

Nasopharynx

a. Anatomy

- i. The nasopharynx is the upper most portion of the throat and connects the nasal cavity to the oropharynx. It contains the adenoids and the eustachian tube openings.
- ii. Squamous cells make up the lining, also known as the mucosa, of the nasopharynx. The adenoids are lymphoid tissue. Therefore, cancers such as squamous cell carcinoma and lymphoma can occur in the nasopharynx.

b. Risk Factors

- i. Some nasopharynx cancers are caused by the Epstein- Barr virus
- ii. Cooking salt-cured fish and meat releases a chemical called nitrosamine, a chemical known to increase the risk of nasopharynx cancer
- iii. This type of cancer occurs more frequently in Southeast Asia and China

c. Symptoms

- i. Patients commonly present with a painless neck mass
- ii. Nasal congestion or ear congestion, hearing loss
- iii. Some people could present with bloody discharge from their nose, or report worsening trouble breathing through the nose.

d. Diagnosis

- i. Once a suspicious growth or lesion is identified within the nasopharynx a biopsy of the suspected area will be required to confirm the diagnosis of nasopharyngeal carcinoma (NPC). Due to the location of the cancer within the back of the throat and nasal cavity; a biopsy at your doctor's office using both topical and local anesthesia may be difficult. To effectively biopsy and evaluate the extent of the nasopharyngeal cancer, you may require biopsy under general anesthesia in the operating room. Prior to scheduling your biopsy under general anesthesia, a complete head and neck exam including possible in-office fiberoptic nasal camera exam may be performed.
- ii. Patients presenting with a neck mass may undergo a needle biopsy in the office with or without ultrasound guidance.
- iii. Your doctor may also order a variety of medical imaging:
 1. Computerized tomogram imaging (CT scan)
 2. Magnetic resonance imaging (MRI)
 3. Positron emission tomography (PET)
 4. Dental imaging (cone beam scan or Panorex).
- iv. These images will help to further determine the extent of the cancer. Different types of imaging help to visualize different anatomic structures. For example, CT scans and dental imaging are particularly useful for visualizing the integrity of the jaw bones. Intravenous contrast enhanced CT scan and MRI can identify abnormalities within tongue, tonsil, palate, throat or neck tissues. PET imaging is a special technology that identifies a glucose (sugar) molecule being absorbed by the cancer cells and help determine whether the tumor has spread to other parts of the body.

Alternatively, CT scan imaging of the chest may be used to visualize any spread of disease to the lungs. Dental imaging may also help identify disease within the jaw bones and assess the health of the teeth in the event that radiation therapy is required.

e. Staging

- i. The American Joint Committee on Cancer (AJCC) has created a staging system (TNM staging) to help guide treatment. The staging system groups cancers into stages based on their anatomic extent. The staging helps to determine the optimal treatment for a specific cancer, and provides information about expected survival rates.
- ii. The staging system includes information about the extent of the original or local tumor (T classification or primary tumor extent), spread to lymph nodes (N-classification or nodal metastases), and spread to distant parts of the body (M classification or distant metastases). Combinations of T, N and M-classifications produce an overall stage. There are four stages (Stage 1-4) reported with increasing disease burden and potentially worse survival as the AJCC stage number increases. Factors that play a role in tumor stage include: destruction of adjacent structures (ie. nasal cavity, brain, orbit/eye, spine or sinuses), involvement of lymph nodes within the neck and distant metastasis.
- iii. Epstein-Barr virus (EBV) DNA testing within the tumor and blood plasma may be performed. In the United States, a minority of nasopharynx cancers are caused by EBV.
- iv. Lymph nodes are small oval shaped structures found within the fat of the neck that harbor specialized immune cells that filter and fight infection and disease. Cancer cells from the nasopharynx detach from the primary tumor and become trapped within the individual nodes where they grow. The number, location and size of lymph nodes affect the tumor stage.

Primary Tumor Stage: Nasopharynx	
Stage	Description
TX	Primary tumor cannot be assessed
T0	No primary tumor identified, but EBV+ lymph node involvement
Tis	Carcinoma <i>in situ</i>
T1	Tumor confined to the nasopharynx, or extension to the oropharynx and/or nasal cavity without parapharyngeal involvement
T2	Tumor with extension to the parapharyngeal space, and/or adjacent soft tissue involvement (medial pterygoid, lateral pterygoid, prevertebral muscles)
T3	Tumor with infiltration of bony structures at skull base, cervical vertebra, pterygoid plates, and/or paranasal sinuses
T4	Tumor with intracranial extension, involvement of cranial nerves, hypopharynx, orbit, parotid gland, and/or extensive soft tissue infiltration beyond the lateral surface of the lateral pterygoid muscle

Regional Nodal Stage: Nasopharynx	
Stage	Description
Nx	Regional lymph nodes cannot be assessed
N0	No regional lymph node metastasis
N1	Unilateral metastasis in cervical lymph node and/or unilateral or bilateral metastasis in retropharyngeal lymph nodes above the lower border of the cricoid cartilage, <= 6 cm
N2	Bilateral metastasis in cervical lymph nodes above the lower border of the cricoid cartilage, <= 6 cm
N3	Unilateral or bilateral metastasis in cervical lymph nodes, > 6 cm, and/or extension below the lower border of the cricoid cartilage

AJCC Prognostic Stage Groups: Nasopharynx	
Stage	Description
0	TisN0M0
I	T1N0M0
II	T0N1M0, T1N1M0, or T2N1M0
III	T0N2M0, T1N2M0, T2N2M0, T3N0M0, T3N1M0, or T3N2M0
IVA	T4N0M0, T4N1M0, T4N2M0, T0N3M0, T1N3M0, T2N3M0, T3N3M0, or T4N3M0
IVB	AnyT, any N, and M1

f. Treatment

- i. The best treatment for nasopharyngeal cancer depends on many factors, including the cancer stage and your general health and other medical conditions. Not everyone receives the same treatment, and you should work with a multidisciplinary team of doctors to tailor an individual treatment plan for you.
- ii. Potential treatment options include clinical trials, radiation, chemotherapy, surgery, or a combination of these.
- iii. Clinical trials study a promising treatment in people. They can allow you to access treatment options that wouldn't otherwise be available. You can ask your oncology team if there is an open clinical trial that you can consider joining.
- iv. Radiation is the main treatment for early stage nasopharyngeal cancer. This treatment uses high-energy rays or particles aimed at the cancer. There are different types of radiation, including Intensity-modulated radiation therapy (IMRT), 3-dimensional conformal radiation therapy and proton therapy. Your radiation therapist will recommend the option they believe is most appropriate for your specific situation. Proton therapy is only available at certain centers.
- v. Chemotherapy, targeted therapies and immunotherapy are drugs that are active against the cancer cells. Some drugs directly destroy cancer cells, while other drugs can stop cancer cells from growing or allow immune cells in your body to destroy the cancer. Most of these drugs are not effective alone, and usually

combined with radiation. They can also be given before or after other treatments as part of a treatment plan.

- vi. Surgery is infrequently used to treat this kind of cancer, because it usually responds to radiation and chemotherapy. Surgery is sometimes used to remove lymph nodes or recurrent cancer after failure of chemotherapy and radiation.

g. Survival

- i. The survival rate for nasopharyngeal cancer is strongly affected by initial tumor stage and patient overall health. Your oncology team can tell you about survival rates based on your cancer stage. However, it is important to remember that these numbers are based on groups of patients in the past, and they don't take into account many individual factors. The American Cancer Society gives 5-year survival rates ranging from 82%-48% depending on the cancer stage. This is based on data collected between 2009 and 2015.¹
- ii. Short term side effects of treatment are common and include skin changes, tiredness, sores in the mouth and nose, loss of appetite, trouble eating or swallowing, and changes in your sense of smell and taste. Long-term side effects can include hearing or vision loss, dental problems, low thyroid hormone levels, and damage to the carotid arteries which can increase your risk of stroke. It is important to discuss the expected and possible side effects of treatment with your care team.
- iii. American Cancer Society "Survival rates for nasopharyngeal cancer." Accessed Oct 10, 2020. <https://www.cancer.org/cancer/nasopharyngeal-cancer/detection-diagnosis-staging/survival-rates.html>

h. Surveillance/Survivorship

- i. Surveillance: As with any cancer, there is a risk that nasopharynx cancer will come back ('recur') after treatment. 'Surveillance' means that your doctor(s) will monitor you after treatment with a combination of physical examination and imaging studies in order to detect disease that has recurred early. Protocol details will vary from institution to institution. Surveillance typically lasts for at least 5 years. Once cleared by the treating physician, patients may transition into a cancer surveillance/survivorship clinic, often run by an advanced practice provider (APP) that is well trained in head and neck cancer surveillance.
 - 1. Physical examination: Your doctor(s) will examine you according to the following schedule:
 - a. Year One: every 1-3 months
 - b. Year Two: every 2-6 months
 - c. Years Three through 5: every 4-8 months
 - d. After 5 years: every 12 months or as needed.
 - 2. Imaging: The timing and type of imaging you have will be based on your doctor's judgment.
 - a. It is recommended to have baseline imaging within 6 months of completing treatment.

- b. Imaging may include CT scans, PET/CT scans, MRI, or sometimes ultrasound.
 - c. Additional imaging will be based on your symptoms, exam, and your doctor's judgment.
 - 3. Blood test: Your doctor may order a blood test to look for Epstein-Barr Virus (EBV) as part of your surveillance.
 - ii. [Survivorship](#): Nasopharynx cancer and its treatment can affect many areas of your health and quality of life. 'Survivorship' refers to caring for your health and well-being from the moment you receive your diagnosis, and for the rest of your life. Depending what your treatment involves, important parts of survivorship for nasopharynx cancer may include:
 - 1. [Speech and swallowing evaluation and therapy](#): This is typically with a Speech-Language Pathologist, or SLP, with expertise in speech and swallowing for head and neck cancer patients.
 - a. Swallowing: Safe swallowing is important for your health and quality of life. Poor swallowing function, called 'dysphagia', can lead to health problems such as pneumonia or malnutrition, and may lead to the need for a feeding tube in order to stay healthy. You may also be referred to a dietician for guidance on how to maintain a nutritious diet even if you have dysphagia.
 - b. Speech: Rehabilitation of your speaking is important for your communication and quality of life.
 - 2. Oral and dental health: Nasopharynx cancer and its treatment can have a major impact on your teeth, taste, saliva and jaw bone.
 - a. [Dental cleaning and care](#): Nasopharynx cancer patients should establish care early after diagnosis, ideally before treatment, with a dentist who has experience in head and neck cancer. Some dental work may be necessary prior to treatment. In the long term, patients should have routine cleaning and examination. Routine fluoride treatments may be recommended.
 - b. Radiation and dental health: Radiation can be detrimental to your dental health. It is especially important for patients who undergo radiation to have regular dental care and excellent dental hygiene.
 - c. [Dry mouth](#): Dry mouth, or 'xerostomia', is common after radiation therapy and can have a significant negative impact on quality of life. There is no cure for xerostomia. If you have xerostomia, you can decrease the symptoms by staying hydrated, using salivary substitutes, and maintaining excellent dental hygiene.
 - d. Trismus: Surgery and radiation can cause difficulty opening your mouth, called trismus. This can interfere with dental care and with eating. Ask your doctor about ways to improve your mouth opening, such as stretching exercises.

3. [Osteoradionecrosis \(ORN\)](#): Patients who undergo radiation are at risk for bone infection of the jaw, spine or skull. This is called osteoradionecrosis (ORN) and may cause pain, ulcerations, exposed bone, and chronic infection.
 - a. ORN is diagnosed with history, examination and imaging.
 - b. Treatment may involve medications, hyperbaric oxygen treatments, or in advanced cases, bone removal and reconstruction.
 - c. ORN of the spine after radiation for nasopharynx cancer is a serious condition. You should tell your doctor if you experience neck pain or any new weakness or numbness.
4. [Thyroid function testing](#): If you have had radiation therapy, you have an increased risk of low thyroid function, or 'hypothyroidism'. Symptoms of hypothyroidism may include fatigue, weight gain, constipation and depression. Your thyroid function should be tested every 6-12 months to determine whether you need treatment with thyroid hormone supplementation.
5. [Tobacco use](#):
 - a. Most head and neck cancers are associated with tobacco use, especially smoking cigarettes.
 - b. Continued tobacco use after treatment is associated with worse survival and higher risk of other cancers, among many other negative health effects.
 - c. Quitting tobacco at any time will improve your overall health and chances of survival from head and neck cancer.
 - d. If you still use tobacco after head and neck cancer treatment, we strongly encourage you to consider quitting. Your doctor can help you find resources, including medications and counseling programs, that have been proven to help.
6. [Lymphedema](#): Lymphedema is swelling of the soft tissue that is common for patients who have had surgery and/or radiation. Specialized Physical Therapy called Lymphedema Therapy, including massage, compression garments, exercises and skin care, is available and can significantly improve lymphedema.
7. [Shoulder dysfunction](#): Many head and neck cancer survivors have shoulder dysfunction, including decreased range of motion, weakness and stiffness as a result of surgery and/or radiation. Physical therapy is very helpful in improving shoulder function. If you have problems with your shoulder, ask your doctor about a referral to a physical therapist.
8. [Obstructive sleep apnea](#): Survivors of head and neck cancer treatment are at risk for obstructive sleep apnea (OSA) because of changes to the upper airway anatomy. Symptoms may include daytime sleepiness, snoring, gasping or choking during sleep, daytime headaches, and irritability. OSA is diagnosed with a sleep study, and there are several

options for treatment. Discuss your risk of OSA with your doctor, to decide whether you should have a sleep study.

9. [Carotid artery stenosis evaluation](#): Radiation therapy to the neck increases the risk of carotid artery narrowing (stenosis) later in life. Carotid artery stenosis increases the risk of stroke. If you have had radiation to your neck, ask your doctor about an ultrasound to look for carotid artery stenosis.
10. Mental and sexual health: Head and neck cancer and its treatment can result in [cognitive dysfunction](#), anxiety, [depression](#), [body image concerns](#), and changes in [sexual function and desire](#). If you suffer from any of these, you are not alone. Ask your doctor about meeting with a mental health professional to determine whether counseling and/or medication may be helpful for you.
11. Hearing evaluation: Head and neck cancer treatments, especially with certain chemotherapy drugs, can cause hearing loss. If you have decreased hearing, you should have a hearing test to evaluate your hearing and determine whether you may benefit from hearing augmentation, such as with a hearing aid.

i. Questions for your doctor

i. Before/during treatment:

1. What types of treatment are recommended (such as surgery, radiation, and/or chemotherapy)?
2. Are there any other treatment options that I should learn about, such as clinical trials?
3. Should I see a dentist before treatment begins?
4. How long will treatment take? How long will it take to fully recover after treatment?
5. What are the risks and side effects of each part of treatment? Which side effects are temporary, and which might be permanent?
6. Will I need a feeding tube or a breathing tube (tracheostomy)? Will they be temporary or permanent?
7. What will my swallowing, speech and breathing be like after treatment?
8. Will I have any other functional problems after treatment?
9. Will I be able to keep doing my job after I've recovered?

ii. After treatment:

1. Should I have any imaging studies?
2. Should I have a blood test for EBV?
3. When should my next follow-up appointment be, and with whom?
4. Should I have my thyroid function tested?
5. Should I be referred to a speech and language pathologist (SLP), and or to a physical or occupational therapist?
6. Am I receiving appropriate dental care?
7. Should I have a sleep study?
8. Do I need a carotid artery ultrasound?

9. Should I have a hearing test?