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# Diagnosis of Pediatric Cholesteatoma

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## ABSTRACT

Cholesteatomas are abnormal collections of squamous epithelium and keratin debris that usually involve the middle ear and mastoid. Although histologically benign, they have the ability to expand and destroy bone. Cholesteatomas are treated surgically. The success of such surgery is highly dependent on the extent of the lesion. This article presents information and images to aid the general pediatrician in the early recognition of cholesteatomas, both congenital and acquired, in hopes of improving the outcome for children with this treatable disorder.

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### Key Words

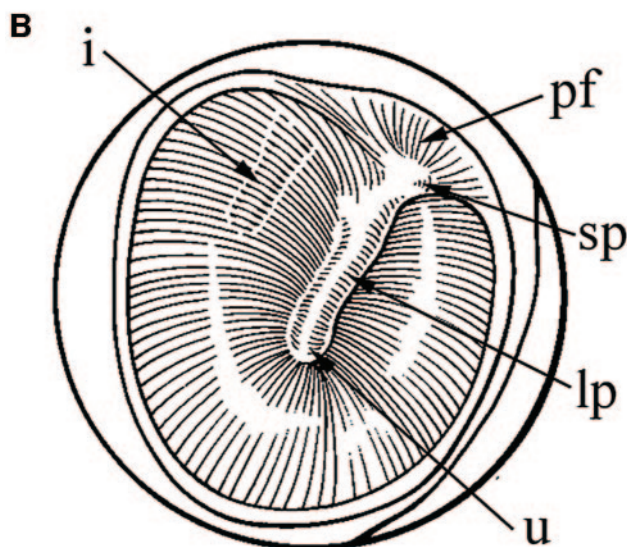
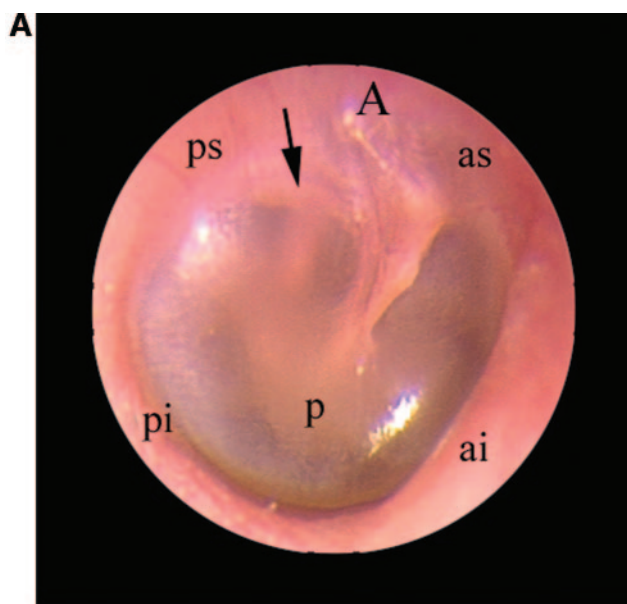
cholesteatoma, chronic otitis media, ear surgery, hearing loss, otorrhea, otoscopy

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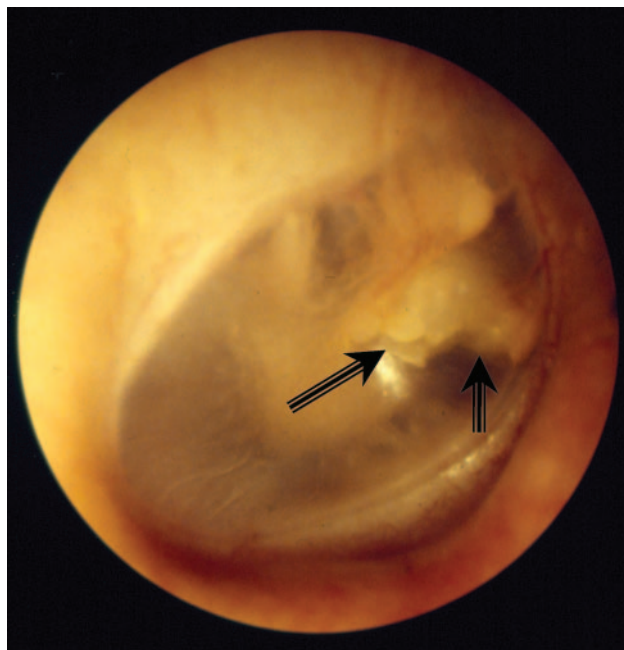
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**S**KIN DOES NOT belong in the middle ear or mastoid. When keratinizing squamous epithelium gets into these air spaces, it can form a progressively enlarging and destructive cystic lesion called a cholesteatoma.<sup>1</sup> In parts of the world with limited access to advanced medical care, cholesteatomas remain a cause of pediatric morbidity and occasionally death.<sup>2</sup> Large erosive lesions that arise in the middle ear can extend through the roof of the temporal bone to compress the brain, and associated infection may cause intracranial abscesses.<sup>3</sup> In the United States, such lethal complications are rare, but late



**FIGURE 1**  
A, Otoscopic view of a normal right tympanic membrane. ps indicates posterosuperior quadrant; as, anteriosuperior quadrant; pi, posteroinferior quadrant; ai, anteroinferior quadrant; A, bone covering the attic or epitympanum; p, promontory seen through the tympanic membrane. The arrow points to the chorda tympani nerve. B, Drawing of a normal right tympanic membrane. i indicates incus long process; sp, malleus short (lateral) process; lp, malleus long process; u, umbo; pf, pars flaccida.



**FIGURE 2**  
Congenital cholesteatoma of the anteriosuperior quadrant. A white mass behind an intact tympanic membrane is shown in the right ear (arrows points to the mass).

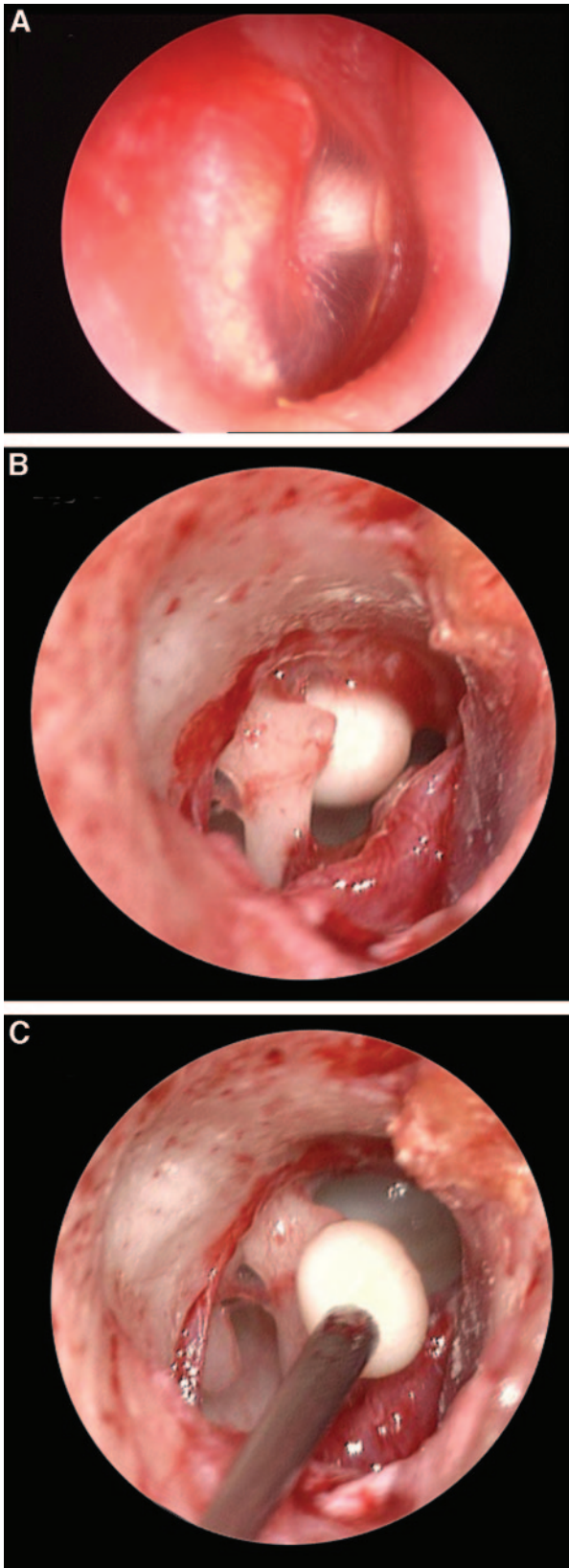
recognition of cholesteatoma remains a major cause of permanent hearing loss.<sup>4</sup>

Most cholesteatomas can be cured by surgery. The extent and effectiveness of such surgery is strongly correlated with the size of the cholesteatoma at presentation.<sup>5</sup> Diagnosis of small lesions by a pediatric otolaryngologist depends on the skills of the referring pediatrician (Fig 1). The focus of this article is on the correct, early identification of cholesteatoma in childhood.

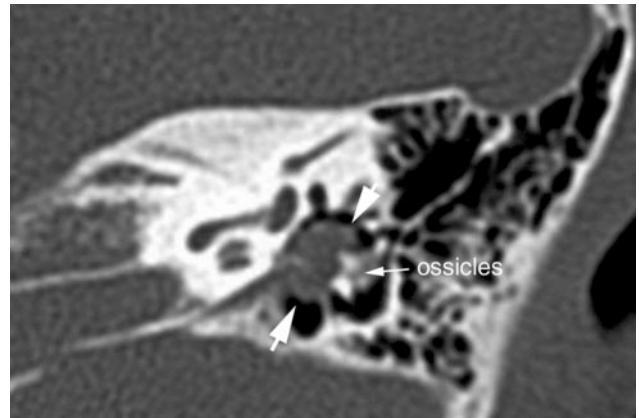
### CONGENITAL CHOLESTEATOMA

Cholesteatomas are either congenital or acquired. Classically, a cholesteatoma is said to be congenital when it presents as a white mass behind an intact eardrum in a child with no previous history of otitis (Fig 2). Because nearly all children have some history of middle-ear disease in infancy, most researchers consider a cholesteatoma congenital if there is no history of previous ear surgery and there is no perforation or retraction of the tympanic membrane.<sup>6</sup>

There is debate about the origin of congenital cholesteatomas. The most popular theory argues that squamous inclusion cysts arise from epithelial rests in the middle ear. These “epidermoid formations” can be demonstrated histologically in fetal temporal bones.<sup>7</sup> They may be single or multiple and typically disappear in the third trimester of gestation. Failed involution leads to cholesteatoma behind an intact tympanic membrane.<sup>8</sup> Alternative theories<sup>9</sup> include seeding of the middle ear by squamous cells in the amniotic fluid<sup>10</sup> or from the



**FIGURE 3**  
A, Congenital cholesteatoma of the anterosuperior quadrant. B, The eardrum is reflected downward to reveal a white spherical lesion. C, Removal of the lesion.

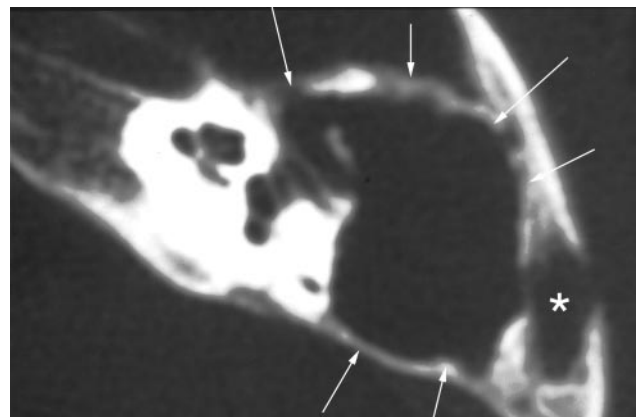


**FIGURE 4**  
Axial computed tomography of moderate-sized congenital cholesteatoma (large arrows surround the lesion) deep to the ossicles in the attic (labeled ossicles).

surface epithelium of the tympanic membrane after infection and microperforation.<sup>11</sup>

Congenital cholesteatomas can occur anywhere in the temporal bone but have a predilection for the anterosuperior quadrant of the middle ear, just above the eustachian-tube opening.<sup>12</sup> Early on, the cyst is hard to appreciate, appearing as a subtle whitish discoloration behind an otherwise normal tympanic membrane (Fig 3A). As the lesion grows, it makes contact with the underside of the eardrum and becomes more obvious. If the cyst obstructs the eustachian tube, the middle ear can fill with effusion, which complicates the diagnosis. As the lesion expands, it can replace the middle-ear space and displace the tympanic membrane outward.

An experienced pediatric otolaryngologist will sometimes operate to remove a small congenital cholesteatoma without additional diagnostic studies. More commonly, preoperative computed tomography confirms the nature of the lesion and its extent (Figs 4 and 5). When an effusion is present, some surgeons prefer to



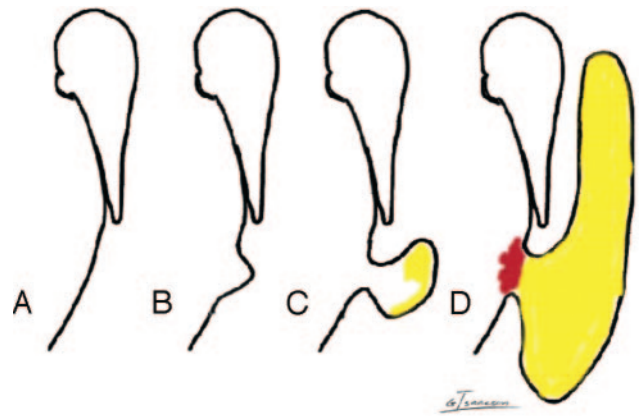
**FIGURE 5**  
Axial computed tomography of a large cholesteatoma of the mastoid. Arrows outline the area of destruction, and the asterisk shows a breach of the mastoid cortex by infection with a subperiosteal abscess.

place a tympanostomy tube and examine the ear under anesthesia as a preliminary step. Tube placement enhances the value of computed tomography by introducing air contrast into the middle ear and mastoid, and it improves the chance of finding a dry, uninfamed middle ear at the time of definitive surgery.

### ACQUIRED CHOLESTEATOMA

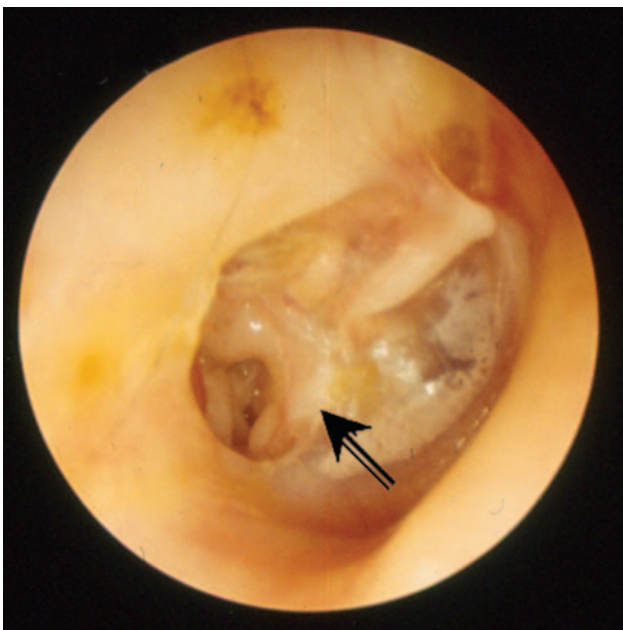
When a cholesteatoma forms after birth, usually as a result of chronic middle-ear disease, it is said to be acquired. Such cholesteatomas form in 3 ways: (1) acquired cholesteatomas arise most commonly from focal retractions of the tympanic membrane (retraction-pocket cholesteatomas); (2) superficial epithelium can enter the middle ear through a perforation of the tympanic membrane (Fig 6) or along a temporal-bone fracture line; and (3) squamous epithelium can be introduced into the middle ear after surgery, as a complication of tympanoplasty, or by creeping in alongside a retained tympanostomy tube (secondary acquired cholesteatoma). These latter 2 etiologies are rare.

Retraction pockets are invaginations of the tympanic membrane. The squamous lining of these pockets is the surface epithelium of the ear tympanic membrane, drawn into the middle-ear space by the vacuum associated with chronic eustachian-tube dysfunction<sup>13</sup> (Fig 7). There is net absorption of oxygen from the middle-ear space into capillaries of the mucosa that lines the middle ear and mastoid. Absorbed gas must be replaced from the nasopharynx via the eustachian tube. Poor tubal function thus leads to chronic negative middle-ear pressure and focal collapse of the drum.<sup>14</sup> Areas of predilec-

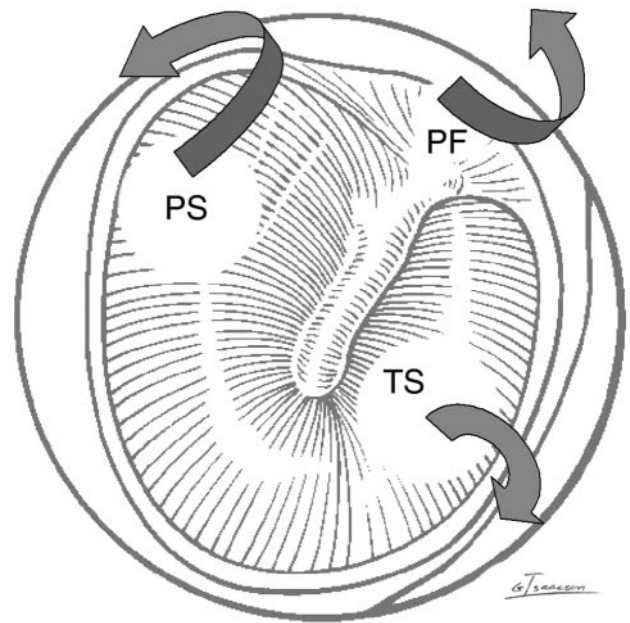


**FIGURE 7**  
Mechanism of acquired cholesteatoma formations: A, normal drum in cross-section; B, shallow focal retraction; C, deep retraction with collection of keratin debris; D, infected cholesteatoma with granulation at its mouth.

tion include old tympanostomy-tube sites, the postero-superior portion of the drum that overlies the entrance to the mastoid, and the pars flaccida<sup>15</sup> (Fig 8). As a retraction pocket is progressively drawn in, it forms a large pouch with a narrow neck that traps desquamating surface epithelium. The cholesteatoma that results is lined by the metabolically active squamous epithelium and filled with dead keratin debris. This epithelium has the ability to erode bone, first the delicate ossicles (Fig 9) and subsequently the solid cortices of the temporal bone (Fig 5). If this debris becomes infected after water exposure or middle-ear infection, chronic purulent drainage results (Fig 10).



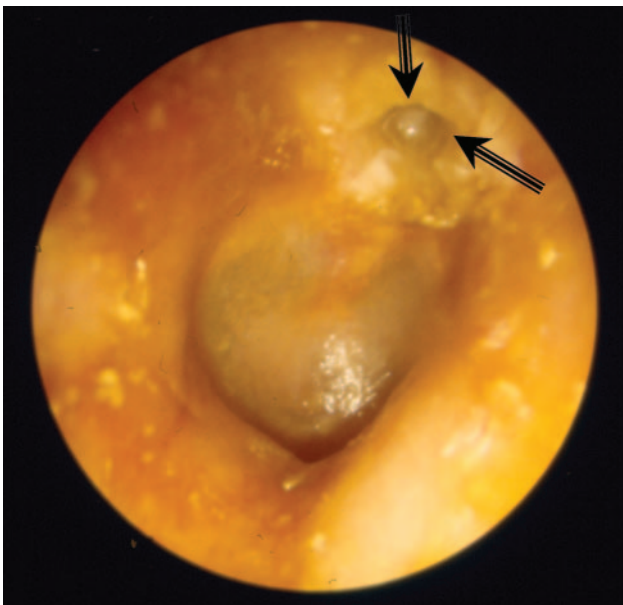
**FIGURE 6**  
A true marginal perforation. The arrow points to perforation.



**FIGURE 8**  
Paths of extension by acquired cholesteatoma. PS indicates posterosuperior quadrant toward mastoid; PF, pars flaccida toward attic; TS, retraction at old tube site toward middle ear.



**FIGURE 9**  
A retraction-pocket cholesteatoma of the posterosuperior quadrant. The incus long process is eroded, which leaves the drum adherent to the stapes head (S). An effusion is present in the middle ear, and squamous debris emanates from the attic.



**FIGURE 10**  
Acquired cholesteatoma of the epitympanum arising in the pars flaccida. This lesion presented with chronic purulent drainage. Arrows point to the pars flaccida retraction.

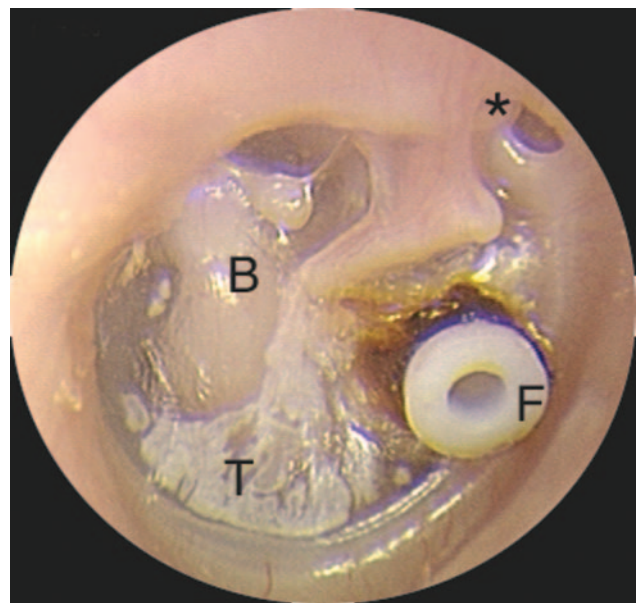
Most children with retraction-pocket cholesteatomas have a history of recurrent acute otitis media and/or chronic middle-ear effusion. Children with cleft palates,<sup>16</sup> craniofacial anomalies, Turner<sup>17</sup> or Down<sup>18</sup> syndromes, or a family history of chronic middle-ear disease and/or cholesteatoma are at increased risk.

## RECOGNIZING A CHOLESTEATOMA

Cholesteatomas are suspected by pediatricians and diagnosed by otolaryngologists using visual examination, palpation under an operating microscope, audiometric testing, computed tomography, and surgical exploration. Careful visual examination is, by far, the most important. As a part of good routine care, it is important to see the entire tympanic membrane. Only by inspecting its outer edges can one see early lesions.

Congenital cholesteatomas are first visible as spherical white cysts behind an intact tympanic membrane (Figs 2 and 3A). As they enlarge, the normal variegated appearance of the middle-ear space disappears, replaced by a bulging white mass. Congenital cholesteatomas often arise above the eustachian-tube orifice and obstruct it early in their course, filling the middle ear with effusion. Perforation of the drum and chronic ear drainage are not typical of early lesions and usually trail the onset of hearing loss by months or years.<sup>12</sup> Other white lesions of the drum are easily confused with congenital cholesteatomas. Tympanosclerosis, white foreign bodies, exostoses, and prosthetic and graft material in operated ears can all mimic cholesteatoma (Fig 11). Squamous inclusion cysts of tympanic membrane occupy the middle layers of the drum but do not extend into the middle ear. Occasional, bulging acute otitis media may be hard to differentiate from a large congenital cholesteatoma.

Retraction pockets are recognized by careful examination of the intact drum. They are said to be “shallow” when the full extent of the pocket is visible by otoscopy (Fig 11). They are “deep” when they disappear into the crevices of the middle ear or into the mastoid air spaces



**FIGURE 11**  
Three white things that are not cholesteatoma. T indicates tympanosclerosis; B, bone (promontory below, stapes head above); F, foreign body (a white tympanostomy tube); \*, a shallow retraction of the pars flaccida.

(Fig 9). Deep pockets can collect debris out of otoscopic view and are assumed to be cholesteatomas until proven otherwise. If a retraction pocket expands into the middle ear, its mouth may be visible near the edge of the drum (Fig 10). The cholesteatoma sac, filled with dead keratin debris, may appear as a white mass behind the drum. Acquired cholesteatomas present more commonly with recurrent or chronic purulent drainage. Any ear with persistent drainage for >2 weeks after an infection or water exposure may represent a cholesteatoma. Careful cleaning of the ear under the operating microscope may reveal focal granulation covering the mouth of an infected cholesteatoma.

### CHOLESTEATOMA WARNING SIGNS

1. a white mass behind an intact ear drum;
2. a deep retraction pocket with or without granulation and skin debris;
3. focal granulation on the surface of the drum, especially at the periphery;
4. an ear that continues to drain for >2 weeks despite treatment; and
5. new-onset hearing loss in a previously operated ear.

When a suspicious lesion is encountered or if an ear continues to drain, referral to a pediatric otolaryngologist is important. Careful cleaning and inspection of the ear under an operating microscope, audiometric testing, and appropriate radiologic examination can aid in the early diagnosis of these destructive, but treatable, lesions.

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