Cholesteatoma is an uncommon condition where a cyst-like growth develops in the ear. It can be a birth defect (congenital problem) but usually occurs as a complication of chronic (long-standing) ear infection. The most common symptoms are loss of hearing and a foul-smelling discharge from the ear. It is not a cancerous (malignant) condition but is important because it can lead to serious complications such as permanent deafness and life-threatening illnesses such as meningitis.

What is the ear like and how do we hear?

![Diagram of the ear and the hearing process]

- Pinna
- Floor of skull
- Middle ear
- Semicircular canals
- Eustachian tube
- Nerve
- Cochlea
- Bone
- Pharynx
- Adenoids
- Hammer (malleus)
- Anvil (incus)
- Stirrup (stapes)
- Sound waves
- Eardrum
- To cochlea and ear nerve which sends sound signals to brain
- Air in middle ear
The ear is divided into three parts - the external ear, the middle ear, and the inner ear. The middle ear, which is behind the eardrum (the tympanic membrane) is filled with air. Air comes from the back of the nose up a thin channel called the Eustachian tube. In the middle ear there are three tiny bones (ossicles) - the hammer (malleus), anvil (incus) and stirrup (stapes). The inner ear includes the cochlea and semicircular canals.

Sound waves come into the external ear and hit the eardrum. The sound waves cause the eardrum to vibrate. The sound vibrations pass from the eardrum to the ossicles. The ossicles then transmit the vibrations to the cochlea in the inner ear. The cochlea converts the vibrations to sound signals which are sent down a nerve from the ear to the brain, allowing us to hear.

The semicircular canals in the inner ear contain a fluid that moves around as we move into different positions. The movement of the fluid is sensed by tiny hairs in the semicircular canals which send messages to the brain down the ear nerve to help maintain balance and posture.

What is a cholesteatoma?

Cholesteatoma is the name given to a collection of skin cells in the ear that form a lump (mass). It is a pearly-white greasy-looking mass that can be seen when examining the ear.

![Diagram of ear with cholesteatoma formation](image)

The cholesteatoma grows. At first it is more likely to grow into (or onto) and destroy structures in the middle ear. In time it may grow and erode into and damage structures in the inner ear or even erode into the bone of the nearby skull and into the brain.

What causes a cholesteatoma?

The cause is not fully understood. It is thought that skin cells from the lining of the ear canal get trapped in the middle ear (which does not normally contain skin cells). Skin cells, including those that line the ear canal, normally multiply regularly to replace those that have died. Usually these skin cells flake off. The dead cells are trapped too and build up. This build-up of dead skin cells over time is what forms the cholesteatoma.

There are two types of cholesteatoma:

- **Congenital cholesteatoma** grows behind the eardrum (the tympanic membrane) from birth. It is thought that some skin cells develop in the wrong place in the ear and develop into a cholesteatoma.
- **Acquired cholesteatoma** develops later, usually in adults. This is often as a result of a long-term (chronic) or recurring ear infection. The infection causes a blockage of the Eustachian tube. This is the tube that connects the middle ear to the back of the nose and throat. The blockage creates a sucking (negative) pressure that draws the eardrum inwards. This can result in a small pocket forming, usually at the very top of the eardrum. In this pocket some skin cells collect, get trapped, but continue to multiply to form the cholesteatoma. The pocket is too deep to allow the dead skin cells to escape, so the cholesteatoma gradually expands.
How common is cholesteatoma?

Cholesteatoma is rare. The true occurrence (incidence) rate is not known. About 1 in 1,000 people with ear problems referred to ear, nose and throat (ENT) clinics have cholesteatoma. It has also been suggested that there is about 1 case per 10,000 population. Most cases are of the acquired type.

What are the initial symptoms of a cholesteatoma?

Cholesteatoma usually affects only one ear (it is unilateral). The most common initial symptoms are a smelly discharge from the ear, and some hearing loss. You are also likely to have had previous problems with ear infections. Other symptoms that may occur include a ringing sound in the ear (tinnitus) and headache.

What are the possible complications and why is it important?

Untreated, a cholesteatoma will slowly grow and expand. As it grows it can eat into (erode) and destroy anything in its path.

Therefore, possible complications that may develop over time include:

- Damage and eventual destruction of the tiny bones of the ear (the ossicles). If these are damaged, permanent deafness can occur.
- Damage to the mastoid bone. This is the thick bony lump you can feel behind the ear. The mastoid bone is normally filled with pockets of air (a bit like a honeycomb). Cholesteatoma can grow into the mastoid bone, causing infection and destroying it.
- Damage to the cochlea and other structures in the inner ear. This can cause permanent deafness on that side, and/or dizziness and balance problems.
- Damage to nearby nerves travelling to the face. This can cause weakness (paralysis) of some of the facial muscles.
- Cholesteatoma is often infected and this infection can spread to nearby body parts. In rare cases a cholesteatoma can erode through the skull next to the ear and into the brain. As a result of spreading infection, conditions such as meningitis and brain abscess can develop. These conditions can cause death.

Cholesteatoma is not cancerous (malignant) and does not spread to distant parts of the body.

How is cholesteatoma diagnosed?

The GP or ENT doctor may suspect cholesteatoma based on the typical symptoms. When the ear is examined with a torch (an otoscope), the cholesteatoma may be seen. Often there is a hole (perforation) in the eardrum (the tympanic membrane) too.

Do I need any tests?

Hearing tests (audiometry) may show deafness or hearing loss and are usually performed in a hospital clinic. Samples (swabs) of the ear discharge may also be taken. The discharge often contains a germ (bacterium) called Pseudomonas which is responsible for the smell. A CT scan might be needed to see the extent of the damage caused by the cholesteatoma, and to plan further treatment.

What is the treatment for cholesteatoma?

Surgery is usually advised to remove the cholesteatoma and prevent further damage. You may be given antibiotic medication to clear up any infection and make the ear dry, prior to surgery.
Surgery
Surgery removes all of the diseased areas, including the cholesteatoma itself:

**Tympanomastoidectomy.** This is removal of the eardrum (the tympanic membrane) and all or part of the mastoid bone of the skull, depending on the extent of the cholesteatoma. It is the procedure often performed and a large hole (cavity) is created in the inner ear region, requiring regular cleaning and removal of earwax (de-waxing) in the ENT clinic. This procedure is the most successful at removing all of the cholesteatoma. Complications of surgery can sometimes occur. These include deafness, dizziness and damage to the facial nerve, leading to weakness (paralysis) of the muscles in the face on the affected side.

**Tympanoplasty.** This is another operation that can be performed on the eardrum. It is a less invasive technique with a better visual (cosmetic) appearance after surgery. However, it is associated with a higher risk of recurrence of cholesteatoma, and a further operation to check is required up to a year later.

Non-surgical treatment
If you are not fit enough to undergo a general anaesthetic or do not want to have surgery, you may have the cholesteatoma treated with the aid of 'aural toilet'. This involves washing out the ear canal and sucking out bits (debris) using a microscope. This has to be done regularly and ultimately will not prevent growth of the cholesteatoma.

Is any follow-up required?
If you have had cholesteatoma, you will be followed up for life in an ENT clinic. You will need to have your ears cleaned regularly at the clinic to remove wax and any dirt that has accumulated. The specialist will need to ensure that the cholesteatoma has not returned. If the ear starts discharging again, further surgery may be required. MRI scans are increasingly being used to replace the need for further check-up surgery.

What is the outlook (prognosis)?
This depends on how much damage has been caused by the cholesteatoma by the time it is found and treated. It is also affected by whether any complications such as meningitis or deafness have occurred. The earlier surgery is done, and attending for regular follow-up, the better the chance of a good outcome.

Further help & information
**Action on Hearing Loss**
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