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About Nasopharyngeal Cancer

Get an overview of nasopharyngeal cancer and the latest key statistics and research in the US.

Overview and Types

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

What Is Nasopharyngeal Cancer?

Research and Statistics

See the latest estimates for new cases of nasopharyngeal cancer in the US and what research is currently being done.

- Key Statistics for Nasopharyngeal Cancer
- What's New in Nasopharyngeal Cancer Research?

What Is Nasopharyngeal Cancer?

- The nasopharynx
- Types of nasopharyngeal tumors

Nasopharyngeal cancer is a type of head and neck cancer. Head and neck cancers can have many different names depending on where the cancer starts. Cancer starts when cells in the body begin to grow out of control.

Nasopharyngeal cancer starts in the nasopharynx, the upper part of the throat behind the nose and near the base of skull.

Since the head and neck area has so many organs in a small place, knowing what type of head and neck cancer you have can be confusing. Ask your doctor to write down the exact kind of cancer you have and show you where it is on a picture. To learn more

Types of nasopharyngeal tumors

Several types of tumors can develop in the nasopharynx. Some of these tumors are benign (not cancer), and others are malignant (cancer). It's important to talk with your doctor about what type of tumor you might have.

Nasopharyngeal carcinoma (NPC)

The most common cancer in the nasopharynx is nasopharyngeal carcinoma (NPC).

Carcinoma is cancer that starts in the cells that line the surfaces of organs in the body, called epithelial cells.

There are different types of NPC. They all start from epithelial cells that line the nasopharynx, but the cells of each type look different when looked at closely in the lab:

- **Keratinizing squamous cell carcinoma** is the most common type in places with low rates of NPC, like the US.
- Non-keratinizing differentiated carcinoma is less common in areas with high rates of NPC and is often associated with the Epstein-Barr Virus (EBV).
- Non-keratinizing undifferentiated carcinoma is the most common type in areas with high rates of NPC and is often associated with EBV.
- Basaloid squamous cell carcinoma is rare and very aggressive.

The treatment is the same for all types of NPC, but the <u>stage of the cancer</u>⁴ (how far it has grown and spread) is often more important than the type in predicting a person's outlook (prognosis). Research is being done to evaluate using EBV DNA levels to also help predict a person's outlook (prognosis), when the NPC is linked to EBV.

Other cancers in the nasopharynx

Other types of cancers can also be found in the nasopharynx:

- About 5% of cancers that start in the nasopharynx are lymphomas They are cancers of immune system cells called lymphocytes, which are found throughout the body, including in the nasopharynx. See <u>Non-Hodgkin Lymphoma</u>⁵ to learn more.
- Sarcomas and melanomas make up less than 5% of cancers that start in the nasopharynx. These cancers don't start in epithelial cells like the other types of NPCs. See <u>Soft Tissue Sarcoma</u>⁶ and <u>Melanoma Skin Cancer</u>⁷ for more about these cancers.

Benign nasopharyngeal tumors

Benign nasopharyngeal tumors are fairly rare and tend to develop in children and young adults. These tumors do not spread to other parts of the body and are usually not life-threatening. They include tumors or malformations of the vascular (blood-carrying) system, such as **angiofibromas**and **hemangiomas**, and benign tumors of minor

salivary glands within the nasopharynx.

Benign nasopharyngeal tumors don't always need treatment. When they do, the treatment is not the same as for nasopharyngeal cancer. If you have a benign tumor, ask your doctor what to expect.

Hyperlinks

- 1. www.cancer.org/cancer/understanding-cancer/what-is-cancer.html
- 2. www.cancer.org/cancer/types/oral-cavity-and-oropharyngeal-cancer.html
- 3. www.cancer.org/cancer/types/laryngeal-and-hypopharyngeal-cancer.html
- 4. www.cancer.org/cancer/types/nasopharyngeal-cancer/detection-diagnosis-staging/staging.html
- 5. www.cancer.org/cancer/types/non-hodgkin-lymphoma.html
- 6. www.cancer.org/cancer/types/soft-tissue-sarcoma.html
- 7. www.cancer.org/cancer/types/melanoma-skin-cancer.html

References

Chang ET, Ye W, Zeng YX, Adami HO. The Evolving Epidemiology of Nasopharyngeal Carcinoma. *Cancer Epidemiol Biomarkers Prev.* 2021;30(6):1035-1047. doi:10.1158/1055-9965.EPI-20-1702.

Leeman JE, Katabi N, Wong RJ, Lee NY and Romesser PB. Ch. 65 - Cancer of the Head and Neck. In: Niederhuber JE, Armitage JO, Doroshow JH, Kastan MB, Tepper JE, eds. *Abeloff's Clinical Oncology*. 6th ed. Philadelphia, Pa. Elsevier; 2020.

Mohanty S, Gopinath M, Subramanian M. Benign tumours of nasopharynxrevisited. *Indian J Otolaryngol Head Neck Surg.* 2013;65(Suppl 1):22-25. doi:10.1007/s12070-011-0384-6.

Thompson L. World Health Organization classification of tumours: pathology and genetics of head and neck tumours. *Ear Nose Throat J.* 2006;85(2):74.

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Key Statistics for Nasopharyngeal Cancer

Nasopharyngeal cancer (NPC) is rare. In most parts of the world (including the United States), there's less than one case for every 100,000 people each year.

This cancer is much more common, however, in certain parts of South Asia, the Middle East, and North Africa. In some parts of China there are as many as 25 to 30 cases per 100,000 men and 15 to 20 cases per 100,000 women. It's also more common among native people in the Arctic.

The risk of NPC increases slowly throughout life, but it can occur in people of any age, including children. In areas where the risk of NPC is high (such as China), the peak age of people being diagnosed is typically between 45 and 59. In areas of low risk of NPC (such as the US), cases can often be seen in young adulthood (ages 15 to 24) followed by a decline in cases until another peak at older ages of 65 to 79.

Overall, cases of NPC have been declining steadily over the past decades, including in high-risk areas. It is thought that this drop might be due to environmental and lifestyle changes.

Men are 2 to 3 times more likely to develop NPC than women.

References

Chang ET, Ye W, Zeng YX, Adami HO. The Evolving Epidemiology of Nasopharyngeal Carcinoma. *Cancer Epidemiol Biomarkers Prev.* 2021;30(6):1035-1047. doi:10.1158/1055-9965.EPI-20-1702.

Chen YP, Chan ATC, Le QT, Blanchard P, Sun Y, Ma J. Nasopharyngeal carcinoma. *Lancet*. 2019;394(10192):64-80. doi:10.1016/S0140-6736(19)30956-0.

Shah AB, Zulfiqar H, Nagalli S. Nasopharyngeal Carcinoma. [Updated 2022 Jan 7]. In: StatPearls [Internet]. Treasure Island (FL): StatPearls Publishing; 2022 Jan-. Accessed at https://www.ncbi.nlm.nih.gov/books/NBK554588/ on May 4, 2022.

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What's New in Nasopharyngeal Cancer Research?

- Causes, prevention, and early detection
- Treatment

Research into the causes, prevention, and treatment of nasopharyngeal cancer (NPC) is being done at many university hospitals, medical centers, and other institutions around the world. Because NPC is rare, it has been hard to study it well. Most experts agree that treatment in a <u>clinical trial</u>¹ should be considered for any type or stage of NPC.

Causes, prevention, and early detection

Many studies are looking at how Epstein-Barr virus (EBV) infection as well as other risk factors cause cells of the nasopharynx to become cancer. These studies may someday lead to vaccines to help prevent some cases of NPC by preventing EBV infection.

Discoveries about EBV, its interaction with nasopharyngeal cells, and the immune system's reaction to EBV have led to new blood tests, such as measuring levels of EBV DNA, that is helping to detect NPC early and being studied as a way to screen people at high risk. Researchers are also finding certain gene changes in the EBV that can make a person more likely to develop NPC . These questions are now being studied in areas of the world where this cancer is more common.

Treatment

Epstein-Barr virus (EBV) DNA

The levels of EBV DNA in the blood are being studied as a way to screen people at high risk of NPC, but are also being looked at as a way to better predict the response to treatment as well as if NPC recurs (comes back after treatment).

Some research is testing if the level of EBV DNA in the blood after chemoradiation or

radiation therapy can help guide treatment: For example, deciding if someone with high levels of EBV DNA after getting chemoradiation should get adjuvant (additional) chemotherapy.

Studies to look for other gene changes linked to NPC are being done in countries where NPC is more common.

New radiation therapy techniques

Most types of radiation therapy use radiation in the form of x-rays (photons). Other types of radiation use protons and carbon ions to kill cancer cells. These are being studied, especially in China, but so far haven't been proven to be better than standard x-ray radiation. Some studies are also investigating **proton** therapy along with **photon** therapy.

Doctors are also studying the best dose of radiation therapy, especially if someone has had a good response to induction (upfront) chemotherapy. Some studies are evaluating if a lower dose of radiation can be given if the NPC has shrunken quite a bit because chemotherapy was given first.

Chemotherapy

Researchers continue to develop new chemo drugs, new drug combinations, and new ways to give drugs that might be more effective against advanced NPC. Several drugs that are already used to treat other cancers, such as capecitabine, oxaliplatin, and gemcitabine, have been studied for use against NPC as well. Clinical trials are looking for the best combination of chemo drugs and how they should be used along with radiation therapy. Studies have shown that for more advanced stages of NPC, giving chemotherapy first, called **induction chemo**, and then chemoradiation, can help some people live longer. Other research is being done on earlier-stage NPC to determine if chemo should be given after radiation therapy, if high levels of EBV DNA are found in the blood.

Immunotherapy

NPC seems to be caused at least in part by infection with the Epstein-Barr virus (EBV). Although patients' immune systems can react against EBV, this doesn't seem to be enough to kill the cancer. Finding and targeting cell proteins linked to EBV and the cells it affects could lead to new, more cancer-focused treatment options. Understanding how EBV interacts with the immune system could also lead to treatments that boost the immune system's response to NPC and maybe even keep it from developing.

Immunotherapy drugs, like PD-1 inhibitors, are approved to treat some people with NPC after the cancer has come back (recurred) or spread (metastasized). Studies are looking to see if adding immunotherapy drugs to chemoradiation upfront might help shrink cancers more than chemoradiation alone.

Predicting treatment outcomes

Researchers are looking for ways to know how well NPC will respond to treatment and understand how likely it is to come back after treatment. This information would allow doctors to tailor treatment for each patient so that the best treatment is used. One area of research is looking for a link between a person's blood EBV DNA levels before treatment, the cancer stage, and overall survival. Results have suggested there is a link between low levels of pre-treatment EBV DNA and early-stage cancers and better outcomes, but much more research is needed. Some studies are trying to find out if using EBV DNA levels after treatment could help predict a person's outcome.

Hyperlinks

1. <u>www.cancer.org/cancer/managing-cancer/making-treatment-decisions/clinical-trials.html</u>

References

Hui EP and Chan A. Epidemiology, etiology, and diagnosis of nasopharyngeal carcinoma. In: Shah S, ed. *UpToDate*. Waltham, Mass.: UpToDate, 2022. https://www.uptodate.com. Accessed May 6, 2022.

Hui EP, Chan A, and Le Quynh-Thu. Treatment of early and locoregionally advanced nasopharyngeal carcinoma. In: Shah S, ed. *UpToDate*. Waltham, Mass.: UpToDate, 2022. https://www.uptodate.com. Accessed May 6, 2022.

Hui EP, Chan A, and Le Quynh-Thu. Treatment of recurrent and metastatic nasopharyngeal carcinoma. In: Shah S, ed. *UpToDate*. Waltham, Mass.: UpToDate, 2022. https://www.uptodate.com. Accessed May 6, 2022.

Li YQ, Khin NS, Chua MLK. The evolution of Epstein-Barr virus detection in nasopharyngeal carcinoma. *Cancer Biol Med.* 2018; 15(1): 1-5.

Outh-Gauer S, Alt M, Le Tourneau C, et al. Immunotherapy in head and neck cancers:

A new challenge for immunologists, pathologists and clinicians. Cancer Treat Rev.