Let's talk about . . .

CHOLESTEATOMA

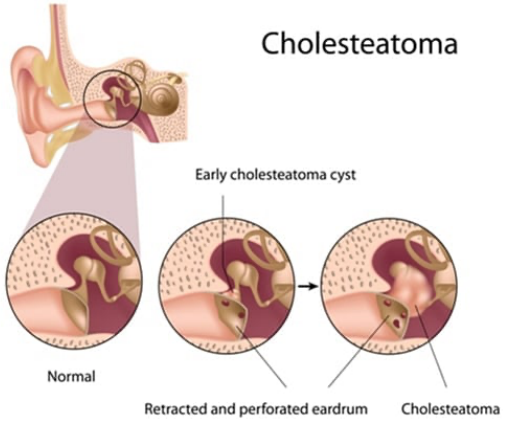
This handout is intended as a general introduction to the topic. As each person is affected differently, speak with your health care professional for individual advice.

Key points

- Skin in the wrong place in the middle ear (behind the eardrum).
- Benign (not cancerous) growth.
- Rare and slow growing.
- Affects people of all ages.
- Hallmark early symptom is a foul-smelling discharge from the inner ear.
- Other early symptoms include conductive hearing loss, pressure, pain, or numbness in or around one ear.
- In children, hearing loss is the most common symptom.
- If not treated, can eventually destroy the parts of the inner ear needed for hearing and balance.
- Surgery is usually needed and successful. Growth rarely comes back.
- Usually, life-long regular ear-cleaning by an ENT is needed after surgery.

What is cholesteatoma?

A cholesteatoma (ko-less-tee-ah-TOME-ah) is a benign (non-cancerous) growth in the middle ear behind the eardrum (tympanum). Simply put, it is skin in the wrong place. People with cholesteatoma usually have a foul-smelling discharge from the ear. Cholesteatoma may lead to conductive hearing loss. In advanced disease, it can extend into the inner ear and cause dizziness. Surgery is needed for most cholesteatomas. Cholesteatoma is sometimes called keratoma.



Cholesteatoma is slow growing. In the early stages, a cholesteatoma may grow to surround and destroy 3 tiny bones (ossicles). This results in conductive hearing loss. Hearing loss also may happen if the cholesteatoma blocks the opening of the Eustachian tube, leading to a build-up of fluid in the space behind the eardrum (middle ear effusion).

Cholesteatoma is rare. It cannot be inherited. Usually only one ear is affected. Each year about 3 in 100,000 children and about 9 in 100,000 adults are diagnosed with cholesteatoma. Males are a little more likely to have cholesteatoma. White people are most affected.

The word cholesteatoma comes from the Greek for "fat" and "tumour." It is a misleading term;

cholesteatoma does not contain fat and it is not a tumour.

What are the causes?

\The cause of cholesteatoma depends on its type:

- **Congenital (present at birth) cholesteatoma** develops when small bits of skin become trapped in the middle ear behind the eardrum before birth. It is the least common type of cholesteatoma.
- **Primary acquired cholesteatoma** develops because of a dysfunctional Eustachian tube. It is the most common type of cholesteatoma. When working normally, the Eustachian tube passes air from the back of the nose into the middle ear to even out pressure in the ear. Sometimes one of the Eustachian tubes malfunctions. This may be because of allergies, sinus infections (sinusitis) or middle ear infections (otitis media). Decreased pressure inside the ear may cause the eardrum to collapse and stretch inward (retract). A pocket-like cyst filled with trapped dead skin cells may form when this happens.
- **Secondary acquired cholesteatoma** develops when skin from the outer part of the eardrum grows through a hole in the eardrum. This usually happens after recurring (chronic) middle ear infection, injury or as a consequence of minor surgery to insert ventilation tubes in the ear. Timely and thorough treatment of recurring middle ear infections may help prevent this type of cholesteatoma.

Scientists do not fully understand the risk factors for developing cholesteatoma. Known risk factors are chronic middle ear infections and Eustachian tube dysfunction. Studies suggest having cleft palate or osteoporosis may also play a role.

What are the symptoms?

Cholesteatoma is slow growing. There may be no symptoms for the first several years. The hallmark symptom of an acquired cholesteatoma is a foul-smelling discharge from the ear (otorrhea).

Other common early symptoms include:

- Gradual conductive hearing loss in the affected ear (early on in secondary acquired cholesteatoma, and later on in primary acquired cholesteatoma).
- A feeling of pressure in the ear (aural fullness).
- Pain or numbness in the ear or around the ear.

See a family doctor (general practitioner) if you have any or all of these symptoms.

If left untreated, over time a cholesteatoma may extend into the inner ear. Complications at this stage may include:

- Persistent ear drainage.
- Vertigo (spinning sensation) caused by damage to one of the semicircular canals.
- Tinnitus (ringing in the ears).
- A breakdown of the facial nerves leading to facial paralysis (not being able to move your face).

In very rare cases, a cholesteatoma may gradually destroy the temporal bone and start to affect the brain. This is a life-threatening complication. Urgent surgery is needed if any of the following develops:

- Mastoiditis – bacterial infection of the mastoid bone (large, bony area on the base of the skull behind the ear connected with the middle ear).
- Brain abscess – a collection of pus and other material in the brain.
- Meningitis – an infection of the membranes covering the brain and spinal cord.

The symptoms of congenital cholesteatoma are somewhat different. Usually, the eardrum looks normal and is unbroken. The child often has not had

a series of pus-forming ear infections, previous ear surgery, or a perforated eardrum. The most common symptom is hearing loss.

How is it diagnosed?

Cholesteatoma may be diagnosed by a family doctor (general practitioner). The doctor will ask about your symptoms, and may examine your ear with an otoscope (an instrument used to look into the ear). It can be hard to confirm a cholesteatoma by looking into your ear. A build-up of pus inside the ear often makes a cholesteatoma hard to see.

If the doctor suspects a cholesteatoma, you will be referred to an otolaryngologist (ear, nose and throat doctor or ENT) for a detailed ear exam and further tests. These may include:

- Audiogram to show the extent of conductive hearing loss.
- Tympanogram to show the condition of the eardrum and middle ear.
- CT (computed tomography) scan of your inner ear and mastoid bone (the bone at the back of the temporal bone just behind the inner ear). CT scans can detect very small defects in bones.
- VEMP test – usually there is no response to this test on the affected side. This would only rarely be done if there is no doubt that the patient has a cholesteatoma.
- MRI (magnetic resonance imaging) in advanced cases if the doctor suspects cholesteatoma has extended into the skull.

Congenital cholesteatoma is usually not diagnosed until a child is 2 to 3 years old when hearing loss is noticed.

Early diagnosis is important to ensure the best outcome for all types of cholesteatoma.

How is it treated and managed?

A small cholesteatoma with few symptoms may be managed by regular, repeated cleaning (debridement) by a doctor. You may be given antibiotic ear drops. This treatment may work for elderly people and those with "anaesthetic risks".

Surgery is usually needed to remove most cholesteatomas. The surgeon may be able to restore some hearing loss caused by the growth.

There are several types of surgeries for treating a cholesteatoma. Which procedure the surgeon chooses will depend largely on how far the cholesteatoma has spread and the amount of hearing loss.

Cholesteatomas can grow aggressively, causing serious and even life-threatening complications. For this reason, the benefits of surgery almost always outweigh any risks. Discuss possible risks before the procedure with your surgeon.

You will be given written post-surgical instructions and scheduled for a follow-up appointment with your surgeon.

Some cholesteatomas come back after surgery.

What to expect in the future

Cholesteatomas usually keep on growing if not removed. Surgery is usually successful, but you will likely need to see your ENT for regular cleaning of your ear for the rest of your life.

More surgery may be needed if the cholesteatoma comes back. You will need to see your ENT regularly to keep an eye on this and may need repeat surgery (revision).

In some cases, hearing improves after surgery. In others, complete removal of the growth also necessitates removal of compromised ossicles. This can make hearing worse. And if the cholesteatoma starts to grow again, hearing may worsen.

Hopefully research will lead to the development of nonsurgical therapies to treat cholesteatoma.

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