About Salivary Gland Cancer

Overview and Types

If you have been diagnosed with salivary gland cancer or are worried about it, you likely have a lot of questions. Learning some basics is a good place to start.

- What Is Salivary Gland Cancer?

Research and Statistics

See the latest estimates for new cases of salivary gland cancers in the US and what research is currently being done.

- What Are the Key Statistics About Salivary Gland Cancer?
- What's New in Salivary Gland Cancer Research and Treatment?

What Is Salivary Gland Cancer?

Salivary gland cancer starts in one of the salivary glands. It's not just a single disease. There are actually several different salivary glands found inside and near your mouth. Many types of cancer and benign (non-cancerous) tumors can develop in these glands.

About the salivary glands

Salivary glands make saliva – the lubricating fluid found in the mouth and throat. Saliva contains enzymes that begin the process of digesting food. It also contains antibodies and other substances that help prevent infections of the mouth and throat.

The 2 main types of salivary glands are the major salivary glands and minor salivary glands.
There are 3 sets of **major salivary glands** on each side of the face:

- The **parotid glands**, the largest salivary glands, are just in front of the ears. About 7 out of 10 salivary gland tumors start here. Most of these tumors are benign (not cancer), but the parotid glands still are where most malignant (cancerous) salivary gland tumors start.
- The **submandibular glands** are smaller and are below the jaw. They secrete saliva under the tongue. About 1 or 2 out of 10 tumors start in these glands, and about half of these tumors are cancer.
- The **sublingual glands**, which are the smallest, are under the floor of the mouth and below either side of the tongue. Tumors starting in these glands are rare.

There are also several hundred **minor salivary glands** that are too small to see without a microscope. These glands are beneath the lining of the lips, tongue, in the roof of the mouth, and inside the cheeks, nose, sinuses, and larynx (voice box). Tumors in these glands are uncommon, but they are more often cancerous than benign. Cancers of the minor salivary glands most often start in the roof of the mouth.

**Benign salivary gland tumors**

Most salivary gland tumors are benign – that is, they are not cancer and will not spread to other parts of the body. These tumors are almost never life threatening.

There are many types of benign salivary gland tumors, with names such as adenomas,
oncocytomas, Warthin tumors, and benign mixed tumors (also known as *pleomorphic adenomas*).

Benign tumors are almost always cured by surgery. Very rarely, they may become cancer if left untreated for a long time or if they are not completely removed and grow back. It’s not clear exactly how benign tumors become cancers.

**Only salivary gland cancers will be discussed further in this document.**

**Salivary gland cancers (malignant salivary gland tumors)**

There are many types of salivary gland cancers. Normal salivary glands are made up of several different types of cells, and tumors can start in any of these cell types. Salivary gland cancers are named according to which of these cell types they most look like when seen under a microscope. The main types of cancers are described below.

Doctors usually give salivary cancers a grade (from 1 to 3, or from low to high), based on how abnormal the cancers look under a microscope. The grade gives a rough idea of how quickly it is likely to grow and spread.

- **Grade 1** cancers (also called *low grade* or *well differentiated*) look very much like normal salivary gland cells. They tend to grow slowly and have a good outcome (prognosis).
- **Grade 2** cancers (also called intermediate grade or moderately differentiated) have an appearance and outlook that is between grade 1 and grade 3 cancers.
- **Grade 3** cancers (also called *high grade* or *poorly differentiated*) look very different from normal cells and often grow and/or spread quickly. The outlook for these cancers is usually not as good as for lower grade cancers.

**Mucoepidermoid carcinoma**

Mucoepidermoid carcinomas are the most common type of salivary gland cancer. Most start in the parotid glands. They develop less often in the submandibular glands or in minor salivary glands inside the mouth. These cancers are usually low grade, but they can also be intermediate or high grade. Low-grade tumors have a much better prognosis than high-grade ones.

**Adenoid cystic carcinoma**
Adenoid cystic carcinoma is usually slow growing and often appears to be low-grade when looked at under the microscope. Still, it's very hard to get rid of completely because it tends to spread along nerves. These tumors tend to come back after treatment (generally surgery and radiation), sometimes many years later. The outlook for patients is better for smaller tumors.

**Adenocarcinomas**

*Adenocarcinoma* is a term used to describe cancers that start in gland cells (cells that normally secrete a substance). There are many types of salivary gland adenocarcinomas.

**Acinic cell carcinoma:** Most acinic cell carcinomas start in the parotid gland. They tend to be slow growing and tend to occur at a younger age than most other salivary gland cancers. They are usually low grade, but how far they have grown into nearby tissue is probably a better predictor of a patient’s prognosis (outlook).

**Polymorphous low-grade adenocarcinoma (PLGA):** These tumors tend to start in the minor salivary glands. They usually (but not always) grow slowly and are mostly curable.

**Adenocarcinoma, not otherwise specified (NOS):** When seen under a microscope, these cancers have enough features to tell that they are adenocarcinomas, but not enough detail to classify them further. They are most common in the parotid glands and the minor salivary glands. These tumors can be any grade.

**Rare adenocarcinomas:** Several types of adenocarcinoma are quite rare. Some of these tend to be low grade and usually have a very good outcome:

- Basal cell adenocarcinoma
- Clear cell carcinoma
- Cystadenocarcinoma
- Sebaceous adenocarcinoma
- Sebaceous lymphadenocarcinoma
- Mucinous adenocarcinoma

Other rare adenocarcinomas are more likely to be high grade and may have a less favorable outcome:

- Oncocytic carcinoma
• Salivary duct carcinoma

Malignant mixed tumors

There are 3 types of malignant mixed tumors:

• Carcinoma ex pleomorphic adenoma
• Carcinosarcoma
• Metastasizing mixed tumor

Nearly all of these cancers are carcinoma ex pleomorphic adenomas. The other 2 types are very, very rare.

Carcinoma ex pleomorphic adenoma develops from a benign mixed tumor (also known as a pleomorphic adenoma). This tumor occurs mainly in the major salivary glands. Both the grade of the cancer and how far it has spread (its stage) are important in predicting outcome.

Other rare salivary gland cancers

Several other types of cancer can develop in the salivary glands.

Squamous cell carcinoma: This cancer occurs mainly in older men. It can develop after radiation therapy for other cancers in the area. This type of cancer tends to have a poorer outlook.

Epithelial-myoepithelial carcinoma: This rare tumor tends to be low grade, but it can come back after treatment or spread to other parts of the body.

Anaplastic small cell carcinoma: The cells in these tumors have nerve cell-like features. These tumors are most often found in minor salivary glands and tend to grow quickly.

Undifferentiated carcinomas: This group of cancers includes small cell undifferentiated carcinoma, large cell undifferentiated carcinoma, and lymphoepithelial carcinoma. These are high-grade cancers that often spread. Overall, the survival outlook tends to be poor. Lymphoepithelial carcinoma, which is much more common in Eskimo and Inuit people, has a slightly better outcome.

Other cancers that can affect the salivary glands
These types of cancer are typically not thought of as true salivary gland cancers, either because they start more often in other parts of the body, or because they start elsewhere and then grow into or spread to the salivary glands.

**Non-Hodgkin lymphoma:** Most non-Hodgkin lymphomas start in lymph nodes. Rarely, these cancers start in immune system cells within the salivary glands. They behave and are treated differently from other types of cancers in the salivary glands. Most lymphomas that start in the salivary glands affect people with Sjogren (Sjögren) syndrome (a disorder that causes the immune system to attack salivary gland cells). For more information on lymphomas, see Non-Hodgkin Lymphoma.

**Sarcomas:** The salivary glands contain blood vessels, muscle cells, and cells that make connective tissue. Cancers that start in these types of cells are called sarcomas. These rarely occur in the salivary gland. For more information on sarcomas, see Sarcoma—Adult Soft Tissue Cancer.

**Secondary salivary gland cancers:** Cancers that start elsewhere and spread to the salivary glands are called secondary salivary gland cancers. These cancers are treated based on where the cancer started.

- References
  See all references for Salivary Gland Cancer

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**What Are the Key Statistics About Salivary Gland Cancer?**

Salivary gland cancers are not very common, making up less than 1% of cancers in the United States. They occur at a rate of about 1 case per 100,000 people per year in the United States.

These cancers can occur in people of almost any age, but they become more common as people get older. The average age at the time of diagnosis is 64.
Overall, about 72% of people diagnosed with salivary gland cancer are still alive at least 5 years after being diagnosed. (This includes people with all types and stages of salivary gland cancer, but the outlook for some people might be better or worse than this.) For more statistics related to survival, see Survival Rates for Salivary Gland Cancer by Stage.

- References

See all references for Salivary Gland Cancer

What’s New in Salivary Gland Cancer Research and Treatment?

Medical centers throughout the world are researching the causes and treatment of salivary gland cancer. This is a challenging disease to study because it’s not common and there are many types of salivary gland cancer. But each year, scientists find out more about what causes the disease and how to better treat it.

Biology of salivary gland cancers

Recent studies have found certain changes in chromosomes and genes of various types of salivary gland cancers. Researchers are learning more about how these changes can cause salivary gland cells to become cancerous. In some salivary gland cancer cells, 2 chromosomes have swapped parts of their DNA, which is known as a translocation. These changes often activate genes that affect cell growth. For example, adenoid cystic carcinomas often have translocations between chromosomes 6 and 9, and mucoepidermoid carcinomas often have translocations between chromosomes 11 and 19. The exact genes involved in these translocations are now being studied.

As scientists learn more about these and other changes in salivary gland cancer cells, they hope to use this information to develop new treatments that work better and cause fewer side effects.
Treatment

Surgery

Advances in surgical techniques now allow teams of head and neck surgeons and neurosurgeons to remove cancers that have spread near the base of the skull. These operations were not thought possible a few years ago but are becoming more common and successful.

Reconstructive surgery is becoming more sophisticated and successful, too. This lets surgeons do more extensive surgery to remove the cancer and improves patients’ quality of life after treatment.

Radiation therapy

Advances in radiation therapy now permit more precise targeting of radiation. Some types of radiation, such as fast neutron beam radiation, have been found to be particularly useful, but they require specialized equipment that’s not available in many hospitals.

Chemotherapy

Advanced salivary gland cancer is rare, so knowledge about treating these cancers with chemotherapy (chemo) is still evolving. Chemo drugs such as gemcitabine, capecitabine, and oxaliplatin are now being tested in clinical trials and may provide more options for people with advanced salivary gland cancer.

Targeted therapy

As researchers have learned more about the changes in cells that cause cancer, they have been able to develop newer drugs that specifically target these changes. These targeted drugs work differently from standard chemotherapy drugs. They sometimes work when chemo drugs don’t, and they often have different (and less severe) side effects.

Studies have identified changes in several proteins in salivary gland cancer cells that help these cancers grow and spread. Some of these changes affect proteins that can be blocked by targeted therapies that are already used to treat other types of cancer. Clinical trials are now looking at whether these targeted therapies can be helpful for
salivary gland cancers.

- **References**
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