral sinus thrombosis, and the pathophysiology is thought to involve obstruction by thrombus of intracranial venous drainage into the neck, producing a rise in cerebral venous pressure and a consequent increase in cerebrospinal fluid pressure. Symptoms are those of increased intracranial pressure. Signs may include, in addition to evidence of OM, paralysis of one or both lateral rectus muscles and papilledema with or without visual acuity loss. MRI can confirm the diagnosis. Treatment measures include the use of antimicrobials and medications such as acetazolamide or furosemide to reduce intracranial pressure, mastoidectomy, repeated lumbar puncture, lumboperitoneal shunt, and ventriculoperitoneal shunt. If left untreated, otitic hydrocephalus may result in loss of vision secondary to optic atrophy.

Physical Sequelae

The physical sequelae of OM consist of structural middle-ear abnormalities resulting from long-standing middle-ear inflammation. In most instances, these sequelae are consequences of severe and/or chronic infection, but some may also result from the noninfective inflammation of long-standing OME. The various sequelae may occur singly or interrelatedly in various combinations.

Tympanosclerosis consists of whitish plaques in the TM and nodular deposits in the submucosal layers of the middle ear. The changes involve hyalinization with deposition of calcium and phosphate crystals. Uncommonly, there may be associated conductive hearing loss. In developed countries, probably the most common cause of tympanosclerosis is tympanostomy tube insertion.

Atelectasis of the TM is a descriptive term applied to either severe retraction of the TM caused by high negative middle-ear pressure or loss of stiffness and medial prolapse of the membrane from long-standing retraction or severe or chronic inflammation. A **retraction pocket** is a localized area of atelectasis. Atelectasis is often transient and usually unaccompanied by symptoms, but a deep retraction pocket may lead to erosion of the ossicles and adhesive otitis and may serve as the nidus of a cholesteatoma. For a deep retraction pocket, and for the unusual instance in which atelectasis is accompanied by symptoms

such as otalgia, tinnitus, or conductive hearing loss, the required treatment is tympanostomy tube insertion and, at times, tympanoplasty. Patients with persisting atelectasis and retraction pockets should have referral to an otolaryngologist.

Adhesive OM consists of proliferation of fibrous tissue in the middle-ear mucosa, which may, in turn, result in severe TM retraction, conductive hearing loss, impaired movement of the ossicles, ossicular discontinuity, and cholesteatoma. The hearing loss may be amenable to surgical correction.

Cholesterol granuloma is an uncommon condition in which the TM may appear to be dark blue secondary to middle-ear fluid of this color. Cholesterol granulomas are rare, benign cysts that occur in the temporal bone. They are expanding masses that contain fluids, lipids, and cholesterol crystals surrounded by a fibrous lining and generally require surgical removal. Tympanostomy tube placement will not provide satisfactory relief. This lesion requires differentiation from bluish middle-ear fluid, which can also rarely develop in patients with the more common OME.

Chronic perforation may rarely develop after spontaneous rupture of the TM during an episode of AOM or from acute trauma, but more commonly results as a sequela of CSOM or as a result of failure of TM closure after extrusion of a tympanostomy tube. Chronic perforations are generally accompanied by conductive hearing loss. Surgical repair of a TM perforation is recommended to restore hearing, prevent infection from water contamination in the middle-ear space, and prevent cholesteatoma formation. Chronic perforations are almost always amenable to surgical repair, usually after the child has been free of OM for an extended period.

Permanent **conductive hearing loss** (see Chapter 677) may result from any of the conditions just described. Rarely, permanent sensorineural hearing loss may occur in association with acute or chronic OM, secondary to spread of infection or products of inflammation through the round window membrane, or as a consequence of suppurative labyrinthitis.

POSSIBLE DEVELOPMENTAL SEQUELAE

Permanent hearing loss in children has a significant negative impact on development, particularly in speech and language. The degree to which OM affects long-term development in children is difficult to assess, and there have been conflicting studies examining this question. Developmental impact is most likely to be significant in children who have greater levels of hearing loss, hearing loss that is sustained for longer periods, or hearing loss that is bilateral and in children who have other developmental difficulties or risk factors for developmental delay (see Table 680.4).

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Chapter 681

Acute Mastoiditis

Brittany Player

Mastoiditis, a suppurative infection of the mastoid air cell system, is the most common infectious complication of acute otitis media (AOM), typically affecting younger children. Coalescent mastoiditis occurs when the suppurative infection leads to bony breakdown of the fine bony septa separating individual mastoid air cells.

ANATOMY

The temporal bone forms a portion of the skull base and has multiple complex anatomic functions. The mastoid process is a pyramid-shaped outgrowth of the temporal bone. The inferior extent is attached to the sternocleidomastoid muscle. The mastoid process borders the middle cranial fossa, posterior cranial fossa, and sigmoid sinus. It is composed of a system of interlinked mucosa-lined air cells that communicate with the middle ear space and contains the fallopian canal, which includes the facial nerve, the chorda tympani supplying taste to the anterior two thirds of the tongue, and the semicircular canal system. Because the mastoid cavity is anatomically adjacent to the meninges, brain, venous sinuses of the brain, facial nerve, and cervical lymph nodes, mastoiditis often accompanies or precedes intracranial complications of AOM (see Chapter 680).

EPIDEMIOLOGY

In the preantibiotic era, acute mastoiditis was a much more common complication of AOM with high rates of intracranial infectious complications, morbidity, and mortality. Currently, mastoiditis is a rare complication, occurring in approximately 1-4 cases per 100,000 population of children <2 years old and less commonly among older children. A multicenter study with 223 consecutive cases of acute mastoiditis reported 28% of patients were younger than 1 year old, 38% of patients were between 1 and 4 years old, 22% of patients were between 4 and 8 years old, and 8% of patients were between 8 and 18 years old. Some studies reported decreased incidence of acute mastoiditis after introduction of the 7-valent pneumococcal conjugate vaccine (PCV7), whereas others reported no change or nominal increases. One study reported a sharp decrease in acute mastoiditis beginning in 2010, which coincided with licensure and widespread use of the 13-valent pneumococcal conjugate vaccine (PCV13). Another study, which included data from eight hospitals, found that the proportion of PCV13 serotypes isolated from cases of mastoiditis decreased from 50% in 2011 to 29% in 2013, with most of the decrease attributable to decreases in serotype 19A. Changes in rates of mastoiditis are likely related to changing incidence of AOM in response to pneumococcal conjugate vaccines. Other factors influencing the occurrence of mastoiditis include rates of antibiotic prescriptions for AOM, access to healthcare, and rates of antimicrobial resistance. Whereas groups in countries such as the Netherlands and Iceland reported adherence to a watchful waiting strategy for treatment of AOM, this resulted in slightly increased rates of acute mastoiditis compared with countries where antibiotics are routinely used to treat AOM. Other studies from several countries have contradicted these findings, demonstrating no increased incidence in acute mastoiditis, with changes in recommendations for more conservative antimicrobial prescribing practices. Despite large differences in antibiotic prescription rates in different countries, because of the overall low incidence of acute mastoiditis, the number of children needed to be treated with antibiotics to prevent one case of acute mastoiditis ranges from 2,500 to 4,800. Some studies have reported an increase in incidence, which has correlated with an increase in infections with drug-resistant bacteria. All-cause mortality among children with mastoiditis is 0.03%.

MICROBIOLOGY

Streptococcus pneumoniae remains the most common pathogen cultured from cases of acute mastoiditis (Table 681.1). After introduction of PCV7, pneumococcal serotype 19A was commonly associated with acute mastoiditis. This serotype is frequently resistant to

Table 681.1 Etiology of Acute Mastoiditis

BACTERIA	FREQUENCY
Streptococcus pneumoniae	10–51%
Streptococcus pyogenes	0–12%
Staphylococcus aureus	2–10%
Pseudomonas aeruginosa	10%
Haemophilus influenzae	2–3%
No growth	20–40%

penicillin and macrolide antibiotics. PCV13 use has been associated with fewer serotype 19A infections overall; its impact on the etiology of mastoiditis is less clear. Other bacteria commonly cultured include *Streptococcus pyogenes, Staphylococcus aureus, Pseudomonas aeruginosa, Fusobacterium necrophorum,* and *Haemophilus influenzae*. *P. aeruginosa* is more likely in patients with chronic otitis media and/or cholesteatoma, older children, and those with previous tympanostomy tubes, though higher rates of *P. aeruginosa* recovery should suggest consideration of the method of sample collection when interpreting this culture result.

CLINICAL MANIFESTATIONS

Acute mastoiditis and AOM present similarly in children. Ninety-seven percent of children with an acute mastoiditis have a coexisting AOM on the affected side. The remaining 3% of children with acute mastoiditis either had a serous middle ear effusion at the time of presentation or had a history of AOM within the past 2 weeks. Other clinical manifestations include protrusion of the ear (87%), retroauricular swelling and tenderness (67%), retroauricular erythema (87%), fever (60%), otalgia, and hearing loss (Tables 681.2 and 681.3). Prolonged symptoms during the treatment of AOM may suggest concurrent mastoiditis. Children with acute mastoiditis were less likely to have bilateral infection. Some children do not have external signs of infection.

DIAGNOSIS AND IMAGING

Acute mastoiditis is usually diagnosed based on history and clinical findings (see Table 681.2). CT scan of the temporal bone can confirm the diagnosis, whereas head CT can identify intracranial complications (see Chapter 680), including epidural abscess or subdural empyema. Findings of acute mastoiditis include bony demineralization, loss of bony septations in the mastoid cavity (Fig. 681.1), and, occasionally, subperiosteal abscess (Fig. 681.2). CT scans have the advantage of being readily available in most emergency rooms, can quickly evaluate for intracranial complications, and can identify whether there is bony destruction or a drainable fluid collection. Contrast-enhanced CT scan or MRI allows evaluation for vascular thrombosis (Fig. 681.3) and abscess formation. MRI is generally reserved for patients in whom there is a suspected intracranial complication (Figs. 681.4 and 681.5). Incidental detection of mastoid air cell opacification occurs in more than 20% of children (and in 40% of children <2 years old) undergoing MRI for other reasons, so imaging findings must be interpreted in the appropriate clinical context.

There is a limited role for ultrasound in the diagnosis of acute mastoiditis. Ultrasound can be used as a screening test when a postauricular subperiosteal abscess is suspected due to clinical findings such as protrusion of the pinna and retroauricular erythema. If there is a fluid collection on ultrasound or concern for a defect in the cranial vault, further imaging with a CT and/or MRI would be recommended. Because ultrasound cannot identify intracranial complications, its use must be limited to a highly selected patient population.

Table 681.2 Diagnosis of Acute Mastoiditis

Fever, otalgia, postauricular swelling plus redness
- Older child: ear up and out
- Infant: ear down and out

Tympanic membrane: acute otitis media
Radiograph: mastoid air cells coalescent or clouded
Computed tomography, magnetic resonance imaging, or bone scan as needed

From Wald ER, Conway JH. Mastoiditis. In: Long SS, Prober CG, Fischer M, eds. *Principles and Practice of Pediatric Infectious Diseases,* 5th ed. Philadelphia: Elsevier; 2018: p. 227.

MANAGEMENT

Acute mastoiditis is a rare complication of AOM, and there is a large degree of overlap between the presentations of children with both disease processes. For the pediatrician confronted with a majority of uncomplicated AOM, it is difficult to decide when to initiate a more extensive evaluation. Any time there is a purulent middle-ear effusion along with postauricular findings, acute mastoiditis needs to be in the differential diagnosis. In general, children with acute mastoiditis will appear sicker than children with uncomplicated AOM, and many of them have already failed to respond to appropriate antibiotic therapy for AOM. Focal neurologic deficits, including facial paresis, in a child with AOM or mastoiditis suggest intracranial spread of infection. In a child with suspected mastoiditis, it is critical to document normal facial nerve function at the time of the initial exam so that if this complication does develop during treatment, the treating team is able to document this complication.

Table 681.3 Differential Diagnosis of Postauricular Involvement of Acute Mastoiditis with Periostitis/Abscess

	POSTAURICULAR SIGNS AND SYMPTOMS					
DISEASE	**CREASE***	**ERYTHEMA**	**MASS**	**TENDERNESS**	**EXTERNAL CANAL INFECTION**	**MIDDLE-EAR EFFUSION**
Acute mastoiditis with periostitis	May be absent	Yes	No	Usually	No	Usually
Acute mastoiditis with subperiosteal abscess	Absent	Maybe	Yes	Yes	No	Usually
Periostitis of pinna with postauricular extension	Intact	Yes	No	Usually	No	No
External otitis with postauricular extension	Intact	Yes	No	Usually	Yes	No
Postauricular lymphadenitis	Intact	No	Yes†	Maybe	No	No

*Postauricular crease (fold) between pinna and postauricular area.
†Circumscribed.
From Bluestone CD, Klein JO, eds. *Otitis Media in Infants and Children,* 3rd ed. Philadelphia: WB Saunders; 2001: p. 333.

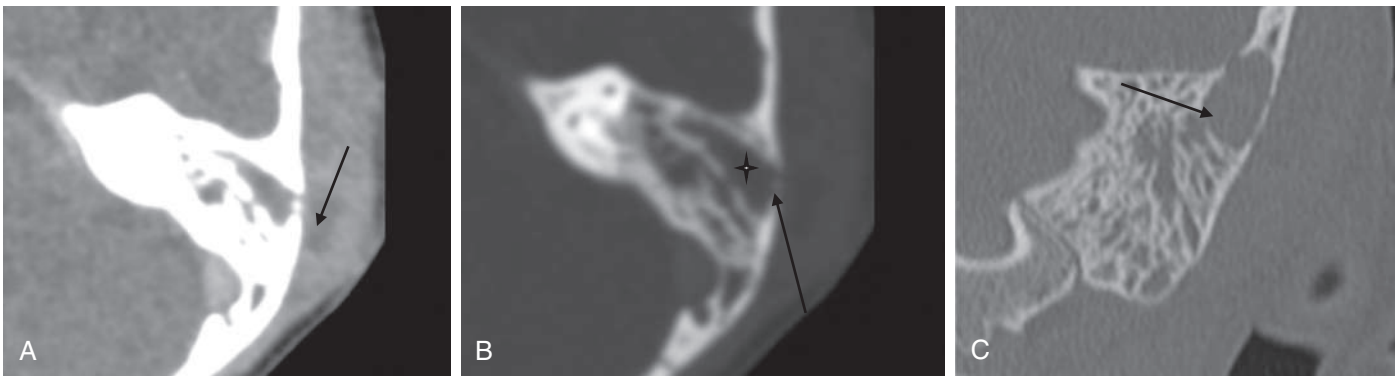

Fig. 681.1 Coalescent mastoiditis with subperiosteal abscess formation. **A** and **B**, Axial CT images with soft tissue windows and bone windows, respectively. *Arrow* in **A** points to the subperiosteal abscess. *Star* in **B** shows the loss of bony septations in the mastoid cavity, and the *arrow* points to the erosion of the bony cortex. **C**, Coronal image shows demineralization of the mastoid tegmen abutting the middle cranial fossa, a precursor to epidural abscess formation.

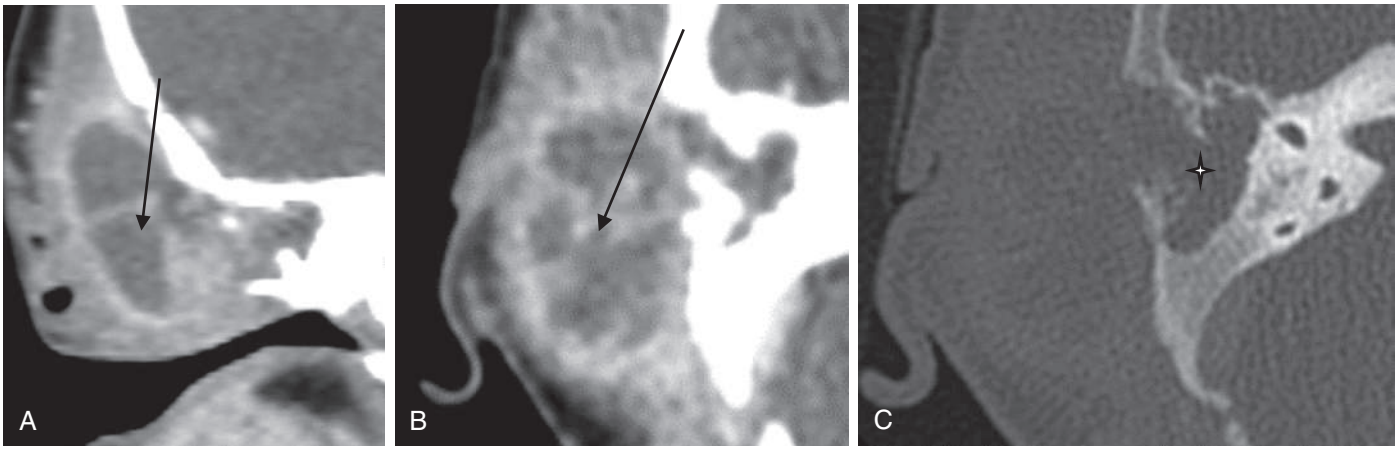

Fig. 681.2 Advanced case of coalescent mastoiditis with subperiosteal abscess formation. Axial (**A**) and coronal (**B**) images; *arrow* points to the subperiosteal abscess **C**, Extensive loss of bony septations in the mastoid cavity (*star).*

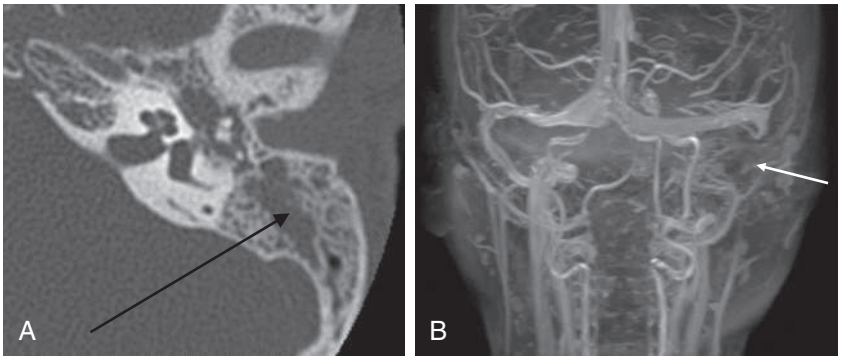

Fig. 681.3 **A**, Axial CT scan with bone windows shows opacification of the mastoid air cells, a small region of coalescence (*arrow),* and opacification of the middle ear space. **B**, CT venogram with a sigmoid sinus thrombosis. The *arrow* points to the area where a patent sigmoid sinus should be present.

Complete blood count typically reveals leukocytosis with neutrophil predominance. C-reactive protein is often highly elevated. If otorrhea is present, implying a perforated tympanic membrane, the fluid should be sent for Gram stain and culture. Blood culture should be considered in any child appearing toxic. For children with postauricular findings consistent with acute mastoiditis, admission to the hospital for intravenous antibiotic therapy and serial exams is recommended.

In highly selected cases, ultrasound may be helpful to differentiate postauricular erythema from a postauricular abscess and avoids the risk of ionizing radiation exposure. However, ultrasound is not as sensitive as CT scanning and will underdiagnose postauricular abscess formation and will provide no information as to whether there is an intracranial complication present such as a brain abscess. Some authors advocate deferring CT scanning in patients with clinically suspected acute mastoiditis and without focal neurologic

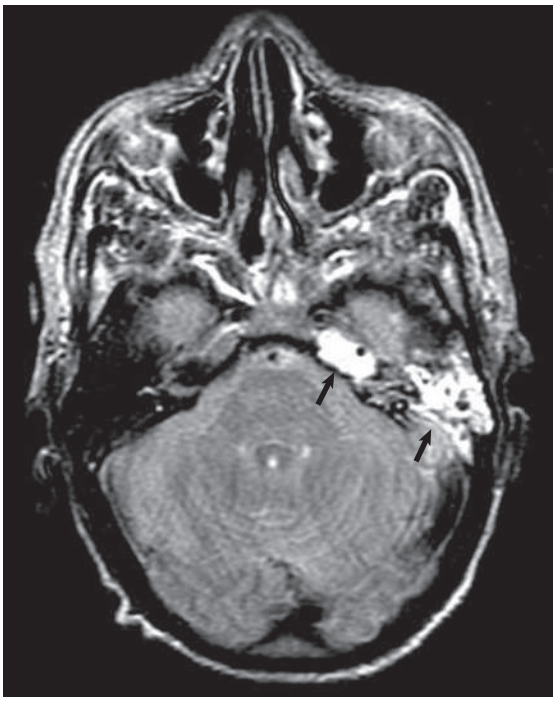

Fig. 681.4 Axial T2-weighted MRI showing left mastoiditis and petrous apicitis *(arrows)* as high signal in mastoid and petrous apex. *(From Budenz CL, El-Kashlan HK. Complications of temporal bone infections. In: Flint PW, Francis HW, Haughey BH, et al, eds. Cummings Otolaryngology Head & Neck Surgery, 7th ed., vol. 2. Philadelphia: Elsevier; 2021: Fig 141.17A.)*

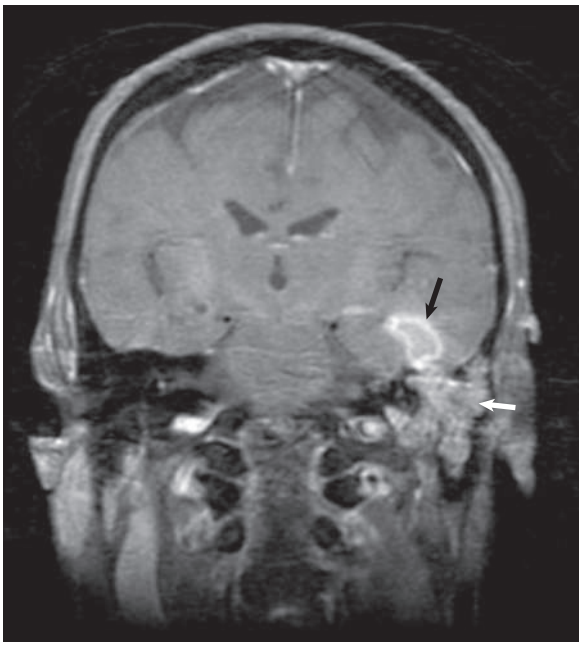

Fig. 681.5 Coronal enhanced T1-weighted MRI of the patient showing enhancing tissue in left mastoid *(white arrow)* and temporal lobe abscess with enhancing capsule *(black arrow)*. *(From Budenz CL, El-Kashlan HK. Complications of temporal bone infections. In: Flint PW, Francis HW, Haughey BH, et al, eds. Cummings Otolaryngology Head & Neck Surgery, 7th ed., vol. 2. Philadelphia: Elsevier; 2021: Fig 141.17D)*

findings to allow for an initial 24- to 48-hour period of inpatient intravenous antibiotic therapy. If there is any concern about the possibility of an intracranial complication, a contrasted CT scan is the most sensitive readily available test and should be ordered upon presentation.

Antibiotic therapy should initially be administered intravenously. Empiric antibiotic selection may include a β-lactam/β-lactamase inhibitor combination (e.g., ampicillin-sulbactam, piperacillin-tazobactam) or third-generation cephalosporin (e.g., cefotaxime, ceftriaxone). In children with chronically draining ears or concern for cholesteatoma, there is an increased incidence of gram-negative infection, and coverage should include antibiotics with activity against *Pseudomonas* spp. (e.g., ceftazidime, cefepime). If intracranial infection is suspected, broader-spectrum antimicrobial coverage (e.g., vancomycin plus a third-generation cephalosporin) should be initiated. In cases of uncomplicated acute mastoiditis (e.g., absence of intracranial complications or localized abscess formation), a 24- to 48-hour trial of intravenous antibiotics may yield clinical improvement without surgical intervention. Therapy should be adjusted when cultures and susceptibilities are available. The total duration of therapy is 4 weeks, with transition from intravenous to oral therapy at discharge for those without intracranial complications. The optimal duration of intravenous therapy is unknown, but some experts recommend a minimum of 7-10 days of intravenous therapy before oral transition, whereas others transition once the patient demonstrates clinical improvement and surgical intervention is no longer required.

Otolaryngology consultation can be helpful to assist with management and to determine whether surgical intervention would be beneficial. Many patients will benefit from tympanostomy tube placement at the time of the acute infection to allow localized ototopical antibiotic treatment and aspiration of middle-ear fluid for culture and sensitivity. In a patient with an additional extracranial complication such as facial paresis, drainage of the middle-ear space with placement of a tympanostomy tube is required and should take place urgently. A small group of patients may necessitate mastoidectomy—surgical removal of diseased bone and granulation tissue in the mastoid cavity. At the time of surgery, a drain is often placed to allow purulent secretions an egress. Indications for mastoidectomy include coalescent mastoiditis, postauricular abscess formation, infectious intracranial complication, and failure to respond to appropriate IV antibiotics. When intracranial complications occur or there are mental status changes, evaluation by otolaryngology and neurosurgery and emergent mastoidectomy are indicated. Most children with mastoiditis make a full recovery. Long-term otologic complications like sensorineural or conductive hearing loss are uncommon. A posttreatment audiogram is often obtained to evaluate the hearing status after an infection.

SPECIAL SITUATIONS

When treating acute mastoiditis, several uncommon situations require particular attention. Selecting empiric antibiotics for unvaccinated and undervaccinated children is challenging, and in this patient population, it is especially important to obtain a sample of middle-ear fluid for Gram stain and culture to guide antibiotic therapy. There is an increased incidence of acute mastoiditis in children with autism spectrum disorder. Immunocompromised patients should be treated with more aggressive and prolonged courses of antibiotic; they may benefit from more aggressive surgical treatment to remove infected tissue. Sigmoid sinus thrombosis can occur secondary to acute mastoiditis. If this does occur, in addition to the standard treatment for acute mastoiditis, consideration should be given to involving hematology and for administering systemic anticoagulation. Otitic hydrocephalus, which is elevated intracranial pressure after middle-ear infection, manifests with emesis, headache, and visual impairment and is associated with lateral or sigmoid sinus thrombosis; management requires antibiotics, anticoagulation, treatment of increased intracranial pressure, and surgical decompression with drainage of the mastoid infection.

Children with **cochlear device implantation** may have a 3.5% incidence of acute mastoiditis. Despite having a foreign body present in the middle ear and inner ear space, the majority of these

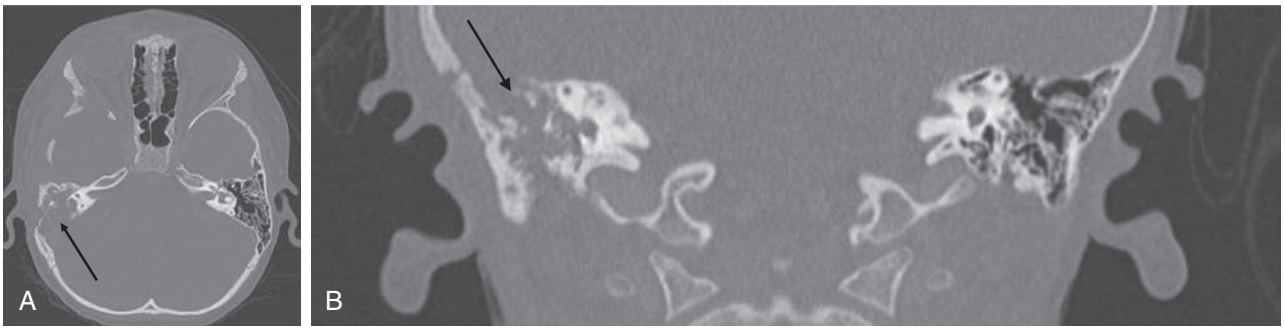

Fig. 681.6 Axial (A) and coronal (B) CT scans of a patient with Langerhans cell histiocytosis of the right temporal bone. A, Opacification of the mastoid with loss of bony septations and erosion of the bone separating the cranial fossa from the mastoid cavity *(arrow)*. B, Bony erosion caused by the tumor and the erosion of the mastoid tegmen *(arrow)*.

acute mastoiditis cases can be managed with tympanostomy tube placement, intravenous antibiotic therapy, and incision and drainage of an abscess without removal of the device.

Although very rare, benign and malignant tumors can affect the temporal bone of children. The presentation mimics that of chronic otitis media and chronic mastoiditis, and this often leads to a delay in diagnosis. Hearing loss, otalgia, and otorrhea are common symptoms. The main differentiating factor is the protracted course of otorrhea and refractory nature of symptoms despite appropriate medical therapy. Aural polyps or a mass lesion may be present on physical exam. Potential causes include rhabdomyosarcoma, nonrhabdomyosarcomatous sarcoma (including chondrosarcoma, chordoma, osteosarcoma, Ewing sarcoma, fibrosarcoma, angiosarcoma, and chloroma), Langerhans cell histiocytosis (formerly histiocytosis X) (Fig. 681.6), lymphoma, and metastasis.

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Chapter 682

The Inner Ear and Diseases of the Bony Labyrinth

Joseph Haddad Jr.

Genetic factors can affect the anatomy and function of the inner ear. Infectious agents, including viruses, bacteria, and protozoa, also can cause abnormal function, most commonly as sequelae of congenital infection (see Table 677.2) or bacterial meningitis (see Chapter 643.1). Other acquired diseases of the labyrinthine capsule include otosclerosis, osteopetrosis, Langerhans cell histiocytosis (see Chapter 556.1), fibrous dysplasia, and other types of bony dysplasia. All of these can cause both conductive hearing loss (CHL) and sensorineural hearing loss (SNHL), as well as vestibular dysfunction.

OTHER DISEASES OF THE INNER EAR

Labyrinthitis (also called **vestibular neuritis**) may be a complication of direct spread of infection from acute or chronic otitis media or mastoiditis and also can complicate bacterial meningitis as a result of organisms entering the labyrinth through the internal auditory meatus, endolymphatic duct, perilymphatic duct, vascular channels, or hematogenous spread. Clinical manifestations of vestibular neuritis can include a sudden onset of rotatory vertigo, dysequilibrium, postural imbalance (furniture walking) with falls to the affected side, deep-seated ear pain, nausea, vomiting, and spontaneous horizontal (occasionally rotary) nystagmus.

The dizziness may last a few days, but balance issues, particularly after rapid head movements toward the affected ear, may last for months. Vestibular neuritis is usually unilateral and is not associated with other neurologic defects; subjective hearing loss is unusual in vestibular neuritis. If hearing loss is present, idiopathic SNHL should be considered, as well as classical labyrinthitis (vestibular and cochlear nerves). Treatment of vestibular neuritis may include prednisone and vestibular rehabilitative exercises. Recurrent episodes should suggest another diagnosis such as vestibular migraine or benign paroxysmal positional vertigo.

In children, viral labyrinthitis is often associated with hearing loss. **Acute serous labyrinthitis**, characterized by mild symptoms of vertigo and hearing loss, most commonly develops secondary to middle-ear infection without direct invasion. **Acute suppurative labyrinthitis**, characterized by abrupt, severe onset of these symptoms, may be caused by bacterial meningitis or acute middle-ear or mastoid infection via a dehiscent horizontal semicircular canal. In these latter cases, a cholesteatoma is almost always present. Treatment of acute infectious labyrinthitis includes antimicrobial agents in cases of bacterial infection or antiviral agents (acyclovir, valacyclovir) in cases of herpes zoster oticus. Oral corticosteroids reduce labyrinthine inflammation and may prevent sequelae. A short course (≤3 days) of vestibular suppressants (dimenhydrinate 1-2 mg/kg) alleviates acute symptoms such as nausea. If it is secondary to otitis media, otologic surgery may be required to remove underlying cholesteatoma or drain the middle ear and mastoid. **Chronic labyrinthitis**, most often associated with cholesteatoma, manifests with SNHL and vestibular dysfunction that develops over time; surgery is required to remove the cholesteatoma. Chronic labyrinthitis also occurs uncommonly secondary to long-standing otitis media, with the slow development of SNHL, usually starting in the higher frequencies, and possibly with vestibular dysfunction. In addition, and more commonly, children with chronic middle-ear fluid often are unsteady or off balance, a situation that improves immediately when the fluid resolves.

Vertigo and dizziness are common among older children and adolescents. Benign **paroxysmal vertigo**, the most common cause of vertigo in pediatric patients, is characterized by short periods of vertigo or dizziness lasting seconds to a few minutes and associated with imbalance and nystagmus; tinnitus or hearing loss is unusual. **Basilar/vestibular migraine** is a common cause of episodic vertigo or dizziness and is associated with headache (50–70% of patients), rotary or to-and-fro nystagmus, and sensitivity to noise and bright light (see Chapter 635.1). **Benign paroxysmal positional vertigo** is less common in young children and more common with increasing age into adulthood. Particles form in the semicircular canals (canalithiasis), most often the

posterior canal; symptoms occur with position changes of the head and may last seconds to minutes. Vertigo and nystagmus may be demonstrated by changing position (sitting to lying down on the right or left). Treatment involves canalith repositioning maneuvers to shift the debris from the canals into the utricle. Additional etiologies of vertigo include vestibular migraine, trauma, superior semicircular canal dehiscence, isolated autoimmune inner ear disease or syndrome-associated autoimmune diseases including Cogan syndrome (interstitial keratitis, vertigo, hearing loss), Vogt-Koyanagi-Harada syndrome (uveomeningitis, vitiligo, vertigo, hearing loss), Susac syndrome (microangiopathy with retinopathy, encephalopathy, deafness), granulomatosis with polyangiitis, mixed connective tissue disease, and, most rarely, CNS tumor.

Otosclerosis, an autosomal dominant disease that affects only the temporal bones, causes abnormal bone growth that can result in fixation of the stapes in the oval window, leading to progressive hearing loss. In one series in North America, otosclerosis was found in 0.6% of temporal bones of children younger than 5 years of age and 4% of those ages 5-18 years. The hearing loss is usually conductive at first, but SNHL can develop. Females are affected most commonly, with onset of otosclerosis in teenagers or young adults, often associated with pregnancy. Corrective surgery to replace the stapes with a mobile prosthesis often is successful.

Osteogenesis imperfecta is a systemic disease that can involve both the middle and inner ears (see Chapter 742). Hearing loss occurs in approximately 20% of young children and as many as 50% of adults by the age of 50 with this disease. The hearing loss most commonly is conductive but can be sensorineural or mixed. Etiologies of hearing loss include otosclerosis, ossicle fractures, or neural degeneration. If the hearing loss is severe enough, a hearing aid may be a preferable alternative to surgical correction of the fixed stapes, because stapedectomy in children with osteogenesis imperfecta can be technically very difficult, and the disease and the hearing loss may be progressive.

Osteopetrosis, a very uncommon skeletal dysplasia, can involve the temporal bone, including the middle ear and ossicles, usually resulting in a moderate to severe CHL. Recurrent facial nerve paralysis also can occur because of excess bone deposition; with each recurrence, less facial function might return (see Chapter 740).

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Chapter 683

Traumatic Injuries of the Ear and Temporal Bone

Joseph Haddad Jr.

AURICLE AND EXTERNAL AUDITORY CANAL

Auricle trauma is common in certain sports. Hematoma, with accumulation of blood between the perichondrium and the cartilage, can follow trauma to the pinna and is especially common in teenagers involved in wrestling or boxing. Prompt drainage of a hematoma can prevent irreversible damage. Immediate needle aspiration or, when the hematoma is extensive or recurrent, incision and drainage and a pressure dressing are necessary to prevent perichondritis, which can result in cartilage loss and a "cauliflower ear deformity." Sports helmets should be worn when appropriate during activities when head trauma is possible.

Frostbite of the auricle should be managed by rapidly rewarming the exposed pinna with warm irrigation or warm compresses.

Foreign bodies in the external canal are common in childhood. Often these can be removed in the office setting without general anesthesia if the child is mature enough to understand and cooperate and is properly restrained; if an adequate headlight, surgical head otoscope, or otomicroscope is used for visualizing the object; and if appropriate instruments such as alligator forceps, wire loops or a blunt cerumen curette or suction are used, depending on the shape of the object. Gentle irrigation of the ear canal with body-temperature water or saline may be used to remove very small objects, but only if the tympanic membrane (TM) is intact. Attempts to remove an object from a struggling child or with poor visualization and inadequate tools result in a terrified child with a swollen and bleeding ear canal and can then mandate general anesthesia to remove the object. Difficult foreign bodies, especially those that are large, deeply embedded, or associated with canal swelling, are best removed by an otolaryngologist and/or under general anesthesia. Disk batteries are removed emergently because they leach a basic fluid that can cause severe tissue destruction. Insects in the canal are first killed with mineral oil or lidocaine and are then removed under otomicroscopic examination. Objects retained in the external auditory canal can lead to complications such as otalgia, conductive hearing loss, infection, and aural drainage.

After a foreign body is removed from the external canal, the TM should be inspected carefully for traumatic perforation, middle-ear effusion, abrasions, and bleeding. If a foreign body has resulted in acute inflammation of the canal, topical otic medications as described for acute external otitis should be instituted (see Chapter 679).

TYMPANIC MEMBRANE AND MIDDLE EAR

Traumatic perforation of the TM usually results from sudden external compression, such as a slap, or penetration by a foreign object such as a stick or cotton-tipped swab. The perforation may be linear or stellate. It is most commonly in the anterior portion of the pars tensa when it is caused by compression, and it may be in any quadrant of the TM when caused by a foreign object. Systemic antibiotics and topical otic medications are not required unless suppurative otorrhea is present. Small traumatic TM perforations often heal spontaneously, but it is important to evaluate and monitor the patient's hearing to ensure that spontaneous healing occurs. If the TM does not heal within several months, surgical graft repair should be considered. If a perforation is present, otorrhea can occur from water entering the middle ear from the ear canal during swimming or bathing; appropriate precautions should be taken. Perforations resulting from penetrating foreign bodies are less likely to heal than those caused by compression. Audiometric examination reveals a conductive hearing loss, with larger air-bone gaps seen in larger perforations. Immediate surgical exploration may be indicated if the injury is accompanied by one or more of the following: vertigo, nystagmus, severe tinnitus, moderate to severe hearing loss, or cerebrospinal fluid (CSF) otorrhea. At the time of exploration, it is necessary to inspect the ossicles, especially the stapes, for possible dislocation or fracture and to clear sharp objects that might have penetrated the oval or round windows. Sensorineural hearing loss results if the stapes subluxates or dislocates into the oval window or if either the oval or round window is penetrated. Children should not be given access to cotton-tipped applicators, because the applicators commonly cause ear trauma. Contact with small objects should be limited to times of parental supervision.

Perilymphatic fistula can occur after barotrauma or an increase in CSF pressure. It should be suspected in a child who develops a sudden SNHL or vertigo after physical exertion, deep water diving, air travel, playing a wind instrument, or significant head trauma. The leak characteristically is at the oval (Fig. 683.1) or the round window and may be associated with congenital abnormalities of these structures or an anatomic abnormality of the cochlea or semicircular canals. Perilymphatic fistulas occasionally close

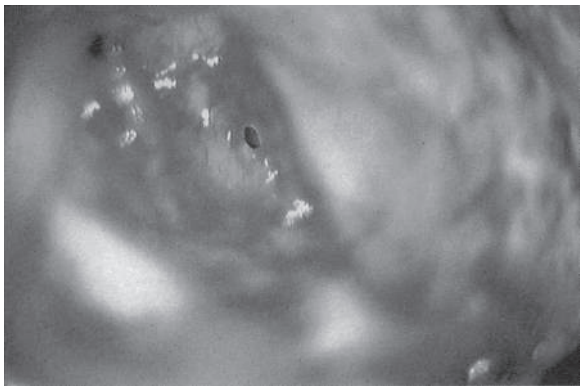

Fig. 683.1 Intraoperative view of traumatic oval window perilymphatic fistula. *(From Kim SH, Kazahaya K, Handler SD. Traumatic perilymphatic fistulas in children: etiology, diagnosis and management. Int J Pediatr Otorhinolaryngol. 2001;60:147–153, Fig. 2.)*

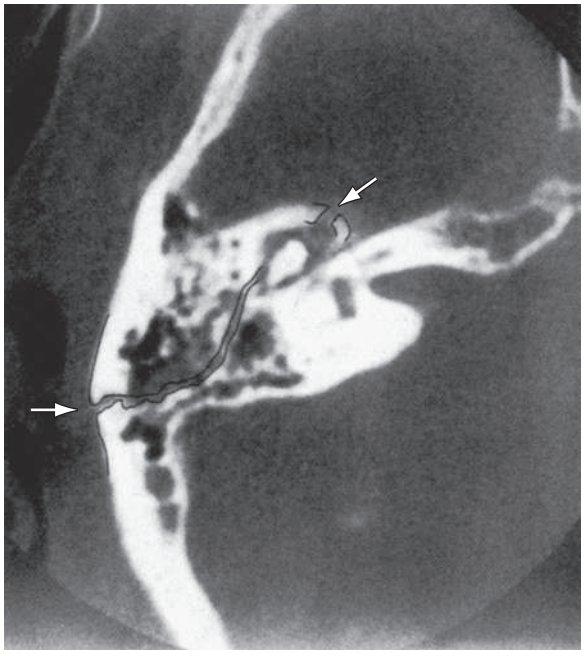

Fig. 683.2 High-resolution axial CT of uncomplicated longitudinal fracture *(arrows)*. A hematoma is present. The course of the fracture has been highlighted. *(From Schubiger O, Valavanis A, Stuckman G, et al. Temporal bone fractures and their complications: examination with high resolution CT. Neuroradiology. 1986;28:93–99.)*

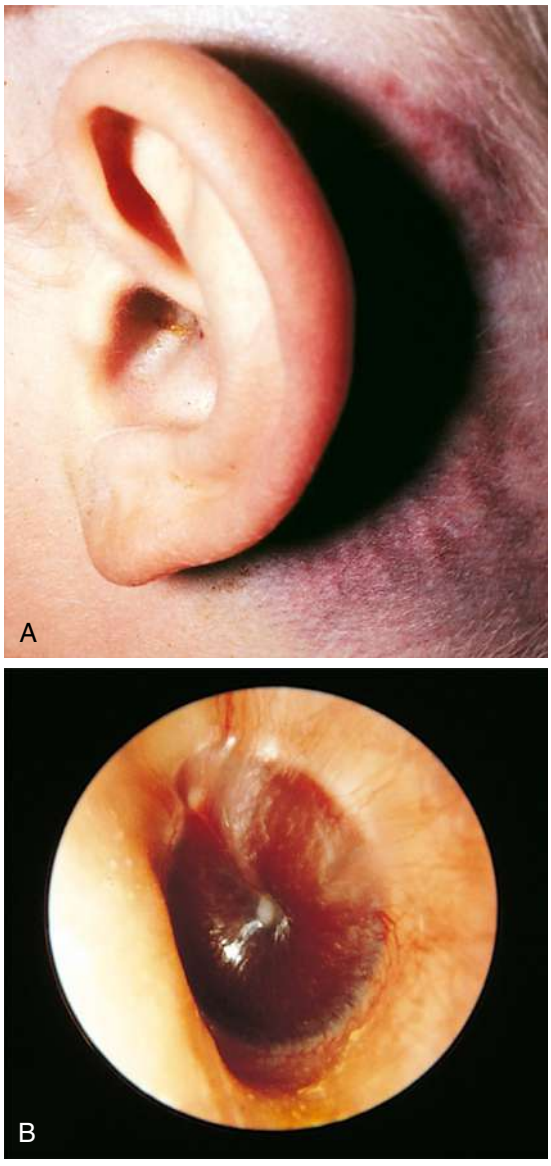

Fig. 683.3 Basilar skull fracture. **A,** Basilar skull fracture involving the temporal bone is often signaled by postauricular ecchymotic discoloration, termed the *Battle sign*. **B,** The force of the blow may also cause tearing of the ear canal or, as shown here, middle ear hemorrhage with hemotympanum. Depending on the timing of examination, this may appear red or blue. *(**B** courtesy Michael Hawke, MD; from Zitelli BJ, McIntire SC, Nowalk AJ, eds. Zitelli and Davis' Atlas of Pediatric Physical Diagnosis, 7th ed. Philadelphia: Elsevier; 2018: Fig. 24-15, p. 874.)*

spontaneously, but immediate surgical repair of the fistula is recommended to control vertigo and to stop any progression of the SNHL; even timely surgery does not usually restore the SNHL. No reliable test is known for perilymphatic fistula, so middle-ear exploration is required for diagnosis and treatment.

TEMPORAL BONE FRACTURES

Children are particularly prone to basilar skull fractures, which usually involve the temporal bone. Temporal bone trauma should be considered in head injuries, and the status of the ear and hearing should be evaluated. Temporal bone fractures are divided into longitudinal (70–80%), transverse (10–20%), and mixed. Longitudinal fractures (Fig. 683.2) are commonly manifested by bleeding from a laceration of the external canal or TM; postauricular ecchymosis **(Battle sign)**; **hemotympanum** (blood behind an intact TM); conductive hearing loss resulting from TM perforation, hemotympanum, or ossicular injury; delayed onset of facial paralysis (which usually improves spontaneously); and temporary CSF otorrhea or rhinorrhea (from CSF running down the eustachian tube) (Fig. 683.3). Transverse fractures of the temporal bone have a graver prognosis than longitudinal fractures and are often associated with immediate facial paralysis and damage to the labyrinth or internal auditory canal. Facial paralysis might improve if caused by edema, but surgical decompression of the nerve is often recommended if there is no evidence of clinical recovery and facial nerve studies are unfavorable. If the facial nerve has been transected, surgical decompression and anastomosis offer the possibility of some functional recovery. Transverse fractures are also associated with severe SNHL, vertigo, nystagmus, tinnitus, nausea, and vomiting associated with

loss of cochlear and vestibular function; hemotympanum; rarely, external canal bleeding; and CSF otorrhea, either in the external auditory canal or behind the TM, which can exit the nose via the eustachian tube.

If temporal bone fracture is suspected or seen on radiographs, gentle examination of the pinna and ear canal is indicated; lacerations or avulsion of soft tissue is common with temporal bone fractures. Vigorous removal of external auditory canal blood clots or tympanocentesis is not indicated, because removing the clot can further dislodge the ossicles or reopen CSF leaks. The effectiveness of prophylactic antibiotics to prevent meningitis in patients with basilar skull fractures and CSF otorrhea or rhinorrhea cannot be determined because studies to date are flawed by biases. If a patient is afebrile and the drainage is not cloudy, watchful waiting without antibiotics is indicated. Surgical intervention is reserved for children who require repair of a nonhealing TM perforation, who have suffered dislocation of the ossicular chain, or who need decompression of the facial nerve. SNHL can also follow a blow to the head without an obvious fracture of the temporal bone (labyrinthine concussion).

ACOUSTIC TRAUMA

Acoustic trauma results from exposure to high-intensity sound (fireworks, gunfire, loud music, heavy machinery) and is initially manifested by a temporary decrease in the hearing threshold, most commonly at 4,000 Hz on an audiometric examination, and tinnitus. If the sound is between 85 and 140 dB, the loss is usually temporary (after a rock concert), but both the hearing loss and the tinnitus can become permanent with chronic noise exposure; the frequencies from 3,000-6,000 Hz are most often involved. Sudden, extremely loud (>140 dB), short-duration noises with loud peak components (gunfire, bombs) can cause permanent hearing loss after a single exposure. Noise-induced hearing loss results from interactions between genes and the environment. A meta-analysis demonstrated that loud music exposure resulted in increased hearing thresholds and decreased otoacoustic emissions in children and adolescents. Ear protection and avoidance of chronic exposure to loud noise are preventive measures. Hearing loss from chronic noise exposure should be entirely preventable. Parents should be made aware of the dangers of acoustic trauma, from the environment and from the use of headphones, and should take measures to minimize exposure. Treatment with high-dose steroids for 1-2 weeks should be considered to treat acute hearing loss related to noise trauma.

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Chapter 684

Tumors of the Ear and Temporal Bone

Joseph Haddad Jr.

Benign tumors of the external canal include osteomas and monostotic and polyostotic fibrous dysplasia. Osteomas are usually unilateral and located lateral in the bony canal; they require removal only if hearing is impaired or external otitis results. Exostoses (see Chapter 550.2), or localized bony hyperplasias, may be confused with osteomas; however, exostoses are usually bilateral and located in the region of the annulus of the tympanic membrane. Masses occurring over the mastoid bone, such as first branchial cysts, dermoid cysts, and lipomas, may be confused with primary mastoid tumors; imaging can help with the diagnosis and treatment plan.

Eosinophilic granuloma, which can occur in isolation or as part of systemic Langerhans cell histiocytosis (see Chapter 556.1), should be suspected in patients with otalgia, otorrhea (sometimes bloody), hearing loss, abnormal tissue within the middle ear or ear canal, and roentgenographic findings of a sharply delineated destructive lesion of the temporal bone. Definitive diagnosis is made by biopsy. Treatment depends on the site of the lesion and histology. Depending on the site, it may be treated by surgical excision, curettage, or local radiation. If the lesion is part of a systemic presentation of Langerhans cell histiocytosis, chemotherapy in addition to local therapy (surgery with or without radiation) is indicated. Long-term follow-up is necessary whether the temporal bone lesion is a single isolated lesion or part of a multisystem disease.

Rhabdomyosarcoma is the most common malignancy of the temporal bone in children. Symptoms and signs of rhabdomyosarcoma (see Chapter 549) originating in the middle ear or ear canal include a mass or polyp in the middle ear or ear canal, bleeding from the ear, otorrhea, otalgia, facial paralysis, and hearing loss. Other cranial nerves also may be involved. Diagnosis is based on biopsy, but the extent of disease is determined by both CT and MRI of the temporal and facial bones, skull base, and brain (Fig. 684.1).

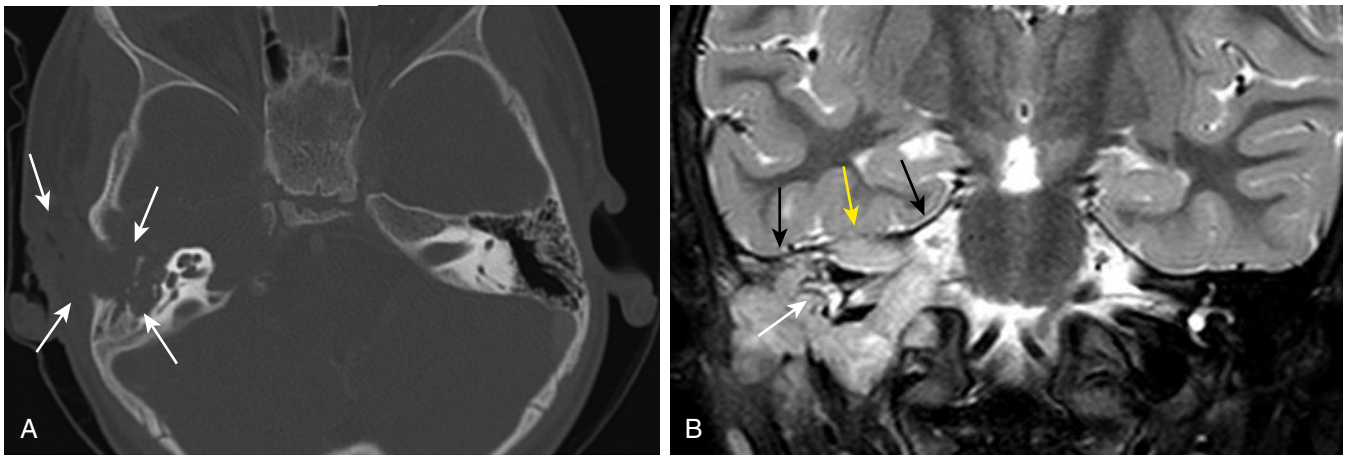

Fig. 684.1 Rhabdomyosarcoma in a 2-yr-old male with hearing loss and right periauricular swelling. A, Axial CT shows a large soft tissue mass of the right temporal bone *(arrows)* with extensive osseous destruction. B, Coronal STIR MR image shows hyperintense mass of the right temporal bone. Focal loss of integrity of the floor of the right middle cranial fossa *(yellow arrow)* and intact dura *(black arrows)* are shown. Destruction of the bony labyrinth *(white arrow)* by the mass. *(Modified from Koral K. Neoplasia. In: Coley BD, ed. Caffey's Pediatric Diagnostic Imaging, 13th ed, Philadelphia: Elsevier; 2019: Fig. 12.1.)*

Management usually involves a combination of chemotherapy, radiation, and surgery.

Non-Hodgkin lymphoma (see Chapter 545.2) and leukemia (see Chapter 544) also occur rarely in the temporal bone. Although primary neoplasms of the middle ear are very uncommon in children, they include adenoid cystic carcinoma, adenocarcinoma, and squamous cell carcinoma. Benign tumors of the temporal bone include glomus tumors. The initial signs and symptoms of the more common nasopharyngeal neoplasms (angiofibroma, rhabdomyosarcoma, epidermoid carcinoma) may be associated with insidious onset of chronic otitis media with effusion (often unilateral). A high index of suspicion is needed for diagnosing these tumors early.

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