Cancer Association of South Africa (CANSA)

Research • Educate • Support

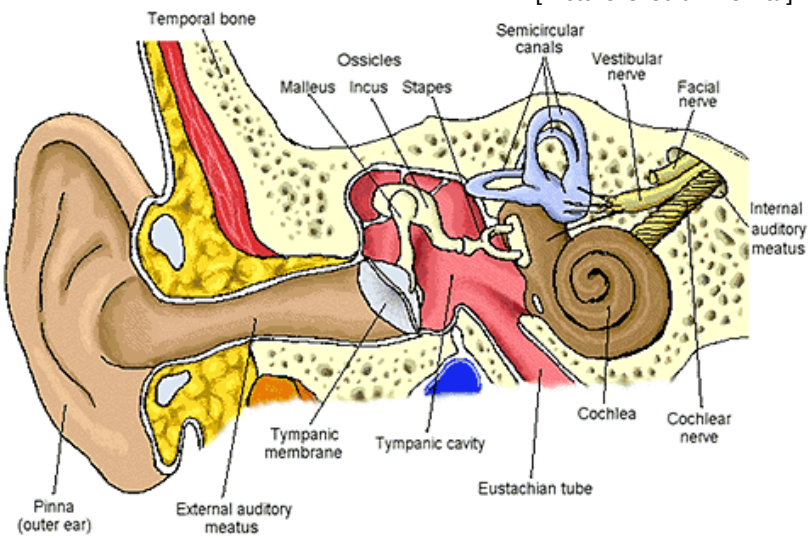
Fact Sheet
on
Cancer of the Ear

Introduction

The ear is part of the auditory system. The ear is the organ that detects sound. It not only receives sound, but also aids in balance and body position. The entire organ is often considered to be the ear, though just the visible portion of the ear may be considered to be the ear. In most mammals, the visible ear is a flap of tissue that is also called the pinna (or auricle in humans) and is plays a role in the first of many steps in hearing. Vertebrates have a pair of ears placed somewhat symmetrically on opposite sides of the head. This arrangement aids in the ability to localise sound sources.

The ear consists of three parts, namely the outer ear, the middle ear and the inner ear.

[Picture Credit: The Ear]



The outer ear - the outer ear is the most external portion of the ear. The outer ear includes the fleshy visible outer ear, called the auricle, the ear canal, and the outer layer of the tympanic membrane, also referred to as the ear drum.

The middle ear - The middle ear is an air-filled cavity behind the tympanic membrane and includes three tiny bones (ossicles): the malleus (or hammer), incus (or anvil), and stapes (or stirrup). The middle ear also connects to the upper throat via the Eustachian tube.

The inner ear - The inner ear is split anatomically into bony and membranous labyrinths. This contains the sensory organs for balance and motion, namely the vestibules of the ear (utricle and saccule), and the semicircular canals. It also contains the sensory organ for hearing, the cochlea. (Wikipedia).

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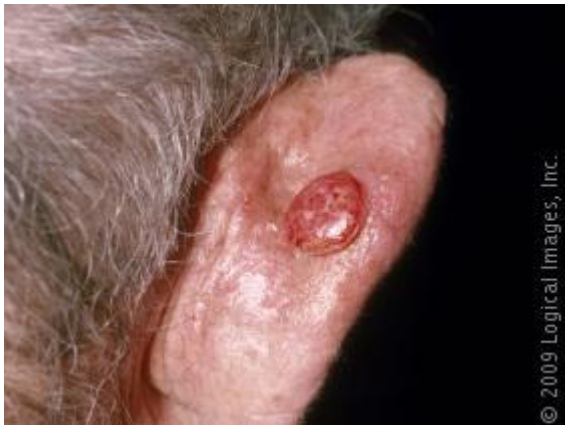
Cancer of the Ear
Cancer of the ear is a rare cancer. Most of these cancers start in the skin of the outer ear. About 5 out of 100 skin cancers develop on the ear. Those that develop inside the ear are very rare. On average, less than 1 in every million people will develop cancer in the middle ear.



[Picture Credit: Squamous Cell Carcinoma on Ear]

Most cancers of the ear are squamous cell carcinomas. Other types of cancer include:

- Basal cell cancer
- Melanoma
- Adenoid cystic carcinoma
- Adenocarcinoma
- Merkel-cell carcinoma

(Cancer Research UK).

Incidence of Cancer of the Ear in South Africa
The National Cancer Registry (2012) does not provide any information regarding the incidence of cancer of the ear in South Africa.

Cause and Symptoms of Cancer of the Ear
The cause of cancers in the middle ear is unknown. People with a history of chronic ear infections have a higher risk of developing cancer in the ear. Chronic, in this instance, means for 10 years or more.

The symptoms of cancer of the ear depend on where the tumour is within the ear. Some people may also have swollen lymph nodes in their neck.

Middle ear – the most common symptom is a discharge from the ear which may be blood stained. Other symptoms include hearing loss and earache. Occasionally people cannot move the face muscles on the side of the affected ear.

Inner ear – pain including a headache, hearing loss, tinnitus (ringing or buzzing in the ear) and dizziness.
(Cancerwise).

Diagnosis of Cancer of the Ear
The only way to confirm a diagnosis of cancer is to take a small amount of tissue from the abnormal area of the ear and examine it under a microscope. Doctors call this a biopsy. Before the doctor takes the biopsy he/she will give the patient a local anaesthetic to numb the area so that he/she will not have any pain. Biopsies of the middle ear can be difficult to take and the patient may need to have a general anaesthetic.
If the biopsy shows a cancer, the patient may also have an MRI scan or a CT scan to help the doctor decide which treatment is needed.

Doctors do not take biopsies of the inner ear. This is because it is very difficult to reach the inner ear without causing problems to other structures around it. The doctor will make a diagnosis using MRI scans and CT scans (Cancer Research UK).

Staging and Grading of Cancer of the Ear

In general, the stages of most cancers break down this way:

- **Stage 0:** Cancer has not spread.
- **Stages I, II, and III:** Cancer has grown or has spread into nearby tissues and perhaps lymph nodes. The higher the stage, the farther the cancer has spread.
- **Stage IV:** Cancer has spread beyond the lymph nodes into other parts of the body (metastasised).

Although there are several methods of staging, most doctors now use the TNM method. The TNM method is based on the size of the tumour (**T**), the spread of the cancer into nearby lymph nodes (**N**), and the spread of the cancer to other body parts (**M**, for metastasis).

T (Tumour)	N 9Lymph Nodes)	M (Metastasis)
TX: Unable to measure tumour. **T0:** No evidence of tumour. **Tis:** Tumour has not grown into nearby tissue. **T1** to **T4:** Tumour has grown into nearby tissue (numbers 1–4 describe how much the tumour has grown).	**NX:** Unable to evaluate lymph nodes. **N0:** No cancer found in lymph nodes. **N1** to **N3:** Cancer has spread into lymph nodes. (Numbers 1–3 are based on how many nodes are involved and how much cancer is found in them.)	**M0:** Cancer has not spread to other parts of the body. **M1:** Cancer has spread to other parts of the body.

(Peoria Sinus, Allergy & Hearing Center of Excellence).

Treatment of Cancer of the Ear

The treatment one has for cancer of the ear depends on:
Where in the ear the cancer is
The type of cancer one has
The size of the tumour
Whether it has spread beyond the area it started in (the stage)
One's general health

The main treatments for cancers that start in the ear canal or middle ear are surgery and radiotherapy. Depending on the stage of the cancer the patient may also have chemotherapy. The treatment the patient has depends on:

- Where in the ear the cancer is
- The type of cancer
- The size of the tumour

- Whether it has spread to
- The general health of the patient

People who have cancers that start in the head and neck usually see a team of specialist doctors and other health professionals. They include:

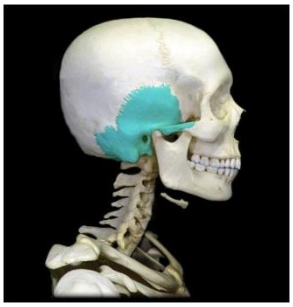
- Head and neck surgeons – including ear, nose and throat surgeons, mouth and facial bone surgeons, and plastic surgeons
- Specialists in cancer drugs and radiotherapy – oncologists
- Dentists
- Specialist nurses, physiotherapists and dieticians

Surgery - the type and amount of surgery a patient needs depends on where the cancer is in the ear and whether it has spread into any of the surrounding tissues, or into nearby structures, such as the bone.

The surgeon will remove the tumour together with an area of tissue surrounding it that is completely free of cancer cells. This is called a clear margin of tissue. It needs to be at least 5mm all-round the cancer. Doing this helps to lower the risk of the cancer coming back.

Surgery may involve having some or all of the following removed:

- The ear canal
- Part or all of the temporal bone
- The middle ear
- The inner ear



[Picture Credit: Temporal Bone]

The temporal bone is the bone at the side of the skull, by the ear. The operation to remove the temporal bone is called a mastoidectomy or temporal bone resection.

Rarely, the surgeon may need to remove the facial nerve. This runs down the side of the face and through the salivary gland. They may also need to remove the lymph nodes nearby in the neck and the salivary gland on that side of the head.

Radiotherapy - radiotherapy uses high energy rays to treat cancer. The patient may have radiotherapy as the main treatment or may have it if the surgeon has not been able to remove a clear margin of tissue from around the tumour. Then radiotherapy can lower the risk of the cancer coming back.

The patient usually has radiotherapy every day (from Monday to Friday) for between 4 and 7 weeks. At the first appointment the radiotherapy doctor (radiation oncologist) plans the treatment. This planning appointment takes a couple of hours but after that each treatment only takes a few minutes.

Chemotherapy - chemotherapy uses anti-cancer (cytotoxic) drugs to destroy cancer cells. Chemotherapy on its own will not cure cancer of the ear but doctors may use it to relieve symptoms if the cancer comes back or when the patient cannot have other treatments.

To help cure ear cancer, researchers have been looking into giving chemotherapy with radiotherapy before or after surgery. More research is needed to find out how well this works and when it is best to have chemotherapy.

The chemotherapy drugs one may have include fluorouracil and cisplatin.
(Cancer Research UK).

Follow-up and Coping with Cancer of the Ear
The patient will have regular check-ups once the treatment has finished. The doctor will examine the affected ear and ask about one's general health. This is the chance to ask any questions one may have and to tell the doctor if anything is causing concern. How often one has check-ups will vary, depending on the situation. They usually start off every 2 or 3 months and become less often as time goes on.

Coping with a diagnosis of cancer can be difficult, both practically and emotionally. It can be especially difficult if one has a rare cancer. Being well informed about the cancer and its treatment can make it easier to make decisions and cope with what happens.

It can also help to talk to other people who have the same condition. But it can be hard to find people who have had a similar rare type of cancer.
(MD Anderson Cancer Center; Cancer Research UK).

About Clinical Trials
Clinical trials are research studies that involve people. These studies test new ways to prevent, detect, diagnose, or treat diseases. People who take part in cancer clinical trials have an opportunity to contribute to scientists' knowledge about cancer and to help in the development of improved cancer treatments. They also receive state-of-the-art care from cancer experts.

Types of Clinical Trials
Cancer clinical trials differ according to their primary purpose. They include the following types:

Treatment - these trials test the effectiveness of new treatments or new ways of using current treatments in people who have cancer. The treatments tested may include new drugs or new combinations of currently used drugs, new surgery or radiation therapy techniques, and vaccines or other treatments that stimulate a person's immune system to fight cancer. Combinations of different treatment types may also be tested in these trials.

Prevention - these trials test new interventions that may lower the risk of developing certain types of cancer. Most cancer prevention trials involve healthy people who have not had cancer; however, they often only include people who have a higher than average risk of developing a specific type of cancer. Some cancer prevention trials involve people who have had cancer in the past; these trials test interventions that may help prevent the return (recurrence) of the original cancer or reduce the chance of developing a new type of cancer

Screening - these trials test new ways of finding cancer early. When cancer is found early, it may be easier to treat and there may be a better chance of long-term survival. Cancer

screening trials usually involve people who do not have any signs or symptoms of cancer. However, participation in these trials is often limited to people who have a higher than average risk of developing a certain type of cancer because they have a family history of that type of cancer or they have a history of exposure to cancer-causing substances (e.g., cigarette smoke).

Diagnostic - these trials study new tests or procedures that may help identify, or diagnose, cancer more accurately. Diagnostic trials usually involve people who have some signs or symptoms of cancer.

Quality of life or supportive care - these trials focus on the comfort and quality of life of cancer patients and cancer survivors. New ways to decrease the number or severity of side effects of cancer or its treatment are often studied in these trials. How a specific type of cancer or its treatment affects a person's everyday life may also be studied.

Where Clinical Trials are Conducted

Cancer clinical trials take place in cities and towns in doctors' offices, cancer centres and other medical centres, community hospitals and clinics. A single trial may take place at one or two specialised medical centres only or at hundreds of offices, hospitals, and centres.

Each clinical trial is managed by a research team that can include doctors, nurses, research assistants, data analysts, and other specialists. The research team works closely with other health professionals, including other doctors and nurses, laboratory technicians, pharmacists, dieticians, and social workers, to provide medical and supportive care to people who take part in a clinical trial.

Research Team

The research team closely monitors the health of people taking part in the clinical trial and gives them specific instructions when necessary. To ensure the reliability of the trial's results, it is important for the participants to follow the research team's instructions. The instructions may include keeping logs or answering questionnaires. The research team may also seek to contact the participants regularly after the trial ends to get updates on their health.

Clinical Trial Protocol

Every clinical trial has a protocol, or action plan, that describes what will be done in the trial, how the trial will be conducted, and why each part of the trial is necessary. The protocol also includes guidelines for who can and cannot participate in the trial. These guidelines, called eligibility criteria, describe the characteristics that all interested people must have before they can take part in the trial. Eligibility criteria can include age, sex, medical history, and current health status. Eligibility criteria for cancer treatment trials often include the type and stage of cancer, as well as the type(s) of cancer treatment already received.

Enrolling people who have similar characteristics helps ensure that the outcome of a trial is due to the intervention being tested and not to other factors. In this way, eligibility criteria help researchers obtain the most accurate and meaningful results possible.

National and International Regulations

National and international regulations and policies have been developed to help ensure that research involving people is conducted according to strict scientific and ethical principles. In these regulations and policies, people who participate in research are usually referred to as "human subjects."

Informed Consent

Informed consent is a process through which people learn the important facts about a clinical trial to help them decide whether or not to take part in it, and continue to learn new information about the trial that helps them decide whether or not to continue participating in it.

During the first part of the informed consent process, people are given detailed information about a trial, including information about the purpose of the trial, the tests and other procedures that will be required, and the possible benefits and harms of taking part in the trial. Besides talking with a doctor or nurse, potential trial participants are given a form, called an informed consent form, that provides information about the trial in writing. People who agree to take part in the trial are asked to sign the form. However, signing this form does not mean that a person must remain in the trial. Anyone can choose to leave a trial at any time—either before it starts or at any time during the trial or during the follow-up period. It is important for people who decide to leave a trial to get information from the research team about how to leave the trial safely.

The informed consent process continues throughout a trial. If new benefits, risks, or side effects are discovered during the course of a trial, the researchers must inform the participants so they can decide whether or not they want to continue to take part in the trial. In some cases, participants who want to continue to take part in a trial may be asked to sign a new informed consent form.

New interventions are often studied in a stepwise fashion, with each step representing a different "phase" in the clinical research process. The following phases are used for cancer treatment trials:

Phases of a Clinical Trial

Phase 0. These trials represent the earliest step in testing new treatments in humans. In a phase 0 trial, a very small dose of a chemical or biologic agent is given to a small number of people (approximately 10-15) to gather preliminary information about how the agent is processed by the body (pharmacokinetics) and how the agent affects the body (pharmacodynamics). Because the agents are given in such small amounts, no information is obtained about their safety or effectiveness in treating cancer. Phase 0 trials are also called micro-dosing studies, exploratory Investigational New Drug (IND) trials, or early phase I trials. The people who take part in these trials usually have advanced disease, and no known, effective treatment options are available to them.

Phase I (also called phase 1). These trials are conducted mainly to evaluate the safety of chemical or biologic agents or other types of interventions (e.g., a new radiation therapy technique). They help determine the maximum dose that can be given safely (also known as the maximum tolerated dose) and whether an intervention causes harmful side effects. Phase I trials enrol small numbers of people (20 or more) who have advanced cancer that cannot be treated effectively with standard (usual) treatments or for which no standard

treatment exists. Although evaluating the effectiveness of interventions is not a primary goal of these trials, doctors do look for evidence that the interventions might be useful as treatments.

Phase II (also called phase 2). These trials test the effectiveness of interventions in people who have a specific type of cancer or related cancers. They also continue to look at the safety of interventions. Phase II trials usually enrol fewer than 100 people but may include as many as 300. The people who participate in phase II trials may or may not have been treated previously with standard therapy for their type of cancer. If a person has been treated previously, their eligibility to participate in a specific trial may depend on the type and amount of prior treatment they received. Although phase II trials can give some indication of whether or not an intervention works, they are almost never designed to show whether an intervention is better than standard therapy.

Phase III (also called phase 3). These trials compare the effectiveness of a new intervention, or new use of an existing intervention, with the current standard of care (usual treatment) for a particular type of cancer. Phase III trials also examine how the side effects of the new intervention compare with those of the usual treatment. If the new intervention is more effective than the usual treatment and/or is easier to tolerate, it may become the new standard of care.

Phase III trials usually involve large groups of people (100 to several thousand), who are randomly assigned to one of two treatment groups, or “trial arms”: (1) a control group, in which everyone in the group receives usual treatment for their type of cancer, or 2) an investigational or experimental group, in which everyone in the group receives the new intervention or new use of an existing intervention. The trial participants are assigned to their individual groups by random assignment, or randomisation. Randomisation helps ensure that the groups have similar characteristics. This balance is necessary so the researchers can have confidence that any differences they observe in how the two groups respond to the treatments they receive are due to the treatments and not to other differences between the groups.

Randomisation is usually done by a computer program to ensure that human choices do not influence the assignment to groups. The trial participants cannot request to be in a particular group, and the researchers cannot influence how people are assigned to the groups. Usually, neither the participants nor their doctors know what treatment the participants are receiving.

People who participate in phase III trials may or may not have been treated previously. If they have been treated previously, their eligibility to participate in a specific trial may depend on the type and the amount of prior treatment they received.

In most cases, an intervention will move into phase III testing only after it has shown promise in phase I and phase II trials.

Phase IV (also called phase 4). These trials further evaluate the effectiveness and long-term safety of drugs or other interventions. They usually take place after a drug or intervention has been approved by the medicine regulatory office for standard use. Several hundred to several thousand people may take part in a phase IV trial. These trials are also known as post-marketing surveillance trials. They are generally sponsored by drug companies.

Sometimes clinical trial phases may be combined (e.g., phase I/II or phase II/III trials) to minimize the risks to participants and/or to allow faster development of a new intervention.

Although treatment trials are always assigned a phase, other clinical trials (e.g., screening, prevention, diagnostic, and quality-of-life trials) may not be labelled this way.

Use of Placebos

The use of placebos as comparison or "control" interventions in cancer treatment trials is rare. If a placebo is used by itself, it is because no standard treatment exists. In this case, a trial would compare the effects of a new treatment with the effects of a placebo. More often, however, placebos are given along with a standard treatment. For example, a trial might compare the effects of a standard treatment plus a new treatment with the effects of the same standard treatment plus a placebo.

Possible benefits of taking part in a clinical trial

The benefits of participating in a clinical trial include the following:

- Trial participants have access to promising new interventions that are generally not available outside of a clinical trial.
- The intervention being studied may be more effective than standard therapy. If it is more effective, trial participants may be the first to benefit from it.
- Trial participants receive regular and careful medical attention from a research team that includes doctors, nurses, and other health professionals.
- The results of the trial may help other people who need cancer treatment in the future.
- Trial participants are helping scientists learn more about cancer (e.g., how it grows, how it acts, and what influences its growth and spread).

Potential harms associated with taking part in a clinical trial

The potential harms of participating in a clinical trial include the following:

- The new intervention being studied may not be better than standard therapy, or it may have harmful side effects that doctors do not expect or that are worse than those associated with standard therapy.
- Trial participants may be required to make more visits to the doctor than they would if they were not in a clinical trial and/or may need to travel farther for those visits.

Correlative research studies, and how they are related to clinical trials

In addition to answering questions about the effectiveness of new interventions, clinical trials provide the opportunity for additional research. These additional research studies, called correlative or ancillary studies, may use blood, tumour, or other tissue specimens (also known as 'biospecimens') obtained from trial participants before, during, or after treatment. For example, the molecular characteristics of tumour specimens collected during a trial might be analysed to see if there is a relationship between the presence of a certain gene mutation or the amount of a specific protein and how trial participants responded to the treatment they received. Information obtained from these types of studies could lead to more accurate predictions about how individual patients will respond to certain cancer treatments, improved ways of finding cancer earlier, new methods of identifying people who have an increased risk of cancer, and new approaches to try to prevent cancer.

Clinical trial participants must give their permission before biospecimens obtained from them can be used for research purposes.

When a clinical trial is over

After a clinical trial is completed, the researchers look carefully at the data collected during the trial to understand the meaning of the findings and to plan further research. After a phase I or phase II trial, the researchers decide whether or not to move on to the next phase or stop testing the intervention because it was not safe or effective. When a phase III trial is completed, the researchers analyse the data to determine whether the results have medical importance and, if so, whether the tested intervention could become the new standard of care.

The results of clinical trials are often published in peer-reviewed scientific journals. Peer review is a process by which cancer research experts not associated with a trial review the study report before it is published to make sure that the data are sound, the data analysis was performed correctly, and the conclusions are appropriate. If the results are particularly important, they may be reported by the media and discussed at a scientific meeting and by patient advocacy groups before they are published in a journal. Once a new intervention has proven safe and effective in a clinical trial, it may become a new standard of care.
(National Cancer Institute).

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