Complications and Sequelae: Intratemporal

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Even though the intracranial suppurative complications of otitis media, such as meningitis and brain abscess, are relatively uncommon today in developed countries of the world, clinicians everywhere still frequently encounter the intratemporal (extracranial) complications and sequelae of otitis media—those that occur within the aural cavity and adjacent structures of the temporal bone. These complications can develop from an antecedent acute otitis media (AOM) or complications or sequelae from acute or chronic middle-ear infections.1–3 The intratemporal complications of otitis media are hearing loss; vestibular, balance, and motor dysfunctions; acute perforation of the tympanic membrane; mastoiditis; petrositis; labyrinthitis; facial paralysis; and external otitis.4 The intratemporal sequelae of otitis media are chronic suppurative otitis media, atelectasis of the middle ear, adhesive otitis media, cholesteatoma, cholesterol granuloma, tympanosclerosis, and ossicular discontinuity and fixation (Figure 1). Hearing loss can be either a complication or a sequela of otitis media.5 Almost all children who have a middle-ear effusion have some degree of hearing loss, but hearing loss can also occur when another suppurative complication (eg, perforation of the tympanic membrane or labyrinthitis) or sequela of otitis media develops, such as perforation of the tympanic membrane, adhesive otitis media, cholesteatoma, tympanosclerosis, or ossicular discontinuity or fixation. Even though there have been some recent clinical trials to the contrary, other studies have suggested that children who have had recurrent episodes of otitis media or persistent middle-ear effusion in early childhood perform less well on tests of speech and language than do their disease-free peers. These data suggest that delay in or impairment of development may be an important sequela of otitis media. Also, there are some reports that otitis media in infants may predispose children to later disturbances of balance and ultrahigh-frequency hearing. Recently, Landis and col-
leagues reported an unusual and apparently not recognized disorder associated with otitis media, significantly lower taste function on the tongue; the more severe the middle-ear inflammation, the greater impairment in gustatory function.6

In this chapter, we discuss the epidemiology, pathogenesis, microbiology, and management of intratemporal complications and sequelae of otitis media. In the next chapter, we present similar information for intracranial complications and sequelae.

The burden of otitis media is particularly heavy for children in areas of the world in which access to medical care is limited. Untreated otitis media may lead to persistent perforation of the tympanic membrane and dysarticulation of ossicles, causing permanent conductive hearing loss. Berman reviewed reports from developing countries indicating high rates of tympanic membrane perforation, persistent otorrhea (consistent with chronic suppurative otitis media), and mastoiditis.7 Hearing loss associated with otitis media is a particular concern in developing countries because of the importance of comprehension of normal speech for those who are illiterate. Necrotizing otitis media leading to permanent perforation of the tympanic membrane and other complications of otitis media are discussed in Chapter 4.

Surgery is indicated for many of the aural and intratemporal complications and sequelae of otitis media. Many of the concepts of the surgical procedures are described in Pediatric Otolaryngology,8 and the surgical techniques are described in the Surgical Atlas of Pediatric Otolaryngology.9

HEARING LOSS

Fluctuating or persistent hearing loss is present in most infants and children with middle-ear effusion; hearing impairment is the most prevalent complication of otitis media.10 The hearing loss is most frequently conductive, but sensorineural hearing loss can also be a consequence.5 Even though the hearing loss is characterized as “mild” and self-limited when otitis media is present, the loss can be substantial in some children, especially when it is associated with a complication or sequela of otitis media.

Conductive Hearing Loss

Audiograms of children with middle-ear effusion usually reveal a mild to moderate conductive loss averaging between 20 and 30 dB, but the range is 0 to 60 dB. Even with mild deficits, the softer speech sounds and voiceless consonants may be missed. The frequency distribution of thresholds for speech stimuli associated with otitis media with effusion, obtained by Fria and colleagues, is shown in Figure 2.11 The average hearing loss in that study was 27 dB at 500, 1,000, and 4,000 Hz; at 2,000 Hz, the loss was only 24.5 dB, which probably accounts for a three-frequency (500, 1,000, 2,000 Hz) pure-tone average better than it truly is. Other studies have reported similar losses, although with differing criteria.12,13 The hearing loss is not influenced by the quality of fluid in the middle ear; ears with thin fluid are impaired to the same degree as are those with fluid of a glue-like consistency.14,15

Ears that are partially filled with fluid (identified
otoscopically by the presence of bubbles or an air-fluid level) have less hearing impairment than do ears that are completely filled with fluid.\textsuperscript{11} The hearing impairment is usually reversed with resolution of the effusion.\textsuperscript{16,17} On occasion, permanent conductive hearing loss is due to irreversible changes resulting from recurrent acute or chronic inflammation (eg, adhesive otitis media or ossicular discontinuity). High negative pressure in the ear, or atelectasis, in the absence of effusion is another cause of conductive loss.\textsuperscript{18}

Hearing in children with chronic suppurative otitis media is usually worse than that reported when otitis media with effusion is present. One study from Sierra Leone evaluated the hearing in children who had perforation with and without suppuration. Of the 37 ears that had dry perforations, 33 (89\%) had a pure-tone average of 26 dB or greater, and of the 100 ears that had chronic suppurative otitis media, 96 (96\%) also had this degree of hearing loss.\textsuperscript{19}

Sensorineural Hearing Loss

Sensorineural hearing loss may also result from AOM or otitis media with effusion.\textsuperscript{20} A reversible hearing impairment is generally attributed to the effect of increased tension and stiffness of the round window membrane. A permanent sensorineural loss may occur, presumably as a result of the spread of infection or products of inflammation through the round window membrane,\textsuperscript{21–26} the development of a perilymphatic fistula in the oval round window,\textsuperscript{27–29} or a suppurative complication, such as labyrinthitis.\textsuperscript{9}

Chronic suppurative otitis media, cholesteatoma, or both can be associated with sensorineural hearing loss.\textsuperscript{30} Sensorineural hearing loss and middle-ear disease frequently coexist. Brookhouser and colleagues evaluated 437 children with bilateral sensorineural hearing loss and found a sufficient degree of otitis media to warrant tympanostomy tubes in 35\%.\textsuperscript{31} In Finland, Rahko and colleagues tested 359 individuals who had a known history of otitis media and failed to detect sensorineural hearing loss.\textsuperscript{32} However, Mutlu and colleagues reviewed the audiograms of 71 children (119 ears) who had otitis media with effusion and found that 9\% had either temporary sensorineural involvement (temporary threshold shift), which improved when the effusion resolved, or possibly permanent sensorineural loss, which they attributed to the effusion.\textsuperscript{33} Ultra-high-frequency hearing (9,000–20,000 Hz) can also be affected in children with otitis media, which would not be detected in routine audiometric testing,\textsuperscript{34,35} and it may be affected for the long term.\textsuperscript{36} It is most likely that otitis media with the presence of bacteria, viruses, inflammatory mediators, enzymes, and possibly even a neurotoxin, such as quinolinic acid,\textsuperscript{37} can affect the inner ear in some children.
Studies of Hearing Loss with AOM

Few studies of hearing have been performed during acute episodes of otitis media. Olmstead and colleagues studied children 2 to 12 years of age with a diagnosis of AOM who were seen in the Outpatient Department of St. Christopher’s Hospital in Philadelphia. Of 82 children enrolled in the study, 33% had no loss of hearing on the initial audiometric test after acute infection; 40% had loss of hearing (up to 15 dB) initially, which disappeared in 1 to 6 months; 12% had loss of hearing throughout the 6-month period of observation; and 15% had loss of hearing initially but were lost to the study between 1 and 4 months after the acute episode of otitis media. The children had no history of hearing difficulty or chronic ear infection. Otoscopic examinations were not performed after the initial diagnosis, and data were not presented concerning the duration of fluid in the middle ear. These data indicate that after a single episode of AOM, many children have prolonged hearing impairment.

Hearing loss has also been identified in children who have apparently recovered from AOM. A longitudinal study of Alaskan Eskimo children showed a statistically significant association between the frequency of episodes of otitis media and hearing loss of greater than 26 dB. Of children who had one or more attacks of otitis media per year, 49% had hearing loss; hearing loss was evident in 15% of children with no diagnosed episodes of otitis media. Other studies with differing criteria have found the incidence of hearing loss associated with AOM to vary between 6 and 30%.

Ryding and colleagues assessed hearing thresholds in children who had had recurrent AOM in the past, compared with those without middle-ear disease, and found no difference in hearing at the traditional levels (ie, 125–8,000 Hz) between the two groups, but children in the otitis media group had hearing levels at high frequencies (8,000–16,000 Hz) and the acoustic middle-ear reflex thresholds were elevated; also middle-ear compliance was higher, and click-evoked otoacoustic emission response levels and middle-ear pressures were lower. They concluded that children with a history of middle-ear disease not only had disturbances of middle-ear mechanics but also cochlear function may have been abnormal as a sequela of the middle-ear disease.

Studies of Hearing Loss with Otitis Media with Effusion

Hearing loss frequently accompanies persistent middle-ear effusion (documented at the time of surgery). Fria and colleagues evaluated hearing in 222 infants (aged 7–24 months) and 540 older children (aged 2–12 years). Both the younger and older children had, on average, thresholds for speech reception and speech awareness of 24.6 and 22.7 dB, respectively (see Figure 2). Not all children with middle-ear effusion have apparent hearing impairment. About one-third of the children had air conduction thresholds of 15 dB, but approximately 25% of children with middle-ear effusion had thresholds of up to 30 dB. The cumulative frequency curves were similar for children of various ages and for the duration of effusion. This large study provides a complete picture of the number of children affected and the extent of the hearing loss when middle-ear effusion is present. Audiometric techniques for children of various ages are discussed in Chapter 7.

In a recent report, Gravel and colleagues found that otitis media with effusion in early childhood, compared with those children without a past experience with otitis media, was associated with disturbances of auditory processing when the children were age 8 years; also, the study revealed hearing sensitivities to be significantly poorer in the children who had had the middle-ear disease.

EFFECTS OF OTITIS MEDIA ON THE DEVELOPMENT OF THE CHILD

The results of many studies of the association of otitis media and development of speech, language, and cognitive abilities have been reviewed by the Agency for Health Care Policy and Research, by Vernon-Feagans, and most recently by the American Academy of
Some studies have identified associations of recurrent otitis media with effusion and lower scores on tests of vocabulary, auditory comprehension, and language skills. Other studies failed to find significant differences among children with and without a history of prolonged time spent with middle-ear effusion. The variables in these studies are many, including documentation of the ear disease, the type of test administered, the socioeconomic class of the parents, the quality and quantity of language in the home and in out-of-home day care, the child’s temperament, and the child’s and the parents’ IQ. The variability of the results of these many studies suggests that the effects of otitis media may be more substantial in some children than in others. The focus of current studies is on the development of criteria for those children who are most likely to be affected by the hearing loss accompanying otitis media.

Sensorineural hearing loss has been associated with impairment in the cognitive, language, and emotional development of children. Children with sensorineural hearing impairment, compared with peers who have normal hearing, are significantly retarded in the development of vocabulary, are placed below their grade level in school, have poorer articulation and auditory discriminatory abilities, and have a high rate of maladjusted behavior patterns and disturbances in psychosocial adjustment.

The first months of life are important in language acquisition. The infant is capable of speech sound discrimination as early as 1 month of age. By the age of 6 weeks, the infant is attracted to human voices more than to environmental sounds and to female more than to male voices. At 5 to 6 months, the infant enters the babble phase and plays with sound-making. The child is putting words together in sentences by the age of 18 months, and by 4 years, the child produces all of the basic syntactic structures that he or she will ever use. Because so much progress in language acquisition is made during infancy, any problems in receiving or interpreting sound signals might have a significant effect on speech and language development. Softer speech sounds and voiceless consonants, in particular, may be missed or confused when effusion is present in the middle ear (Figure 3). Important differences have been identified in the early patterns of vocalization of the hearing-impaired infant compared with hearing infants. It is unknown how these data about differences in infant babble relate to the ultimate development of speech and language when the hearing impairment is mild and fluctuating.

The current hypothesis for the effects of otitis media on the child’s development is presented in Figure 4. Children with severe or recurrent otitis media spend prolonged time with middle-ear effusion. Hearing impairment (average loss approximately 25 dB) accompanies the effusion in most children, and if the hearing impairment occurs at a time of rapid intellectual growth, the result may be impaired development of speech, language, and cognitive abilities.

Many investigators have studied the effects of otitis media on the development of the child.
Although most studies indicate that children with a history of recurrent episodes of acute otitis media score lower on tests of speech and language than do their disease-free peers, some important recent studies fail to show a difference in long-term developmental outcomes. A brief resume of the results of selected studies follows:

1. One of the earliest and most widely cited studies is that of Holm and Kunze of Seattle. Children aged 5 to 9 years who had a history of chronic otitis media with onset before the age of 2 years were compared with children in a control group matched for age, sex, and socioeconomic background. Children with a history of ear disease were delayed in all language skills requiring the receiving or processing of auditory stimuli, but the groups were similar in tests measuring visual and motor skills (Figure 5). The diagnosis of otitis media was made on the basis of history; otoscopic and audiometric examinations were not performed, and the sample size was small (16 children in each group).

2. Eskimo children were observed prospectively during the first 4 years of life and had tests of hearing, intelligence, and assessment of school performance at age 10 years. Children with recurrent episodes of otitis media (defined as the presence of a draining ear) during the first 2 years of life and with loss of hearing of 26 dB or more had lower scores in tests of reading, mathematics, and language than did children who had little or no disease in infancy. Otorrhea was the sole criterion for otitis media; data were not available about the presence or duration of middle-ear effusions or episodes of AOM that did not result in otorrhea.

3. Lewis studied Aboriginal children from Brisbane, Australia. Children aged 7 to 9 years who “failed otoscopic examinations” and had hearing deficits measured by audiometry or tympanometry during a 4-year period were compared with age-matched control children who had consistently passed the audiometric tests and were assumed to be disease free. Children with ear disease had mean scores for speech and language development that were significantly lower than those of the children without ear disease. The sample size was small: 14 children with disease and 18 control subjects.

4. Needleman evaluated 20 children aged 3 to 8 years with a history of recurrent otitis media and a first episode before the age of 18 months. Twenty control subjects who had no history of hearing problems or recurrent
ear infections were matched with the patients for age, grade, and socioeconomic status. The children were evaluated for their ability to use speech sounds expressively and receptively. Children with a history of ear disease had poorer phonologic abilities than did the control children. The diagnosis of ear disease was based on the history alone.

5. Sak and Ruben used a sibling control for children with a history of otitis media. Children received tests of speech and language between the ages of 8 and 11 years. One sibling of each pair had a documented history of persistent otitis media beginning before the age of 5 years, whereas the other sibling had had no middle-ear problem. The children who had had otitis media had a lower verbal IQ, poorer auditory reception, and lower spelling achievement than their matched sibling controls. More of the siblings with otitis media were boys, and more minor middle-ear abnormalities identified by audiometry or tympanometry were prevalent among the otitis media siblings than among the control siblings, suggesting the possibility that deficits were associated with recent, rather than earlier, middle-ear disease.

6. Friel-Patti and colleagues examined the association of otitis media early in life with language development at 12, 18, and 24 months. The infants had been selected from intensive care units of low birth weight nurseries and were predominantly from low socioeconomic groups. Frequent episodes of otitis media were correlated with a higher prevalence of language delay, but no correlation was found between hearing impairment measured by auditory brainstem response testing and language delay.

7. Teele and colleagues studied 190 white children of varying socioeconomic strata from greater Boston to determine the association between time spent with middle-ear effusion and development of speech, language, and cognitive abilities. The children were selected from a cohort of children in five health centers who were observed from birth with regular examinations of the middle ear at each visit to the office or clinic, whether for illness or for routine care. The study was prospective, used uniform criteria to diagnose otitis media and middle-ear effusion, and tested children from all socioeconomic strata. Tests of speech and language administered at the third birthday included the Peabody Picture Vocabulary Test (a test of both early receptive and expressive language), the Fisher-Logemann and Goldman-Fristoe tests of articulation (tests of production of speech sound), and other measurements of language structure complexity and estimates of intelligibility.

Children who had spent fewer than 30 days with middle-ear effusion during the first 3 years of life were compared with those who had spent 30 to 129 days with middle-ear effusion during the first 3 years of life and with those who had spent 130 or more days with middle-ear effusion from birth to age 3 years. A summary of the results indicated the following:

- Test scores were lower for the total number of children tested, but significant differences were present only in the test scores of children from the high socioeconomic group. No significant differences were found for children from low socioeconomic groups (Tables 1 and 2). The basis for the difference in the results for

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<tr>
<th>Table 1. COGNITIVE ABILITY AT AGE 7 YEARS BY ESTIMATED DAYS WITH MIDDLE-EAR EFFUSION DURING THE FIRST 3 YEARS OF LIFE</th>
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<td>Estimated Days with MEE</td>
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<td>Verbal</td>
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<td>Performance</td>
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Adapted from Teele DW et al. MEE = middle-ear effusion. Cognitive ability is expressed as mean IQ by the Wechsler Intelligence Scale for Children-Revised after adjusting for socioeconomic status and gender (general linear model procedure SAS-GLM). Time spent with MEE is a natural log of time with effusion (days, 1). Intervals with MEE were selected to produce three groups of about equal size. p = *.007, .001, .026, .008, not significant (.07).
The authors followed up 207 children randomly selected from the same cohort who were observed from birth to the age of 7 years and administered tests of intellectual ability, school achievement, speech, and language.63 After confounding variables were controlled for, estimated time spent with middle-ear effusion during the first 3 years of life was significantly associated with lower scores on tests of cognitive ability, speech and language, and school performance at age 7 years. The adjusted mean full-scale Wechsler Intelligence Scale for Children-Revised (WISC-R) score was 113.1 for those with the least time with middle-ear effusion and 105.4 for those with the most time (see Table 1). Similar significant differences were found for verbal and performance IQ scores. For the Metropolitan Achievement Test, we found that middle-ear disease in the first 3 years of life was associated with significantly lower scores in mathematics and reading. Similar differences were found for articulation and use of morphology markers. After time spent with middle-ear effusion during the first 3 years of life was considered, time spent after the first 3 years was not a significant predictor of scores on any of the tests administered.

8. Hubbard and colleagues in Pittsburgh evaluated two matched pairs of children with repaired palatal clefts.64 The treatment of the children had been equivalent, with the exception that one group had undergone early myringotomy with tympanostomy tube placement (mean age 3 months) and the other group had undergone initial myringotomy later (mean age 30.8 months) or not at all. Hearing acuity and consonant articulation were significantly less impaired in the group having undergone early myringotomy. Mean verbal performance and full-scale IQs and scores on psychosocial indices were normal in both groups and did not differ significantly between the groups.

9. Watanabe and colleagues studied total speaking time in infants and children with and without middle-ear effusion.65 To support the premise that improvements in the child’s performance occurred when hearing improved with the resolution of middle-ear effusion, the authors developed a technique to identify the time of vibration of the vocal cords. The duration of speaking time was measured in children with otitis media with effusion before and after tympanostomy tube placement. Preoperative speaking time was found to be 8 minutes and 2 seconds per measured hour when middle-ear effusion was present and 10 minutes per hour when the effusion cleared after tympanostomy tube placement. The implications of this innovative study are uncertain but suggest that hearing improvement increases speaking time and causes the child’s ordinary behavior to be more animate.

10. Chase studied the early development of children with and without experience with otitis media in the first year of life (one episode within 6 months or two episodes in the first

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<th>Table 2. ACUTE OTITIS MEDIA AND DEVELOPMENTAL OUTCOME AT AGE 1 YEAR</th>
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<td>Mental and motor development of children with AOM</td>
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<td>motor scores</td>
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<td>Behavior of children with AOM</td>
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<tr>
<td>Less attentive and persistent (rated by examiner)</td>
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<td>More irregular in sleeping and eating (rated by parents)</td>
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<td>Less response in structural interaction with parent</td>
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<td>Behavior of parents of children with AOM</td>
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<td>Less skilful at providing effective teaching</td>
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Adapted from Chase C.66

AOM = acute otitis media.
There were no differences in overall mental and motor development. There were, however, clear behavioral differences between 1-year-old children who experienced recurrent otitis media in the first year and those who did not. Children who experienced otitis media were rated by their parents as less attentive and less persistent by the examiner during testing and more irregular in their patterns of sleeping, eating, and elimination. In addition, children who had experienced otitis media were less responsive and less attentive in working with their mothers in a learning situation. These findings suggest that a child who has experienced ear infection and mild hearing loss may show signs of attentional difficulty early in life.

The Pittsburgh studies by Paradise and colleagues reported on an extensive, prospective study, 6,350 infants followed from soon after birth to the age of 3 years were systematically examined for the presence or absence of middle-ear disease. The design of the study included randomization of subjects who developed sufficient otitis media to warrant myringotomy and tympanostomy tube placement. To determine the effect of a relatively short period of middle-ear effusion versus a more prolonged course, the children were randomly assigned to either early (prompt) or delayed insertion of the tubes, and then the two groups had assessment of developmental outcomes at 3 years of age and later at age 6 months of age. The investigators concluded in the first report that “in children younger than three years of age who have persistent otitis media, prompt insertion of tympanotomy tubes does not measurably improve outcomes at age three years.” This conclusion was challenged by Rosenfeld, who asked for more details, which was responded to by Paradise and colleagues. Then Paradise and colleagues concluded in the later assessment that “in otherwise healthy children younger than three years of age who have persistent middle-ear effusion within the duration that we studied, prompt insertion of tympanotomy tubes does not improve developmental outcomes at age six years of age.” Even though the relatively short difference in the period between the “early” tube insertion versus the “delayed” placement was a substantial design flaw. These results are an important contribution to the discussion of the effects of otitis media on the development of the child.

Other studies of otitis media and language performance demonstrated the following results:

- A study of children in Montreal, aged 3 to 5 years, identified significant differences among the children with a history of otitis media and matched control children.
- A study of Danish children aged 3 to 9 years did not show an effect of otitis media with effusion early in life on reading achievement.
- A study by Lous demonstrated a small but significant correlation between flat tympanograms (type B) in first-grade children and silent work reading at the beginning of the second grade.
- An evaluation of Apache Indian children aged 6 to 8 years with a contrasting history of otitis media showed no significant difference in language performance.
- A retrospective study suggested that middle-ear disease in school-aged children was associated with hyperactivity or inattention, independent of learning ability. Also, a recent brief report by Rinkel and colleagues found that early-life otitis media with effusion negatively influenced the development of speech-in-noise understanding, which was postulated to subsequently affect speech, language, and learning development.
- Roberts and colleagues prospectively followed 83 black children, primarily from low-income families, who were examined periodically from 6 months to 4 years of age for the presence of middle-ear effusion, and then assessed the children’s later academic skills in reading and word recognition during the early years of elementary school. The children who had had otitis media and hearing loss scored lower in mathematics and expressive language at
young ages but caught up in math on entering school and in expressive language by the second grade. The investigators concluded that the child’s home environment was more related to these outcomes than the presence of a past experience with otitis media with effusion but cautioned about interpreting these findings when generalizing to other populations. In a follow-up study with a similar design and methodology that included children from New York, outcomes were comparable to those of the first report.78

These reports are disturbing, but many have one or more flaws in study design: reliance on a retrospective history of AOM; uncertain validity of the diagnosis of otitis media; a lack of information or documentation about middle-ear effusion; the presence of significant hearing impairment in subjects at the time of tests of speech and language; small numbers of subjects; special populations tested (eg, Australian Aborigines, Alaskan Eskimos, children with a cleft palate); and inadequate criteria for selection of children without disease used for comparison.

Hignett summarized the study design issues and evaluated the effectiveness of 10 early studies of otitis media and speech, language, and behavior.79 The studies of Teele and colleagues and Hubbard and colleagues represent significant advances in study design compared with previous investigations,62,64 but both of these studies have been subject to criticism because of perceived limitations and deficiencies.80,81 Roberts and colleagues conducted a meta-analysis of the studies reported on the effect of otitis media on speech and language and found a small effect but also that the evidence was neither combinable nor generalizable.82 Vernon-Feagans and colleagues, in a similar review, arrived at a similar conclusion.83 These inadequacies of study design limit general application of these results to planning care for young children, but they do not prevent concern that many children may suffer from the sequelae of otitis media with persistent middle-ear effusion in infancy.

Factors of Importance in Analysis of Studies of Otitis Media and Development of Speech, Language, and Cognitive Abilities

Parents Also Suffer

The working parent who has spent a sleepless night attending to a child who is fretful because of ear pain may have work- and home-related stress. Chase noted that parents of 1-year-old children who had experienced otitis media were less effective teachers in structured interactive tasks.66 They were less effective in gaining the child’s attention, less able to respond effectively when the child was distracted from the task, and less able to help the child understand and perform the task. The parent must contend with the child’s acute pain, persistent irritability and inattentiveness, and the expense and inconvenience of frequent visits to the physician. Although some families can accept and cope with the stress of a child’s recurrent illnesses, other families cannot. A constellation of disturbances in psychosocial development may result. Paradise and colleagues noted that stress ratings were highest among those parents whose baseline stress scores were the highest.84

Focus groups of parents described chronic otitis media as a condition that affects not only the child who is ill but the entire family. These findings were reported by the Functional Outcomes Project of the American Academy of Pediatrics, whose goals were to assess various aspects of the physical, social, and emotional well-being of children with chronic illnesses and their families.85 Parents reported the influence of the disease on the child’s behavior, such as aggressiveness, whining, or excessive clinging. Parents worried about the cost of visits to the physician, prescription medication, and tests and expressed frustration over inconsistently effective medications. Siblings often demanded more attention when another was ill, and parents expressed feelings of guilt over spending so much time with one child to the exclusion of siblings. The implications for the practitioner include providing support groups and teaching materials or information sessions for parents and encoura-
ging office-based research in the area of child and family well-being to better understand the impact of chronic illnesses, such as otitis media, on the family.

**Recurrent and Persistent Otitis Media as a Chronic Disease**

The systemic effects of illness, including irritability, malaise, lethargy, and local or generalized pain, may be sufficiently distracting to affect development. These effects of a chronic illness must be distinguished from the specific effects of otitis media (ie, hearing loss) in the interpretation of the sequelae of the disease. Is the child treated differently by the parents, siblings, peers, or teachers because of the recurrent illnesses? Is the child vulnerable to effects unassociated with the specific morbidity of the disease (kept indoors, away from peers, or out of exercise and athletic programs)?

**Critical Ages for Effects of Otitis Media**

Otitis media of a similar duration may affect children differently at different ages. There may be critical periods of perception of language when the child is most vulnerable to mild, fluctuating, or persistent hearing loss. The results of the Boston study suggested that the children were most affected by middle-ear effusion when disease occurred during the first year of life.62 During early stages of language development, the child learns the sounds of the language; different or changing auditory signals resulting from persistent or fluctuating hearing deficits may impede the child’s abilities to form linguistic categories.86

**Auditory Deprivation**

Studies in birds and rodents indicate that deprivation of sound early in life leads to identifiable changes in auditory sectors of the brain. A decrease in the size and number of neurons in the auditory nuclei of mice occurred when the animals were deprived of auditory stimuli during early development.87 In the normal postnatal development of the mouse, the neurons of the auditory brainstem reach adult size by the age of 12 days, the time of onset of hearing. Mice that underwent auditory deprivation by experimentally produced conductive hearing loss 4 to 45 days after birth had auditory brainstem neurons that were significantly smaller than normal. If the mice that underwent induced hearing loss early in life were returned to normal hearing after 45 days, the smaller-than-normal neurons were retained. The size of the neurons was not altered in mice raised in a normal sound environment until 45 days and then deprived of sound until 90 days of age.

These data demonstrate that a period exists in the development of mice during which adequate sound stimulation is needed to establish the normal size of neuronal cells in the auditory brainstem. These experimental data in animals raise concerns about irreparable damage from temporary conductive hearing loss in infants. Webster pointed out, however, that the factors in the experimental model differ from the mild to moderate hearing losses of otitis media with effusion in humans; in the experimental model, the conductive loss is approximately 50 dB, greater than the loss in most cases of otitis media with effusion.87 The loss is persistent rather than fluctuating, and the impairment starts at the inception of hearing in the mouse, whereas inception of sound occurs prenatally in the human. The author concluded that although the restrictions of the animal model must be kept in mind, “the fact that early auditory restriction has a profound effect on the central nervous system in one mammal must arouse concern about possible related effects in humans.”

**Unilateral Hearing Loss**

Unilateral hearing loss has not been considered a handicap for children. Data indicate, however, that children with unilateral hearing impairment score less well on auditory, linguistic, and behavioral tests than do children without hearing impairment.88 Although the children studied had sensorineural hearing deficits, the data suggest that we should no longer accept a unilateral hearing loss as benign. Children with unilateral conductive loss may also suffer during critical
periods of language perception because of confused speech signals.

**Effects of Group Day Care**
Respiratory infections are readily spread among children in day care, and children in day care are likely to have more episodes of otitis media than will children in home care. In relation to development of language, the quantity and quality of the speech sounds around the infants in group care differ from those presented to the child in home care. The factors of increased number of infections and differences in the speech environment in group day care will need to be considered in future studies.

**Child Behavior and Quality of Life Outcomes**
Disturbances in children’s behavior associated with otitis media have been reported to include restlessness, frequent disobedience, impaired task orientation in the classroom, inattention, short attention span and distractibility, and restricted social interaction. Only selected children may be most affected. Paradise and colleagues found that parent-child stress and behavior problems were highest among children from the most socioeconomically disadvantaged homes. Gray suggested that inconsistencies in the child’s ability to hear may have a lasting effect on the child’s motivation to achieve and may cause strained relationships with teachers and parents. A review of the various studies of the impact of otitis media on the quality of life of the child’s behavior was recently prepared by Haggard and colleagues. They concluded that limitations of study design hampered interpretation of the results of the studies.

**Other Variables that May Relate to Early Childhood Language Development**
Future study designs must also consider these factors: quality of the diagnosis of ear disease; visual status; physical and motor development; intellectual, social, and emotional development; nutritional status; history of medications; dialect exposure; birth order; and number of siblings.

Other factors to be considered are the social and economic class of the parents and the quality and quantity of language in the home and day-care center.

**Test Results and Functional Significance**
Do a few percentage points of one or more standard tests of speech, language, or cognitive abilities affect the child’s capability to function in the school, play, and home settings? Some investigators question whether these are statistical differences of limited importance to the child’s development, but there are reasons for concern. Because the data are expressed here in terms of mean differences, some children will be close to or better than the norm, whereas others will have scores that are much lower. Otitis media is so common in early childhood that even if a small percentage of children are adversely affected in terms of development, the number of children who suffer is large. Of the 3.7 million children born in the United States each year, more than one-third will have recurrent episodes of otitis media (three or more) by the age of 3 years. If only 10% of the children with recurrent episodes are affected adversely, the national impact is greater: more than 100,000 of each year’s newborn infants would be involved.

Because the tests measure the child’s potential for achievement, it is possible that the loss suffered by the child with frequent and recurrent episodes of otitis media accompanied by hearing loss in early infancy is never perceived by the parents, teachers, or physicians. The child is not obviously slow or behind his or her peers. The failure of the child to reach his or her potential is a loss for the child and the family, and because the number of these children is large each year, it must be considered a national concern.

**Summary: Role of Otitis Media in Infant Development**
The accumulated results of the various studies of otitis media and development of speech, language, and cognitive abilities suggest that children do
suffer long-term effects from otitis media early in life. The scientific evidence, however, remains incomplete. As recently concluded by Roberts, “the issue of whether recurrent otitis media with effusion affects later acquisition of speech, language, and academic skills continues to be controversial.”94 Some experts are skeptical about available data.92,93 Ventry concluded that no causal link had been established (by 1982) between early recurrent middle-ear effusion and language delay or learning problems.93 Rapin noted that no studies published by 1977 “met the standards of rigor needed to provide a definitive answer to this question, although the burden of the evidence is that a persistent and mild hearing loss, especially if present since infancy, probably has a measurably deleterious effect on the language of most, but not all, children.” Rapin’s statement is as applicable today as it was in 1977.

The difficulties in study design needed to resolve the issues and account for many of the variables are formidable. The optimal design will need to include frequent otoscopic observations beginning soon after birth to develop a chronology of time spent with middle-ear effusion. Hearing assessments will need to be performed in infants when they have effusion and are free of effusion to measure the duration and severity of hearing deficits. All of this will need to be done in the first years of life, when hearing assessments are more difficult and less precise than in the older child. The study will need to be cross-sectional and prospective from birth and should be performed by validated otoscopists. Tests of speech, language, and cognitive abilities will need to be selected that are accurate and standardized for the populations to be tested. The tests should be performed at least annually to define the time of onset or the effect of otitis media on development. The previous section identified the other variables that will need to be considered, including the quality of parenting, the effect of siblings, and the time spent in group day care.

Concern about the association of middle-ear disease and development of speech and language was expressed in a policy statement of the American Academy of Pediatrics.95 Although recognizing the validity of criticism of published studies, the Committee on Early Childhood, Adoption and Dependent Care concluded that “there is growing evidence demonstrating a correlation between middle-ear disease with hearing impairment and delays in the development of speech, language, and cognitive skills... When a child has frequently recurring AOM and/or middle-ear effusion persisting for longer than 3 months, hearing should be assessed and the development of communicative skills must be monitored.” Until definitive answers are available from studies of appropriate design to evaluate the sequelae of otitis media in early infancy, the physician must decide, for each child in his or her care, the optimal management of persistent middle-ear effusion. Chapter 8 provides guidelines for such care.

**VESTIBULAR, BALANCE, AND MOTOR DYSFUNCTIONS**

The most common cause of vestibular disturbance in children is otitis media.96 Many parents of infants and children report balance problems, such as clumsiness, when a middle-ear effusion is present.97 We now have evidence from studies of labyrinthine function in children with and without middle-ear effusion to confirm that the vestibular system is adversely affected, and after tympanostomy tube placement, these dysfunctions return to normal.98–101 Test results of motor proficiency have also been demonstrated to be abnormal in children when middle-ear effusion is present.102–104 Most recently, a study revealed that children who had a history of otitis media but no middle-ear effusion at the time of the vestibular testing also had abnormal function, which indicates that there may be some residual effect of otitis media on the labyrinth.105 Also, a recent case report described a child who had otitis media with effusion and vestibular neuritis.106 Disturbances in balance associated with otitis media may also be due to labyrinthitis, as described later in this chapter.107 Furman and Casselbrant have provided a detailed description...
of the methods to evaluate children with vestibular disorders.108

**PERFORATION OF THE TYPANIC MEMBRANE**

Perforation of the tympanic membrane secondary to otitis media (and certain related conditions, such as atelectasis of the tympanic membrane) can be either acute or chronic; otitis media may or may not be present; and when otitis media is present, otorrhea may or may not be evident.1,10 The perforation can be classified according to the following:

**Site:** pars tensa (anterosuperior, anteroinferior, posterosuperior, or posteroinferior quadrants) or pars flaccida

**Extent:** limited to one quadrant (less than 25%); involving two or more quadrants, but not total; or total perforation (all quadrants).

**Duration:** acute or chronic

A perforation may also be due to a complication of a surgical procedure for management of otitis media, such as myringotomy, tympanostomy tube insertion, or tympanoplasty (ie, an iatrogenic complication). An acute perforation is most frequently secondary to an episode of AOM; if it persists for more than 3 months, it is considered chronic. The defect may involve almost the entire pars tensa or be so small as to be detectable only with the otomicroscope or when the immittance testing measures a volume larger than the expected ear canal volume (see Chapter 7). Otitis media (with or without discharge) may be present or absent; when chronic otitis media is present, the condition is called chronic suppurative otitis media, which is described in detail in a later section. Likewise, a perforation may be associated with some of the other complications and sequelae described in this chapter. In the past, perforations have been classified into central and marginal types. Regardless of size, if there is a rim of tympanic membrane remaining at all borders, the perforation is classified as central. When any part of the perforation extends to the annulus, it is termed marginal.

A defect in the pars flaccida has commonly been called an attic perforation. The so-called marginal perforation of the pars tensa, which usually occurs in the posterosuperior portion, and the “perforation” are, in reality, either a deep retraction pocket or a cholesteatoma, which are described later in this chapter (Figure 6). For the attic perforation and marginal perforation, there is usually no continuity between the defect in the membrane and the middle ear until late in the disease process, when infection erodes the membrane of the pocket or the matrix of the cholesteatoma. Therefore, the terms marginal perforation and attic perforation are misnomers; they were applied on the basis of observations made before the availability of the otomicroscope, modern middle-ear surgery, advances in temporal bone histopathologic techniques, the use of immittance testing, and a better under-

![Figure 6. Examples of defects in the tympanic membrane. A, A small “central” perforation in the anteroinferior portion of the pars tensa of the tympanic membrane. B, A “central” perforation that involves approximately half of the pars tensa. C, A deep retraction pocket in the posterosuperior portion of the pars tensa that has been incorrectly called a marginal perforation. D, A deep retraction pocket in the pars flaccida that has been inappropriately called an attic perforation.](image-url)
standing of the pathogenesis of a retraction pocket and cholesteatoma.

**Acute Perforation**

An acute perforation of the tympanic membrane is usually caused by an episode of AOM that is complicated by rupture of the eardrum. However, an acute perforation can be present in which otitis media is absent, such as when the perforation is secondary to trauma or occurs as a complication of ear surgery (eg, after tympanostomy tube insertion).

**Classification**

Acute perforations can be classified (see Chapter 1) as acute perforation without otitis media and acute perforation with otitis media, with or without otorrhea (AOM with perforation).

**Acute Perforation without Otitis Media**

When an acute perforation is not a complication of otitis media, otitis media may be absent, such as after spontaneous extrusion or removal of a tympanostomy tube or secondary to trauma. Management of such perforations is “watchful waiting” because most will heal within 2 to 3 months. In the two clinical trials conducted in Pittsburgh that evaluated the efficacy of tympanostomy tube placement for chronic otitis media with effusion in 215 children who were observed for 2 to 3 years after the tubes were inserted, 32 (14.8%) had a perforation at the tube site after extrusion, but only 3 children (1.4%) developed a chronic perforation requiring tympanoplasty. Not only can an acute perforation persist into a chronic perforation, but a child with an acute perforation may develop an AOM; both are discussed in the following.

**Acute Perforation with Otitis Media**

An acute perforation (not due to trauma) is usually secondary to AOM but may also occur during the course of chronic otitis media with effusion. Boswell and Nienhuys described spontaneous perforations developing in Australian Aborigines who had chronic middle-ear effusions. Because a spontaneous perforation commonly accompanies an episode of acute middle-ear infection, it may be considered part of the disease process rather than a complication. Because such a perforation allows purulent material to drain into the external canal and enhances drainage of pus down the eustachian tube (owing to the effects of an opening in the eardrum), a perforation of the eardrum may prevent further spread of infection within the temporal bone or, more important, into the intracranial cavity. Infants and children of certain racial groups, such as Alaskan natives (Eskimos) and some Native American tribes, have a high incidence of spontaneous perforation with discharge; the eardrum is perforated spontaneously with almost every episode of AOM. In 711 Australian Aboriginal children 6 to 30 months of age, 7% had an AOM with perforation. The disease runs a similar course in certain other children not belonging to these high-risk populations (see “Chronic Perforation”).

**Pathogenesis.** Because of the presence of a patulous or semipatulous eustachian tube, the perforation may occur in high-risk populations, such as the White River Apache American Indian tribe. A eustachian tube with low resistance would permit a larger bolus of bacteria-laden purulent material from the nasopharynx to enter (by reflux, aspiration, or insufflation) the middle ear, causing a more fulminating infection than would occur if the eustachian tube had either normal or high resistance. An alternative explanation of why some children seem to suffer a perforated eardrum with each episode of AOM whereas others do not could be that there are differences in the virulence of the bacteria or decreased resistance of the host. Factors that predispose children to acute perforation are most likely similar to those described later for chronic perforation, especially in high-risk populations.

The possible outcomes of an acute perforation associated with an attack of AOM are as follows:
The otitis media and otorrhea completely resolve, and the tympanic membrane perforation heals.

The otitis media and otorrhea completely resolve, but the perforation persists and becomes chronic.

Otitis media and perforation persist into the chronic stage, which is termed chronic suppurative otitis media.

A suppurative complication develops.

**Microbiology.** The organisms most frequently cultured from an aural discharge when AOM is present are the same as those that have been cultured from acute middle-ear effusions when a tympanocentesis has been performed (eg, *Streptococcus pneumoniae*, *Haemophilus influenzae*, and *Moraxella catarrhalis*). *Streptococcus pyogenes*, when present and untreated, has been associated with acute perforation of the tympanic membrane. These organisms have also been cultured from children who have tympanostomy tubes in place and have an episode of acute otorrhea. *Pseudomonas aeruginosa* can also be cultured from children. More recently, methicillin-resistant *Staphylococcus aureus* (MRSA) has been isolated from discharging ears, which emphasizes the importance of obtaining a culture and studies for susceptibility, especially when the infection does not respond to the more traditional, routine antimicrobial treatment. A report from Australia described the development of a perforation of the tympanic membrane in 22 patients who had fungal external otitis, but most perforations closed following successful treatment of the external otitis. Also, tuberculosis may be involved in acute and chronic middle-ear infections with perforation and otorrhea.

**Management.** Antimicrobial therapy for children with acutely perforated eardrums should be the same as or similar to that recommended for those with AOM when a perforation is not present (see Chapter 8). The current trend toward not actively treating AOM with an antibiotic agent is not acceptable when the attack is complicated by rupture of the tympanic membrane with otorrhea because the episode can be characterized as severe; thus, active treatment is indicated. When an aural discharge is present, it is desirable to culture the drainage. The antimicrobial regimen can then be adjusted according to the results of the Gram stain, culture, and susceptibility testing. The most effective method for obtaining a sample of the discharge is to remove as much of the purulent material as possible from the external canal by suction or a cotton-tipped applicator and then aspirate pus directly at or through the perforation, using a spinal needle attached to a tuberculin syringe or an Alden-Senturia trap and suction. Some experts would argue against using ototopical medication when a perforation is present because of the potential danger of ototoxicity. Some children, however, benefit when otic drops are instilled into the external canal.

In addition to the systemic antibiotic therapy, it is advisable to add an ototopical medication, which will not only aid in eliminating the organisms from the external auditory canal, but if the ear drops are pumped into the canal by gently applying pressure on the tragus (eg, four to five times), the medication can enter the middle ear through the perforation and potentially aid in curing the middle-ear infection. Also, it is important to treat the initial canal infection to prevent potentially pathogenic bacteria from the external ear canal entering the middle ear, such as *Pseudomonas* and *S. aureus*, causing a secondary infection of the middle ear. If the suppurative process in the external canal and middle ear is not effectively treated in the acute stage, the infection can progress into chronic suppurative otitis media; chronic infection is prevented by effective treatment of acute infection. The most common bacterium cultured from the middle ear in chronic suppurative otitis media that develops after an acute otorrhea is *P. aeruginosa* and not the common bacteria isolated initially (eg, *S. pneumoniae* and *H. influenzae*) from AOM with or without perforation. Many clinicians advocate applica-
tion of an antibiotic-corticosterone otic medication whenever a discharge is present despite the possibility of ototoxicity because the topical medication may treat or prevent an external canal infection and hasten the resolution of the middle-ear infection. Even though the new ototopical antimicrobial agent ofloxacin (Floxin Otic, Daiichi Pharmaceutical Corp, Montvale, NJ) is approved for treatment of AOM and otorrhea in children only when a tympanostomy tube is in place, this medication should be equally effective when a perforation is present.128,129 This agent is currently the only ototopical medication that has been demonstrated to be safe and effective and is approved for use when a tympanostomy tube is present.128 A recent clinical trial has shown that topical ciprofloxacin-dexamethasone otic solution (Ciprodex, Alcon, Inc, Fort Worth, TX) may be superior to ofloxacin for post-tympanostomy tube otorrhea, which is probably due to the addition of the corticosteroid; also, ciprofloxacin-dexamethasone is effective when granulation tissue on the tympanic membrane is present.130,131 Some have advocated the use of other ototopical agents, but only these two agents cited above are approved for use when the tympanic membrane is not intact. Thus, ototopical agents such as Burow’s solution (aluminum acetate, 13%), even though relatively inexpensive and shown to be effective for the offending bacteria,132 are not currently indicated when the middle ear is exposed to the external canal. Likewise, the other agents listed in Table 3 are not currently indicated, such as the still commonly prescribed combination of neomycin, polymyxin B sulfate, and hydrocortisone otic suspension (Pediotic Suspension, Monarch Pharmaceuticals, Bristol, TN), owing to their potential ototoxicity.133,134 Therefore, it is likely that applying an ototopical antibiotic medication during the acute episode would prevent the secondary chronic infection.

The discharge, especially when it is profuse, should be prevented from draining onto the pinna and adjacent areas because this usually results in dermatitis (see the section on external otitis). The parent should be instructed to keep cotton in the external auditory meatus and change it as often as necessary to keep the canal dry. Cotton-tipped applicators should not be used by the child or parent. Prevention of water entering the ear canal is advisable; swimming is not recommended until the infection is cured and the tympanic membrane is healed.

The tympanic membrane will frequently heal after the suppurative process in the middle ear ends. The defect usually closes within a week after onset of infection. When persistent discharge lasts longer than the initial 10-day course of antibiotic treatment, the child requires more intensive evaluation and aggressive management. In addition to obtaining a culture of the purulent material from the middle ear and adjusting antimicrobial agents, frequent cleaning of the canal, followed by instillation of ototopical drops, may also be required.

The presence of acute mastoiditis with peristeitis or acute mastoid osteitis should be suspected if the child has persistent otalgia, tenderness of the ear to touch, erythema, and

<table>
<thead>
<tr>
<th>Generic Name</th>
<th>Trade Name (Company)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Acetic acid (2%) otic solution</td>
<td>VoSol (Wallace Laboratories, Cranbury, NJ)</td>
</tr>
<tr>
<td>Acetic acid (2%) and hydrocortisone (1%) otic solution</td>
<td>VoSol HC (Wallace Laboratories, Cranbury, NJ)</td>
</tr>
<tr>
<td>Acetic acid 2% in aqueous aluminum acetate otic solution</td>
<td>Otic Domeboro (Bayer Corporation, West Haven, CT)</td>
</tr>
<tr>
<td>Ciprofloxacin hydrochloride and hydrocortisone otic suspension</td>
<td>Ciprofloxin HC Suspension (Alcon, Humacao, PR)</td>
</tr>
<tr>
<td>Colistin sulfate–neomycin sulfate–thonzonium bromide–hydrocortisone acetate otic suspension</td>
<td>Cortisone-TC Otic Suspension (Monarch Pharmaceuticals, Bristol, TN)</td>
</tr>
<tr>
<td>Neomycin, polymyxin B sulfate, and hydrocortisone otic suspension</td>
<td>Pediotic Suspension (Monarch Pharmaceuticals, Bristol, TN)</td>
</tr>
<tr>
<td>Ofloxacin otic solution 0.3%</td>
<td>Floxin Otic (Daiichi Pharmaceutical Corp., Montvale, NJ)</td>
</tr>
<tr>
<td>Ciprofloxacin-dexamethasone otic suspension</td>
<td>Ciprodex (Alcon Laboratories, Fort Worth, TX)</td>
</tr>
</tbody>
</table>

*Agents listed are commonly used in children and adults; only Floxin Otic and Ciprodex have US Food and Drug Administration approval for this indication, but only for adults.
swelling in the postauricular area. Computed tomography (CT) of the mastoids may be helpful but is not always diagnostic of mastoid osteitis (see the section on mastoiditis). CT scans can also help diagnose spread of infection outside the middle ear and mastoid. Even if an intratemporal (or intracranial) complication is not readily apparent, if the aural discharge persists 2 or 3 weeks after onset of AOM that has been treated with appropriately administered oral antibiotics, the patient may require hospitalization. The child should be evaluated again thoroughly in a search for an underlying illness that would interfere with resolution of the infection. The otologic assessment should include an examination of the entire external canal and tympanic membrane, with use of the otomicroscope to determine whether another otologic condition, such as a cholesteatoma or neoplasm, is present. If an adequate examination cannot be performed with the child awake, it should be carried out under general anesthesia, at which time a culture can be obtained directly from the middle ear. If no condition other than the perforation and subAOM is found, parenteral antimicrobial agents should be administered. The selection of both the systemic and topical antimicrobial agents should be based on the results of cultures. A gram-negative organism (eg, P. aeruginosa) is frequently present at this stage, and management is essentially as recommended later under “Chronic Perforation with Chronic Otitis Media (Chronic Suppurative Otitis Media).”

With this method of management, the infection will usually subside. If the discharge persists, an exploratory tympanostomy and complete simple mastoidectomy are indicated even if no signs and symptoms of mastoid osteitis are present and the CT scans fail to show osteitis (ie, coalescence). During surgery on the middle ear and mastoid, a thorough search for another cause of the persistent infection must be made. On occasion, a cholesteatoma that could not be visualized through the otomicroscope will be found. Resolution of infection in the middle ear and mastoid will invariably follow the surgery because mastoid osteitis is the usual cause of this complication of AOM.

Fortunately, the occurrence of such cases is now uncommon, and the perforation usually heals rapidly. However, the defect will frequently remain open without evidence of otitis media (with or without discharge). If the perforation remains free of infection, it will frequently close in a few months. At this stage, no attempt at surgical closure of an uncomplicated perforation is indicated. If there is no sign of progressive healing after 3 months or more, management should be as described later (see “Chronic Perforation”), such as tympanoplasty.

**Prevention.** In patients who are prone to recurrent AOM and who perforate their tympanic membrane, prevention is desirable (described in detail in Chapter 8). In certain populations at high risk of AOM and perforation, which subsequently progresses to chronic suppurative otitis media, the most effective method of prevention of the chronic infection is to prevent development of an acute perforated tympanic membrane during an attack of AOM. This is most effectively accomplished by treating the child early, appropriately, and adequately with an antimicrobial agent at the onset of the episode of AOM (see “Chronic Perforation”).

**Chronic Perforation**

A perforation of the tympanic membrane may remain open after an episode of AOM or after spontaneous extrusion (or removal) of a tympanostomy tube. When an acute perforation is present with no signs of healing and there are no signs of otitis media for 3 months or longer, the perforation is considered chronic and possibly permanent. If chronic suppurative otitis media is present, the perforation may close spontaneously after appropriate treatment if the infection resolves. However, healing of the perforation may not occur for the same reason that chronic perforations, without chronic infection, fail to heal: the presence of squamous epithelium at the edges of the perforation prevents spontaneous
repair. The effect on hearing of a small chronic perforation is not significant, regardless of its location and in the absence of other middle-ear abnormalities. A large perforation, however, can be associated with an appreciable conductive hearing loss (eg, 20–30 dB).

**Classification**

Chronic perforation of the tympanic membrane can be classified as chronic perforation without otitis media and chronic perforation with otitis media. Chronic perforation with otitis media can be further classified into chronic perforation with AOM, with or without otorrhea, and chronic perforation with chronic otitis media (chronic suppurative otitis media), with or without otorrhea. The last disease stage is invariably associated with chronic mastoiditis (see Chapter 1).

**Chronic Perforation without Otitis Media**

Chronic perforation without otitis media has been inappropriately termed chronic otitis media inactive. This terminology is confusing and incorrect in many instances. The patient may never have an attack of AOM—other than possibly the one that originated the perforation—or an episode of chronic suppurative otitis media (see Chapter 1).

**Epidemiology.** The incidence of chronic perforation in the pediatric population has not been formally studied, but chronic perforation is a frequent reason for referral to an otolaryngologist. In a study of tympanoplasty for chronic perforation—not associated with infection, cholesteatoma, or ossicular involvement—in children from Finland, Vartiainen and Vartiainen operated on 60 children during a 15-year period. Caylan and colleagues described a similar number of children who had surgical repair of a chronic perforation in an Italian center during a 10-year period. They also reviewed the literature from 1974 to 1991 and found that 640 children had tympanoplasty as reported in 12 studies. Another review of the literature from 1979 to 1995 reported that 870 had tympanoplasty, and still another review found 1,741 cases reported between 1985 and 1998. However, many reports combine children and adults in the study, which often prevents assessment of the number of children included. Many reports in the literature also combine children who have chronic perforation without active infection with those patients who have chronic suppurative otitis media (see later, “Chronic Perforation with Chronic Otitis Media [Chronic Suppurative Otitis Media]”). In addition, the incidence of tympanoplasties performed in children would not accurately reflect the true incidence of chronic perforation because many physicians elect to withhold surgery until later in the child’s life. More important, most otolaryngologists do not report their results of tympanoplasty in children. Thus, the number of children being operated on for repair of a chronic perforation is a great deal larger than is reported in the literature. Next to myringotomy, with or without tympanostomy tube insertion, tympanoplasty is the most common ear operation performed in children.

Chronic perforation, as a complication of otitis media, is more prevalent in certain racial groups that also have a high prevalence and incidence of perforations associated with acute middle-ear infection. In 1970, new cases of chronic perforation (with or without chronic suppurative otitis media) were reported in 8% of the native population of Alaska, although this rate appears to be dropping. Similar rates have been reported in Native American populations. Zonis reported that of 207 Apache Indian children examined in Canyon Day, Arizona, 17 (8%) had chronic perforations as their only sign of otitis media, whereas Todd and Bowman returned to the same village 16 years later in 1983, examined 145 Indian children living there at the time, and found only 1 child who had a perforation of the tympanic membrane but 12 (8%) children who had other evidence of otitis media. Even though chronic perforation with chronic suppurative otitis media is common in young Australian Aboriginal children, chronic perforation without otitis media is less common.
Indeed, Morris and colleagues reported that 15% of 711 of these children, aged 6 to 30 months, had chronic suppurative otitis media, but only 2% had a perforation, without otitis media.116 Because many of these chronic perforations are associated with chronic middle-ear and mastoid infection, the epidemiology of this complication of otitis media is presented in more detail under “Chronic Perforation with Chronic Otitis Media (Chronic Suppurative Otitis Media).”

Chronic perforation of the tympanic membrane is a recognized complication of tympanostomy tube insertion.143 Of 1,062 ears of children who received tympanostomy tubes in one study reported from West Germany, 26 ears (2.5%) had a persistent perforation.144 This figure, however, depends on the site of the tube placement and the type of tube used. The rate of chronic perforation as a complication of tube placement has been reported to be as low as 0.5% and as high as 25%. The conventional tubes are associated with the lowest rate and the permanent tubes with the highest rate. Of the 215 children prospectively observed for at least 2 to 3 years in the three clinical trials of efficacy and safety of tympanostomy tube insertion conducted in Pittsburgh, 2.4% had to eventually have tympanoplasty for chronic perforation (see “Myringotomy and Tympanostomy Tube Placement” in Chapter 8).111,112,145

Management. The management of so-called dry chronic perforation (more appropriately termed chronic perforation without otitis media) in children is both difficult and controversial. On the one hand, the perforation provides ventilation and drainage of the middle ear. On the other hand, the physiologic protective function of the eustachian tube–middle-ear system is impaired; the middle ear is too open. The middle ear and mastoid gas cells no longer have a gas cushion to prevent nasopharyngeal secretions from entering the ear, which can then result in reflux otitis media (Figures 7 and 8).127 In addition, the open tympanic membrane can permit contaminated water to enter the middle ear during bathing and swimming. Therefore, the dilemma of when to

Figure 7. A perforation of the tympanic membrane may promote the reflux of secretions into the middle ear from the nasopharynx because the middle-ear air cushion is not present (see also Figure 8).

Figure 8. Flask model showing how a perforation of the tympanic membrane may result in reflux of nasopharyngeal secretions into the middle ear. The nasopharynx–eustachian tube–middle-ear–mastoid air cell system is likened to a flask with a narrow neck. When the system is intact, liquid is prevented from entering the body of the flask, but when the body of the flask is not intact (ie, a perforation is present), liquid can readily flow through the system.
close such a perforation is comparable to that regarding the most appropriate time to remove a tympanostomy tube: a small, uncomplicated chronic perforation and a tympanostomy tube have similar benefits and risks. Like a tympanostomy tube, a perforation may be beneficial for a child who had had recurrent AOM or chronic otitis media with effusion before the perforation developed, but recurrent acute reflux otitis media with discharge, which can progress into chronic suppurative otitis media, may become a problem, making repair of the eardrum defect a consideration. However, recurrent AOM that results in otorrhea through a chronic perforation can be effectively treated and even prevented without repair of the tympanic membrane (or removal of a tympanostomy tube).

When the episodes are infrequent, the treatment of each bout should be the same as recommended for an acute perforation associated with AOM. If the episodes of acute infection are frequent and the interval between bouts is short, preventive measures are recommended. The options appropriate today are the same as those recommended for infants and children whose tympanic membranes are intact but who have frequently recurrent AOM (see Chapter 8). Reducing the risk of exposure to viral infections (eg, eliminating day care or choosing a day-care setting with as few children as possible) may prove helpful. The currently available pneumococcal vaccine (after the age of 2 years) and the influenza vaccine can be administered. A search for an underlying problem, such as an immune disorder, adenoid hypertrophy, paranasal sinusitis, or allergy, may also help. If none of these recommendations prove beneficial and an underlying problem is not uncovered, prophylaxis with an antimicrobial agent during the risk months for upper respiratory tract infections (late fall, winter, and spring) can be initiated. This method was demonstrated to be effective in reducing recurrent otorrhea in Alaskan natives.\textsuperscript{146} A prolonged course of a prophylactic antimicrobial agent (eg, amoxicillin, 20 mg/kg, given in one dose before bedtime) may be considered to prevent recurrent middle-ear infection and discharge. The selection of the agent should be based on the results of the cultures obtained from previous episodes of discharge. The dosage and duration of treatment should be the same as those recommended for children who have had frequently recurrent AOM without a perforation. Today, however, with the ever-increasing rate of antibiotic-resistant otogenic bacteria that has been attributed to the overuse of antibiotics for treatment and prolonged low-dose prophylaxis, other preventive measures should be attempted before systemic antimicrobial prophylaxis is recommended. One method that can be helpful is to give the child a course of an ototopical ofloxacin during the duration of an upper respiratory tract infection. This method is more appealing than systemic administration of an antimicrobial agent because drug-resistant bacteria apparently do not develop during topical treatment. Also, consideration should be given to repairing the perforation. Children in whom an attack of acute middle-ear infection and discharge persists despite adequate medical treatment and in whom the infection is thought to be chronic should be evaluated and managed as described in the section on chronic suppurative otitis media.

Most children who have a defect in the eardrum that is thought to be preventing otitis media can be watched until the risk of recurrence of infection is low enough to consider surgical closure of the perforation. A perforation, such as a tympanostomy tube, may prevent development of a retraction pocket and, subsequently, a cholesteatoma. If the eustachian tube function is poor, it is desirable to delay such surgery as long as possible to prevent such complications. Surgery for a chronic perforation should not necessarily be withheld in children because of recurrent episodes of otitis media and discharge; the perforation may be causing the middle-ear infection (as a result of reflux from the nasopharynx) rather than preventing disease.\textsuperscript{127} Also, perforation of the tympanic membrane is associated with some degree of hearing loss.\textsuperscript{147}
Tympanoplasty. Tympanoplasty is not as successful in children as it is in adults. These results may be due to the higher incidence of upper respiratory tract infection leading to otitis media in children and the unpredictability of their eustachian tube function. However, the success rate of tympanoplasty is still high, as shown in Table 4. The study by Bluestone and colleagues had the lowest success rate compared with later studies because a medial graft technique was used initially and the criteria for success versus failure were rigid during a prolonged follow-up period. Since then, a variety of surgical techniques have been used, with success rates comparable to those reported in the recent literature.

Optimal ages at which to perform tympanoplasty have variously been stated to be from 3 years to puberty. Lau and Tos reviewed the outcome of 124 tympanoplasties performed on 116 children between the ages of 2 and 14 years and found a 92% success rate of grafts; these authors recommend tympanoplasty in children of all ages, as do Paparella, Vartiainen and Vartiainen, Bajaj and colleagues, and Denoyelle and colleagues. In the Lau and Tos study, however, the initial 92% success rate of the procedure fell to 64% over time; 14% required tympanostomy tubes, 5% had persistent middle-ear effusion, and 9% had postoperative atelectasis. Tos updated these outcomes in the young child. Similar outcomes were reported by Manning and colleagues in a study of 56 children (63 ears). Even though 78% of the grafts healed, only 52% of the children had a healed graft and adequate middle-ear function during the postoperative period. Sheehy and Anderson do not recommend elective tympanic membrane grafting for children who are younger than 7 years because of the possibility of postoperative otitis media. Other surgeons reported tympanoplasty outcomes that agree with the recommendation of Sheehy and Anderson and not with Tos and Lau. In general, infants and children younger than 7 years have less favorable outcomes after tympanoplasty than do older children and adults. The controversy over which age is most appropriate is due to definitions of failure and the duration of follow-up, as well as the intervals between observations. Graft take should not be the only outcome measure. Additional outcomes should be recurrence of middle-ear effusion, high negative middle-ear pressure, atelectasis, subsequent reperforation, the need for tympanostomy tube placement, and serial assessment of hearing during a long period. When children aged 7 years and older are operated on, their outcomes are relatively good. A meta-analysis of tympanoplasty in children from 1966 to 1997 by Vrabec and colleagues revealed that the success rate increased with advancing age and that none of the other parameters studied were shown to be significant predictors of success.

The level of experience of the surgeon has also been called into question. Whereas a teaching institution has a responsibility to train young surgeons, that is not the case with the private practitioner or in areas where only the attending surgeon performs the surgery. These factors are also probably related to the variation in post-

### Table 4. OUTCOME OF TYMPANOPLASTY, TYPE I (MYRINGOPLASTY), IN CHILDREN WHO HAD CHRONIC PERFORATION OF THE TYMPANIC MEMBRANE AS REPORTED FROM 20 CENTERS FROM 1979 TO 2005

<table>
<thead>
<tr>
<th>Study (Year)</th>
<th>Number of Ears</th>
<th>Success Rate (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Bluestone et al (1979)</td>
<td>51</td>
<td>35</td>
</tr>
<tr>
<td>Cohn et al (1979)</td>
<td>21</td>
<td>81</td>
</tr>
<tr>
<td>Raine and Singh (1983)</td>
<td>114</td>
<td>81</td>
</tr>
<tr>
<td>Adkins and White (1984)</td>
<td>30</td>
<td>87</td>
</tr>
<tr>
<td>François et al (1985)</td>
<td>150</td>
<td>81</td>
</tr>
<tr>
<td>Lau and Tos (1986)</td>
<td>155</td>
<td>92</td>
</tr>
<tr>
<td>Ophir et al (1987)</td>
<td>172</td>
<td>79</td>
</tr>
<tr>
<td>Koch et al (1990)</td>
<td>64</td>
<td>73</td>
</tr>
<tr>
<td>Prescott and Robartes (1991)</td>
<td>114</td>
<td>84</td>
</tr>
<tr>
<td>Kessler et al (1994)</td>
<td>200</td>
<td>92</td>
</tr>
<tr>
<td>Black and Wormald (1995)</td>
<td>100</td>
<td>75</td>
</tr>
<tr>
<td>Chandrasekar et al (1995)</td>
<td>268</td>
<td>81</td>
</tr>
<tr>
<td>Mitchell et al (1997)</td>
<td>342</td>
<td>80</td>
</tr>
<tr>
<td>Vartiainen and Vartiainen (1997)</td>
<td>60</td>
<td>90</td>
</tr>
<tr>
<td>Caylan et al (1998)</td>
<td>51</td>
<td>82</td>
</tr>
<tr>
<td>Bajaj et al (1998)</td>
<td>45</td>
<td>91</td>
</tr>
<tr>
<td>Denoyelle et al (1999)</td>
<td>231</td>
<td>83</td>
</tr>
<tr>
<td>Carr et al (2001)</td>
<td>89</td>
<td>75</td>
</tr>
<tr>
<td>Collins et al (2003)</td>
<td>72</td>
<td>82</td>
</tr>
<tr>
<td>Couloigner et al (2005)</td>
<td>29</td>
<td>71</td>
</tr>
</tbody>
</table>
operative outcomes reported in the literature. Persistent perforation is a problem in some patients even in the best of circumstances.\(^\text{167}\)

Some surgeons advocate removing the adenoids before repairing a chronic tympanic membrane perforation, but Varthainen and Varthainen reported that their graft failures occurred in patients who had had a previous adenoidectomy.\(^\text{135}\) They suggest that children who had had adenoidectomy in the past most likely had more middle-ear disease and were more likely to have failure of tympanoplasty. Indications for adenoidectomy in children who have a chronic perforation should be similar to the indications for children in whom the tympanic membrane is intact (see Chapter 8). Also, on occasion, a tympanostomy tube has been recommended as part of the procedure.\(^\text{168}\)

The addition of a mastoidectomy to the tympanoplasty, when a chronic perforation without otitis media is present, has been advocated by some,\(^\text{169}\) but the evidence that the benefits of mastoidectomy outweigh the potential risks (eg, facial paralysis, injury to the inner ear, increased time of general anesthesia) has not been demonstrated, and we do not advocate this additional surgical procedure.\(^\text{9}\)

Studying eustachian tube function before the patient with a chronic perforation of the tympanic membrane is operated on may be helpful in determining the potential results of tympanoplasty surgery because the function of the tube can affect outcome.\(^\text{170}\) Bluestone and colleagues, in a study of 45 children with chronic perforation, found that the assessment of eustachian tube function was an aid in preoperative assessment.\(^\text{151}\)

In a subsequent study at the same institution, Manning and colleagues evaluated eustachian tube function preoperatively in 56 children and found that normal function was associated with a good outcome, whereas poor function did not predict a poor outcome.\(^\text{159}\) Kumazawa and colleagues reported that they consider preoperative evaluation with eustachian tube function tests to be helpful in prognosis.\(^\text{171}\) An alternative method to assess eustachian tube function when actual testing is unavailable is to observe the contralateral ear, if the tympanic membrane is intact, for four or more seasons to determine if middle-ear disease recurs, for example, otitis media, atelectasis, and symptoms of eustachian tube dysfunction. This predictor of outcome has not been formally tested, but it does not make sense to repair the tympanic membrane perforation in the face of recurrent middle-ear disease in the opposite ear; in children, eustachian tube function is usually similar in both ears. Collins and colleagues agree that the status of the contralateral ear is an important factor related to outcome.\(^\text{172}\)

Thus, children, especially those who are younger than 7 years, are uncertain candidates for tympanoplasty surgery because as a group, their eustachian tube function is not as good as that of adults. Improvement may occur in some children, indifferent results are obtained in others, and there are more problems with middle-ear infections in a third group. The problem for the clinician is deciding which child should have the perforation repaired. The development of an improved method of testing the eustachian tube, a method more indicative of the actual function available for clinical use, could possibly help in this decision-making process. Currently, we recommend tympanoplasty for children after the age of 6 years because the incidence of middle-ear disease declines after that age, probably owing to maturation of the structure (length) and function of the eustachian tube and maturation of the child’s immunity. In general, the indications for repairing a perforation are similar to the indications for removing a retained tympanostomy tube; both procedures attempt to restore an intact tympanic membrane.

The indications for removing a tympanostomy tube are listed in the section “Myringotomy and Tympanostomy Tube Placement” in Chapter 8. There have been many surgical techniques proposed to repair the defect in the ear-drum.\(^\text{173–175}\) The surgical techniques performed by us are described in detail in Bluestone.\(^\text{9}\)

**Chronic Perforation with Otitis Media**

When an acute perforation of the tympanic membrane is associated with AOM, the infection
can progress into a chronic perforation and chronic suppurative otitis media. This is termed chronic suppurative otitis media. A chronic perforation may also be present with no middle-ear infection but is susceptible to an episode of otitis media. This is initially AOM, which can progress to the chronic stage (ie, chronic suppurative otitis media).

**Classification.** Chronic perforation with otitis media can be classified as follows:

- Chronic perforation with AOM, with or without otorrhea
- Chronic perforation with chronic otitis media, with or without otorrhea

The term chronic suppurative otitis media implies that a chronic perforation of the tympanic membrane is present, as in chronic mastoiditis, but otorrhea may or may not be present.

**Chronic Perforation with AOM.** A chronic perforation of the tympanic membrane can develop AOM, which can then have the following possible outcomes:

- The AOM and otorrhea completely resolve, and the chronic perforation heals.
- The AOM and otorrhea completely resolve, but the chronic perforation persists.
- AOM persists into the chronic stage, that is, chronic suppurative otitis media.
- A suppurative complication, such as acute mastoiditis, develops.

The presumed reason that a chronic perforation occasionally heals after an episode of AOM is that the rim (margins) of the perforation is denuded of the epithelium by the infection; the epithelium at the margins prevented healing before the infection.

When an episode of AOM develops in a child who has a chronic perforation of the tympanic membrane, initial management should be similar to that described earlier when an acute perforation develops during an attack of AOM. If the AOM resolves but the chronic perforation persists, the decision-making process to close the perforation, or not to close it, is the same as described before for a chronic perforation without otitis media.

**Chronic Perforation with Chronic Otitis Media (Chronic Suppurative Otitis Media).** Chronic suppurative otitis media is the stage of ear disease in which there is chronic inflammation of the middle ear and mastoid and in which a nonintact tympanic membrane (chronic perforation or tympanostomy tube) is present. Otorrhea may or may not be evident; a discharge may be present in the middle ear, mastoid, or both, but otorrhea is not evident through the perforation—or tympanostomy tube—or in the external auditory canal. There is no consensus regarding the duration of otitis media to be designated chronic suppurative otitis media. Even though 3 months or longer appears to be appropriate, some clinicians consider a shorter duration of otitis media to be chronic, especially when the causative organism is *Pseudomonas.*

**Terminology.** Chronic suppurative otitis media is the term commonly used when a chronic perforation is associated with chronic otitis media. Mastoiditis is invariably part of the pathologic process. The condition has also commonly been called chronic otitis media, but this term can be confused with chronic otitis media with effusion, which is not a complication of otitis media and in which no perforation of the tympanic membrane is present. Chronic otitis media is also an inappropriate term when a chronic perforation of the tympanic membrane is present but the middle ear and mastoid are free of infection. The proper term for this condition is chronic perforation without otitis media or, more simply, chronic perforation. Some clinicians also inappropriately use the terms chronic otitis media inactive and chronic otitis media active; chronic perforation associated with infection is active, and when infection is absent, it is inactive. Other terms for chronic suppurative otitis media are chronic suppurative otitis media and mastoiditis, chronic purulent otitis media, and chronic
The most descriptive term is **chronic otitis media with perforation, discharge, and mastoiditis**, but this is not commonly used. When a cholesteatoma is also present, the term **cholesteatoma with chronic suppurative otitis media** is appropriately used. However, because an acquired aural cholesteatoma does not have to be associated with chronic suppurative otitis media, cholesteatoma is not part of the pathologic features of the type of ear disease described in this section and is presented as a separate entity in this chapter.\textsuperscript{176,179}

Despite the strict definition of chronic suppurative otitis media presented here, a review of the literature reveals that many reports describing various aspects of chronic suppurative otitis media, such as epidemiology and pathogenesis, improperly include chronic perforation without otitis media in this disease entity.

### Epidemiology

Chronic suppurative otitis media is a major health problem in many populations around the world, affecting diverse racial and cultural groups living not only in temperate climates but in climate extremes ranging from the Arctic Circle to the equator. From a review of approximately 50 reports published during the past 30 years, the disease appears to be prevalent in four groups of populations (Table 5).\textsuperscript{115} The prevalence of chronic otitis media (defined here as chronic perforation with and without suppuration) has been reported to be the highest in the Inuits of Alaska (30–46%), Canada (7–31%), and Greenland (7–12%); Australian Aborigines (12–25%); and certain Native Americans, such as Apache and Navajo tribes (4–8%). Apparently, these North American Indian tribes have higher rates than others.\textsuperscript{180} One study from the Eastern Canadian Arctic compared the rates in Cree

#### Table 5. PREVALENCE OF CHRONIC SUPPURATIVE OTITIS MEDIA*

<table>
<thead>
<tr>
<th>Population</th>
<th>Prevalence (%)</th>
<th>References</th>
</tr>
</thead>
<tbody>
<tr>
<td>Highest</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Inuits</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Alaska</td>
<td>30–46</td>
<td>Brody et al,\textsuperscript{563} Kaplan et al,\textsuperscript{57} Tschopp\textsuperscript{564}</td>
</tr>
<tr>
<td>Canada</td>
<td>7–31</td>
<td>Ling,\textsuperscript{565} Schaefer,\textsuperscript{186} Baxter and Ling,\textsuperscript{566} Timmermans and Gerson,\textsuperscript{567} Baxter,\textsuperscript{568} Baxter et al,\textsuperscript{181,569}</td>
</tr>
<tr>
<td>Greenland</td>
<td>7–12</td>
<td>Pederson and Zachau-Christiansen,\textsuperscript{570} Homoe and Breltau,\textsuperscript{571} Homoe et al,\textsuperscript{172}</td>
</tr>
<tr>
<td>Australian Aborigines</td>
<td>12–25</td>
<td>Stuart et al,\textsuperscript{572} McCafferty et al,\textsuperscript{419} Lewis et al,\textsuperscript{574} Moran et al,\textsuperscript{575} Dugdale et al,\textsuperscript{576} Hudson and Rockette,\textsuperscript{577} McCafferty et al,\textsuperscript{578} Foreman,\textsuperscript{579} Leach et al,\textsuperscript{188} Boswell and Nienhuys,\textsuperscript{113} Morris et al,\textsuperscript{116}</td>
</tr>
<tr>
<td>Native Americans</td>
<td>4–8</td>
<td>Johnson,\textsuperscript{580} Zonia,\textsuperscript{141} Jaffe,\textsuperscript{462} Mortimer,\textsuperscript{581} Wiet,\textsuperscript{463} Todd and Bowman\textsuperscript{142}</td>
</tr>
<tr>
<td>High</td>
<td></td>
<td></td>
</tr>
<tr>
<td>South Pacific Islands</td>
<td>4–6</td>
<td>Eason et al,\textsuperscript{582} Tonkin,\textsuperscript{583} Giles and O’Brien,\textsuperscript{584}</td>
</tr>
<tr>
<td>New Zealand Maori</td>
<td>4</td>
<td>Elango et al,\textsuperscript{585} Dever et al,\textsuperscript{586,587} Chan et al,\textsuperscript{188}</td>
</tr>
<tr>
<td>Malaysia</td>
<td>4</td>
<td></td>
</tr>
<tr>
<td>Micronesia</td>
<td>4</td>
<td></td>
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<tr>
<td>Africa</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Sierra Leone Gambia</td>
<td>6</td>
<td>Seeley et al,\textsuperscript{179} McPherson and Holborow,\textsuperscript{489} Hatcher et al,\textsuperscript{190}</td>
</tr>
<tr>
<td>Nigeria</td>
<td>4</td>
<td>Miller et al,\textsuperscript{183} Okeowa,\textsuperscript{184} Manni and Lema,\textsuperscript{591} Bastos et al,\textsuperscript{592} Minja and Machemba,\textsuperscript{593}</td>
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<tr>
<td>Tanzania</td>
<td>2–3</td>
<td></td>
</tr>
<tr>
<td>Low</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Korea</td>
<td>2</td>
<td>Kim et al,\textsuperscript{594} Kapur,\textsuperscript{595}</td>
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<tr>
<td>India</td>
<td>2</td>
<td></td>
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<tr>
<td>Saudi Arabia</td>
<td>1.4</td>
<td>Muhaimeid et al,\textsuperscript{596}</td>
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<tr>
<td>United States</td>
<td>&lt; 1</td>
<td>Casselbrant et al,\textsuperscript{597,598} Zeisel et al,\textsuperscript{599} Paradise et al,\textsuperscript{600}</td>
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<td>United Kingdom</td>
<td>&lt; 1</td>
<td>Mawson and Ludman,\textsuperscript{190} Williamson et al,\textsuperscript{191} Fiellau-Nikolaesen,\textsuperscript{602} Marttila,\textsuperscript{603}</td>
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<tr>
<td>Denmark</td>
<td>&lt; 1</td>
<td></td>
</tr>
<tr>
<td>Finland</td>
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*Some reports included patients who had chronic perforation but without otitis media.*
Indian schoolchildren with the rates in Inuit children living in the same area and found the rate to be 22% in the Inuit but only 1% in the Cree. In a recently reported prevalence study of 711 Aboriginal children from 29 remote communities in Australia, 118 (15%) had chronic suppurative otitis media, which verifies that this stage of middle-ear disease is still a major health problem in this special population.

Populations with moderately high rates are certain natives of the South Pacific islands, such as natives of the Solomon Islands (4–6%), New Zealand Maori (4%), natives of Malaysia (4%), and natives of Micronesia (4%) (in contrast to these high rates in some islands of the South Pacific, natives of Melanesia have an extremely low rate, less than 1%); and some African populations, such as those of Sierra Leone (6%), Gambia (4%), Kenya (4%), and Tanzania (2–3%). However, not all reports from Africa have documented these relatively high rates. One study from Nigeria reported less than 1% with the disease; another study found a 4% rate. One study of South African rural blacks also found a rate of less than 1%. A study from south India found the rate to be 6% in rural children.

Populations with relatively low rates of chronic otitis media are those of Korea (2%), India (2%), and Saudi Arabia (1.4%). Studies from highly industrialized nations have reported the lowest rates (none or less than 1%), such as the United States, Finland, the United Kingdom (in one adult population, the rate has been reported to be 3.1%), and Denmark. However, with the widespread use of tympanostomy tubes in these countries, chronic suppurative otitis media occurs as a not uncommon complication in infants and children in whom these tubes have been inserted.

Risk factors that have been attributed to the high rates of chronic suppurative otitis media in these populations are a lack of breast-feeding, overcrowding, poor hygiene, poor nutrition, passive smoking, high rates of nasopharyngeal colonization with potentially pathogenic bacteria, and inadequate and unavailable health care.

**Pathogenesis.** The etiology and pathogenesis of chronic suppurative otitis media are multifactorial, involving one or more of the risk factors noted before, but chronic suppurative otitis media begins with an episode of AOM. Thus, the factors that have been associated with AOM may initially be involved, such as upper respiratory tract infection; anatomic factors, such as eustachian tube dysfunction; host factors, such as young age; immature or impaired immunologic status; the presence of an upper respiratory allergy; familial predisposition; the presence of older siblings in the household; male sex; race; method of feeding (bottle versus breast); and environmental (eg, smoking in the household) and social factors. Probably the most important factors related to the onset of AOM in infants and young children are immaturity of the structure and function of the eustachian tube and immaturity of the immune system.

AOM with perforation (or when a tympanostomy tube is present) usually precedes chronic suppurative otitis media, but in certain high-risk populations, such as the Australian Aborigines, chronic otitis media with effusion is initially present. Factors most likely related to the progression of AOM into the chronic stage have been noted before, but most likely the process, if it is long-standing, results in a chronic osteitis of the middle-ear cleft. Figure 9 shows the possible outcomes of an episode of AOM that may result in chronic suppurative otitis media. Because a spontaneous perforation commonly accompanies an episode of AOM that is untreated with an antimicrobial agent, and less commonly despite adequate treatment, it may be part of the natural history of the disease process rather than a complication. Figure 10 also shows the sequence of events after an attack of AOM that can lead to a chronic perforation or chronic suppurative otitis media.

When a chronic perforation or tympanostomy tube is present and there is no evidence of infection, reinfection probably occurs in one of two ways:

1. Bacteria from the nasopharynx gain access to the middle ear by reflux or insufflation of
nasopharyngeal secretions (owing to the infant crying, nose blowing, or swallowing when a nasal obstruction is present, that is, the Toynbee phenomenon\textsuperscript{191}) through the eustachian tube because the middle-ear air cushion is lost; an episode usually occurs with an upper respiratory tract infection. In most instances, these bacteria are initially the same as those isolated when AOM occurs behind an intact tympanic membrane, such as \textit{S. pneumoniae} and \textit{H. influenzae}.\textsuperscript{118} After the acute otorrhea, \textit{P. aeruginosa}, \textit{S. aureus}, and other organisms from the external ear canal enter the middle ear through the nonintact tympanic membrane, which results in secondary infection and acute otorrhea and chronic suppurative otitis media (Figure 11). Bacteria commonly isolated from ears with chronic suppurative otitis media (eg, \textit{P. aeruginosa}, \textit{S. aureus}) are isolated as normal flora from the

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**Figure 9.** Possible outcomes of acute otitis media. CSOM = chronic suppurative otitis media; OME = otitis media with effusion.

**Figure 10.** One possible sequence of events after an episode of acute otitis media that can result in a chronic perforation of the tympanic membrane or chronic suppurative otitis media in a right ear (1). During an upper respiratory tract infection (URI), nasopharyngeal secretions with viruses and bacteria can gain entrance into the middle ear (2), causing acute otitis media, perforation of the tympanic membrane, and otorrhea (3), after which bacteria from the ear canal can enter the middle ear (4), resulting in chronic suppurative otitis media (5). The infection resolves, but the perforation persists and becomes chronic (5a), or there is both resolution and healing of the perforation (5b).
external auditory canal and thus can be readily available as secondary invaders into the middle ear.

2. Chronic suppurative otitis media also occurs when the middle-ear cleft is contaminated by organisms (e.g., *P. aeruginosa*) present in water that enters the nonintact eardrum during bathing and swimming (Figure 12). As described before, certain populations are at high risk of development of chronic perforation of the tympanic membrane with and without chronic suppurative otitis media. From this review, it appears that there are special populations who are at highest risk of development of chronic suppurative otitis media who live in diverse geographic regions in the world (see Table 5), which would make climate an unlikely explanation for their disease. In these racial groups, it is likely that the pathogenesis of their disease is due to genetic differences in their eustachian tube function. The tube is most likely hyperpatent, which may be due to a semipatulous or patulous lumen, the tube is too short, or both. (Histopathologic temporal bone specimens of young individuals who had cleft palate and those who had Down syndrome—both conditions of high risk of otitis media—had statistically shorter eustachian tubes than age-matched specimens from individuals without these disorders.) Nevertheless, environmental and behavioral factors and the availability of adequate health care are also important in these racial groups. Thus, these other factors (i.e., other than abnormalities of the eustachian tube) would make other populations also at risk. What other host factors contribute to their high rate of disease? One important possible explanation is that these groups have eustachian tubes that make them more prone than others to middle-ear infection. Indeed, a study identified anatomic differences in the bony segment of the eustachian tube in the

Figure 11. One of two proposed sequences of events in the pathogenesis of recurrence of otorrhea when the tympanic membrane is not intact and active infection is absent in a right ear (1). After an upper respiratory tract infection (URI), nasopharyngeal organisms are refluxed or insufflated into the middle ear (2), which results in an acute otitis media and otorrhea (3). Organisms from the external canal can then enter the middle ear (4), which can result in chronic suppurative otitis media (5). TVP = tensor veli palatini muscle (see Figure 12).
craniofacial structures of Eskimo, American Indian, white, and black crania. In a clinical study, White Mountain Apache Indians were also found to have eustachian tubes that were semipatulous (of low resistance) compared with those of a group of white subjects. Similar findings have been reported in Canadian Inuits. Ratnesar calibrated the eustachian tube with ureteric catheters in Canadian Inuits and white individuals and found the tube to be larger in Inuits than in whites.

One study reported that environment plays a role in a high-risk population but that genetic differences are probably more important in those who are at high risk. Adopted Apache children had more episodes of AOM than did their non-Apache siblings and had an illness rate similar to that of Apache children who remained on the reservation. In a study from Greenland of 591 children aged 3, 4, 5, and 8 years, 9% had chronic perforation, with and without chronic supplicative otitis media, which was statistically most common in children whose parents were both native Greenlanders.

It is likely that eustachian tube dysfunction is involved in the process even in individuals who are not members of high-risk populations or who have an obvious craniofacial abnormality. In a study of eustachian tube function in the ears of Japanese children and adults who had chronic perforations, Iwano and colleagues found impaired active opening function of the tube. They concluded that the tube was functionally obstructed; however, organic (ie, mechanical or anatomic) obstruction was also considered to be involved in the pathogenesis in adults.

One possible mechanism that can cause chronic supplicative middle-ear infection is an allergic reaction from a tympanostomy tube, as described in one child by Dohar. But this is an extremely unusual case. Most presumed similar cases are not truly an allergic reaction to the components of a tympanostomy tube but acute and chronic infection owing to the middle ear.

Figure 12. One of two proposed sequences of events in the pathogenesis of recurrence of otorrhea when the tympanic membrane is not intact and active infection is absent in a right ear (1). The middle ear is contaminated by organisms in water that enter the middle ear through the perforation (2), which can result in acute otitis media and otorrhea (3) and, if persistent, chronic supplicative otitis media (4). TVP = tensor veli palatini muscle (see Figure 11).
being nonintact and bacterial contamination from the external canal or reflux of nasopharyngeal secretions into the middle ear.\textsuperscript{127}

**Microbiology.** The bacteria that cause the initial episode of AOM and perforation or acute otorrhea through a tympanostomy tube are usually not those that are isolated from chronic suppurative otitis media.\textsuperscript{118} The most common organism isolated from around the world is *P. aeruginosa*; *S. aureus* is also found, but less commonly.\textsuperscript{201–206} Table 6 shows the frequency of bacteria isolated from children with chronic suppurative otitis media at the Children’s Hospital of Pittsburgh.\textsuperscript{207} More recently, MRSA has been isolated from draining ears.\textsuperscript{119} Anaerobic bacteria were isolated infrequently in this study. Anaerobic bacteria have been isolated from ears with chronic suppurative otitis media, but whether they are true pathogens remains to be demonstrated. Brook isolated *Bacteroides melaninogenicus* in 40% and *Peptococcus* species in 35% of middle-ear exudates; the collection of exudate was performed through the perforation in the tympanic membrane with use of an 18-gauge needle covered by a plastic cannula.\textsuperscript{208}

There have also been reports of isolation of unusual organisms, such as *Mycobacterium tuberculosis*, *Mycobacterium chelonae*, *Mycobacterium avium-intracellulare*, *Blastomyces dermatitidis*, *Actinomyces*, *Alcaligenes piechaudii*, and *Candida* species.\textsuperscript{120,121,209–216}

**Pathology.** It is important to understand the pathologic process of chronic suppurative otitis media because the decision for or against surgical intervention may depend on the pathologic changes in the middle ear and mastoid. These include edema, submucosal fibrosis, and infiltration with chronic inflammatory cells, which together cause thickening of the mucous membrane.\textsuperscript{217} Polyps may result from excessive mucosal edema; in the more advanced stage, there may be polypoid tissue and granulation tissue and osteitis of the mastoid bone, ossicles, and labyrinth. Adhesive otitis media and sclerosis of bone may occur with healing. Typanosclerosis may also be present and is commonly associated with this disease in Alaskan (Eskimo) natives.\textsuperscript{140} If intensive medical treatment is instituted early, these pathologic changes may be reversible without surgery. When long-standing chronic disease has led to irreversible changes, however, middle-ear and mastoid surgery is usually indicated to eradicate the infection.

**Diagnosis.** A purulent, mucoid, or serous discharge through a “central” perforation of the tympanic membrane for at least 2 or 3 months is evidence of chronic suppurative otitis media. A polyp will frequently be seen emerging through the perforation or tympanostomy tube (Figure 13), which must be distinguished from a tumor in the external canal.\textsuperscript{218} The size of the perforation has no relation to the duration or severity of the disease, but the defect frequently involves most of the pars tensa. There is no otalgia, mastoid or pinna tenderness, vertigo, or fever. When any of

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**Table 6. BACTERIOLOGY OF OTORRHEA IN 51 CHILDREN (80 EARS) WITH CHRONIC SUPPURATIVE OTITIS MEDIA**

<table>
<thead>
<tr>
<th>Bacteria Isolated</th>
<th>Number of Isolates*</th>
</tr>
</thead>
<tbody>
<tr>
<td><em>Pseudomonas aeruginosa</em></td>
<td>56</td>
</tr>
<tr>
<td><em>Staphylococcus aureus</em></td>
<td>18</td>
</tr>
<tr>
<td>Diphtheroids</td>
<td>8</td>
</tr>
<tr>
<td><em>Streptococcus pneumoniae</em></td>
<td>7</td>
</tr>
<tr>
<td><em>Haemophilus influenzae</em></td>
<td>6</td>
</tr>
<tr>
<td>(nontypeable)</td>
<td></td>
</tr>
<tr>
<td><em>Bacteroides species</em></td>
<td>3</td>
</tr>
<tr>
<td><em>Candida albicans</em></td>
<td>2</td>
</tr>
<tr>
<td><em>Candida parapsilosis</em></td>
<td>2</td>
</tr>
<tr>
<td><em>Enterococcus</em></td>
<td>2</td>
</tr>
<tr>
<td><em>Acinetobacter</em></td>
<td>2</td>
</tr>
<tr>
<td><em>Staphylococcus epidermidis</em></td>
<td>1</td>
</tr>
<tr>
<td><em>Morganella morganii</em></td>
<td>1</td>
</tr>
<tr>
<td><em>Providencia stuartii</em></td>
<td>1</td>
</tr>
<tr>
<td><em>Klebsiella species</em></td>
<td>1</td>
</tr>
<tr>
<td><em>Proteus species</em></td>
<td>1</td>
</tr>
<tr>
<td><em>Serratia marcescens</em></td>
<td>1</td>
</tr>
<tr>
<td><em>Moraxella</em></td>
<td>1</td>
</tr>
<tr>
<td><em>Pseudomonas cepacia</em></td>
<td>1</td>
</tr>
<tr>
<td><em>Providencia rettgeri</em></td>
<td>1</td>
</tr>
<tr>
<td><em>Pseudomonas maltophilia</em></td>
<td>1</td>
</tr>
<tr>
<td><em>Achromobacter xylosoxidans</em></td>
<td>1</td>
</tr>
<tr>
<td><em>Eikenella</em></td>
<td>1</td>
</tr>
</tbody>
</table>

*Adapted from Kenna MA et al.\textsuperscript{207}*

*Number exceeds 80 because more than 1 organism was isolated in 38 ears.*
these signs or symptoms are present, the examiner should look for a possible suppurative intratemporal complication, such as mastoiditis or labyrinthitis, or an intracranial complication (see Chapter 10). A search for the underlying cause of the infection may reveal paranasal sinusitis, which must be actively treated because the ear infection may not respond to medical treatment until the sinusitis resolves. An upper respiratory tract allergy or a nasopharyngeal tumor may also contribute to the pathogenesis of chronic suppurative otitis media and will need to be managed appropriately.

The discharge should be appropriately examined by a Gram-stained smear and cultured as described earlier (see “Acute Perforation with Otitis Media”).

One of the most important parts of the evaluation is a complete examination of the ear canal, the tympanic membrane, and, if the perforation is large enough, the middle ear. This should be done with the aid of the otomicroscope. If a satisfactory examination cannot be performed with the child awake, an examination under general anesthesia will be necessary. At this time, the discharge can be aspirated and a specimen for culture can be obtained from the middle ear. A search for a polyp or unsuspected cholesteatoma or neoplasm should also be conducted.

A conductive hearing loss usually accompanies chronic suppurative otitis media. If a hearing loss greater than 20 to 30 dB is found, the ossicles may be involved. However, the patient may also have a sensorineural component, which is most likely due to a serous labyrinthitis. Impedance testing may be helpful if purulent material in the ear canal prevents adequate visualization of the eardrum to identify a possible perforation. If a perforation is present, the measured volume of the external canal will be larger than expected. However, the tympanometric pattern may be flat, despite the presence of a perforation, if the volume of air in the middle ear and mastoid is small. When this is suspected, the pressure on the pump-manometer of the impedance audiometer can be increased in an attempt to force open the eustachian tube. If the tube can be opened with positive air pressure from the pump-manometer, a perforation must be present.

CT scans of the middle ear and mastoid should be obtained when intensive medical treatment (including intravenous antimicrobial therapy) fails, the child has an early recurrence, or cholesteatoma is suspected initially. In the typical case, the sclerotic or undeveloped mastoid appears “cloudy.” If a defect in the bone owing to osteitis is present, however, the area will appear on the CT scans. Discontinuity of the ossicular chain, if present, may be visualized on the CT scans.

Unusual causes of a chronic draining ear, including neoplasms and eosinophilic granuloma, must be considered in the differential diagnosis of chronic suppurative otitis media, and in such cases, CT scans should be obtained initially. Erosion of portions of the temporal bone, especially the labyrinth, on the CT scan is suggestive of tumor.

Management. Treatment of chronic suppurative otitis media is initially medical and directed toward eliminating the infection from
the middle ear and mastoid. Because the bacteria most frequently cultured are gram-negative, antimicrobial agents should be selected to be effective against these organisms.\textsuperscript{122}

**Ottotopical Medications.** Ottotopical medications can be safe and effective, but not all still being prescribed are.\textsuperscript{125} A suspension containing polymyxin B, neomycin sulfates, and hydrocortisone (Pediotic) and one that has neomycin, polymyxin E, and hydrocortisone (Coly-Mycin) have been used in the past but are no longer in use in the United States. Caution is also advised owing to the concern over the potential ototoxicity of these agents.\textsuperscript{220–223} In addition, an in vitro susceptibility study showed that only 18% of middle-ear isolates were sensitive to topical neomycin.\textsuperscript{187} Some clinicians use topical tobramycin (with dexamethasone) (TobraDex)\textsuperscript{224} or gentamicin (Garamycin) ophthalmic drops instilled into the ear when *Pseudomonas* is isolated, but, again, these agents are aminoglycosides and may therefore be ototoxic.\textsuperscript{134,225–229} A recently convened consensus panel concluded that there is relatively no indication to use these potentially ototoxic agents today.\textsuperscript{230} More important, none of these popular medications are approved for use when there is a nonintact tympanic membrane.\textsuperscript{129} Nevertheless, these ottotopical agents are used widely and appear to be effective for treating chronic suppurative otitis media.\textsuperscript{231} Clinicians who have employed them with apparent success think that if the infection is not eliminated, it, too, may cause damage to the inner ear. The bacteria, or their by-products, may enter the inner ear through the round window during a middle-ear infection.\textsuperscript{22,24}

The US Food and Drug Administration (FDA) has approved ofloxacin otic solution (Floxin Otic), an ottotopical agent, for use in children when AOM with otorrhea occurs when a tympanostomy tube is in place. At present, it is the only topical antimicrobial agent that has been demonstrated to be safe and effective\textsuperscript{126,128,232,233} and approved for this indication in children. It is also approved for adults who have chronic suppurative otitis media, but it is currently not approved for this indication in children,\textsuperscript{234} even though it has been reported to be effective in this age group.\textsuperscript{235} Topical ofloxacin has been shown to be more effective than the combination of neomycin–polymyxin B–hydrocortisone otic drops in adults with chronic supplicative otitis media.\textsuperscript{236}

Even though ciprofloxacin is not approved, it appears to be effective for chronic suppurative otitis media.\textsuperscript{237–241} One study showed that topical ciprofloxacin was more effective than topical gentamicin for chronic supplicative otitis media in adults,\textsuperscript{242} and another showed that this antibiotic and tobramycin are equally effective in adults with this infection.\textsuperscript{243} No apparent ototoxicity has occurred after use of this ottotopical agent in patients with chronic suppurative otitis media.\textsuperscript{244} In addition, topical ciprofloxacin did not cause ototoxicity in the monkey model of chronic suppurative otitis media.\textsuperscript{245} There is still no consensus about the potential efficacy of adding a corticosteroid component to the antimicrobial agent, but the addition of a steroid may hasten resolution of the inflammation.\textsuperscript{246}

Recently, the combination of ciprofloxacin–dexamethasone otic suspension (CiproDex) has become available for treatment of otorrhea. Roland and colleagues reported success in treating children who had AOM with otorrhea through a tympanostomy tube,\textsuperscript{130} and it is effective when granulation tissue is present on the eardrum and around the tube.\textsuperscript{131} The consensus panel concluded that these two new fluoroquinolone agents that are now available provide a safe and effective alternative to the older, potentially ototoxic agents.\textsuperscript{133} Thus, the lack of reported clinical trials in children with chronic suppurative otitis media notwithstanding, it seems reasonable today to use ofloxacin or ciprofloxacin–dexamethasone initially in children who have uncomplicated chronic suppurative otitis media, especially since they are not ototoxic and are effective when the tympanic membrane is not intact (ie, following insertion of a tympanostomy tube).\textsuperscript{125}

Other ottotopical agents have been used for treatment of chronic suppurative otitis media...
although not approved for this use by the Food and Drug Administration. Ciprofloxacin with hydrocortisone (Cipro HC) has been used for treatment of external otitis in both children and adults but is not indicated when the tympanic membrane is not intact. As an alternative to an antibiotic topical agent, some clinicians recommend antiseptic drops. An antiseptic ototopical agent (aluminum acetate) was found to be as effective as topical gentamicin sulfate for otitis media in a randomized clinical trial reported from the United Kingdom. Thorp and colleagues evaluated the in vitro activity of acetic acid and aluminum subacetate (Burow’s solution) and found both to be effective against the major pathogens causing chronic suppurative otitis media. More recently, Kashiwamura and colleagues also found Burow’s solution to be effective. Burow’s solution was somewhat more effective than acetic acid. Antiseptic drops (eg, acetic acid) are commonly used in underdeveloped countries and are reputed to be effective. Because of cost and availability, antibiotic ototopical agents are used when antiseptic drops are ineffective. But, again, these agents are currently not recommended in the face of an open eardrum.

Table 3 provides a list of ototopical agents currently (2006) approved by the FDA for ear infections. As stated before, only ofloxacin otic solution and ciprofloxacin-dexamethasone otic suspension solution are approved for children when the tympanic membrane is nonintact and then only when a tympanostomy tube is present. Nevertheless, other agents, such as ciprofloxacin with hydrocortisone, may be beneficial for use in children with this middle-ear and mastoid infection. The advantage of quinolone topical agents is that there is no evidence of ototoxicity in animal models, which had been reported with use of the aminoglycosides. With the growing concern regarding the emergence of multidrug-resistant bacterial otic pathogens, an ototopical agent is desirable because it is hoped that the use of a high concentration of the drug directed at the site of infection will prevent emergence of resistant organisms; to date, we have not seen resistance developing following use of these new quinolone ototopical agents.

If topical antibiotic medication is elected, the child should ideally return to the outpatient facility daily to have the discharge thoroughly aspirated or swabbed (ie, *aural toilet, ear mopping*) and to have the ototopical medication directly instilled into the middle ear through the perforation or tympanostomy tube by use of an otoscope or otomicroscope. Frequently, the discharge rapidly improves within a week with this type of treatment, after which the eardrops may be administered at home until there is complete resolution of the middle-ear–mastoid inflammation. When daily administration by the physician is not feasible, the parent or caregiver can administer the drops.

**Oral Antimicrobial Agents.** Oral antibiotics that are approved for treatment of AOM may be effective if the bacterium is susceptible, but because the organism is usually *P. aeruginosa*, agents that are currently approved for children will usually not be effective; approximately 40% of *Pseudomonas* is susceptible in vitro to trimethoprim-sulfamethoxazole. Despite these potential drawbacks, many clinicians administer a broad-spectrum oral antibiotic in the hope that the underlying infection is caused by the usual bacteria that are isolated from ears with AOM. In a report of a randomized clinical trial conducted in Kenya, Smith and colleagues compared (1) oral amoxicillin-clavulanate, dry mopping of the ear, and ototopical antibiotic-cortisone drops; (2) dry mopping alone; and (3) no treatment. They found the combination of oral antibiotic and topical agents to be statistically more effective than dry mopping alone or no treatment. They found the combination of oral antibiotic and topical agents to be statistically more effective than dry mopping alone or no treatment. However, a randomized clinical trial found that topical ofloxacin was more effective than systemic amoxicillin-clavulanate (without eardrops) in adults with chronic suppurative otitis media. At present, we recommend for initial treatment an approved ototopical quinolone and an oral agent if the bacteria are susceptible.
Orally administered ciprofloxacin has been shown to be effective in adults and Israeli children who had chronic suppurative otitis media, but this agent is currently not approved for patients younger than 17 years in the United States. In addition to aural toilet, topical ciprofloxacin was effective management in Australian Aboriginal children. Some clinicians advocate a course of an oral quinolone for chronic suppurative otitis media when the causative organism is \textit{Pseudomonas} and obtain oral consent from the parent or guardian. Safety concerns are of less concern today given the lack of serious side effects in children who have cystic fibrosis and who have taken these drugs for many years.

When ototopical agents, oral antimicrobial agents, or both are used, the child should be reexamined in about 1 week, when any adjustments can be made in the medications after the results of the microbiologic studies are available. After approximately 1 week, there should be cessation of the discharge or marked improvement. If, indeed, the otorrhea is improving, the child is reexamined periodically thereafter until resolution occurs. If there is no improvement after 1 to 2 weeks, other treatment options should be considered, such as parenteral antimicrobial therapy. As noted above, a systemic course of a quinolone (with appropriate parental consent) may be preferable to initiating parenteral treatment when \textit{Pseudomonas} is the offending bacterium.

\textbf{Parenteral Antimicrobial Agents.} If treatment of the child with the administration of ototopical agents, with or without an oral antimicrobial agent, has failed, the patient should receive a parenteral \(\beta\)-lactam antipseudomonal drug, such as ticarcillin, piperacillin, or ceftazidime; empirically, ticarcillin-clavulanate is usually selected because \textit{Pseudomonas}, with and without \textit{S. aureus}, is frequently isolated; the results of the culture and susceptibility studies dictate the antimicrobial agent ultimately chosen (see Chapter 8). Dagan and colleagues in Israel and Arguedas and colleagues in Costa Rica reported excellent results with the use of ceftazidime. In Finland, Vartiainen and Kansanen also recommended a trial of intravenous antimicrobial therapy before considering mastoid surgery. The regimen can be altered when the results of culture and susceptibility tests are available. Also, the external canal purulent material and debris (and middle ear, if possible) are aspirated and the ototopical medication is instilled daily. This method of treatment is usually performed with the child hospitalized, but it can be carried out on an ambulatory basis. The intravenous line can be inserted as an operative procedure or in the radiology suite.

In about 90\% of children, the middle ear will be free of discharge and the signs of chronic suppurative otitis media will be greatly improved or absent within 5 to 7 days. Kenna and colleagues conducted a study of 36 pediatric patients with chronic suppurative otitis media in which all received parenteral antimicrobial therapy and daily aural toilet. Medical therapy alone resolved the infection in 32 patients (89\%); 4 children required tympanomastoidectomy. The investigators later increased the study group to 66 children and reported similar short-term results; 89\% had dry ears after intravenous antibiotic therapy. In a follow-up of that study, 51 of the original 66 were evaluated for their long-term outcomes. Of these 51 children, 40 (78\%) had resolution of their initial or recurrent infection after medical treatment and 11 (22\%) had to eventually have mastoid surgery. Failure was associated with older children and an early recurrence. Englender and colleagues did serotyping and pyocin typing of \textit{P. aeruginosa} of 142 patients, including children, and found that if the patient had a recurrence with a different type, medical treatment was frequently successful. If the patient had recurrence of the otorrhea with the same type, medical therapy usually failed and the patient required mastoid surgery. Leiberman and colleagues found that when children had an early recurrence, they were less likely to benefit from either medical treatment, including intravenous antibiotics, or surgical management. (For details of the surgical technique of mastoi-
dectomy for chronic suppurative otitis media, see Bluestone.152)

If resolution occurs during hospitalization, the child can be discharged and receive the parenteral antibiotic and eardrops (by the parent or caregiver) for a period of 10 to 14 days at home. The patient should be observed at periodic intervals to watch for signs of spontaneous closure of the perforation, which frequently happens after the middle ear and mastoid are no longer infected. Appropriate intensive medical treatment should be attempted before major ear surgery is recommended because the outcome of surgery is not as favorable when medical treatment is withheld.205

Surgery. When chronic suppurative otitis media fails to respond to intensive medical therapy (ie, intravenous antibiotics, aural toilet, and ototopical medications) within several days, surgery on the middle ear and mastoid (ie, tympanomastoidectomy) may be required to eradicate the infection. A CT scan should be obtained (see earlier). Failures usually occur when there is an underlying blockage of the communication between the middle ear and mastoid (ie, aditus ad antrum), irreversible chronic osteitis, cholesteatoma (or tumor), or early recurrence with the same causative organism.207

Insertion of a tympanostomy tube can be helpful if the chronic suppurative otitis media is associated with a perforation that is too small to permit adequate drainage or if the perforation frequently closes, only to reopen with episodic drainage. On the other hand, if the chronic infection is related to a tympanostomy tube (ie, the middle-ear air cushion is absent), some clinicians advocate removal of the tube, hoping that the infection will subsequently subside. However, the recurrent and chronic ear infections for which the tube was originally inserted frequently recur. There may be some merit in attempting this approach in a child who has had a long-standing retained tube.

Prevention of Recurrence. With an understanding of the pathogenesis of chronic suppura-
Complications and Sequelae. The most common sequela of chronic suppurative otitis media is chronic hearing loss, with the potential deficits related to this disability. The chronic infection may result in permanent conductive hearing loss because of damage to the ossicles, and sensorineural loss may also occur. Chronic suppurative otitis media can also progress into one or more of the intratemporal (extracranial) suppurative complications (acute mastoiditis, acute labyrinthitis, facial paralysis) or an intracranial suppurative complication, which will require immediate surgical intervention. Intracranial complications caused by chronic suppurative otitis media occur with a frequency equal to or greater than that of those due to cholesteatoma. Not only is this finding important in underdeveloped countries where chronic suppurative otitis media is common, it has also been reported from industrialized nations (see Chapter 10).

A child who has a chronic suppurative aural discharge and who develops one or more of the intratemporal suppurative complications (such as acute mastoid osteitis, labyrinthitis, and facial paralysis) or an intracranial suppurative complication requires immediate surgical intervention. It is not uncommon for a cholesteatoma to be present when an ear with chronic suppurative otitis media fails to respond to intensive medical treatment, even though no preoperative evidence for the presence of cholesteatoma was identified by otomicroscopy or imaging. The cholesteatoma is usually found in the middle ear (and mastoid) following migration of the squamous epithelium through the perforation in the tympanic membrane. When a cholesteatoma is found, surgical removal is indicated as outlined later in this chapter.

Role of the Primary Care Physician. Many children who have chronic suppurative otitis media can be effectively managed by the primary health care provider with ototopical antimicrobial medications, oral antibiotic agents, or both. When these measures fail, the child requires parenteral administration of an antimicrobial agent. Referral to an otolaryngologist is advisable in the following situations:

- A suppurative complication, cholesteatoma, or tumor is present or suspected.
- An otomicroscope is necessary to completely examine the ear.
- An examination under general anesthesia is required to completely evaluate the ear (and child, eg, the nose and pharynx).
- A child does not respond to intensive medical therapy because tympanomastoidectomy is usually required.
- Prevention of recurrence involves possible adenoidectomy, surgery for chronic or recurrent paranasal sinusitis, surgical repair of a tympanic membrane perforation (ie, tympanoplasty), or removal of a tympanostomy tube.

MASTOIDITIS

Mastoiditis is a potential complication of acute and chronic middle-ear infections. In the preantibiotic era, acute mastoiditis was the most common infection for which infants and children were hospitalized. Since the widespread use of antimicrobial agents, the incidence has fallen dramatically, but the clinician should always be aware that acute mastoiditis remains the most common suppurative complication of acute otitis media. In addition, there is some evidence that the incidence of acute mastoiditis has increased in certain geographic areas and may be related to withholding antimicrobial agents in children who have AOM. But some studies report that acute mastoiditis may be the first evidence of ear infection and that initial treatment with an antimicrobial agent would not have prevented the mastoiditis. Also, some reports concluded that there has been no increase in the incidence of acute mastoiditis during the recent past. The incidence of acute mastoiditis appears to be greater in southern Israel than the rates reported elsewhere and was related to S. pyogenes as a common causative bacterium but not penicillin-nonsusceptible S. pneumoniae. But in Dallas, a recent report implicated
resistant S. pneumoniae as the cause of an increase in suppurative complications of AOM.273

Chronic mastoiditis that develops after an episode of acute mastoiditis has also decreased during the past 50 years for similar reasons. However, chronic mastoiditis is still a major problem when chronic suppurative otitis media is present, especially in racial groups and geographic areas in which this disease is common.115 Both acute and chronic mastoiditis can occur in the presence of cholesteatoma.274 Thus, both acute and chronic mastoiditis still occur and may be responsible for significant morbidity and life-threatening infection, especially from intracranial extension of the disease.275–279

**Anatomy, Pathogenesis, and Pathology**

At birth, the mastoid consists of a single cell, the antrum, connected to the middle ear by a small channel, the aditus ad antrum. Pneumatization of the mastoid bone takes place soon after birth and is usually extensive by the age of 2 years. The process may continue throughout life. The clinical importance of the mastoid is related to contiguous structures, including the posterior cranial fossa, the middle cranial fossa, the sigmoid and lateral sinuses, the canal of the facial nerve, the semicircular canals, and the petrous tip of the temporal bone. The mastoid air cells are lined with modified respiratory mucosa, and all are interconnected with the antrum (Figure 14A).

Infection in the mastoid proceeds after middle-ear infection through the following stages:

- Hyperemia and edema of the mucosal lining of the pneumatized cells
- Accumulation of serous and then purulent exudates in the cells
- Demineralization of the cellular walls and necrosis of bone owing to pressure of the purulent exudate on the thin bony septa and ischemia of the septa, caused by a decrease in blood flow
- Formation of abscess cavities owing to coalescence of adjacent cells after destruction of cell walls
- Escape of pus into contiguous areas

This process may halt at any stage with subsequent resolution. However, when infection persists for more than a week or 10 days, inflammatory granulation tissue forms in the pneumatic cavity. A hypertrophic osteitis develops that results in thickening and sclerosis of cellular walls and a reduction in the size of the cellular space. There may be repeated cycles of absorption and deposition of bone. If the infection remains chronic but low grade, there is thickening of the mucosa caused by a fibrinous exudate, which may become organized and lead to permanent adhesions. Columnar metaplasia
with new gland formation may result in extensive production of mucus in the former cells.

Mastoiditis can be classified into acute and chronic types. Acute mastoiditis is further subdivided according to the pathologic stage present, which has clinical significance because management depends on the stage of the disease. Unfortunately, because of failure to appreciate the natural history and pathologic features of acute mastoiditis, there is confusion in the minds of clinicians, as well as in the current literature, regarding the most appropriate management of each stage.

**Mastoiditis without Periosteitis or Osteitis**

Because the mastoid gas cell system is connected to the distal end of the middle ear, most likely all episodes of otitis media are associated with some inflammation of the mastoid. Thus, mastoiditis can be a natural extension and part of the pathologic process of middle-ear inflammation; the mastoid may be involved in AOM or otitis media with effusion (of any duration).

**Diagnosis**

The diagnosis is commonly made after CT scans or magnetic resonance imaging (MRI) studies are obtained for another problem (eg, sinusitis, head trauma) in a child who has no signs or symptoms referable to the ears, in which case, it is an incidental finding. No specific signs or symptoms of mastoid infection (eg, protrusion of the pinna, postauricular swelling, tenderness, pain, and erythema) are present in this most common stage of mastoiditis. Imaging of the mastoid area frequently reveals cloudy mastoids, which indicates inflammation, but no mastoid osteitis (ie, bony erosion of the mastoid air cells) is evident (Figure 15).

**Management and Outcome**

The process is usually reversible as the middle-ear–mastoid infection resolves, either as a natural process or as a result of medical management (eg, antimicrobial therapy). Thus, with no periosteal involvement of the postauricular region, osteitis of the mastoid, or subperiosteal abscess, this stage of mastoiditis is not a suppurative complication of otitis media.

When this stage of mastoiditis is diagnosed, such as by CT scan, in a child who has an episode of AOM, the management is the same as recommended for AOM because the involvement of the mastoid is a natural extension of the middle-ear infection (see Chapter 8). Indications for tympanocentesis (diagnostic aspiration of the middle ear) are the same as when AOM is diagnosed and the status of the mastoid is undetermined (by CT or MRI), such as when the patient is severely ill or toxic, fails to improve rapidly while receiving appropriate and adequate antibiotic treatment, develops otitis media while receiving antimicrobial agents, develops otitis media in the newborn period, is immunologically deficient, or has a suppurative complication. Myringotomy is indicated when drainage of the middle ear is desirable, such as when the child...
has severe otalgia or when a supplicative complication is suspected or is present (see Chapter 7).

When acute infection in the mastoid (and usually middle ear) does not resolve at this stage, the disease can rapidly progress to acute mastoiditis with periostitis. The next stage is acute mastoid osteitis, which can occur with or without the presence of a subperiosteal abscess.

When imaging of the head reveals the incidental finding of inflammation of the mastoid gas cells in a child who has an episode of otitis media with effusion (regardless of the duration of the effusion), management is the same as recommended for this middle-ear inflammation when the status of the mastoid is unknown, that is, CT or MRI is not obtained (see Chapter 8).

### Acute Mastoiditis with Periostitis

When infection within the mastoid spreads to the periosteum covering the mastoid process, periostitis can develop. The route of infection from the mastoid cells to the periosteum is by venous channels, usually the mastoid emissary vein. The condition should not be confused with a subperiosteal abscess, the management of which usually requires a mastoidectomy. Periostitis frequently responds to medical treatment and tympanocentesis or myringotomy.

### Epidemiology

Before the antibiotic era, the incidence of acute mastoiditis as a complication of AOM was high. In 1938, the rate was 20%, whereas it was 2.8% 10 years later (with an almost 90% reduction in the mortality rate during that period), and in some studies, it dropped almost to zero. At the Children’s Hospital of Pittsburgh between 1980 and 1995, there were 72 infants and children with acute mastoiditis, but only 18 (25%) required a mastoidectomy; the remaining 54 (75%) did not have a mastoidectomy. The majority of children who develop acute mastoiditis today most likely have acute mastoiditis with periostitis. The growing enthusiasm for withholding antimicrobial therapy today in some countries will most likely result in an increase in this supplicative complication.

### Pathogenesis

Infection in the mastoid air cells, which is frequently part of an episode of AOM, usually resolves spontaneously or after effective antimicrobial treatment. The effusion within the mastoid air cells drains into the middle ear, which, in turn, drains down the eustachian tube (i.e., the clearance function of the tube). The narrow communication between the middle ear and the mastoid air cells is the aditus ad antrum (see Figure 9–14A). Physiologically, the mastoid air cell system is most likely a reservoir of gas for the middle ear, and this narrow passage makes it more difficult for middle-ear infection to enter the mastoid. When infection is in the mastoid, however, the aditus ad antrum can become obstructed by edema and granulation tissue (Figure 9–14B). This obstruction has been called the bottleneck and has been implicated in the pathogenesis of acute mastoiditis. If the obstruction persists, acute mastoiditis with periostitis can develop, which can then progress into acute mastoid osteitis (with and without subperiosteal abscess) and further progress into another supplicative complication in the temporal bone or the intracranial cavity.

### Clinical Presentation and Diagnosis

The child has the classic signs and symptoms of AOM, such as fever and otalgia, but also postauricular erythema; mild tenderness and some edema may also be present in the postauricular area. The pinna may or may not be displaced inferiorly and anteriorly, with loss of the postauricular crease. Subperiosteal abscess is absent. Examination of the eardrum typically reveals evidence of AOM. However, the middle ear may be effusion free in the presence of acute mastoiditis if there is an obstruction of the aditus ad antrum. The middle-ear effusion drains down the eustachian tube, but the infection in the mastoid cannot drain into the middle ear. Table 7 shows the age and frequency of the presenting signs and symptoms of 72 infants and children.
who had acute mastoiditis at the Children’s Hospital of Pittsburgh from 1980 to 1995 (some of these patients had acute mastoid osteitis with and without subperiosteal abscess). In this series, half were 3 years of age or younger; two-thirds were younger than 7 years. Also, only 28% of these children were diagnosed as having a middle-ear effusion, which is clinically important because this finding emphasizes the point made earlier that mastoiditis can be present despite the lack of otitis media; the middle-ear effusion resolves, but the infection in the mastoid persists.

Table 7. AGE, PRESENTING SYMPTOMS, AND SIGNS OF 72 INFANTS AND CHILDREN WITH ACUTE MASTOIDITIS

<table>
<thead>
<tr>
<th>Age (yr)</th>
<th>Patients (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>0–3</td>
<td>35 (50.0)</td>
</tr>
<tr>
<td>4–6</td>
<td>12 (16.7)</td>
</tr>
<tr>
<td>7–12</td>
<td>17 (23.6)</td>
</tr>
<tr>
<td>13–18</td>
<td>7 (9.7)</td>
</tr>
<tr>
<td>Symptoms</td>
<td></td>
</tr>
<tr>
<td>Otalgia</td>
<td>62 (86.1)</td>
</tr>
<tr>
<td>Postauricular pain</td>
<td>58 (80.6)</td>
</tr>
<tr>
<td>Fever</td>
<td>51 (70.8)</td>
</tr>
<tr>
<td>Otorrhea</td>
<td>29 (40.3)</td>
</tr>
<tr>
<td>Upper respiratory infection</td>
<td>17 (23.6)</td>
</tr>
<tr>
<td>Irritability</td>
<td>16 (22.2)</td>
</tr>
<tr>
<td>Hearing loss</td>
<td>6 (8.3)</td>
</tr>
<tr>
<td>Signs</td>
<td></td>
</tr>
<tr>
<td>Postauricular tenderness</td>
<td>58 (80.6)</td>
</tr>
<tr>
<td>Pinna protrusion</td>
<td>51 (70.8)</td>
</tr>
<tr>
<td>Postauricular erythema</td>
<td>51 (70.8)</td>
</tr>
<tr>
<td>Middle-ear effusion</td>
<td>48 (66.7)</td>
</tr>
<tr>
<td>Tympanic membrane erythema</td>
<td>42 (58.3)</td>
</tr>
<tr>
<td>Contralateral middle-ear abnormalities</td>
<td>35 (48.6)</td>
</tr>
<tr>
<td>Middle-ear effusion</td>
<td>20 (27.8)</td>
</tr>
<tr>
<td>Tympanic membrane bulging</td>
<td>8 (11.1)</td>
</tr>
<tr>
<td>Tympanic membrane erythema</td>
<td>6 (8.3)</td>
</tr>
<tr>
<td>Tympanic membrane perforation</td>
<td>1 (1.4)</td>
</tr>
<tr>
<td>Tympanic membrane bulging</td>
<td>34 (47.2)</td>
</tr>
<tr>
<td>Otorrhea</td>
<td>24 (33.3)</td>
</tr>
<tr>
<td>Fever ≥ 38.3°C</td>
<td>24 (33.3)</td>
</tr>
<tr>
<td>Tympanic membrane perforation</td>
<td>13 (18.1)</td>
</tr>
<tr>
<td>Postauricular mass</td>
<td>13 (18.1)</td>
</tr>
<tr>
<td>Cervical adenopathy</td>
<td>12 (16.7)</td>
</tr>
<tr>
<td>Postauricular abscess</td>
<td>9 (12.5)</td>
</tr>
<tr>
<td>Sagging of posterior auditory canal wall</td>
<td>2 (2.8)</td>
</tr>
</tbody>
</table>

Table 8. HEARING STATUS IN 41 INFANTS AND CHILDREN WITH ACUTE MASTOIDITIS

<table>
<thead>
<tr>
<th>Audiologic Findings</th>
<th>Patients (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Normal range</td>
<td>21 (51)</td>
</tr>
<tr>
<td>Abnormal</td>
<td>20 (49)</td>
</tr>
<tr>
<td>Conductive loss (dB)</td>
<td></td>
</tr>
<tr>
<td>30–39</td>
<td>4 (10)</td>
</tr>
<tr>
<td>40–49</td>
<td>8 (20)</td>
</tr>
<tr>
<td>50–59</td>
<td>2 (5)</td>
</tr>
<tr>
<td>Mixed loss (dB)</td>
<td></td>
</tr>
<tr>
<td>60–69</td>
<td>2 (5)</td>
</tr>
<tr>
<td>80–99</td>
<td>2 (5)</td>
</tr>
<tr>
<td>Sensorineural (dB)</td>
<td></td>
</tr>
<tr>
<td>40–49</td>
<td>1 (2)</td>
</tr>
<tr>
<td>Profound</td>
<td>1 (2)</td>
</tr>
</tbody>
</table>

Adapted from Goldstein NA et al.262

For this stage of acute mastoiditis, a CT scan of the temporal bones (and intracranial cavity) should be obtained to determine whether osteitis of the mastoid is present. A CT scan is not mandatory if the infection is limited to the middle ear and mastoid, the child is not severely ill or toxic, and improvement is rapid after tympanocentesis-myringotomy and parenterally administered antimicrobial therapy.

Differential Diagnosis

The differential diagnosis of a child who has a suspect acute mastoiditis with periosteitis includes perichondritis of the pinna and external otitis. The typical presentation of a child who has a perichondritis of the pinna is swelling and erythema of the outer ear, which may spread to the periosteum of the postauricular area, but the postauricular crease is usually obliterated and external otitis and middle-ear effusion are absent unless a concurrent, unrelated otitis media is present. External otitis can also spread to the postauricular area; like perichondritis, it will obliterate the postauricular crease. In contrast to these two infections, the postauricular crease is usually—but not always—present.

When the acute mastoiditis has progressed to a subperiosteal abscess, the postauricular swel-
ling may be misdiagnosed as an enlarged lymph node. Lymph node enlargement in the postauricular area is typically due to a scalp infection, dermatitis, or an insect bite above the affected ear, and middle-ear effusion is absent. The lymph node is well circumscribed, usually freely movable in all four directions, and most often not tender to touch. On the other hand, a subperiosteal abscess, secondary to acute mastoiditis, does not move readily, is not well circumscribed, and is tender to touch (Table 9).

**Microbiology**

Middle-ear aspirates reveal bacterial pathogens similar to those isolated from children’s ears that have uncomplicated acute middle-ear infection, such as *S. pneumoniae*, *S. pyogenes*, or *H. influenzae*, but there may be an unusual organism, such as *P. aeruginosa*, if there has been otorrhea.286,287 Indeed, the leading pathogen in Israel today is *Pseudomonas* when the child has had recurrent episodes of AOM.288 Also, tuberculosis has been reported to be the causative bacterium in mastoiditis, not only in Korea,289 but also in Chicago.290 Table 10 shows the type and frequency of bacteria isolated from 65 infants and children who had acute mastoiditis (acute mastoiditis with periostitis and acute mastoid osteitis) diagnosed at the Children’s Hospital of Pittsburgh between 1980 and 1995.262 Cultures were obtained from one or more of the following sites: the external auditory canal when otorrhea was present, the middle ear by tympanocentesis, or the middle ear, mastoid, or both at the time of mastoidectomy. The most common pathogenic bacteria were *S. pneumoniae*, *P. aeruginosa*, and *S. pyogenes*.

**Management**

The patient may be managed on an ambulatory basis if the infection is not severe. However, hospitalization is usually necessary because parenteral antimicrobial therapy is frequently needed, and most patients require an immediate tympanocentesis (for aspiration and microbiologic assessment of the middle-ear–mastoid effusion) and myringotomy to drain the middle ear. In the absence of an aditus ad antrum “block,” this should also drain the mastoid. If the child

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**Table 9. DIFFERENTIAL DIAGNOSIS OF POSTAURICULAR INVOLVEMENT OF ACUTE MASTOIDITIS WITH PERIOSTEITIS OR ABSCESS**

<table>
<thead>
<tr>
<th>Disease</th>
<th>Postauricular Signs and Symptoms</th>
<th>Tenderness</th>
<th>External Canal Infection</th>
<th>Middle-Ear Effusion</th>
</tr>
</thead>
<tbody>
<tr>
<td>Acute mastoiditis with periostitis</td>
<td>May be absent</td>
<td>Absent</td>
<td>Usually</td>
<td>No</td>
</tr>
<tr>
<td>Acute mastoiditis with subperiosteal abscess</td>
<td>Absent</td>
<td>Maybe</td>
<td>Yes</td>
<td>No</td>
</tr>
<tr>
<td>Periosteitis of pinna with postauricular extension</td>
<td>Intact</td>
<td>Yes</td>
<td>Usually</td>
<td>No</td>
</tr>
<tr>
<td>External otitis with postauricular extension</td>
<td>Intact</td>
<td>Yes</td>
<td>Usually</td>
<td>Yes</td>
</tr>
<tr>
<td>Postauricular lymphadenitis</td>
<td>Intact</td>
<td>No</td>
<td>Yes (circumscribed)</td>
<td>No</td>
</tr>
</tbody>
</table>

*Postauricular crease (fold) between pinna and postauricular area.

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**Table 10. BACTERIOLOGY OF EFFUSIONS, OTORRHEA, AND MASTOIDS IN 65 INFANTS AND CHILDREN WITH ACUTE MASTOIDITIS**

<table>
<thead>
<tr>
<th>Organism</th>
<th>Number of isolates (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td><em>Streptococcus pneumoniae</em></td>
<td>21 (32.3)</td>
</tr>
<tr>
<td><em>Pseudomonas aeruginosa</em></td>
<td>19 (29.2)</td>
</tr>
<tr>
<td><em>Streptococcus pyogenes</em></td>
<td>12 (18.5)</td>
</tr>
<tr>
<td>Diphtheroids</td>
<td>9 (13.9)</td>
</tr>
<tr>
<td>Anaerobes</td>
<td>7 (10.8)</td>
</tr>
<tr>
<td>Others*</td>
<td>39 (60.0)</td>
</tr>
<tr>
<td>No growth</td>
<td>6 (9.2)</td>
</tr>
<tr>
<td><strong>Total</strong></td>
<td><strong>113</strong></td>
</tr>
</tbody>
</table>

*Coagulase-negative staphylococcus (23), β-hemolytic streptococcus (4), Haemophilus influenzae (3), Micrococcus species (2), Moraxella catarrhalis (1), Staphylococcus aureus (1), Staphylococcus species (1), Enterobacter cloacae (1), Neisseria species (1), Enterococcus species (1), Citrobacter diversus (1).

Total of isolates is greater than 100% because some cultures were polymicrobial.
has had recurrent attacks of AOM or has the current episode of AOM superimposed on chronic otitis media with effusion, insertion of a tympanostomy tube is indicated. Insertion of a tympanostomy tube is desirable and will enhance drainage for a longer time than myringotomy alone.\textsuperscript{193} Even though there have been reports revealing that antibiotic treatment was successful in curing some children, without the benefit of tympanocentesis or myringotomy, aspiration of the middle ear is an important diagnostic (and therapeutic) procedure today. An antibiotic-resistant bacterial pathogen, such as multidrug-resistant \textit{S. pneumoniae}, may be the causative organism, which may require an antimicrobial agent not frequently used for AOM–mastoiditis, such as vancomycin.\textsuperscript{262,286,287,291,292}

Cultures for bacteria from the middle ear are required to identify the causative organism. Antimicrobial susceptibility studies are important for selecting the most effective antibiotic agent. For empiric parenteral antimicrobial therapy, cefuroxime sodium, ticarcillin disodium with clavulanate potassium, or ampicillin-sulbactam can be initiated until the results of Gram stain, culture, and susceptibility studies of the middle-ear aspirates are available. If penicillin-resistant \textit{S. pneumoniae} is the possible pathogen, some clinicians today also add vancomycin while awaiting the culture and susceptibility report.

The periosteal involvement should resolve within 24 to 48 hours after the tympanic membrane has been opened for drainage and adequate and appropriate antimicrobial therapy has begun. Some recent reports indicate that needle aspiration of the subperiosteal abscess, in conjunction with intravenous antibiotic therapy and myringotomy, is effective and thus avoids mastoidectomy.\textsuperscript{293–295} A mastoidectomy should be performed if the symptoms of the acute infection (such as fever and otalgia) persist, the postauricular involvement does not progressively improve, or a subperiosteal abscess develops.

A CT scan can be helpful in the decision to surgically intervene.\textsuperscript{296} A mastoidectomy is also indicated if another intratemporal (extracranial) supplicative complication (facial paralysis, labyrinthitis, petrous apicitis) or intracranial complication (meningitis, lateral sinus thrombosis, or abscess of the epidural or subdural space or brain) of otitis media is present.

**Outcome**

In the review by Goldstein and colleagues of the 72 infants and children with acute mastoiditis at the Children’s Hospital of Pittsburgh, 54 (75%) were managed conservatively with broad-spectrum intravenous antibiotics and myringotomy, with and without tympanostomy tube insertion.\textsuperscript{262} The other 18 (25%) required mastoidectomy. Of these 18 children, 14 (78%) had one or more of the following: mastoid osteitis, subperiosteal abscess, cholesteatoma, or another supplicative complication (eg, facial paralysis). A review from Australia by Harley and colleagues related approximately the same experience as in Pittsburgh.\textsuperscript{286} Between 1982 and 1993, 58 infants and children were admitted to the Royal Children’s Hospital of Melbourne, and of these, 45 (78%) were treated conservatively with intravenous antimicrobial therapy, with and without tympanostomy tube insertion. The remaining 13 patients required mastoidectomy. Others have also reported that the majority of children with acute mastoiditis can be managed conservatively, but other centers have reported that most of their patients required a mastoidectomy.\textsuperscript{287,297–302} The reasons for these conflicting reports are most likely the lack of a uniform definition of the disease, dissimilarity in the presentation of cases, and variation in management. Our opinion is that most patients with acute mastoiditis with only periostitis recover without the need for mastoidectomy.

Immediate treatment at this stage of acute mastoiditis is mandatory. Failure to provide immediate treatment may result in development of acute mastoid osteitis (with or without a subperiosteal abscess) or, potentially more life-threatening to the child, a supplicative complication such as meningitis or brain abscess.

In the absence of mastoid osteitis (with or without subperiosteal abscess), the primary care physician or pediatric infectious disease specialist
can provide the initial medical care for patients with acute mastoiditis with periostitis. However, tympanocentesis-myringotomy is required, and an otolaryngologist will be needed if the medical specialists are untrained in this procedure. Referral to an otolaryngologist is appropriate if a mastoidectomy is indicated. Immediate referral for surgical evaluation and management is indicated when acute mastoid infection develops in a child with chronic suppurative otitis media, cholesteatoma, or both.

Acute Mastoiditis with Osteitis (with and without Subperiosteal Abscess)

Acute mastoiditis with osteitis has also been called *acute coalescent mastoiditis* or *acute surgical mastoiditis*, but the pathologic process is really *acute mastoid osteitis*. A *subperiosteal abscess* may or may not be present.

Epidemiology

A review during a 20-year period (1977–1996) in one Danish county revealed that 79 children had a mastoidectomy performed for acute mastoiditis; a subperiosteal abscess was present in 66%. In the review during a 15-year period (1980–1995) of 72 children who had acute mastoiditis by Goldstein and colleagues, 31% had either a postauricular mass or subperiosteal abscess. In a report from Rome, Italy, in which 39 children had mastoidectomy for acute mastoiditis, all had a subperiosteal abscess.

Pathogenesis and Pathology

When infection within the mastoid progresses, rarefying osteitis can cause destruction of the bony trabeculae that separate the mastoid cells so that there is a “coalescence” of the cells. At this stage, a mastoid empyema is present. The pus may spread in one or more of the following directions:

- Anterior to the middle ear through the aditus ad antrum, in which case spontaneous resolution usually occurs
- Lateral to the surface of the mastoid process, resulting in a postauricular subperiosteal abscess (Figure 16)
- Anterior into the zygomatic cells, with development into an abscess in the anterior and superior portion of the pinna and preauricular area
- Inferior through the tip of the mastoid, burrowing beneath the skin to form a soft tissue abscess below the pinna or behind the attachment of the sternocleidomastoid muscle in the neck, which is known as *Bezold’s abscess* (Figures 17 and 18)
Medial to the petrous air cells, resulting in petrositis

- Posterior to the occipital bone, which can result in osteomyelitis of the calvarium, or Citelli’s abscess

The acute mastoid infection may be associated with an episode of AOM, cholesteatoma, or chronic suppurative otitis media. In the review of 39 Italian children who had this complication, 23 (59%) had AOM, and most of the remaining 16 had an associated cholesteatoma.

Infection may also spread medial to the labyrinth, involve the facial nerve, or extend into the intracranial cavity, causing one or more suppurative complications, such as dural sinus thrombosis.

**Clinical Presentation**

The child usually has the same signs and symptoms as those associated with AOM, such as fever and otalgia, although the fever may be low grade, with occasional temperature spikes. Some patients may have toxic symptoms. The signs and symptoms referable to the mastoid infection are swelling, erythema, and tenderness to touch over the mastoid bone; displacement of the pinna outward and downward (Figure 19); and swelling or sagging of the posterosuperior external auditory canal wall (see Table 9). A fluctuant subperiosteal abscess or even a draining fistula from the mastoid to the postauricular area may be present (Figure 20). The subperiosteal abscess can be in any of the anatomic sites described.

Examination of the tympanic membrane usually reveals middle-ear effusion, or there may be a purulent discharge and a perforation. In the 20-year review of 79 Danish children who developed acute mastoiditis and had a mastoidectomy, 92% had a purulent middle-ear effusion. Conversely, the tympanic membrane and middle ear may appear almost normal for the reasons described when mastoiditis with periostitis occurs. Acute mastoid osteitis (without otitis media) may also be the focus of infection when a child has a fever of unknown origin, in which case the disease in the mastoid is occult; as described later, the term subacute or masked mastoiditis has been used to describe this entity.

**Diagnosis**

The diagnosis should be suspected on the basis of clinical signs and symptoms. CT scans of the mastoid area usually reveal one or more of the following: haziness, distortion, or destruction of the mastoid outline; loss of sharpness of the shadows of cellular walls owing to demineralization, atrophy, and ischemia of the bony septa (Figure 21); and a decrease in density and cloudiness of the areas of pneumatization owing to inflammatory swelling of the air cells. In longstanding cases, a chronic osteoblastic inflammatory reaction may obliterate the cellular struc-
ture. Small abscess cavities in sclerotic bone may be confused with pneumatic cells.

Antonelli and colleagues reviewed CT scans of 21 patients with acute coalescent mastoiditis (with osteitis) or acute noncoalescent mastoiditis (without osteitis) and 12 patients with chronic mastoiditis. They found that erosion of the cortical plate overlying the sigmoid sinus was the most sensitive and specific CT finding to distinguish osteitis from nonosteitis acute disease.

As part of the diagnostic workup, cultures to determine the causative bacterial organisms should be obtained, either before or at the time of mastoid surgery. When otorrhea is present, cultures for bacteria from the ear drainage must be taken with care to discern fresh drainage from debris in the external canal. As described in detail earlier (see “Chronic Perforation with Chronic Otitis Media [Chronic Suppurative Otitis Media]”), the canal must be initially cleaned. Then, if fresh pus is exuding through a perforation in the tympanic membrane, the discharge is cultured at the point of exit from the tympanic membrane with a cotton-tipped wire swab or, preferably, a needle and syringe under direct view. A Gram stain of the pus provides...
immediate information about the responsible organisms. When the mastoidectomy is performed, portions of mucosa and bone of the mastoid should be sent for Gram stain, culture, and antibiotic susceptibility testing.

The differential diagnosis between this stage of mastoiditis and other disease entities (eg, acute external otitis with postauricular periostitis or postauricular lymphadenitis) that involve the postauricular area is described in the section “Acute Mastoiditis with Periostitis” (see Table 9).

**Microbiology**

The microbiology of mastoid osteitis is the same as that of acute mastoiditis with periostitis, as described before (see Table 10). In the review by Petersen and colleagues of children who had a mastoidectomy for acute mastoiditis, *S. pneumoniae*, *S. pyogenes*, and *H. influenzae* were the most common organisms isolated. Unusual organisms, such as *M. avium* and *M. chelonae*, have been reported.

**Management**

An otolaryngologist should be consulted if a child has a diagnosis of acute mastoid osteitis. Parenteral antimicrobial therapy should be instituted as described before for acute mastoiditis with periostitis. To ensure that drainage of the middle ear and mastoid is adequate, in the absence of a large perforation and otorrhea, a wide-field large myringotomy should be done immediately. Insertion of a tympanostomy tube,
in addition to a large myringotomy incision, can provide more prolonged drainage from the middle-ear–mastoid than myringotomy alone. The tympanostomy tube placement will also help prevent AOM (and mastoiditis) from recurring.

A cortical (simple) mastoidectomy is usually required when there is evidence of acute mastoid osteitis, especially when the mastoid empyema has extended outside the mastoid bone and a subperiosteal abscess is present. The procedure should be considered an emergency, but the timing of the operation depends on the status of the child. Ideally, sepsis should be under control, and the patient must be able to tolerate a general anesthetic. The principle is to clean out the mastoid infection, to drain the mastoid air cell system into the middle ear by eliminating any obstruction caused by edema or granulation tissue in the aditus ad antrum, and to provide external drainage (Figure 22). If another suppurative intratemporal or intracranial complication is also present, surgical intervention for these conditions may also be required.

Summary of Recommended Treatment Plan Related to the Stage of Acute Mastoiditis

Table 11 shows the clinical features of acute mastoiditis and findings on a CT scan on presentation to the physician related to our recommended management. From this summary, it is apparent that we recommend consultation with an otolaryngologist for possible tympanocentesis and myringotomy (and tympanostomy tube insertion) whenever the primary physician lacks the expertise (or qualifications for tympanostomy tube placement) to perform these procedures. Consultation with the otolaryngologist is also indicated for possible mastoidectomy whenever a child has the diagnosis of acute mastoiditis with subperiosteal abscess, regardless of the findings on a CT scan, and the CT scan...
reveals osteitis, regardless of the other clinical features. Surgical procedures for mastoidectomy have been described in detail elsewhere.9

An otolaryngologist should be immediately consulted whenever acute mastoiditis is associated with another possible suppurative complication of otitis media and mastoiditis and when acute mastoiditis develops in a child who has chronic suppurative otitis media, cholesteatoma, or both.

Subacute Mastoiditis (Masked Mastoiditis, Occult Mastoiditis)

A condition has been described called masked mastoiditis, for which a complete simple mastoidectomy has been advocated.150 The disease appears to be a subacute stage of otitis media and mastoiditis (without osteitis) characterized by the same signs and symptoms as AOM (such as fever and ear pain), except that they are persistent and less severe. The progression to this stage is attributed to failure of the initial antimicrobial agent to resolve the middle-ear and mastoid infection within a short period. Persistence of otalgia and fever in a patient receiving an antimicrobial agent is an indication for tympanocentesis and myringotomy to identify the causative organism and promote drainage. In selected children, especially those who have had frequently recurrent episodes of AOM, insertion of a tympanostomy tube (in addition to the appropriate antimicrobial therapy) will resolve the problem. Mastoid surgery is not indicated unless insertion of the tympanostomy tube and intravenous antimicrobial therapy are ineffective.193

Denoyelle and colleagues described 165 children, identified during 2 years, with subacute mastoiditis, which was defined as an attack of AOM that did not resolve with 10 days of antibiotic treatment, despite intravenous therapy.316 Middle-ear aspiration (or cultures of otorrhea) revealed H. influenzae (28%), P. aeruginosa (23%), and S. pneumoniae (16%). Of these 165 children, 31 (19%) had mastoidectomy. It is likely that tympanostomy tube insertion in
these children would have prevented mastoidectomy in most if not all.

In contrast to this condition, which is usually a failure of initial antimicrobial treatment that frequently resolves after adequate middle-ear drainage and identification of the causative organism followed by administration of a culture-directed antibiotic, infants and children may have a suppurative process in the mastoid that is not clinically obvious (ie, occult). This mastoid infection may even result in an intratemporal or intracranial complication, in which the middle ear may not appear to be diseased and the patient lacks the classic signs and symptoms of otitis media and mastoiditis. This condition can be called masked mastoiditis. The diagnosis is usually made by CT scan or by obtaining a bone scan.317 Children who have intracranial suppurative disease or disease of the temporal bone that could possibly be due to mastoid infection should have CT of the temporal bones included in the workup, even though there is no evidence of middle-ear disease on otoscopy. Although rare, children who have fever of unknown origin may have masked mastoiditis.

On occasion, older children or adolescents will complain of persistent or recurrent postauricular pain, but the middle ear appears to be free of disease. The communication between the middle ear and the mastoid air cells (ie, aditus ad antrum) may be blocked, causing mastoiditis. These children usually have had a history of recurrent AOM, recurrent or chronic otitis media with effusion, or chronic suppurative otitis media, and when a CT scan is obtained, mastoiditis is diagnosed. Medical treatment (eg, antimicrobial therapy) is indicated, but if the symptoms are severe or a trial of medical management fails, a mastoidectomy is indicated to relieve the aditus ad antrum blockage and eliminate the infected cells. More rarely, a child with these symptoms may have relatively normal mastoid cellular architecture on a CT scan, in which case, the patient could have negative pressure within the mastoid because of an aditus ad antrum obstruction. Again, mastoidectomy may be the only method of eliminating the blockage and relieving the symptoms.

**Chronic Mastoiditis (with and without Chronic Suppurative Otitis Media or Cholesteatoma)**

Today, chronic mastoiditis is most commonly associated with chronic suppurative otitis media (see earlier).318 Also, chronic mastoiditis can be involved in a child’s ear that has a cholesteatoma that extends to the mastoid gas (air) cells, which is associated with chronic suppurative otitis media.179 However, an occasional child will develop acute mastoiditis that is either untreated or inappropriately treated, or the child is neglected, and the infection progresses to a chronic stage in which no perforation (or tympanostomy tube) is present in the tympanic membrane; no chronic suppurative otitis media, with or without otorrhea, is present. Nevertheless, the disease in the mastoid progresses to the chronic stage, and otitis media may or may not be present.193 These children can present with a fever of unknown origin or chronic or recurrent otalgia and tenderness over the mastoid process. Like the child who has subacute or masked mastoiditis, the patient with chronic mastoiditis may have another intratemporal or intracranial complication. A child with an intracranial infectious process, such as a brain abscess, who has no clinical evidence of otitis media or mastoiditis, may have chronic mastoiditis as the focus of the intracranial infection.193

**Diagnosis**

Examination of the tympanic membrane shows evidence of middle-ear effusion, but the eardrum may appear normal if the chronic infection is localized only to the mastoid. The diagnosis that the mastoid is affected is made by CT. The mastoid may be poorly pneumatized or sclerotic, or it may show evidence of bone destruction, with opacification of the mastoid.
Management
The chronic infection, if it is reversible, may be brought under control by medical treatment with antimicrobial agents (similar to those recommended for acute mastoiditis without periosteitis or osteitis). A tympanocentesis (for Gram stain, culture, and susceptibility studies) and myringotomy (for drainage) should be performed. When there are extensive amounts of granulation tissue and osteitis in the mastoid (ie, irreversible mastoid disease) or the child fails to improve with medical therapy, referral to an otolaryngologist is needed because a tympanomastoidectomy is required to eliminate the chronic mastoid osteitis. When another suppurative complication is present in addition to the chronic mastoiditis, such as brain abscess, dural sinus thrombosis, or otitic hydrocephalus, mastoidectomy is indicated.

Chronic mastoiditis can also be caused by a cholesteatoma, which is usually manifested by chronic otorrhea through a defect in the tympanic membrane. A cholesteatoma requires definitive surgical treatment.

Petrositis
Petrositis is a relatively rare suppurative complication secondary to an extension of infection from the middle ear and mastoid into the petrous portion of the temporal bone. All of the inflammatory and cellular changes described in the mastoid can also occur in the pneumatized petrous pyramid. Only about 30% of individuals have well-pneumatized petrous bones. Petrositis may be more frequent than is appreciated by clinical and radiographic signs because there is communication of the petrosal air cells with the mastoid–middle-ear system. Pneumatization usually does not occur before the age of 3 years. For a more complete and detailed discussion of petrositis, the reader is referred to Bluestone.

Petrositis may be either acute or chronic. It has also been found to be recurrent. In the acute form, there is extension of AOM and mastoiditis into the pneumatized petrous air cells. The condition, like acute mastoiditis, is usually self-limited, with resolution of the acute middle-ear and mastoid infection, but the infection in the petrous portion of the temporal bone occasionally does not drain owing to mucosal swelling or because granulation is obstructing the passage from the petrous air cells to the mastoid and middle ear. This results in acute petrous osteomyelitis. The widespread use of antimicrobial agents has made this a rare complication. Chronic petrous osteomyelitis, however, can be a complication of chronic suppurative otitis media or cholesteatoma, and it is much more common than the acute type. Pneumatization of the petrous portion of the temporal bone does not have to be present because the infection can invade the area by thrombophlebitis, by osteitis, or along fascial planes. The infection may persist for months or years with mild and intermittent signs and symptoms, or it may spread to the intracranial cavity and result in one or more of the suppurative complications of ear disease, such as an extradural abscess or meningitis.

At the Children’s Hospital of Pittsburgh during the 15-year period from 1980 to 1995, only four children were admitted with this diagnosis, and three of these patients had simultaneous intracranial suppurative complications, such as dural sinus thrombosis. All of these children had this complication as a result of spread of an attack of AOM.

Microbiology
The organisms that cause acute petrositis are the same as those that cause acute mastoid osteitis: S. pneumoniae, H. influenzae, and group A streptococci. Chronic petrous osteomyelitis, however, may be caused by the bacteria associated with chronic suppurative otitis media and cholesteatoma, such as P. aeruginosa or Proteus species. Tuberculosis has also been isolated from these ears.

Diagnosis
The disease is characterized by pain behind the eye, deep ear pain, persistent ear discharge, and
sixth nerve palsy. However, in the four patients who were admitted to Children’s Hospital of Pittsburgh during a 15-year period with this complication of otitis media, eye pain, deep ear pain, and persistent otorrhea were not consistently present, which has been the experience in other reviews. Eye pain is due to irritation of the ophthalmic branch of the fifth cranial nerve. On occasion, the maxillary and mandibular divisions of the fifth nerve are involved, and pain occurs in the teeth and jaw. A discharge from the ear is common with acute petrositis but may not be present with chronic disease. Paralysis of the sixth cranial nerve leading to diplopia is a late complication. Acute petrous osteomyelitis should be suspected when persistent purulent discharge follows a complete simple mastoidectomy for mastoid osteitis. The triad of pain behind the eye, aural discharge, and sixth nerve palsy is known as Gradenigo’s syndrome. The diagnosis of acute petrous osteomyelitis is suggested by the unique clinical signs. Standard radiographs of the temporal bones may show clouding with loss of trabeculation of the petrous bone. The visualization is uncertain, however, because of normal variation in pneumatization (including asymmetry) and the obscuring of the petrous pyramids by superimposed shadows of other portions of the skull. CT scans of the temporal bones can lead to diagnosis and should be obtained if there might be a possibility of an extension of infection into the cranial cavity. This complication must be distinguished from destructive lesions of the petrous apex owing to such conditions as cholesteatoma, cholesterol granuloma, and arachnoid cysts; CT and MRI can be diagnostic in distinguishing these diseases.

**Management**

Management of acute petrositis is similar to that described for acute mastoiditis with osteitis; at this stage, it can be considered further spread of the mastoid infection into the pneumatized petrous portions of the temporal bone. All four of the patients who had this diagnosis at the Children’s Hospital of Pittsburgh (see earlier) were successfully treated with high-dose broad-spectrum intravenous antibiotic therapy and cortical (simple) mastoidectomy without entering the petrous apex; the petrous apex disease most likely drained into the mastoid cavity during the postoperative period. The three patients with intracranial complications also had specific management of that complication. In more severe cases of acute petrous osteomyelitis and acute mastoid osteitis, a more aggressive surgical approach to management may be required. A transcanal approach to the petrous apex has been reported, but the more traditional approaches have been described by Coker and Jenkins.

**LABYRINTHITIS**

This complication of otitis media occurs when infection spreads into the cochlear and vestibular apparatus. The usual portal of entry is the round window or, less commonly, the oval window, but invasion may take place from an infectious focus in an adjacent area, such as the mastoid antrum, the petrous bone, or the meninges, and as a result of bacteremia. Schuknecht classified labyrinthitis into three types:

1. Serous (toxic) labyrinthitis, in which there may be bacterial toxins or biochemical involvement, but no bacteria are present
2. Suppurative (acute and chronic otogenic suppurative) labyrinthitis, in which bacteria have invaded the otic capsule
3. Meningogenic suppurative labyrinthitis, which is the result of invasion of bacteria from the subarachnoid space into the labyrinth. Labyrinthitis ossificans (labyrinthine sclerosis), in which there is replacement of the normal labyrinthine structures by fibrous tissue and bone, is the end stage of this complication if it is arrested.

An acceptable classification of labyrinthitis today is acute labyrinthitis, subacute labyrinthitis, chronic labyrinthitis, and labyrinthitis ossificans.
Acute Labyrinthitis

Acute labyrinthitis can be classified as serous or suppurative, and each of these entities can be either localized or generalized. Acute Serous Labyrinthitis (with or without Perilymphatic Fistula)

The acute serous (toxic) type of labyrinthitis is considered to be one of the most common suppurative complications of otitis media. Paparella and colleagues described the histopathologic evidence of serous labyrinthitis in most of the temporal bone specimens from patients who had otitis media. Bacterial toxins from the infection in the middle ear may enter the inner ear, primarily through an intact round window or through a congenital defect between the middle ear and inner ear. The portal of entry may also be through an acquired defect of the labyrinth, such as from head trauma or previous middle-ear or mastoid surgery. Biochemical changes within the labyrinth have also been found. The cochlea is usually more severely involved than the vestibular system. Paparella and colleagues reviewed the audiograms of 232 patients who had surgery for chronic otitis media and found a significant degree of bone conduction loss in the younger age groups. In addition, there was a marked difference in the presence and degree of sensorineural hearing loss in the affected ear, compared with the normal ear, in patients of all age groups who had unilateral disease. They postulated that the high-frequency sensorineural hearing loss frequently accompanying this disease is due to a pathologic insult to the basal turn of the cochlea. In the review of intratemporal complications of otitis media from the Children’s Hospital of Pittsburgh between 1980 and 1995, three children were admitted with a diagnosis of acute serous labyrinthitis that was a complication of an attack of AOM. Fluctuating sensorineural hearing loss has been described in patients with otitis media and has been thought to be due to either endolymphatic hydrops or a perilymphatic fistula. However, fluctuating or progressive sensorineural hearing loss can be due to a variety of other hereditary and acquired causes.

Clinical Presentation

The signs and symptoms of serous labyrinthitis (especially when a perilymphatic fistula is present) are sudden, progressive, or fluctuating sensorineural hearing loss and vertigo in association with otitis media or one or more of its complications or sequelae, such as mastoid osteitis. The loss of hearing is usually mixed, that is, there are both conductive and sensorineural components, when serous labyrinthitis is a complication of otitis media. In some children who have recurrent middle-ear infections, the hearing may be normal between episodes. In other children, only a mild or moderate sensorineural hearing loss will be present at all times. The presence of vertigo may not be obvious in children, especially infants. Older children may describe a feeling of spinning or turning; younger children may not be able to verbalize the symptoms but manifest the dysequilibrium by falling, stumbling, or being clumsy. The vertigo may be mild and momentary, and it may tend to recur during a period of months or years. Onset of vertigo, progressive sensorineural hearing loss, or both in a patient who has a preexisting hearing loss is frequently due to a fistula. Spontaneous nystagmus may also be present, but the signs and symptoms of acute suppurative labyrinthitis, such as nausea, vomiting, and deep-seated pain, are usually absent. Fever, if present, is usually due to a concurrent upper respiratory tract infection or AOM.

If congenital perilymphatic fistula is present, nystagmus may be present during the course of the episode of AOM, in addition to the mixed hearing loss. In a review of 47 infants and children who had exploratory tympanotomy for possible fistula at the Children’s Hospital of Pittsburgh, 30 children (64%) had a history of otitis media, and of these 30 patients, 28 (93%) had a fistula diagnosed at surgery (Table 12).
**Diagnosis**

A labyrinthine fistula may be identified by performing a fistula test with a Siegle pneumatic otoscope or by applying positive and negative external canal pressure with use of the pump-manometer system of an impedance audiometer. The fistula test result is considered positive if nystagmus or vertigo is produced by the application of the pressures. Electronystagmography is an objective way of documenting the presence or absence of the nystagmus; the findings of the fistula test may be misleading because there can be false-positive and false-negative results. The test can be done in the presence of a perforation of the tympanic membrane or tympanostomy tube. Fistulae are frequently associated with congenital or acquired defects in the temporal bone, such as the Mondini malformation. CT scans may be helpful in identifying such defects, such as a dilated vestibular aqueduct, as shown in Figure 23. Weissman and colleagues assessed the CT scans of 10 children (15 ears) with fistula, confirmed at surgery, and were able to identify an abnormality of the inner ear, middle ear, or both in 53% of the ears. Electrocochleography has also been advocated for diagnosis of a fistula, but this method has not proved to be as sensitive and specific as desired. Currently, the most effective method for diagnosis of a perilymphatic fistula is exploratory middle-ear surgery.

Weber and colleagues proposed a relatively new test for the presence of perilymph in the middle ear as a way to confirm the presence of a perilymphatic fistula. A sample of the suspect fluid in the middle ear must be obtained at the time of exploratory surgery, and then it is assessed by an immunopathologic assay. The test appears to be specific but not sensitive. A study by Buchman and colleagues found that the test did not identify perilymph, but the result was positive when cerebrospinal fluid was present. Thus, when the test result is positive, it most likely confirms a cerebrospinal fluid leak from the inner to the middle ear. A more appropriate term for these malformations of labyrinthine windows is congenital perilymphatic/cerebrospinal fistula because the fluid emanating from the defect may be cerebrospinal fluid.

**Management**

When this complication occurs during an attack of AOM, a tympanocentesis and myringotomy should be performed for microbiologic assessment of the middle-ear effusion and drainage. If possible, a tympanostomy tube should also be inserted for more prolonged drainage and in an attempt to ventilate the middle ear. Antimicrobial agents with efficacy against S.
pneumoniae, H. influenzae, and M. catarrhalis, such as parenteral amoxicillin-sulbactam, should be administered; other organisms, such as S. aureus and Pseudomonas, have also been isolated from the middle ears of children who have acute labyrinthitis. After resolution of the otitis media with effusion, the signs and symptoms of the labyrinthitis should rapidly disappear; however, sensorineural hearing loss may persist. If the diagnostic assessment indicates a possible congenital or acquired defect of the labyrinth, an exploratory tympanotomy should be performed as soon as the middle ear is free of infection. The most common malformations are an abnormal round window and niche (such as a laterally facing round window), deformities of the stapes superstructure and footplate, deformed long process of the incus, or some combination of these congenital defects. More rarely, a congenital fissure between the round and oval windows is present. If a perilymphatic fistula is found, it should be repaired by use of temporalis muscle grafts. Even when no defect of the oval or round window is identified but a fistula is still suspected, the stapes footplate and round window should be covered with connective tissue because a leak may not be present at the time of the tympanotomy but may recur. A tympanostomy tube should be reinserted if recurrent otitis media persists.

When acute mastoid osteitis, chronic suppurative otitis media, or cholesteatoma is present, definitive medical and surgical management of these conditions is essential in eliminating the labyrinthine involvement. A careful search for a labyrinthine fistula must be performed when mastoid surgery is indicated. However, a labyrinthine fistula is not indicated for serous labyrinthitis. The surgical procedure to repair a perilymphatic or cerebrospinal fluid fistula is described in detail by Bluestone.

Conclusion

Any child with sensorineural hearing loss (with or without vertigo) who also has recurrent acute or chronic otitis media with effusion should be carefully evaluated for the possible existence of serous labyrinthitis, which can be secondary to a perilymphatic fistula. This combination appears to be common, and failure to identify this complication can result in irreversible severe to profound hearing loss, making early diagnosis and prevention imperative. Because prevention of sensorineural hearing loss owing to other causes (such as congenital and viral) is not yet possible, our goal should be to prevent this loss of function in those children in whom it can be prevented. In addition, serous labyrinthitis may develop into acute suppurative labyrinthitis.

Acute Suppurative Labyrinthitis

Suppurative (purulent) labyrinthitis may develop as a complication of otitis media or be one of its complications and sequelae when bacteria migrate from the middle ear into the perilymphatic fluid through the oval or round window, a preexisting temporal bone fracture, an area where bone has been eroded by cholesteatoma or chronic infection, or a congenital defect, such as a congenital perilymphatic (cerebrospinal) fistula, as described before. The most common way that bacteria enter the labyrinth is from the meninges, but migration by this route is usually not a complication of otitis media.

Epidemiology and Pathogenesis. The incidence of suppurative labyrinthitis as a complication of otitis media is unknown, but it is rare because of the widespread use of antibiotics. In a series of 96 cases of suppurative intratemporal and intracranial complications of acute and chronic otitis media that were treated from 1956 to 1971, there were only 5 cases of suppurative labyrinthitis, and all were secondary to cholesteatoma that had caused a labyrinthine fistula. Nonetheless, suppurative labyrinthitis still occurs. When it occurs in children who have an episode of AOM and who are apparently treated appropriately and adequately, a congenital (or acquired) perilymphatic fistula must be ruled out to prevent further hearing loss and recurrence, which can be life-threatening because of meningitis. Conversely, when a child develops bacterial meningitis, especially recurrent epi-
sodes, a congenital defect of the inner and middle ear must be ruled out. A congenital or acquired defect between the paranasal sinuses and the anterior cranial cavity can also cause meningitis. In a review of children who had intratemporal complications of otitis media, Goldstein and colleagues found two patients who had a suppurative labyrinthitis during a recent 15-year period; one child had a congenital defect of the labyrinthine windows, which was considered to be a perilymphatic or cerebrospinal fluid fistula (Figure 24).²⁶²

Meningitis that has even resulted in death was reported to be caused by otitis media progressing to labyrinthitis in children who have had cochlear implants (one offending implant has been removed from the market), but in most cases, the pathogenesis is most likely due to a congenital defect between the middle and inner ears. The appropriate vaccines have been recommended for prevention, and prompt treatment of AOM in children who have congenital malformations of the ear, and repair of the defect is desirable.⁹

Clinical Presentation and Diagnosis. The sudden onset of vertigo, dysequilibrium, deep-seated pain, nausea and vomiting, and sensorineural hearing loss during an episode of AOM or an exacerbation of chronic suppurative otitis media indicates that labyrinthitis has developed. The hearing loss is severe, and there is loss of the child’s ability to repeat words shouted in the affected ear, with masking of sound in the opposite ear. Spontaneous nystagmus and past-pointing can often be observed. Initially, the quick component of the nystagmus is toward the involved ear, and there is a tendency to fall toward the opposite side. However, when there is complete loss of vestibular function, the quick component is toward the normal ear.

Today, radiographic imaging can be an invaluable diagnostic aid. MRI can be diagnostic of the labyrinthitis, and CT can identify congenital or acquired defects of the inner and middle ear that may have predisposed the child to spread of the infection from the middle ear to the labyrinth (and subarachnoid space). In the absence of associated meningitis, the cerebrospinal fluid pressure and cell count are normal. The onset of suppurative labyrinthitis may frequently be followed by facial paralysis, meningitis, or both. In later stages, cerebellar abscess can develop. Thus, suppurative labyrinthitis is a serious complication of otitis media. The development of purulent labyrinthitis means that infection has spread to the fluid of the inner
ear, and infection can then spread to the subarachnoid space through the cochlear aqueduct, the vestibular aqueduct, or the internal auditory canal. Figure 25 shows the radiographic image of a child in whom acute suppurative labyrinthitis owing to otitis media spread to the labyrinth through a congenital perilymphatic fistula and spread to the meninges, resulting in death.

**Management.** The management of suppurrative labyrinthitis in the absence of meningitis consists of otologic surgery combined with intensive antimicrobial therapy. A congenital defect between the middle and inner ears should be investigated using imaging. If this complication is due to AOM, immediate tympanocentesis and myringotomy with tympanostomy tube insertion are indicated, as described when serous labyrinthitis is present. If acute mastoid osteitis is present, a cortical (simple) mastoidectomy should be performed. However, because this complication can be secondary to cholesteatoma, a radical mastoidectomy or modified radical mastoidectomy may be required. A modified radical mastoidectomy may also be required when chronic suppurative otitis media is present without cholesteatoma.

If meningitis has also occurred in association with suppurative labyrinthitis, otologic surgery other than a diagnostic and therapeutic tympanocentesis-myringotomy may have to be delayed until the meningitis is under control and the child is able to tolerate a general anesthetic, but it is important to control the source of the infection in the middle ear and labyrinth as soon as possible. A labyrinthectomy should be performed only if there is complete loss of labyrinthine function or if the infection has spread to the meninges in spite of adequate antimicrobial therapy. Parenteral antimicrobial agents appropriate for management of the primary middle-ear and mastoid disease should be administered initially. However, because cholesteatoma and chronic suppurative otitis media are frequent causes of suppurative labyrinthitis, antimicrobials effective for the gram-negative organisms (*P. aeruginosa* and *Proteus*) are frequently required. The results of culture of the middle-ear effusion, the purulent discharge, or the cerebrospinal fluid may alter the selection of the antibiotics.

**Chronic Labyrinthitis**

The most common cause of chronic labyrinthitis as a complication of middle-ear disease is a cholesteatoma that has eroded the labyrinth, resulting in a fistula. Osteitis may also cause bone erosion of the otic capsule. The fistula most commonly occurs in the lateral semicircular canal and is filled by squamous epithelium of a cholesteatoma, granulation tissue, or fibrous tissue entering the labyrinth. The middle ear and mastoid are usually separated from the inner ear by the soft tissue at the site of the fistula, but when there is continuity, acute suppurative labyrinthitis may develop. However, chronic labyrinthitis may be caused by chronic suppurative otitis media or even chronic otitis media with effusion, especially if the child has a congenital
defect between the middle and inner ear (congenital perilymphatic fistula).

Clinical Presentation and Diagnosis
The signs and symptoms of chronic labyrinthitis are similar to those of the acute forms of the disease (eg, sensorineural hearing loss and vertigo), except that their onset is more subtle. The disease is characterized by slowly progressive loss of cochlear and vestibular function during a prolonged period. The fistula test may be helpful in diagnosis of a labyrinthine fistula, MRI may reveal labyrinthitis, and CT may reveal a bony defect. When there is complete loss of function, there may be no signs or symptoms of labyrinthine dysfunction.

Management
Because a cholesteatoma is the most common cause of this type of labyrinthitis, middle-ear and mastoid surgery must be performed. For children with a labyrinthine fistula owing to a cholesteatoma, a modified radical mastoidectomy may be required. When labyrinthine function is still present, the cholesteatoma matrix overlying the fistula should be left undisturbed because removal can result in total loss of function. Even though there are advocates of performing an intact canal wall procedure and surgeons who prefer to remove the cholesteatoma matrix, either during the initial surgery or in a second-stage procedure, the approach that is the safest is recommended when a cholesteatoma has caused a labyrinthine fistula in a child.

Failure to diagnose this complication and perform the surgery as soon as possible may result in complete loss of cochlear and vestibular function with possible development of labyrinthine sclerosis or an acute suppurative labyrinthitis. Suppurative labyrinthitis can cause a life-threatening intracranial complication, such as meningitis.

Labyrinthitis Ossificans (Labyrinthine Sclerosis)
Labyrinthitis ossificans is caused by fibrous replacement or new bone formation (labyrinthitis ossificans) in part or all of the labyrinth, with resulting loss of labyrinthine function. Today, this end stage of labyrinthitis is most commonly the result of meningitis, not of otitis media. As found in the review of CT scans by Weber and colleagues, however, one child had labyrinthitis ossificans in association with a congenital perilymphatic fistula, presumably secondary to otitis media because meningitis had not occurred. This condition is the end stage of healing after acute or chronic labyrinthitis, and prevention of disease of the middle ear is the most effective way to avoid this complication.

FACIAL PARALYSIS
Facial paralysis, as a complication of otitis media and related diseases, is still a common problem in infants and children. Facial paralysis most commonly occurs in children during an episode of AOM because of exposure of the facial nerve from a congenital bony dehiscence in its tympanic portion within the middle ear (Figures 26 and 27). It can also be a complication of acute mastoiditis with osteitis or chronic suppurative otitis media. When it occurs as a complication of chronic suppurative otitis media,
A cholesteatoma is also frequently present. On rare occasions, it can occur in children as a complication of otitis media with effusion. Also, on rare occasions, it can be bilateral after the onset of AOM or as a complication of acute mastoiditis. Not infrequently, when facial paralysis develops during an attack of otitis media, an underlying disease, such as leukemia, may be present. It is not uncommon for a child who has an acute facial paralysis to be examined in an emergency center and to be misdiagnosed as having Bell’s palsy. All infants and children who have an acute onset of a facial paralysis should have a complete ear, nose, and throat examination, which should include pneumatic otoscopy to rule out an otogenic cause of the paralysis.

Facial paralysis is a relatively frequent complication of AOM in infants and children. In the preantibiotic era, facial paralysis was estimated to occur in 0.5% of patients with AOM, whereas the current rate is 0.005%, as reported in a study from Denmark. A review by Goldstein and colleagues of 22 infants and children who had facial paralysis in association with otitis media or related infections at the Children’s Hospital of Pittsburgh between 1980 and 1995 revealed that paralysis occurred most frequently in children aged 6 years and younger; 50% were younger than 4 years (Table 13).

Table 13. Age, Presenting Symptoms, and Signs in 22 Infants and Children Who Developed Acute Facial Paralysis as a Complication of Otitis Media or Related Infection: Children’s Hospital of Pittsburgh, 1980–1995

<table>
<thead>
<tr>
<th>Characteristic</th>
<th>Number of Patients (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age (yr)</td>
<td></td>
</tr>
<tr>
<td>0–3</td>
<td>11 (50)</td>
</tr>
<tr>
<td>4–6</td>
<td>6 (27)</td>
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<tr>
<td>7–12</td>
<td>3 (14)</td>
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<tr>
<td>13–18</td>
<td>2 (9)</td>
</tr>
<tr>
<td>Signs and symptoms</td>
<td></td>
</tr>
<tr>
<td>Facial weakness</td>
<td>22 (100)</td>
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<tr>
<td>Middle-ear effusion</td>
<td>16 (73)</td>
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<tr>
<td>Otalgia</td>
<td>15 (69)</td>
</tr>
<tr>
<td>Otorrhea</td>
<td>8 (36)</td>
</tr>
<tr>
<td>Perforation of tympanic membrane</td>
<td>8 (36)</td>
</tr>
<tr>
<td>Upper respiratory tract infection</td>
<td>8 (36)</td>
</tr>
<tr>
<td>Fever</td>
<td>7 (32)</td>
</tr>
<tr>
<td>Hearing loss</td>
<td>2 (9)</td>
</tr>
<tr>
<td>Irritability</td>
<td>2 (9)</td>
</tr>
<tr>
<td>Postauricular cellulitis/mass</td>
<td>2 (9)</td>
</tr>
</tbody>
</table>

Adapted from Goldstein NA et al. Facial weakness, otalgia, otorrhea, concomitant upper respiratory tract symptoms of infection, and fever were the most common symptoms. The mean duration of ear symptoms and of facial weakness on presentation to the hospital was 6 and 4 days, respectively. Two children had acute mastoiditis with periosteitis, and one patient had acute mastoiditis with osteitis and subperiosteal abscess. In 18 patients (80%), initial treatment consisted of antimicrobial therapy, and in all but 1 of these children, a myringotomy, with or without tympanostomy tube placement, was also performed. However, of these 22 children, 4 patients (18%) had further surgery: 2 patients had a cortical (simple) mastoidectomy for acute mastoiditis; 1 child who had lymphoblastic leukemia underwent facial nerve decompression, radical mastoidectomy, and a labyrinthectomy; and 1 child who had had a previous mastoidectomy for cholesteatoma later developed an acute mastoiditis with subperiosteal abscess and a facial paralysis and had a revision tympanomastoidectomy.

In a smaller series of 10 patients who developed facial paralysis after the onset of AOM in Washington, DC, Elliott and colleagues
reported that 8 had an incomplete paralysis, which resolved with only myringotomy and intravenous administration of an antibiotic. Two children who had complete paralysis and persistent fever and otorrhea despite antibiotic treatment had a mastoidectomy. None required decompression of the facial nerve.

Most children who have facial paralysis that is not associated with acute mastoiditis or a concomitant underlying disease can be treated successfully with parenteral antimicrobial therapy, tympanocentesis, and myringotomy with or without tympanostomy tube insertion. When it occurs as an isolated complication, tympanocentesis and a myringotomy should be performed, and parenteral antibiotics effective for *S. pneumoniae* and *H. influenzae* should be administered. Insertion of a tympanostomy tube is indicated when the child has had recurrent episodes of AOM, when the attack AOM is superimposed on preexisting chronic otitis media with effusion, and whenever prolonged drainage of the middle ear and mastoid is desirable. The paralysis will usually improve rapidly without requiring further surgery (eg, mastoidectomy or facial nerve decompression). Mastoidectomy is not indicated unless acute mastoiditis with osteitis (acute “coalescent” mastoiditis), chronic suppurative otitis media, or cholesteatoma is present. However, if there is complete loss of facial function and electrophysiologic testing indicates the presence of degeneration or progressive deterioration of the nerve, facial nerve decompression may be necessary to achieve complete return of function.

Immediate surgical intervention is indicated when a facial paralysis develops in a child who has chronic suppurative otitis media, cholesteatoma, or both.

**EXTERNAL OTITIS**

Otitis media with perforation (also a patent tympanostomy tube) can be associated with an infection of the external auditory canal secondary to a discharge from the middle ear and mastoid. This is *external otitis*; the terms *acute diffuse external otitis* and *acute infectious eczematoid dermatitis* are also used. The infection can be acute or chronic. When acute, it usually follows an episode of AOM in which the tympanic membrane perforates, or a chronic perforation (without otitis media) is present and an episode of AOM occurs. The otorrhea that follows is then complicated by acute infection of the external auditory canal. Similarly, when a tympanostomy tube is in place, an attack of AOM develops with otorrhea followed by infection of the external canal. An infection in the mastoid may also erode the bone of the ear canal or the postauricular area, resulting in a dermatitis.

**Diagnosis**

The ear canal skin is erythematous, edematous, and filled with purulent drainage and yellow-crusted plaques, and a perforation of the eardrum (or tympanostomy tube) is present. It is frequently difficult for the clinician to distinguish between external otitis caused by otitis media with otorrhea secondary to a perforation or tympanostomy tube and acute diffuse external otitis in which middle-ear disease is absent. However, severe inflammatory stenosis of the external auditory meatus owing to middle-ear infection and otorrhea is uncommon; therefore, it can be differentiated from the extreme tenderness and pain that are so commonly present in acute external otitis not secondary to middle-ear infection. When the differential diagnosis is in doubt, the most effective way in which to make the distinction is to clean the products of the infection from the external auditory canal with a suction or cotton-tipped applicator to determine the status of the tympanic membrane. However, the external auditory canal may be too edematous and tender to be cleaned adequately for visualization of the entire drum. In such a circumstance, the immediate history (eg, recent episode of AOM) may be valuable. If a significant hearing loss is present, in the absence of extreme canal stenosis, it is also likely that the patient has otitis media in association with the
external otitis. A tympanogram can be helpful if the canal is not too tender for insertion of the probe tip of the instrument because a large volume indicates a nonintact tympanic membrane. The presence of a tympanometric peak in the normal pressure zone, even though somewhat lower than normal, suggests a normal middle ear; a flat pattern (with normal canal volume) suggests the presence of a middle-ear effusion without a perforation or patent tympanostomy tube. Also included in the differential diagnosis are impetigo contagiosa and a secondary infection associated with contact, or seborrheic, dermatitis.

The external canal infection may be associated with a mass in the canal, on the tympanic membrane, or both that is difficult to distinguish between an aural polyp, which may be due to a middle-ear cholesteatoma, versus a true tumor of the ear canal. When a canal mass is present, thorough cleaning of the canal and elimination of the external canal otorrhea are critical in confirming the diagnosis. When this procedure is difficult in the young child or one who is uncooperative, the patient may have to be taken to the operating room for an examination under general anesthesia (see Chapter 7). Appropriate imaging of the temporal bone, biopsy, or both may be required for a definitive diagnosis.

When the external otitis owing to otitis media involves the pinna and postauricular area, the condition may be misdiagnosed as acute mastoiditis with periostitis. As described in Table 9, however, the postauricular crease is commonly obliterated when the swelling is secondary to external otitis, whereas the crease is maintained when acute mastoiditis with periostitis is present.

**Microbiology**

The organisms involved are usually the same as those found in the middle-ear–mastoid infection, but the flora of the external canal may contribute to the infectious process. *P. aeruginosa*, *S. aureus*, and *Proteus* are frequently present. Also, MRSA may be the causative organism. Fungi may also be found in chronic cases—most commonly, *Aspergillus niger* or *Aspergillus alba*. These are bacteria similar to those isolated from the external canals of individuals who have acute external otitis in which the tympanic membrane is intact.

A culture of the external auditory canal should be obtained and the results compared with those of a needle aspiration of the middle-ear discharge through the tympanic membrane perforation or the tympanostomy tube. This will aid in determining the offending organisms. Antimicrobial therapy can then be selected by the results of the culture and susceptibility testing. The infection may spread to the auricle, periauricular area, or other parts of the body, possibly as a result of direct implantation of the organisms or as an autosensitivity phenomenon. Coagulase-positive *S. aureus* is the most frequently involved bacterium in this type of reaction.

**Management**

Management should be directed toward resolving the middle-ear–mastoid infection. This may require medical treatment, surgery, or both. But the principles of treatment for otitis externa should be followed if the skin of the ear canal and the tympanic membrane are involved, but it should be remembered that the tympanic membrane is not intact when this complication occurs secondary to a middle-ear infection. The ototopical medications listed in Table 3 can be beneficial, but ofloxacin otic solution (Floxin) and ciprofloxacin-dexamethasone suspension (Ciprodex) are currently the only ototopical agents approved by the FDA for use in children who have a tympanostomy tube in place and develop otorrhea. The other ototopical drops used in the past, such as the aminoglycosides, are potentially toxic to the inner ear when the tympanic membrane is not intact and are not indicated. Ofloxacin is safe and effective, and it is approved for children (and adults) who have external otitis. These topical ear drops are usually equally effective when a
perforation is present and an episode of acute otitis occurs. It is also approved for use in adults who have chronic suppurative otitis media. Other combinations of an antibiotic with hydrocortisone otic drops, such as ciprofloxacin hydrochloride with hydrocortisone, may be effective when external otitis is present and are approved for children when the tympanic membrane is intact and external otitis is present. 365

Analgesics are usually required to control the otalgia until the infection is under control. When canal stenosis is present and too severe to permit adequate application of ototopical medications, a cotton or Mericel Pope Ear Wick may have to be inserted into the ear canal to enhance treatment. When the infection is severe, such as when the patient has a high fever or is toxic, or when the infection spreads to the pinna, periauricular areas, or cervical lymph nodes, intravenous antimicrobial therapy effective against the offending bacterial organism is required. Even though the oral quinolones would be a reasonable initial treatment alternative to parenteral antibiotics at this stage, these agents are not approved for children younger than 18 years. Since the orally administered quinolones, such as ciprofloxacin, have been used for many years in children who have cystic fibrosis and appear to be safe, these agents are a reasonable alternative to parenteral therapy, but appropriate informed consent should be obtained from the parent or guardian prior to being given. But intravenous antimicrobial therapy may have to be instituted, which can be administered on an outpatient basis. 254

If a fungal infection is present, m-cresyl acetate eardrops (Cresylate) may need to be prescribed. Irrigation of the ear canal with 2% acetic acid or frequent suctioning of the ear canal may also hasten the resolution of the external ear canal infection, but, again, acetic acid is not approved for use when the tympanic membrane is not intact. Likewise, other topical antifungal agents, such as terbinafin hydrochloride, are not approved when the eardrum is open. An alternative is to paint the external auditory canal with gentian violet, being careful not to contaminate the middle ear with this medication, but this method is also problematic owing to a lack of approval for use for this ear infection.

When the skin adjacent to the auricle or on other parts of the body is involved, the skin should be cleaned with saline solution or aluminum acetate and treated with a local antibiotic-corticosteroid cream. The child should be cautioned about spread of the infection from the ear canal to other parts of the body and should refrain from putting his or her finger in the ear or scratching the infected skin. Cotton in the external ear canal can be helpful if profuse drainage is present but should be changed as frequently as necessary. One recently reported randomized trial found that medicated ribbon gauze inserted into the external canal facilitates cure more effectively and less costly than the customary insertion of ear wicks, but this study was conducted in patients with external otitis and presumably an intact eardrum. 366

The acute infection can become chronic, which requires a thorough investigation of the pathogenesis and etiology to rule out chronic middle ear and mastoid disease. 122

Strategies to prevent recurrence of this complication of otitis media are similar to those described for when AOM or chronic suppurative otitis media occurs in the presence of a perforation of the eardrum or tympanostomy tube (see “Perforation of the Tympanic Membrane”).

ATELECTASIS OF THE MIDDLE EAR (WITH AND WITHOUT A RETRACTION POCKET)

Atelectasis of the middle ear—which includes the tympanic membrane—is a sequela of eustachian tube dysfunction. The condition, as the term implies, occurs when there is recurrent or chronic underpressure in the middle ear, which results in retraction or collapse of the tympanic membrane. Retraction of the tympanic membrane may be attributed to the presence of high negative middle-ear pressure; retraction of the tympanic membrane can also occur in the absence of middle-ear negative pressure. A flaccid, atelectatic tympanic membrane may or may not be
associated with high negative intratympanic pressure; the abnormal negative pressure may have been the original cause of such a condition of the membrane but may no longer be present. Also, a middle-ear effusion may or may not be present. The term atelectasis has been borrowed from the pulmonary disorder atelectasis of the lung.

Unfortunately, there have been no reports of studies of the incidence of atelectasis and retraction pockets. However, a retraction pocket is a common sequela of atelectasis of the tympanic membrane with or without otitis media with effusion.\(^{367}\) As a sequela of tympanostomy tube insertion, retraction pockets at the site of the extruded tube are common and are more prevalent in older children, adolescents, and adults as a long-term aftermath of tubes than in young children.\(^{368}\) The incidence in individuals with cleft palates must be greater than that of cholesteatoma in this population (7.1%) because a retraction pocket precedes the development of a cholesteatoma in children with cleft palate.\(^{369}\)

**Pathogenesis**

Atelectasis of the middle ear is the result of transient or chronic underpressure within the middle ear, which, in turn, is caused by eustachian tube dysfunction. The dysfunction is most commonly failure of opening of the eustachian tube, anatomic obstruction, or both; in some individuals, it may be secondary to habitual sniffing in the presence of a tube that fails to close.\(^{370}\) Because the eustachian tubes of individuals with chronic atelectatic middle ears can be successfully inflated, the tube is not anatomically obstructed.\(^{371}\) Thus, the tube is more likely to be functionally obstructed, that is, there is a failure of the opening mechanism.\(^{369,372}\)

An alternative but less convincing hypothesis for the pathogenesis is related to the persistence of mesenchyme or an inflammatory reaction, secondary to otitis media, in the middle ear, especially in the posterosuperior quadrant of the pars tensa and the pars flaccida, two sites that commonly develop partial atelectasis with a retraction pocket.\(^{373}\) However, these two portions of the tympanic membrane are the most compliant areas of the eardrum\(^{374}\) and thus most likely to retract when there is significant under-pressure within the middle ear. In some children, these deformations of the tympanic membrane are seen when chronic otitis media with effusion is present.\(^{375}\) Retraction pockets are also not an uncommon sequela after extrusion of tympanostomy tubes. The pocket occurs when the tympanic membrane has a dimeric membrane at the tube site, and if eustachian tube dysfunction persists, the membrane retracts because it is more compliant than before the tube was inserted. Thus, retraction pockets occur at naturally floppy portions of the tympanic membrane (posterosuperior quadrant and pars flaccida) or at the site of an acquired defect, such as a dimeric membrane from a previously inserted tube or the site of a healed spontaneous perforation.

In studies of children who have retraction pockets, the eustachian tube was determined to have a failure of the opening mechanism, that is, functional obstruction (see Chapter 3).\(^{376}\) This sequela of eustachian tube dysfunction or otitis media increases with advancing age.\(^{368,377}\) If the atelectasis is persistent and progressive, it can lead to sequelae such as hearing loss, ossicular chain discontinuity, and cholesteatoma. All are described in this chapter.

Also, an atelectatic middle ear can have positive pressure in the middle ear,\(^{378}\) but this phenomenon may be due to insufflation of gas from the nasopharynx during closed-nose swallowing (ie, Toynbee phenomenon) or from forced inflation (eg, Valsalva’s maneuver) of the eustachian tube-middle ear in an effort to improve hearing and treat middle-ear pressure sensation; even though positive pressure is within the middle ear, dysfunction of the eustachian tube is still the underlying pathogenic mechanism.

**Diagnosis and Classification**

The diagnosis of atelectasis is only by visual inspection with the aid of either an otoscope or otomicroscope; tympanometry is not diagnostic.
The tympanic membrane is deformed. Atelectasis of the middle ear can be acute or chronic; partial (localized), which may or may not be a retraction pocket, or total (generalized); and mild, moderate, or severe. Partial atelectasis, with or without a retraction pocket, may also be visualized in an area of a healed perforation or at the site where a tympanostomy tube had been inserted (“atrophic scar” or dimeric membrane). A retraction pocket in the posterosuperior quadrant of the pars tensa or a pars flaccida retraction pocket is more frequently associated with the development of more serious sequelae (ossicular discontinuity, cholesteatoma, or both) than is a retraction pocket in other areas of the tympanic membrane. As described later, these variations should be kept in mind in deciding how to manage atelectasis.

It is appropriate to classify, grade, and stage atelectasis of the middle ear, as visualized by the clinician, according to its extent, severity, and duration because these factors are related to management decisions. The following is a proposed classification:

**Partial atelectasis** is a localized area of the tympanic membrane that is atelectatic, which may or may not be a retraction pocket because the depth of the retraction can be mild, moderate, or severe. When partial, in the absence of a retraction pocket, it may be in one or more of the four quadrants of the pars tensa (ie, anterosuperior, anteroinferior, posterosuperior, posterosinfe-rior), in the pars flaccida, or in both the pars tensa and the pars flaccida.

A **retraction pocket** is characterized by a partial area of atelectasis of the tympanic membrane in which there is indrawing of the membrane forming borders (ie, an edge or margin), most frequently at the site of an osseous anatomic structure (eg, notch of Rivinus or scutum) or the malleus. A retraction pocket can be in one or more of the four quadrants of the pars tensa, in the pars flaccida, or both and can be acute or chronic or reversible or irreversible. Sade proposed a classification of a posterosuperior retraction pocket that is helpful but does not include duration, the presence or absence of adhesive changes (which relates to reversibility), or other sites. Tos and Poulsen classified attic retraction pockets in relation to their extent and severity.

The stages of retraction pockets are divided into acute (less than 3 months in duration) and chronic (3 months or longer in duration). Key factors that affect the progression of a retraction pocket from stage 1 to stage 4 are the following:

1. **Relation to middle-ear structures:** The pocket does or does not approximate (touch) or is not adherent (ie, adhesive otitis media) to an ossicle (ie, incus, incudostapedial joint, stapes, head of malleus, or incudomallear joint) or other middle-ear structure, such as the promontory of the cochlea.

2. **Expands with pressure:** The entire pocket does or does not easily expand to the normal position when negative pressure is applied with a pneumatic otoscope or with the Bruening otoscope with a nonmagnifying lens under the otomicroscope, or when positive pressure is applied when the patient is anesthetized with nitrous oxide.

3. **Extent visualized:** The entire pocket is visualized or parts are not seen even after pressure is applied. This is because the pocket extends beyond the visible portion of the middle-ear space (eg, sinus tympani, facial recess, epi- tympanum, or medial to other parts of the tympanic membrane).

4. **Self-cleansing and free of infection:** Epithelial debris, crusting, or purulent material is or is not within the pocket.

**Total atelectasis** can be acute (less than 3 months in duration) or chronic (3 months or longer in duration). It involves all four quadrants of the pars tensa, with or without involvement of the pars flaccida, and can be staged as follows:

Stage 1a, **acute total mild atelectasis:** middle ear aerated

Stage 1c, chronic total mild atelectasis: same as stage 1a but chronic

Stage 2a, acute total severe atelectasis: middle ear not aerated (ie, no apparent middle-ear space)

Stage 2c, chronic total severe atelectasis: same as stage 2a but chronic

Management

When atelectasis of the tympanic membrane is of relatively recent onset, a middle-ear effusion may or may not be present, and a retraction pocket is absent, current management options are of uncertain benefit and are controversial. There have been no clinical trials reported that have addressed the efficacy of the various treatments compared with no treatment. The decisions related to actively treat or not to treat and the treatment options when an effusion of short duration is present are discussed in detail in Chapter 8.) Total atelectasis, or even a partial area that is retracted for only a short time (acute retraction), is usually caused by transient high negative middle-ear pressure associated with an acute upper respiratory tract infection or barotrauma. This condition is common in children and adolescents and is usually self-limited. No specific treatment should be directed toward the middle ear unless the child complains of severe otalgia, hearing loss, tinnitus, or vertigo. The atelectasis (and high negative intratympanic pressure) and associated symptoms, if present, will usually subside when the acute upper respiratory tract infection disappears. Treatment at this time should be directed toward relief of the nasal symptoms. Topical or systemic nasal decongestants may provide relief of these symptoms and may also relieve congestion of the eustachian tube, although their effectiveness in this latter area has not been demonstrated. If the symptoms become severe, myringotomy may be necessary to provide relief by returning middle-ear pressure to ambient levels, such as when the atelectasis is secondary to barotrauma (see Chapter 8). Middle-ear inflation through the eustachian tube has been advocated, but as described later, when atelectasis progresses to the chronic stage, this method of management is of uncertain benefit. Thus, watchful waiting is appropriate.

When the atelectasis is chronic (with or without the presence of chronic otitis media with effusion) and there is no evidence of a deep retraction pocket in the posterosuperior quadrant or pars flaccida, a thorough search should be made for an underlying origin, such as hypertrophied adenoids, nasal allergy, or paranasal sinusitis. If none is found, the management options are either watchful waiting or active treatment. (See also Chapter 8, when chronic otitis media with effusion is present.) The decision for or against treatment should rest on the presence or absence of other associated symptoms and whether there is abnormal negative pressure within the middle ear. The presence of persistent or transient otalgia, hearing loss, vertigo, or tinnitus that is troublesome to the patient warrants active treatment. For chronic atelectasis in this case, a trial with a topical or systemic nasal decongestant with or without an antihistamine may be helpful. However, this type of treatment is often disappointing.

Inflation of the eustachian tube–middle-ear system by use of the Valsalva or Politzer method has been advocated when middle-ear effusion is present. However, studies in animals indicate that these methods will not return the middle-ear pressure to normal for a sustained period when the eustachian tube is functionally obstructed. A clinical trial conducted by Chan and Bluestone also failed to show efficacy of middle-ear inflation in 41 children who had chronic middle-ear effusion with use of a specially designed inflation device. Kaneko and colleagues also failed to show consistent and sustained improvement after inflation in children who had middle-ear effusion.

Inflation of the eustachian tube and middle ear may provide temporary relief of atelectasis, in which no effusion is present, but must usually be repeated for permanent control of the symptoms.
and to maintain the tympanic membrane in a more normal position. However, Luntz and Sade inflated the middle ears of individuals with chronic atelectasis with either air or nitrogen for up to 5 consecutive days and found that the eardrum returned to its former atelectatic position within 15 minutes to 5 hours after the inflation. Yung used nasal continuous positive airway pressure in patients with atelectasis and reported a short-term benefit but did not study the long-term effect. Doyle and Alper explained this phenomenon using a mathematical model.

When a chronic retraction pocket or total atelectasis is present and nonsurgical methods of management have failed, myringotomy and insertion of a tympanostomy tube should be performed to prevent possible irreversible changes in the middle ear. After insertion of a tympanostomy tube, the tympanic membrane in the area of the retraction pocket should return to a more neutral position within several weeks or months. If the retraction area remains adherent to the ossicles, middle ear, or both (Figure 28), adhesive otitis media is present (see "Adhesive Otitis Media"), and tympanoplasty should be considered to prevent further progression of the disease process (such as ossicular discontinuity, cholesteatoma formation, or both). To prevent a recurrence of the retraction pocket (or total atelectasis), in addition to tympanostomy tube insertion, a portion of cartilage (from the pinna) placed over the affected area is effective. Even though this method of management has not been tested in appropriately controlled clinical trials and the natural history of retraction pockets in these areas has not been adequately studied, this method of management appears to be reasonable.

When a flaccid tympanic membrane is passively collapsed on the ossicles and middle ear and high negative middle-ear pressure is not present, the nonsurgical and surgical management options described before may not be effective in restoring the tympanic membrane to a more normal position. Fortunately, symptoms of high negative middle-ear pressure and eustachian tube obstruction are frequently absent, so no treatment may be necessary. Even myringotomy and tympanostomy tube insertion may not be beneficial because the tympanic membrane is no longer actively being retracted by high negative middle-ear pressure. In addition, at this stage, adhesive otitis media may also be present, and portions of the tympanic membrane may be adherent to the middle ear. The posterior or epitympanic (attic) portions of the middle ear may become separated from the anterior portion by adhesions, and, subsequently, ventilation from the eustachian tube or a tympanostomy tube does not aerate the affected area. In such cases, there are two management options: tympanoplasty or periodic observation (once or twice a year).

Tympanoplasty

In selected cases in which severe atelectasis is present, a tympanoplastic procedure may be indicated. The surgical procedures recommended
The most compelling indication for such a procedure is the presence of a deep retraction pocket in the posterosuperior portion of the pars tensa that is unresponsive to nonsurgical and other surgical methods of management previously described for this defect. For example, if a tympanostomy tube had been inserted previously but the retraction pocket did not return to the neutral position after several months of equalization of the intratympanic pressure, tympanoplasty should be considered because adhesive otitis media is most likely binding the drum to the ossicles and surrounding structures within the middle ear. Even though the natural history of such deep retraction pockets has not been formally studied, the risk of erosion necrosis of the incus, formation of a cholesteatoma, or both appears to be high. It is frequently difficult to determine whether there is only a retraction pocket present or if a cholesteatoma has already developed; therefore, a thorough examination of the entire external canal and tympanic membrane should be performed with the otomicroscope. An examination under general anesthesia will be required for all infants and children in whom the examination is unsatisfactory without general anesthesia. At the time of the examination under anesthesia, a thorough examination of the retraction pocket, employing a curved, blunt probe, should be performed to determine the extent of the pocket. In addition, the continuity of the incus and stapes should be assessed because erosion of the long process of the incus may require surgical correction. When nitrous oxide is employed as one of the anesthetic agents, the retraction pocket can frequently be seen to balloon laterally, as visualized through the otomicroscope. When this occurs, insertion of the tympanostomy tube will usually be sufficient to prevent recurrence of the retraction pocket. Reinsertion of the tube may be needed, however, if the retraction pocket recurs after spontaneous extubation.

Many techniques are advocated for repair of a severely atelectatic tympanic membrane, many of which have been shown to be successful. The surgeon should be cautioned, however, that even though the graft “takes,” the child will most likely have persistent eustachian tube dysfunction with sustained fluctuating or negative intratympanic pressure after the procedure, which could result in recurrence of the retraction pocket months or years later. Therefore, a tympanostomy tube should be considered at the time of the tympanoplasty surgery and reinserted if atelectasis begins to recur after the tympanostomy tube is spontaneously extruded. Some surgeons prefer use of tragal or conchal cartilage attached to its perichondrium to cover the area of the retraction pocket so that recurrence of an attic or posterosuperior retraction pocket can be prevented.

All children who require tympanoplasty for severe atelectasis must be observed at relatively frequent intervals for the first year after the procedure and at appropriate intervals for several succeeding years. Recurrence of the atelectasis should always be anticipated.

**ADHESIVE OTITIS MEDIA**

Adhesive otitis media is a sequela of recurrent and chronic inflammation of the middle ear and mastoid and is a result of the healing process. The mucous membrane is thickened by proliferation of fibrous tissue, which frequently impairs (binds) the movement of the ossicles, resulting in a conductive hearing loss.

Adhesive otitis media can be classified according to three stages related to the severity, extent, and presence or absence of functional impairment:

**Stage 1:** adhesive otitis media within the middle ear, mastoid, or both, with no functional deficit secondary to the adhesive changes (ie, hearing loss). The middle ear remains aerated.

**Stage 2:** adhesive otitis media within the middle ear (with or without mastoid involvement) with mild hearing loss secondary to an adhesive pathologic process, such as involvement of the ossicular chain (fixation, disconti-
nuity, or both; see “Ossicular Discontinuity
and Fixation”), limitation of tympanic mem-
brane compliance, or both. The middle ear
remains aerated.

Stage 3: similar to Stage 2 but with maximal
conductive hearing loss secondary to an
ossicular pathologic process. No middle-ear
space is present. Both conditions are due to
extensive adhesive otitis media.

Pathology and Pathogenesis

Schuknecht has described the pathologic process
as a proliferation of fibrous tissue within the
middle ear and mastoid and has termed the
condition fibrous sclerosis.217 When cystic spaces
are present, it is called fibrocystic sclerosis; when
there is new bone growth in the mastoid, he has
classified it as fibro-osseous sclerosis.

The etiology and pathogenesis have not been
extensively studied in the past. Caye-Thomasen
and colleagues, in experiments in animals,
proposed that adhesive otitis media is a patho-
logic phenomenon caused by infection in six
pathogenic stages394:

1. Localized epithelial rupture
2. Prolapse of subepithelial tissue
3. Epithelialization of the prolapse, resulting in
   a polypoid, fold-like prominence
4. Growth and elongation of the prominence
5. Fusion of the end tip of the prominence with
   another part of the mucosa
6. Formation of an adhesion

Management

No data are currently available on the prevalence
of adhesive otitis media in children, but the
condition is commonly encountered in the pediatric
age group when recurrent or chronic middle-ear
disease has been a long-standing problem. Unfortu-
nately, we likewise have no data from which to
establish the probability with which a child who
has recurrent or chronic disease of the middle ear
might develop adhesive otitis media. However, the
possible occurrence of adhesive changes when
recurrent or chronic inflammation—including
atelectasis of the middle ear—is present in the
middle ear and mastoid must be seriously con-
sidered in selecting the most appropriate medical or
surgical treatment of children who have recurrent
acute and chronic middle-ear disease.

In addition to fixation of the ossicles, adhe-
sive otitis media may result in ossicular disconti-
uity and conductive hearing loss owing to
rarefying osteitis, especially of the long process
of the incus. When there is a retraction pocket
(severe partial atelectasis) in the posterosuperior
portion of the pars tensa of the tympanic
membrane, adhesive changes may bind the ear-
drum to the incus, stapes, and other surrounding
middle-ear structures and cause resorption of the
ossicles. Once adhesive changes bind the tympanic
membrane in this area, a cholesteatoma may also
develop. Timely ventilation of the middle ear and
mastoid before the adhesive changes may return
the tympanic membrane to the normal position,
thus preventing ossicular damage. If medical
treatment fails, a myringotomy should be per-
formed, and a tympanostomy tube should be
inserted in an attempt to reverse the potentially
progressive pathologic condition (see Figure 28).
If the tympanic membrane is still attached to the
ossicles after tympanostomy tube insertion,
adhesive otitis media is present. In children,
tympanoplasty should be considered to prevent
further structural damage; the process may pro-
gress because of persistent eustachian tube
obstruction.

When ossicular fixation occurs, ossicu-
loplasty may be performed to restore function,
but, unfortunately, this procedure is not always
successful. When the middle ear and mastoid are
bound by adhesive otitis media, the results of
ossiculoplasty are frequently not permanent
because the adhesive process recurs. However,
surgery should be considered.

The best method of management for adhesive
otitis media is prevention. This involves treating
its precursors: recurrent AOM, chronic otitis
media with effusion (see Chapter 8), and atelec-
tasis (see earlier).
CHOLESTEATOMA

Cholesteatoma is keratinizing stratified squamous epithelium and an accumulation of desquamating epithelium of keratin within the middle ear or other pneumatized portions of the temporal bone. This disease has also been called a keratoma, but cholesteatoma is the more commonly used term.\(^{395}\) Cholesteatoma is histologically similar to epithelium of the skin of the external auditory canal.

Classification

Aural cholesteatoma can be classified as congenital and acquired. Cholesteatomas can be acquired as a sequela of middle-ear disease or from implantation, which can be due to either trauma or related surgery of the middle ear (including the tympanic membrane), external auditory canal, or mastoid (ie, iatrogenic).

A congenital cholesteatoma has been defined as a congenital rest of epithelial tissue and appears as a white, cyst-like structure within the middle ear (intratympanic) or temporal bone. The tympanic membrane is intact, and it is apparently not a sequela of otitis media or eustachian tube dysfunction.\(^ {396,397}\) Rosenfeld and colleagues reviewed the records of 232 children in Pittsburgh who had cholesteatoma and identified 43 (18%) patients in whom the cholesteatoma was thought to be congenital in origin.\(^ {398}\) Possibly more would have been diagnosed as congenital, but in 46% of the children, the distinction between acquired and congenital origin could not be made owing to the advanced stage of the disease.

Acquired cholesteatoma may be secondary to implantation, or it may be a sequela of otitis media or a retraction pocket. Implantation cholesteatoma may develop from epithelium that has migrated through a traumatic perforation of the tympanic membrane (or the site of a tympanostomy tube) or that has inadvertently been overlooked in the middle ear or mastoid during surgery of the ear (iatrogenic).\(^ {399}\) Iatrogenic cholesteatoma can develop in the ear canal after middle-ear and mastoid surgery at the site of the incisions made in the canal and can occur after any of the various incisions.

The most common cholesteatoma is the acquired type, which is a sequela of middle-ear disease. In a study of 1,024 patients (adults and children), a cholesteatoma was found in 42% in the attic, in 31% in the posterosuperior quadrant, in 18% when there was a “total” perforation, in 6% when there was a “central” perforation, and in 3% when there was no perforation.\(^ {400}\) However, it is possible that the patients in whom the cholesteatoma was associated with a total perforation originally had involvement of the posterosuperior portion of the pars tensa. In children, the most common defect in the tympanic membrane begins developing in the posterosuperior quadrant of the pars tensa or, somewhat less commonly, in the pars flaccida. The term marginal perforation has been used to describe the defect in the posterosuperior quadrant, and the defect in the pars flaccida has been called an attic perforation, but in reality, these are most frequently not perforations but are either retraction pockets or cholesteatomas that appear otoscopically to be perforations (Figure 29). No continuity between the defect and the middle ear occurs until later in the disease process.

Staging of Acquired Cholesteatoma

It is appropriate to stage cholesteatomas for management, reporting, and research. Staging can be extremely helpful in determining the outcomes of surgery, such as residual and recurrence rates, from center to center around the world.\(^ {401,402}\) In staging of cholesteatoma, the presence or absence of infection should be noted and, if present, the duration of the otitis media as follows\(^ {10,320}\):

Cholesteatoma without infection is a cholesteatoma that is not associated with infection, either within the cholesteatoma itself or in any other portion of the middle-ear cleft.
Cholesteatoma with infection is a cholesteatoma that is associated with infection, which can be either acute infection (with or without otorrhea) or chronic. The most common infection associated with cholesteatoma is chronic suppurative otitis media.

Cholesteatoma can be further classified by its site and extent:

Stage 1: Cholesteatoma is confined to the middle ear (hypoepitympanum and mesoepitympanum), without erosion of the ossicular chain

Stage 2: Same as stage 1 but with erosion of one or more ossicles

Stage 3: Middle ear and mastoid gas cell system are involved without erosion of the ossicles

Stage 4: Same as stage 3 but with erosion of one or more ossicles

Stage 5: Extensive cholesteatoma of the middle ear, mastoid, and other portions of the temporal bone, the extent of which is not totally accessible to surgical removal (e.g., medial to labyrinth), with one or more ossicles involved. Fistula of the labyrinth may or may not be present.

Stage 6: Same as Stage 5, but cholesteatoma extends beyond the temporal bone

Epidemiology, Complications, and Sequelae

Cholesteatoma has persisted as a sequela of middle-ear disease in adults and children despite the advent of the widespread use of antimicrobial agents and tympanostomy tubes during the past half-century. However, the mortality associated with cholesteatoma has fallen dramatically during this period in the developed countries of the world.

In an epidemiologic study from Michigan, Ritter compared 152 adults and children who had cholesteatoma identified during the period from 1965 to 1970 with 303 cases from a similar study in Massachusetts that were identified during a period before the use of antimicrobial agents (1925–1936). He found in both series that about 45% of cases of cholesteatoma were operated on before the patient was 20 years old; that in approximately 65% of the patients, the aural discharge had begun by the age of 11 years; and that the distribution of sites on the tympanic membrane where the defect was located was about the same. He concluded that antimicrobial agents had not altered the incidence and natural
history of cholesteatoma during the 40 years between the two studies. However, Rigner and colleagues concluded that in the Gothenburg area of Sweden, the incidence declined during a 10-year period between 1977 and 1986; they attributed the fall in cases to better accessibility to specialists and improved methods to diagnose middle-ear disease earlier in that community compared with other parts of their country. The epidemiology of cholesteatoma in children has been reported by several investigators during the past several decades. Harker and Koontz, in a study of the general population in Iowa, reported the overall incidence of cholesteatoma to be 6 per 100,000; in children up to 9 years of age, the incidence was 4.7 per 100,000, whereas in children 10 to 19 years old, the incidence of 9.2 per 100,000 was the highest for all age groups. Karma and colleagues in Finland and Tos in Denmark estimated the annual incidence of cholesteatoma to be 4.5 and 15 per 100,000 children, respectively. In a more recent report from Finland, the mean annual incidence of cholesteatoma during the period from 1982 to 1991 in one region for all age groups was 9.2 per 100,000 inhabitants (range 3.7–13.9).

Cholesteatoma is a common sequela in children with a cleft palate. Severeid reviewed the records of 160 children and young adults with a cleft palate (70% were aged 10 to 16 years), all of whom had had a history of ear disease, and found the incidence of cholesteatoma to be 7.1%. The posterosuperior portion of the pars tensa was the most common site. A later review from the same institution reported that almost 10% of children with a cleft palate developed cholesteatoma. During a recent 10-year period in Finland, of 500 patients who had cholesteatoma, 8% had a cleft palate.

In contrast to this high incidence of cholesteatoma in the cleft palate population is the rare occurrence of cholesteatoma in Alaskan (Eskimo) natives, American Indians, and Australian Aboriginal children, in whom other middle-ear disease is common. This remarkable difference in the incidence of cholesteatoma in children with a cleft palate and in certain racial groups, both of which have a high prevalence and incidence of otitis media, is most probably related to differences in the pathogenesis and natural history of the respective middle-ear disease processes (see Chapter 3).

Before the widespread use of antimicrobial agents and modern otologic surgery, complications of cholesteatoma were common. For many children, the result was death when infection involved the intracranial cavity. Now, serious complications of cholesteatoma in children are uncommon in the developed countries of the world. In a study of 181 children who had cholesteatoma, 8 (4.4%) developed a labyrinthine fistula and 1 suffered facial paralysis, but none had intracranial complications. However, in the same study, which also included 843 adults, the incidence of both intratemporal and intracranial complications increased the longer the cholesteatoma was present. Because most of the adults could date the onset of their disease to childhood and because the best way of preventing serious complications of cholesteatoma is by diagnosis and surgery, physicians dealing with ear problems in children should treat suspected cholesteatoma early and aggressively. Nevertheless, intratemporal complications and sequelae are still a problem in even the highly industrialized nations of the world.

The rate of cholesteatoma, and especially its complications, in developing countries is relatively high compared with that in industrialized nations. Prescott reported his experience in treating 81 children who had cholesteatoma from 1988 to 1996; 24 (30%) presented with mastoiditis, and 7 (9%) had an intracranial complication. In that study, 56 (70%) of the cholesteatomas were from a retraction pocket, and 21 (25%) were from a central perforation or total atelectasis; 3 were congenital (see Chapter 10).

Sequelae, such as ossicular involvement, which not only contribute to the associated conductive hearing loss but are related to the residual and recurrence rates, are common. Cholesteatoma with chronic suppurative otitis
media has also been shown to be associated with sensorineural hearing loss. 414

**Pathogenesis**

The pathogenesis of acquired middle-ear cholesteatoma is a subject of continuing controversy, but most likely there is more than one pathogenetic mechanism. Of the many hypotheses proposed to explain the pathogenesis of cholesteatoma, the most popular are (1) metaplasia of the middle ear and attic owing to infection; 415, 416; (2) invasive hyperplasia of the basal layers of the meatal skin adjoining the upper margin of the tympanic membrane; 417–421; (3) invasive hyperkeratosis of the deep external auditory canal; 422; and (4) retraction or collapse of the tympanic membrane with invagination secondary to eustachian tube dysfunction. 423–425 In addition, there are those who consider the condition not to be acquired but to be an embryonic epidermal rest occurring in the attic. 426–428 Thus, it is apparent that there are probably several ways in which cholesteatoma can develop. 429

In a study from Pittsburgh, varying degrees of functional rather than mechanical (anatomic) obstruction of the eustachian tube were found in 13 children and adults who had a retraction pocket or an acquired cholesteatoma. 376 Subsequently, from the same center, the findings in 12 children with acquired cholesteatoma, all of whom had functional obstruction of the eustachian tube, were also reported by the same group. 430 Children were specifically studied because the development of an acquired cholesteatoma with its attendant irreversible changes was thought to occur early in life and because the function of the eustachian tube might improve with growth and development. In these children, the function of the eustachian tube was assessed by the modified inflation-deflation technique (after Ingelstedt and colleagues). 431 Bluestone and colleagues and Chan and colleagues performed two other studies to further clarify the cause of this functional obstruction by employing a new test of eustachian tube function, the forced-response test, 433 and to evaluate a larger group of children who had either a cholesteatoma or a retraction pocket. 369, 372 In addition, children with an apparent congenital cholesteatoma were also studied, and the results obtained in both groups were then compared with the results of testing children who had traumatic perforation of the tympanic membrane but who were otherwise considered otologically normal. Another goal of these studies was to determine whether there were any differences in eustachian tube function among ears with a posterosuperior or pars flaccida retraction pocket or cholesteatoma, ears with a central perforation and a cholesteatoma, and ears with congenital cholesteatoma.

In a study from Sweden, Lindeman and Holmquist showed similar findings in adults with acquired cholesteatoma. 433 Compared with adults with traumatic perforations (ie, control subjects), 20 adults with cholesteatoma had poor eustachian tube test results and smaller mastoid air cell areas as measured on radiographs. It appears from these studies that the basic problem in children with acquired cholesteatoma is a failure of the opening mechanism of the eustachian tube. This results in a functional obstruction of the tube during swallowing rather than normal dilation of the tube. (This type of functional obstruction of the eustachian tube was present in subjects with a retraction pocket or cholesteatoma irrespective of the site.) In the absence of obstruction of the middle-ear end of the tube by cholesteatoma, anatomic (mechanical) obstruction is not involved in the pathogenesis. Abnormal functioning of the tube then results in impaired ventilation of the middle-ear–mastoid air cell system, which, in turn, results in fluctuating or sustained high negative middle-ear pressure. Periodic, rather than regular, ventilation could result in wide variations in middle-ear pressure that would produce greater than normal excursions of the tympanic membrane. The membrane would then lose elasticity and would become flaccid and, eventually, atelectatic. The most flaccid parts of the tympanic membrane are the posterosuperior and pars flaccida areas. 374 When the atelectasis becomes severe and localized in these sites, a
retraction pocket forms. Inflammation between the medial portion of the retracted or collapsed tympanic membrane could then result in adhesive changes and could fix the pocket to the ossicles, surrounding structures, or both. The next stage in this series of events would be discontinuity of the ossicles, cholesteatoma formation, or both. Figure 30 shows the progression from the stage of a retraction pocket with atelectasis to adhesive otitis media and, finally, to cholesteatoma in the posterosuperior quadrant of the pars tensa and pars flaccida.

Even with the aid of the otomicroscope, it is frequently difficult to make the distinction between a deep retraction pocket and a cholesteatoma in either the posterosuperior quadrant of the pars tensa or the pars flaccida (Figure 31). The transition between the two conditions usually follows a progressive change from a retraction pocket to cholesteatoma; however, the factors involved in this transition are currently obscure, although infection within the retraction pocket–sac appears to be important.

A cholesteatoma can also develop at the site of a central perforation. Most likely this is due to migration of epithelium from the tympanic membrane through the perforation and into the middle ear. However, it must be stressed that in children, this type of acquired cholesteatoma is less common than the posterosuperior or attic type.

Congenital cholesteatoma is classically differentiated from acquired when the tympanic membrane is intact and the child does not have a history of otorrhea or has had middle-ear surgery.\(^{434,435}\) But it is often difficult to distinguish between acquired and congenital cholesteatoma, especially when the cholesteatoma is extensive and the tympanic membrane is involved.\(^{398,436}\) There is some uncertainty as to the origin of congenital cholesteatomas. Some authorities consider them to be truly congenital in origin because there is epidermoid formation in the fetus middle ear, which then presumably disappears prior to birth.\(^{437}\) But children who have an intratympanic cholesteatoma may have had otitis media. On the one hand, it could be argued that intratympanic cholesteatoma is the result of metaplasia secondary to middle-ear inflammation and that it is not congenital.\(^{438,439}\) Also, a cholesteatoma behind an intact tympanic membrane could have been due to a retraction pocket that subsequently resolved, leaving the tympanic membrane without evidence of the

Figure 30. Chain of events in the pathogenesis of acquired aural cholesteatoma in the posterosuperior portion of the pars tensa of the tympanic membrane.
On the other hand, otitis media, when present, may be unrelated to a congenital rest. The fact that children who have congenital cholesteatomas tend to be younger than those who present with a retraction pocket or acquired cholesteatoma would support the congenital origin of a cholesteatoma medial to an intact tympanic membrane. In any event, most acquired cholesteatomas not due to implantation are secondary to otitis media, a retraction pocket, or both, and some children who have an apparently congenital cholesteatoma may have developed the disease in the same way. Levenson and colleagues believe that the pathogenesis of congenital cholesteatoma is an epithelial rest, which is stimulated to grow by otitis media. Some surgeons believe that since congenital cholesteatomas commonly occur in two different sites (i.e., anterosuperior and posteroinferior mesotympanum), they have a different pathogenesis.

Studies reveal that inflammatory factors, such as cytokines, adhesion molecules, tumor necrosis factor \( \alpha \), and the nuclear phosphoprotein \( TP53 \) tumor suppressor gene, may play a role in invasion, migration, and proliferation of congenital and acquired cholesteatoma. Also, erbB-2 protein, nitric oxide, bone morphogenetic protein 2 messenger ribonucleic acid, telomerase activity, angiogenesis and angiogenic growth factors, and changes in noncollagenous protein content in the mastoid bone may play roles in the pathogenesis.

A common sequela of middle-ear disease in patients with a cleft palate is cholesteatoma. It has been shown that all infants with an unrepaired cleft palate have otitis media with effusion and that they have functional obstruction of the eustachian tube because of impairment of the tubal opening mechanism. Studies of infants, children, and adolescents with a cleft palate demonstrate constriction of the eustachian tube during the forced-response test. Cholesteatoma is a common sequela of middle-ear disease in patients with a cleft palate. Therefore, the child with a cleft palate represents an in vivo model of the type of functional eustachian tube obstruction that can result in an acquired cholesteatoma. Because this type of dysfunction also occurs commonly in whites who have otitis media or atelectasis but who do not have a cleft palate, they are also at risk of development of cholesteatoma.

In contrast to the frequency of cholesteatoma in the cleft palate population, cholesteatoma is uncommon in American Indian populations. Jaffe reported that attic perforations are rarely found in Navajo children; in more than 200
tympanoplasties performed to repair central perforations, no cholesteatoma was found.\textsuperscript{462} Wiet, in a study of 600 White Mountain Apache Indians, also reported a low incidence of cholesteatoma; the few cases he found were mostly of the attic type.\textsuperscript{463} In a subsequent study by Beery and colleagues, otoscopic examination of 25 Apache Indians revealed no cholesteatomas.\textsuperscript{117} The eustachian tube function was tested in these Indians by the inflation-deflation and forced-response tests, which revealed a eustachian tube that had low resistance to airflow (was semipatulous) but active muscle function. This type of tube probably precludes the development of high negative middle-ear pressure, a retraction pocket, or cholesteatoma. The Apache Indian appears to have a eustachian tube that allows easier passage of gas and liquid than does that of the white subject with or without a cleft palate. The middle ear of the Apache individual is easily ventilated and, consequently, is not protected from unwanted secretions from the nasopharynx. It appears that the structure of the eustachian tube of the Apache Indian of the White Mountain Reservation is conducive to the development of reflux otitis media, perforation, and discharge.

Some American Indian tribes appear to be in vivo models of the semipatulous eustachian tube that actively dilates during swallowing. Cholesteatoma formation is rarely seen in such ears because the middle ear is aerated either by the eustachian tube, a central perforation, or both. By studying these in vivo models, we can gain a clearer perspective of the whole spectrum of eustachian tube dysfunction (see Chapter 3).

**Microbiology**

Cholesteatomas may or may not be associated with infection, and when infection is present, it can be either acute or chronic.\textsuperscript{122} An acute infection can also be superimposed on a pre-existing chronic infection. A cholesteatoma is frequently present in the middle ear (and mastoid) without any signs of acute or chronic infection; but when infection is present, it can involve the entire middle-ear cleft (ie, eustachian tube, middle ear, and mastoid), or only the cholesteatoma is infected and the rest of the middle-ear cleft is apparently free of infection.

When infection is present, the organisms cultured from the discharge are similar to those identified from ears with chronic suppurative otitis media; \textit{P. aeruginosa} and \textit{Proteus} are the most commonly identified aerobic bacteria, and \textit{Bacteroides} and \textit{Peptococcus-Peptostreptococcus} are the most commonly seen anaerobic organisms. Multiple bacteria were cultured from the discharges of more than half of 30 patients with cholesteatomas studied by Harker and Koontz (Table 14).\textsuperscript{405} Karma and colleagues reported that when they cultured 18 infected cholesteatomas, they found both aerobic and anaerobic bacteria in half of the cultures.\textsuperscript{464} From the results of these studies, it seems that the most appropriate ototopical medication\textsuperscript{126} and systemic antimicrobial therapy for patients who have an infected cholesteatoma are agents effective against gram-negative organisms and anaerobic bacteria; however, the results of culture of the discharge will aid in selecting the proper antimicrobial therapy. A recent study found microbial biofilms in human and experi-

<table>
<thead>
<tr>
<th>Organism</th>
<th>Number of Cases</th>
</tr>
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<tbody>
<tr>
<td><strong>Aerobes</strong></td>
<td></td>
</tr>
<tr>
<td>\textit{Pseudomonas aeruginosa}</td>
<td>11</td>
</tr>
<tr>
<td>\textit{Pseudomonas fluorescens}</td>
<td>2</td>
</tr>
<tr>
<td>\textit{Proteus} species</td>
<td>4</td>
</tr>
<tr>
<td>\textit{Escherichia coli}</td>
<td>4</td>
</tr>
<tr>
<td>\textit{Klebsiella, Enterobacter, and Serratia species}</td>
<td>4</td>
</tr>
<tr>
<td>\textit{Streptococcus} species</td>
<td>8</td>
</tr>
<tr>
<td>Alcaligenes and Achromobacter species</td>
<td>3</td>
</tr>
<tr>
<td>\textit{Staphylococcus} aureus</td>
<td>1</td>
</tr>
<tr>
<td>\textit{Staphylococcus epidermidis}</td>
<td>2</td>
</tr>
<tr>
<td><strong>Anaerobes</strong></td>
<td></td>
</tr>
<tr>
<td>\textit{Bacteroides} species</td>
<td>13</td>
</tr>
<tr>
<td>\textit{Peptococcus} and \textit{Peptostreptococcus} species</td>
<td>11</td>
</tr>
<tr>
<td>\textit{Propionibacterium} acnes</td>
<td>8</td>
</tr>
<tr>
<td>\textit{Fusobacterium} species</td>
<td>4</td>
</tr>
<tr>
<td>\textit{Bifidobacterium} species</td>
<td>3</td>
</tr>
<tr>
<td>\textit{Clostridium} species</td>
<td>3</td>
</tr>
<tr>
<td>\textit{Eubacterium} species</td>
<td>2</td>
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</tbody>
</table>

Adapted from Harker LA, and Koontz FP.\textsuperscript{405
mental cholesteatoma, which could explain the persistent infection in many patients who have cholesteatoma despite apparently appropriate and adequate medical treatment. These considerations may be lifesaving when an intratemporal or intracranial complication of cholesteatoma is present. In addition, preoperative and postoperative antimicrobial therapy for patients with profuse otorrhea may also be necessary to prevent a postoperative infection from developing.

Pathology

Cholesteatoma is characterized by keratinizing stratified squamous epithelium, with accumulation of desquamating epithelium or keratin within the middle-ear cleft or other pneumatized portions of the temporal bone. A cyst-like structure is usually produced by the keratinizing squamous epithelium. Laminated keratin from its inverted surface accumulates within the cavity, which may also contain necrotic tissue and purulent material. If the pocket is dry, the rate of exfoliation may be slow. A cholesteatoma may or may not be infected or associated with chronic suppurative otitis media. Sheehy and colleagues reported that of 1,024 children and adults with cholesteatoma, 26% had no history of aural discharge in the past; in 53%, it had been intermittent, and in only 21% of the patients was discharge reported as being continuous. When these patients had surgery, almost half had no evidence of discharge.

Cholesteatoma usually causes bone resorption, which is thought to be secondary to pressure erosion as the mass enlarges or possibly due to the activity of collagenase. Erosion can occur anywhere in the temporal bone, although the ossicles are commonly involved. Ossicular erosion can result in discontinuity (usually erosion of the long process of the incus) and a conductive hearing loss or fistulization of the labyrinth. (The lateral semicircular canal is the most common site of erosion.) Alternatively, the epidermis may invade the aerated space of the temporal bone and form an incomplete surface lining into which the desquamated keratin debris overflows. This process may give the impression that the mucous membrane is converted by metaplasia to keratinizing squamous epithelium. However, there apparently is no histopathologic support for this hypothesis.

In children, cholesteroloma is considered to be more aggressive (invasive) than the disease that occurs in adults for two reasons: (1) extensive disease is found at the time of surgery more frequently in children than in adults and (2) higher rates of residual (persistent) and recurrent cholesteroloma after surgery have been found in children compared with the rates in adults. Palva and colleagues compared 65 children with cholesterolomas with 65 adults with the same disease and found that whereas 22% of the children had extensive disease that filled the middle ear and mastoid, only 6% of adults had such extensive disease. In children, in contrast to adults, cholesteroloma frequently extends into the cell tracts of the temporal bone because pneumatization is usually more extensive in children than in adults. However, despite the finding that cholesterolomas in children tend to be more extensive than those occurring in adults, childhood cholesteroloma may still be confined to the mesotympanum or epitympanum. Schuring and colleagues compared the extent of and outcomes after surgery for cholesteroloma in 228 adults and teenagers with those in 38 children and concluded that children had more recurrent disease, greater ossicular necrosis, and poorer hearing postoperatively.

Diagnosis

Because there may be a lack of signs and symptoms of ear disease, cholesteroloma may go undetected for many years in children and adults. In children, this is an even greater problem because they are frequently unaware of slowly developing ear problems. Most adults have a history of hearing loss, which is usually progres-
sive and associated with recurrent ear discharge. However, children rarely complain of hearing loss, especially if the disease is unilateral. There is frequently no discharge, and otalgia may be absent in most children and adults. In addition, children are usually unaware of the more subtle symptoms associated with the disease, such as fullness in the ear, tinnitus, mild vertigo, and the foul smell of the discharge when it is present. Fever is not a sign of cholesteatoma; when it accompanies this disease, and especially when otalgia is also present, a search for an intratemporal or intracranial complication must be made. Other signs and symptoms, such as facial paralysis, severe vertigo, vomiting, and headache, should also alert the physician to the presence of a suppurative complication. In children, the attic type of cholesteatoma appears to be less symptomatic than a cholesteatoma in the posterosuperior quadrant; in the posterosuperior quadrant, cholesteatoma is frequently preceded by symptomatic recurrent or chronic otitis media with effusion and an early onset of ossicular discontinuity with a significant hearing loss. However, in both types, the preceding atelectasis and retraction pocket may not be associated with significant symptoms in children. The intratympanic congenital cholesteatoma, which may be secondary to otitis media, is even more obscure because hearing loss may be a late sequela and discharge is not present.

The diagnosis of cholesteatoma is most effectively made with an otoscope or, more accurately, with the otomicroscope. However, otoscopic findings may be equivocal, otoscopy may be difficult to adequately perform if infection is present in the ear canal, or the child may be uncooperative. If otoscopy is not successful and a cholesteatoma is suspected, the child may have to be examined with use of a general anesthetic. A CT scan may be diagnostic. Recurrent or chronic otorrhea that is unresponsive to medical management can be a sign that a cholesteatoma is the underlying pathologic process.

In a defect in the posterosuperior portion of the pars tensa or the attic or through a large perforation, otoscopy will usually reveal the presence of white, shiny, greasy flakes of debris, which may or may not be associated with a foul-smelling discharge. A polyp may be seen coming through the defect, which, like a crust, can prevent adequate visualization of the tympanic membrane. A crust overlying the area of the posterosuperior quadrant or the pars flaccida must be removed because a retraction pocket or cholesteatoma may be present. The size of the defect in the tympanic membrane may not indicate the extent of the cholesteatoma because a small defect, especially in the attic, may be associated with extensive cholesteatoma. On the other hand, the cholesteatoma may be confined only to the attic or middle ear despite the presence of a large defect. If an adequate examination of the child’s ear is not possible with the child awake, an examination with the patient under anesthesia is indicated. Every child who must be given a general anesthetic for myringotomy (with or without a tympanostomy tube insertion) should have the entire tympanic membrane examined to identify a possible cholesteatoma or its precursor, a retraction pocket. In addition, an intratympanic cholesteatoma may be visualized through the tympanic membrane or through the incision after a myringotomy.

It is frequently difficult to determine whether the defect is a retraction pocket or a “dry” cholesteatoma. However, even though this distinction cannot be made, the management of the defect is usually the same (see “Atelectasis of the Middle Ear”).

Unfortunately, there is no tympanometric pattern for diagnosis of a cholesteatoma. An abnormal tympanogram should alert the clinician to the presence of middle-ear disease, but the tympanogram may be normal even when a cholesteatoma is present. Impedance testing may reveal a perforation of the tympanic membrane, but this occurs less commonly in children than in adults. Likewise, audiometric testing may reveal a conductive hearing impairment or possibly a mixed conductive and sensorineural deficit, but a cholesteatoma may be present without a loss of
hearing. A sensorineural hearing loss is presumably due to serous labyrinthitis or possibly a labyrinthine fistula. CT scans of the temporal bone are helpful in diagnosis and management when a cholesteatoma is suspected. The CT scans should be studied carefully to identify the extent of the cholesteatoma, possible ossicular involvement, and any complication that might be present, such as a labyrinthine fistula. The CT scans should be restudied preoperatively in planning the surgical procedure; decisions concerning the surgical approach and the extent of surgery can be aided by CT scans.

When aural discharge associated with a cholesteatoma is profuse, microbiologic assessment of the discharge is indicated so that the infection can be controlled by administering the most appropriate antimicrobial agent preoperatively and, when indicated, intraoperatively and postoperatively.

**Management**

Medical management of cholesteatoma is indicated only when infection is coexistent and should be directed at the organisms listed above (see “Microbiology”). Otherwise, the primary method of management is surgical. Aural toilet, obtaining a Gram stain, and culture and susceptibility studies are key to culture-directed antibiotic treatment. Ototopical agents that are nontoxic to the inner ear are recommended. As discussed above for chronic suppurative otitis media, there are ototopical agents that are potentially ototoxic and not indicated today with the availability of the two new fluoroquinolone options, ofloxacin otic drops and ciprofloxacin-dexamethasone. Systemic antibiotics may also be beneficial but should be culture directed. *Pseudomonas* is the most common; thus, orally administered quinolones are restricted in children, but with appropriate parental informed consent, oral administration of a quinolone, such as ciprofloxin, is more feasible than intravenous potentially toxic agents, such as the aminoglycosides.

Surgical management of cholesteatoma and conditions that may be causally related to this disease should be based on an understanding of its pathogenesis. A deep retraction pocket in the posterosuperior or pars flaccida area of the tympanic membrane, if persistent, should be managed promptly by insertion of a tympanostomy tube in an effort to return the tympanic membrane to the neutral position and to prevent adhesions from forming between the tympanic membrane and the middle-ear structures (see Figure 28). If the pocket is not treated, a cholesteatoma may develop. In children, the retraction pocket may be seen (through the otomicroscope) to distend during inhalation anesthesia; this is a promising sign that the tympanic membrane will return to the normal position after the insertion of a tympanostomy tube. On the other hand, if the retraction pocket does not distend during anesthesia, the surgeon should carefully examine the depth and extent of the pocket, probing gently with a blunt right-angled hook. Mirrors may also help visualize the extent of the pocket. We prefer a Hopkins rod-lens telescope to determine the exact borders of the pocket. Also, we have found the telescope to be invaluable in removing cholesteatoma at the time of middle-ear (and mastoid) surgery. Others have also now advocated its use in reducing postoperative residual disease.

A retraction pocket can extend into any area of the middle ear, but it is found most frequently extending into the epitympanum, facial recess, and sinus tympani. If the retraction pocket persists after the middle ear has been ventilated by a tympanostomy tube that has been in place for several weeks, a tympanoplasty procedure to prevent ossicular discontinuity, the development of a cholesteatoma, or both should be recommended. Heermann and colleagues advocate the use of cartilage to support the tympanic membrane graft to prevent recurrence of the retraction pocket. Other surgeons have reported good results using cartilage to prevent postoperative retraction. Simple excision of the retraction pocket has also been advocated, but this technique is not as effective as...
reconstruction of the defect and reinforcing the tympanic membrane with cartilage. In children, a tympanostomy tube should be inserted into the tympanic membrane remnant because eustachian tube function will most probably remain poor postoperatively.

**Tympanotomy** is a surgical procedure that opens the middle-ear space. In an **exploratory tympanotomy**, a tympanomeatal flap is elevated so that the middle ear and its structures can be viewed directly. An exploratory tympanotomy is indicated when it is suspected that there is an abnormality, such as intratympanic cholesteatoma or ossicular chain abnormality, or it is a planned second-stage procedure after tympanoplasty with or without mastoidectomy has been performed to manage cholesteatoma.

**Myringoplasty** is the surgical repair of a defect in the tympanic membrane with no attempt made to explore the middle ear. A perforation (or retraction pocket) is commonly repaired by use of an autogenous connective tissue graft (temporalis fascia or compressed adipose tissue from the earlobe) as a lattice onto which epithelial cells can migrate from the edges of the existing perforation. The procedure is employed to manage a simple uncomplicated tympanic membrane perforation without cholesteatoma.

**Tympanoplasty** is the surgical reconstruction of the tympanic membrane–ossicle transformer mechanism. If a perforation is present, it is repaired with a connective tissue graft, but unlike in a myringoplasty, the middle ear is explored. Ossicles can be repositioned (ossiculoplasty) to restore ossicular chain continuity. Traditionally, tympanoplasty operations are characterized according to the degree to which the reconstructed ossicular chain approximates the anatomic juxtaposition of ossicles in the normal middle ear (see “Perforation of the Tympanic Membrane” and “Ossicular Discontinuity and Fixation”).

**Mastoidectomy** involves the surgical exposure and removal of mastoid air cells. There are several types of mastoidectomy (Figure 32). In a **complete simple (cortical) mastoidectomy** (see Figure 32A), the mastoid air cell system is exenterated, including the epitympanum, but the canal wall is left intact. The operation is performed when acute or chronic mastoid osteitis is present and is frequently part of the surgical procedure advocated by some surgeons for cholesteatoma. A **posterior tympanotomy** or **facial recess tympanotomy** (see Figure 32B) involves exenteration of mastoid air cells followed by formation of an opening between the mastoid and middle ear created in the posterior wall of the middle ear lateral to the facial nerve and medial to the chorda tympani. This procedure is an extension of the complete simple mastoidectomy that allows better visualization of the facial recess without removal of the canal wall and is primarily advocated for ears in which a cholesteatoma is present. A **modified radical mastoidectomy** (see Figure 32C) is an operation in which a portion of the posterior ear canal wall is removed and a permanent mastoidectomy cavity is created, but the tympanic membrane and some or all of the ossicles are left. The procedure is usually performed when a cholesteatoma cannot be removed without removal of the canal wall; some function may be preserved. **Radical mastoidectomy** (see Figure 32D) involves exenteration of all mastoid air cells, opening of the epitympanum, and removal of the posterior ear canal wall along with the tympanic membrane, the malleus, and the incus. Only the stapes, or the footplate of the stapes, remains. No attempt is made to preserve or improve function. By the removal of the posterior ear canal wall, the exenterated mastoid cellular area, middle ear, and external auditory canal communicate, forming a common single cavity. The procedure is indicated when there is extensive cholesteatoma in the middle ear and mastoid that cannot be removed by a less radical procedure. In addition, the operation may be indicated when a suppurative complication of otitis media is present.

**Tympanomastoidectomy** is the term used when a tympanoplasty operation is performed in conjunction with a mastoidectomy. Mastoidectomy operations that leave the
posterior ear canal wall intact are termed closed cavity, canal wall up, or intact canal wall procedures, whereas those in which the posterior canal is partially removed are called open cavity or canal wall down procedures. For more complete details of the surgical procedures, the reader is referred to Bluestone and Bluestone and Stool.

The type of surgery chosen for management of cholesteatoma in children should be selected on the basis of the site and extent of the cholesteatoma and other factors, such as the patient’s age, presence or absence of otitis media, eustachian tube function, and availability of health care. The operation must be tailored for each child. Today, every effort should be made to maintain the posterior ear canal in children as opposed to performing canal wall down procedures, that is, modified or radical mastoidectomy. However, when the tympanic membrane is preserved or reconstructed ( tympanoplasty), a second operative procedure is indicated in most children, usually 6 months later, to determine whether there is residual cholesteatoma. Unfortunately, CT and MRI are not sensitive enough to replace exploratory surgery; the disease must be far advanced for these scans to be an effective diagnostic tool. Some have advocated use of a telescope to detect residual cholesteatoma in an effort to avoid the traditional exploratory tympanotomy (and possible mastoidotomy). We advocate exploratory tympanotomy (ie, “second look”) and possible mastoidotomy in children in approximately 6 months in a search for possible residual disease from the initial procedure. Other surgeons also recommend a second-stage procedure.

Preventing the recurrence of cholesteatoma in these children is important because the same underlying pathogenetic conditions may still be present. Thus, effective management of medical
conditions that may interfere with adequate eustachian tube function, such as adenoids, allergy, or chronic sinusitis, may prove to be beneficial. However, because many of these children have a basic eustachian tube dysfunction, placement of cartilage grafts, tympanostomy tubes, or both is usually required to prevent a new cholesteatoma. Long-term follow-up (eg, 5–7 years) after initial control of cholesteatoma in children is required because recurrence has been reported to be as high as 50 to 70%.\textsuperscript{398,549–501}

**CHOLESTEROL GRANULOMA**

Cholesterol granuloma is a sequela of chronic otitis media with effusion. It has been described as *idiopathic hemotympanum* because, clinically, the tympanic membrane appears to be dark blue, a so-called blue eardrum.\textsuperscript{502} However, this term is a misnomer because there is no evidence that bleeding within the middle ear or the presence of fresh blood or microscopic amounts of old blood is related to the etiology of this disease.\textsuperscript{16,503} The condition is rare in all age groups but does occur in children and is most likely due to long-standing changes associated with chronic otitis media with effusion.\textsuperscript{504,505} In a series of 17 cases of cholesterol granuloma of the petrous apex reported by Brodkey and colleagues, 3 (18%) occurred in adolescents and the rest in adults.\textsuperscript{506}

The blue color of the tympanic membrane as visualized through the otoscope is probably due to the reflection of light from the thick liquid (granuloma) within the middle ear. The condition must be differentiated from an uncovered high jugular bulb and a glomus tumor, either tympanicum or jugulare,\textsuperscript{507} and more commonly chronic otitis media with effusion or barotitis. Because cholesterol granuloma is relatively rare in children, the presence of a blue color of the tympanic membrane, as visualized by otoscopy or otomicroscopy, is most commonly middle-ear effusion, and the blue appearance is related to how the light is reflected off the drum.

Chronic granulations, with foreign body giant cells and foam cells within the middle ear, mastoid, or both are characteristic of the tissue. Cholesterol crystals are usually present. The condition is similar to a chronic middle-ear effusion except that a soft brownish material that contains shining golden-yellow specks is present. The pathologic process present with cholesterol granuloma should not be confused with that of a cholesteatoma.\textsuperscript{217} Similar granulomas have been described in other parts of the body: atheromatous and dermoid cysts, periaxial and follicular cysts of the jaw, old infarcts, and hematomas.\textsuperscript{508} When the granulomas are stained, prominent iron deposits or hemosiderin may be found,\textsuperscript{509,510} but not in quantities sufficient to account for the otoscopic appearance of the blue tympanic membrane.

The condition has been reproduced in experimental animals by injection of foreign material into the middle ears of guinea pigs\textsuperscript{511} and rabbits,\textsuperscript{512} by obstruction of the long bones of birds,\textsuperscript{513–515} and after chronic obstruction of the eustachian tube in monkeys.\textsuperscript{516} The pathogenesis described in the last experimental model is similar to that known to occur in humans when the eustachian tube was obstructed by a muscle pedicle flap\textsuperscript{517} or by a tumor. In addition to occurring as an isolated pathologic entity, cholesterol granuloma can be associated with chronic supplicative otitis media with or without cholesteatoma, any inflammation that may obstruct portions of the middle ear or mastoid, or both.

CT may be helpful in diagnosis and in determining the extent of the disease, but MRI is probably more specific as a diagnostic aid.\textsuperscript{518}

**Management**

Cholesterol granuloma will not respond to medical treatment, middle-ear inflation, or myringotomy with tympanostomy tube insertion. However, when a child is observed to have a tympanic membrane that has a dark blue appearance and is unresponsive to nonsurgical management, a myringotomy under general
anesthesia should be performed because, on occasion, chronic otitis media with effusion may also be associated with a blue tympanic membrane (again, probably as the result of the way light from the otoscope is reflected from the middle-ear effusion). However, if a thick, brown liquid is found during the procedure, successful aspiration of the material will not be possible, and if a tympanostomy tube is inserted, it will become occluded immediately. Takahashi and colleagues reported success in treating five patients, aged 6 to 19 years, with a short course of oral prednisone for a 10- to 14-day period and tympanostomy tube insertion.519 Despite the small number of patients evaluated, this method of treatment may be helpful because the traditional method of management has been middle-ear and mastoid surgery.

When tympanostomy tube insertion and possibly a trial of cortisone fail, the treatment is middle-ear and mastoid surgery.520 The granuloma in the mastoid can be removed by performing a complete simple mastoidectomy, and the middle-ear portion can be removed by use of a tympanomeatal approach. There is no reason to remove the canal wall unless a cholesteatoma is present. A tympanostomy tube should be inserted into the tympanic membrane at the time of the procedure and reinserted as often as needed, that is, until the middle ear remains normally aerated after spontaneous extubation.

When cholesterol granuloma involves the petrous apex, conservative management is recommended unless the patient has severe symptoms, such as vertigo and otalgia. When surgery is indicated, a transmastoid drainage procedure is recommended, as opposed to resection of the petrous apex.506 However, recurrence after a drainage procedure has a rate as high as 60% and requires an extended middle fossa approach or petrosal approach.521

It appears from what is known of the pathogenesis and pathology of cholesterol granuloma that the best management is prevention, which should consist of active treatment and prevention of chronic otitis media with effusion. **TYMPANOSCLEROSIS**

Tympanosclerosis may be a sequela of chronic middle-ear inflammation or the result of trauma. It is characterized by whitish plaques in the tympanic membrane and nodular deposits in the submucosal layers of the middle ear.522,523 When the disease is limited to the tympanic membrane, the term myringosclerosis is applied. Conductive hearing loss may occur if the ossicles become embedded in the deposits.

Tympanosclerosis was first described by von Troltsch, who called it sclerosis,524 but it was Zollner who called the disorder tympanosclerosis and differentiated it from otosclerosis.525 Schuknecht preferred the term hyalination to tympanosclerosis because the histopathologic condition is that of hyalin degeneration, which is the result of a healing reaction characterized by fibroblastic invasion of the submucosa, followed by thickening and fusion of collagenous fibers into a homogeneous mass.217 He also described the hyalinized collagen around the ossicles. The pathologic condition in the tympanic membrane occurs in the lamina propria, whereas the pathologic condition within the middle ear is in the basement membrane; in both sites, there is hyalinization followed by deposition of calcium and phosphate crystals.

**Epidemiology and Pathogenesis**

Tympanosclerosis (myringosclerosis) of the tympanic membrane is a common sequela in children who have or have had recurrent AOM or chronic otitis media with effusion. It is also common at the site of a healed, spontaneous perforation or after myringotomy and tympanostomy tube placement. In children who have had tympanostomy tube insertion for otitis media, tympanosclerosis increases with advancing age as the incidence of otitis media with effusion declines.368,526 The middle ear may have tympanosclerosis without the presence of clinically evident myringosclerosis.527 It is not commonly a sequela of myringotomy, and when due to the
aftermath of tympanostomy tube insertion, it is not linked to hearing loss.\textsuperscript{528} The chalky patch seen in the tympanic membrane of children may be due to inflammation, trauma, or both. In the pediatric age group as a whole, however, the condition is not common in the middle ear, especially in infants and young children. In particular, ossicular involvement is rare in very young children. Of 311 cases of tympanosclerosis studied by Kinney, only 20\% occurred in individuals 30 years of age or younger.\textsuperscript{529} This implies that the condition in the middle ear takes many years to develop. An expression of macrophages is an early event.\textsuperscript{530} Mattsson and colleagues, in experiments in rats, reported that the formation of oxygen free radicals contributes significantly to the development of myringosclerosis.\textsuperscript{531} This finding supported their hypothesis that the condition is secondary to a hyperoxic condition of the middle ear, such as when the tympanic membrane is not intact (eg, perforation or tympanostomy tube). In experiments in rats, nitrous oxide was found to be involved in this pathologic condition.\textsuperscript{532} A classification-related site, severity, and middle-ear involvement have been proposed by Bluestone and colleagues.\textsuperscript{10}

Schiff and colleagues hypothesized that tympanosclerosis has an immune component that occurs in the middle ear after an insult or mucosal disruption.\textsuperscript{533} They also proposed that there is a genetic component, which would explain the low incidence of the condition in children who have such a high prevalence and incidence of middle-ear inflammation.

This hypothesis may be the explanation for the relatively high rate of tympanosclerosis among children who are Alaskan natives (Eskimos) and American Indians.\textsuperscript{462,463,534} Other factors may predispose these children to the disease, such as differences in eustachian tube function. Wiet and colleagues reported that tympanosclerosis affected a higher percentage of Alaskan native children than children of a similar age in his New Hampshire private practice; tympanosclerosis of the tympanic membrane, the ossicles, or both was found in 78 (68\%) of 114 Alaskan native children who had tympanoplasty surgery, whereas only 7 such cases were diagnosed in 377 consecutive tympanoplasties performed on children in his practice.\textsuperscript{140} In addition, they also found that far-advanced tympanosclerosis that resulted in fixation of the ossicular chain occurred at an early age in the Alaskan native children but not in children in his practice.

Inflammatory mediators have been uncovered as being involved in the pathogenesis in humans\textsuperscript{535,536} and in animal models.\textsuperscript{537}

**Management**

Currently, no surgical correction, such as tympanoplasty, is indicated when tympanosclerosis of the tympanic membrane (ie, myringosclerosis), even though extensive, is the only abnormality of the middle ear. Medical treatment and prevention are still in the laboratory stage of research.\textsuperscript{538} However, surgical intervention can be indicated on an individualized basis.\textsuperscript{523} If a middle-ear effusion is present and a myringotomy, with or without tympanostomy tube insertion, is indicated, the incision should be placed, if possible, in an area without involvement, leaving the affected area untouched. Removal of large tympanosclerotic plaques may result in a permanent perforation of the tympanic membrane. When an incision must be made in an area of tympanosclerosis, only the amount necessary to perform the procedure should be removed. When a tympanoplasty is being performed to repair a perforation of the tympanic membrane and tympanosclerosis is present in the drum remnant, removal of the plaque is optional; the plaque may remain if the area of tympanosclerosis does not interfere with the surgical procedure and is not impeding function. When tympanosclerosis is the cause of ossicular fixation and a tympanoplasty procedure is elected, the methods of removing the plaques and ossiculoplasty described by Glasscock and Shambaugh\textsuperscript{539} are appropriate for the rare child with this advanced stage of tympanosclerosis. A two-stage stapedectomy has
been advocated to correct the conductive hearing loss with some success. However, refixation of the ossicles is not uncommon even after apparently adequate surgical removal of the plaques and ossiculoplasty. A hearing aid should be considered if surgery is not performed or is not successful in restoring the hearing loss. Nevertheless, long-term hearing success was reported after ossicular reconstruction in older children and adults.

Even though the pathogenesis of tympanosclerosis is not completely understood, it seems most probable that appropriate management of recurrent and chronic middle-ear inflammation in infants and children is the best method of prevention. Because it also occurs after trauma to the tympanic membrane, myringotomy with tympanostomy tube placement should be performed with tympanosclerosis in mind as one of the potential complications and sequelae. In general, tympanosclerosis involving the tympanic membrane does not appreciably affect function, although when the ossicles are involved, the patient may have a significant conductive hearing loss. Thus, tympanostomy tubes may increase the incidence of tympanosclerosis of the tympanic membrane, but their placement may decrease the frequency of ossicular fixation owing to this disease later in life.

Some clinicians believe that tympanosclerosis, as a sequela of tympanostomy tube insertion, causes conductive hearing loss. For most children who have tympanosclerosis that affects the tympanic membrane secondary to recurrent or chronic otitis media, tympanostomy tube placement, or both, the condition is cosmetic and does not affect hearing. Stenstrom and Ingvarsson observed 88 otitis-prone children for up to 8 years and concluded that the level of hearing, as assessed by audiometry, was a poor indicator of tympanic membrane disease. Of course, the more ear disease and the more surgical procedures the child has can eventually involve the conductive hearing mechanism (see also “Myringotomy and Tympanostomy Tube Placement” in Chapter 8).

**OSSICULAR DISCONTINUITY AND FIXATION**

Discontinuity and fixation of the ossicles are possible sequelae of recurrent acute and chronic middle-ear disease and, in rare instances, complications of surgery. A staging system related to site and pathology has been proposed by Bluestone and colleagues, and Dornhoffer and Gardner have provided a statistical staging system related to prognostic factors in ossiculoplasty to correct the defects. Staging that is agreed on by consensus is important when reporting the outcome of ossiculoplasty.

**Pathology and Pathogenesis**

Ossicular interruption is the result of rarefying osteitis secondary to chronic inflammation of the middle ear. A retraction pocket or cholesteatoma may also cause resorption of the ossicles. The long process of the incus is most commonly involved, which results in incudostapedial disarticulation. The commonly accepted reason for erosion of this portion of the incus is its poor blood supply. However, because the tympanic membrane frequently becomes attached to this part of the incus when a posterosuperior retraction pocket is present, adhesive otitis media may be the cause of the osteitis and subsequent erosion. Cholesteatoma is also commonly found in the same area. The stapes (or, more specifically, its crural arches) is the second most commonly involved ossicle. Erosion of the stapes is more likely to be associated with a retraction pocket or cholesteatoma than with decreased vascular supply. Less commonly, the body of the incus and the manubrium of the malleus may also be eroded. The ossicles may become fixed by fibrous tissue secondary to adhesive otitis media or, more rarely in children, secondary to tympanosclerosis. Neither the incidence of ossicular discontinuity and fixation nor the natural history of the pathologic conditions that precede these abnormalities has been formally studied in children. However, ossicular discontinuity is commonly associated with a deep retraction...
pocket or cholesteatoma in the posterosuperior portion of the tympanic membrane. Disarticulation or fixation of the ossicles may also occur when there is a central perforation of the tympanic membrane with or without chronic suppurative otitis media and, more rarely, when the tympanic membrane is intact.

**Diagnosis**

Ossicular chain abnormalities that are secondary to otitis media and its related conditions can frequently be diagnosed by visualization of the defect through the otoscope or, more accurately, the otomicroscope. Erosion of the long process of the incus can usually be seen when a deep posterosuperior retraction pocket is present. A significant conductive hearing loss (eg, >30 dB) when a perforation of the tympanic membrane is present is evidence of ossicular involvement. When the tympanic membrane is normal, a significant conductive loss may be due to inflammatory ossicular involvement that has occurred in the past. However, congenital ossicular abnormalities and otosclerosis must be part of the differential diagnosis.

In addition to the history, otoscopic examination, and conventional audiometric testing, immittance audiometry may aid in the diagnosis. A tympanogram showing high compliance is evidence of ossicular chain discontinuity when there is a significant conductive hearing loss. If the compliance is low, ossicular fixation is more probable. However, the accuracy with which tympanometry can differentiate between ossicular discontinuity and fixation is not high because several other parameters in the middle ear affect the shape of the tympanogram, such as the mobility of the tympanic membrane. CT may also aid in identifying ossicular discontinuity but is usually of diagnostic benefit only when a large defect is present. The most accurate way to diagnose these defects is exploration of the middle ear, either during exploratory tympanotomy, when the tympanic membrane is intact, or by inspecting the entire ossicular chain when surgery of the middle ear and mastoid is indicated, such as tympanoplasty.

Conductive hearing loss is usually present when the ossicular chain is affected, and the degree depends on the site and the degree of involvement of the ossicles and on the presence or absence of associated conditions, such as a perforation of the tympanic membrane. When there is a discontinuity of the incudostapedial joint and the tympanic membrane is intact, a maximal conductive hearing loss may be present, that is, 50 to 60 dB. However, when the same ossicular pathologic condition is present and a perforation is also present, the hearing loss may be less severe. Erosion of the manubrium of the malleus is usually associated with a perforation of the tympanic membrane but does not contribute to the hearing loss. However, erosion of the ossicles may not be associated with any significant degree of hearing loss if the defect is partial and the continuity of the ossicular chain is present or the tympanic membrane, retraction pocket, or cholesteatoma connects a disarticulation. When a cholesteatoma creates this artificial connection, this phenomenon is commonly termed hearing through cholesteatoma.

**Management**

Ossicular deformities in children can be managed like those in adults, with some notable exceptions. Most adults who have ossicular discontinuity or fixation are no longer at risk of development of otitis media with effusion or high negative pressure within the middle ear owing to eustachian tube dysfunction, but many children still have or will have these conditions. These conditions could interfere with the success of reconstructive middle-ear surgery generally and ossiculoplasty specifically. Therefore, the indications for timing and the type of middle-ear surgery may be different for children.

When an ossicular deformity is suspected and the tympanic membrane is intact, such as when the child has a conductive hearing loss, and there is no evidence of otitis media or any of its other complications or sequelae (eg, retraction pocket
or cholesteatoma), the decision to perform an exploratory tympanotomy to diagnose and possibly repair the ossicular deformity depends on several considerations. First, and most important, is the child still at risk of developing a middle-ear effusion, atelectasis (retraction pocket), or both? As a general rule, if neither condition has occurred in either ear for 1 year or longer, the risk is low. However, the younger the child, the higher the risk. If further middle-ear disease may still occur, the operation should be delayed. The second consideration is the degree of hearing loss and whether the defect is unilateral or bilateral. A child who has a maximal conductive hearing loss in both ears is a probable candidate for surgical intervention, whereas the child who has only a unilateral mild conductive loss should not be operated on. Another important consideration is the need for general anesthesia to perform the surgery in all children. The benefit of surgery must be weighed against the risk of general anesthesia. For the child who has a bilateral maximal conductive hearing loss, the benefit of hearing improvement may outweigh the risk of general anesthesia, whereas the risk of anesthesia may not override the potential chance of improving the hearing in a child with only a unilateral mild to moderate hearing loss. Withholding the reconstructive surgery until the child is able to tolerate a local anesthetic (adolescence) is a preferred option when the hearing loss is unilateral and mild to moderate in degree. Whenever surgical intervention is not planned or the operation is delayed until the child is older, a hearing aid evaluation should be considered, even when the hearing loss is unilateral.

When surgery of the middle ear is indicated because a perforation of the tympanic membrane is present, with or without chronic suppurative otitis media, a cholesteatoma, or a retraction pocket, the very fact that the patient is a child still affects the decision to perform an ossiculoplasty. This is because children are at increased risk of suffering future episodes of otitis media and of developing atelectasis or adhesive otitis media. When these conditions are a possibility, the surgeon should consider staging the surgery and performing the ossiculoplasty when the child is older. If the patient has a cholesteatoma, and a canal wall up procedure followed by a planned “second-look” tympanotomy is elected to determine whether residual cholesteatoma is present, the ossiculoplasty can be performed when the middle-ear cleft is free of disease.

There are various ways in which ossicles that are either eroded or fixed may be reconstructed. However, the type of ossiculoplasty chosen for a child may be different from that performed in an adult. We believe, and most experts agree, that middle-ear ossicular implants should, in general, be used only in selected children who have ossicular defects as a result of middle-ear disease because neither the long-term safety nor the efficacy of these prostheses has been proved. However, if the child’s middle ear is free of disease and the patient is unlikely to have future middle-ear problems, insertion of a prosthesis is an option.549

Some surgeons advocate the use of homograft ossicles in children. Whenever possible, only the child’s own tissue should be used to reconstruct the ossicular chain. For the most common discontinuity encountered, that of the incudostapedial joint, an incus transposition or insertion of a fitted incus is the ideal procedure. When the stapes crura are missing, the shaped incus can usually be inserted between the mobile footplate of the stapes and the malleus handle. For all age groups, whenever the stapes is fixed, a stapedectomy should not be performed unless the tympanic membrane is intact; and in children who have had otitis media in the recent past, stapedectomy should rarely, if ever, be performed, even when the tympanic membrane is intact because a recurrence of otitis media with suppurative labyrinthitis as a complication is an ever-present risk. Freeing other fixed ossicles can be attempted in children, but refixation often occurs because adhesive otitis media, which is the most frequent cause of fixation, commonly leads to further fibrosis. There are surgical procedures in which no prosthesis is used,550 but for some defects, a prosthesis is necessary, the placement
of which should be as optimal as possible to optimize the chance of as good a hearing result as possible.\textsuperscript{531} For further details of surgical techniques for children, see Batti and Bluestone.\textsuperscript{548}

The most effective method of managing ossicular discontinuity and fixation is preventing the diseases that cause these ossicular abnormalities. Of special note is the early diagnosis and management, usually by insertion of a tympanostomy tube, of a posterosuperior retraction pocket in which the tympanic membrane is lying on the incus and stapes (see the section on atelectasis of the middle ear).

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