**PARATHYROID CANCER**

ALTERNATIVE NAMES: The alternative name for parathyroid cancer is parathyroid carcinoma.

**DEFINITION / DESCRIPTION**

Parathyroid cancer is a rare cancer that develops in the tissue of your parathyroid glands. Your parathyroid glands are usually located behind your thyroid — a butterfly-shaped gland in the front of your neck.

Healthcare professionals don’t use the four-stage naming system for parathyroid cancer that’s commonly used for other types of cancers. Instead, parathyroid cancer is categorized in one of three ways:

* Localized: Localized parathyroid cancer means the cancer is found in your parathyroid gland and may have spread to surrounding tissues.
* Metastatic: Metastatic parathyroid cancer means the cancer has spread to other parts of your body, such as your lymph nodes, liver, lungs, bone or pancreas. Approximately 10% to 30% of people who are diagnosed with parathyroid cancer will have metastatic cancer at the time of diagnosis. Most commonly, the cancer will have spread to your lung, bone or liver.
* Recurrent: Recurrent parathyroid cancer means the cancer comes back after initial treatment — typically, surgical removal of the cancerous gland. More than half of people with parathyroid cancer will experience recurrence. Usually, the cancer comes back two to five years after the first surgery, but it could take over 20 years before it returns.

***Parathyroid glands***

Most people have four pea-sized parathyroid glands located behind their thyroid gland. Like the thyroid, the parathyroid glands are part of your endocrine system.

Sometimes, your parathyroid glands can be found along your esophagus or in your chest. These are known as ectopic (in an abnormal place) parathyroid glands.

Your parathyroid glands are in charge of controlling the amount of calcium in your blood by producing parathyroid hormone (PTH). Sometimes, one or more of your parathyroid glands can release (secrete) too much parathyroid hormone. This is known as primary hyperparathyroidism, or pHPT. Too much parathyroid hormone causes the levels of calcium in your blood to rise, a condition known as hypercalcemia.

***Parathyroid cancer and parathyroid adenoma***

A parathyroid adenoma is a benign (noncancerous) growth in a parathyroid gland. Parathyroid cancer is when malignant (cancer) cells form in your parathyroid tissue. Within the scope of parathyroid disorders, parathyroid adenomas are much more common than parathyroid cancer.

Both parathyroid cancer and parathyroid adenomas often cause your parathyroid to become overactive and release too much parathyroid hormone (PTH), which causes hypercalcemia.

Hypercalcemia can be harmful to your body and health. Because of this, the most common treatment for both parathyroid cancer and parathyroid adenomas is surgical removal of your overactive parathyroid gland.

It can be challenging for healthcare professionals to tell the difference between a benign parathyroid adenoma and parathyroid cancer because their cells look similar.

Further testing is needed after the overactive parathyroid gland is surgically removed to determine if it’s an adenoma or cancer.

***What is hypercalcemia and how is it related to parathyroid cancer?***

Hypercalcemia means that there are higher than normal levels of calcium in your blood. People with parathyroid cancer often have hypercalcemia because the cancer causes your parathyroid gland to become overactive and release large amounts of parathyroid hormone (PTH). Parathyroid hormone regulates the amount of calcium in your blood, so too much parathyroid hormone means you’ll have too much calcium in your blood.

Hypercalcemia can be harmful to your health and body. Because of that, it’s just as important to treat hypercalcemia caused by parathyroid cancer as it is to treat the cancer itself.

***Types of parathyroid cancer***

There are two kinds of parathyroid cancer. The more common of the two types is when the cancer cells cause your parathyroid gland to produce and release too much parathyroid hormone (PTH), which increases calcium levels in your blood.

The other form of parathyroid cancer is non-functioning (also called non-secreting) parathyroid carcinoma (cancer).

It differs from the more common form of parathyroid cancer in that people with non-functioning parathyroid cancer don’t have increased levels of parathyroid hormone and calcium in their blood.

Non-functioning parathyroid carcinoma accounts for less than 10% of parathyroid cancer cases, making it extremely rare.

After parathyroid cancer has been diagnosed, further tests are done to determine if the cancer is localized (found in one place), metastatic (the cancer cells have spread to another part of your body) or recurrent (the cancer comes back after treatment) based on testing findings.

Healthcare professionals don’t use the four-stage naming system for parathyroid cancer that’s commonly used for other types of cancers.

***Who is likely to have parathyroid cancer?***

Parathyroid cancer affects males and females equally. It generally occurs in people over the age of 30.

Parathyroid cancer is very rare. It accounts for 0.005% of all cancers and 0.5% to 1% of all parathyroid conditions.

**CAUSES**

There isn’t a known cause of parathyroid cancer.

The following rare genetic (inherited by a family member) disorders are considered risk factors for parathyroid cancer:

* Multiple endocrine neoplasia type I (MEN1).
* Familial isolated hyperparathyroidism (FIHP).
* Hyperparathyroidism-jaw tumor syndrome.

Past radiation therapy for your head or neck can also increase your risk of parathyroid cancer.

**RISK FACTORS**

The risk factors for parathyroid cancer include several genetic conditions and other factors. Certain inherited disorders significantly increase the risk, such as hyperparathyroidism-jaw tumor (HPT-JT) syndrome, which is caused by a mutation in the CDC73 gene.

Up to about one-third of people with HPT-JT syndrome develop parathyroid cancer. Familial isolated hyperparathyroidism (FIHP), which can be caused by changes in the MEN1, CASR, CGM2, or CDC73 genes, also increases the risk, although most cases involve non-cancerous tumors.

Multiple endocrine neoplasia type 1 (MEN1), caused by changes in the MEN1 gene, is another inherited condition that increases the risk of parathyroid cancer, though it is rare.

Other potential risk factors include secondary hyperparathyroidism, which is often associated with long-term kidney disease, and multiple endocrine neoplasia type 2A (MEN2A), which is caused by changes in the RET gene.

Exposure to high doses of radiation, particularly to the neck area, may also increase the risk of developing parathyroid tumors, although this is more commonly linked to thyroid cancer.

Additionally, a history of hyperparathyroidism with chronic kidney failure, thyroid cancer, and previous radiation therapy on the neck may contribute to the risk.

It is important to note that most cases of parathyroid cancer are not caused by inherited genetic factors, and the exact cause is often unknown. Somatic mutations, which are acquired during a person's lifetime, are more commonly associated with parathyroid cancer.

However, in some cases, germline mutations, which are inherited from a parent, can increase the risk. The CDC73 gene is found to be mutated in up to 70% of parathyroid cancer cases, and in approximately one-third of these cases, the mutation is inherited.

**SIGNS / SYMPTOMS**

Most of the symptoms of parathyroid cancer are actually symptoms of hypercalcemia that develop because of it.  
Symptoms and signs of hypercalcemia include:

* Having to pee more often than usual (frequent urination).
* Being thirstier than usual.
* Nausea and vomiting.
* Not feeling as hungry as usual.
* Constipation.
* Feeling really tired.
* Feeling depressed.
* Forgetfulness or memory loss.
* Muscle aches, weakness and/or cramping.

Other possible symptoms of parathyroid cancer include:

* Having a lump in your neck.
* Experiencing voice changes or hoarseness.
* Difficulty swallowing.

**DIAGNOSIS METHODS**

Parathyroid cancer can be challenging to diagnose. One reason for this is that the cells of a benign (noncancerous) parathyroid adenoma and the cells of parathyroid cancer look similar.

A diagnosis of parathyroid cancer is most often made after your abnormal, overactive parathyroid gland has been surgically removed (parathyroidectomy) and further testing is done with the tissue. Sometimes, the surgeon is able to tell during the surgery that it’s parathyroid cancer.

***What tests will I have to diagnose parathyroid cancer?***

Parathyroid cancer is most often diagnosed after your abnormal, overactive parathyroid gland has been surgically removed (parathyroidectomy) because you were diagnosed with primary hyperparathyroidism.

Sometimes, the surgeon is able to diagnose parathyroid cancer during the surgery.

Prior to the surgery to remove your overactive parathyroid gland, you may undergo the following tests and procedures:

* Blood calcium test.
* Blood PTH (parathyroid hormone) test.
* Parathyroid scan (typically a radioactive sestamibi scan and/or CT scan).

If you’ve been diagnosed with parathyroid cancer, you may undergo the following imaging tests so your healthcare provider can determine if the cancer has spread (metastasized) to other parts of your body:

* CT (computerized tomography) scan: A CT scan uses X-rays and a computer to produce many 3D (three-dimensional) images of your body.
* MRI (magnetic resonance imaging): MRI uses a large magnet, radio waves and a computer to produce detailed images of your body. It doesn’t use X-rays (radiation).

**TREATMENT OPTIONS**

Surgery to remove the cancerous parathyroid gland (en bloc resection) is the main option for treatment of parathyroid cancer. Your surgeon may also need to remove tissue around your parathyroid gland or cancerous tissues elsewhere in your body if the parathyroid cancer has spread (metastasized).

The following surgical procedures may be used for parathyroid cancer:

* En bloc resection: Your surgeon will remove your entire parathyroid gland and the capsule around it. Your surgeon may need to also remove half of your thyroid gland that’s on the same side as the cancerous parathyroid gland and surrounding tissues, muscle and nerves.
* Tumor debulking: Your surgeon will remove as much of the tumor as possible. Some tumors cannot be completely removed.
* Metastasectomy: Your surgeon will remove any cancer that has spread (metastasized) to other tissues and/or organs in your body, such as your lung.

***Will I receive chemotherapy or radiation for parathyroid cancer?***

Chemotherapy and radiation therapy aren’t commonly used to treat parathyroid cancer. Your healthcare provider will determine if there’s a chemotherapy and/or radiation therapy option that’ll work for you.

***Will I take medications for parathyroid cancer?***

There aren’t currently any known medications for parathyroid cancer specifically. Most people with parathyroid cancer experience symptoms of hypercalcemia. Since hypercalcemia can be harmful, your healthcare provider may have you take one or more of the following hypercalcemia medications:

* Cinacalcet (Sensipar®).
* Bisphosphonates.
* Denosumab (XGEVA®).

If you’re experiencing severe symptoms of hypercalcemia, your healthcare provider may recommend immediate hospitalization for IV (intravenous) fluids and other treatments.

1. Chemotherapy Drugs Used (generally for metastatic or recurrent disease)

* There are no standard chemotherapy drugs approved specifically for parathyroid cancer, but some agents have been used alone or in combination:
  + Dacarbazine (DTIC)
  + 5-Fluorouracil (5-FU)
  + Cyclophosphamide
  + Methotrexate
  + Doxorubicin
  + Lomustine
* These chemotherapies may be considered in cases where surgery is not possible or cancer has spread.
* Side effects include:
  + Low white blood cell count (immunosuppression)
  + Nausea and vomiting
  + Diarrhea
  + Hair loss
  + Fatigue and general malaise

2. Hypercalcemia Control Medications

Since parathyroid cancer often causes severe hypercalcemia due to excess parathyroid hormone (PTH), drugs to lower calcium levels are crucial:

* Cinacalcet (a calcimimetic agent): Helps lower calcium by increasing the sensitivity of calcium-sensing receptors, reducing PTH secretion.
* Bisphosphonates (e.g., pamidronate): Inhibit bone resorption to lower serum calcium.
* Calcitonin: Temporarily lowers calcium levels by inhibiting bone resorption.
* Side effects of hypercalcemia drugs may include:
  + Gastrointestinal symptoms (nausea, vomiting)
  + Flushing (especially with calcitonin)
  + Rarely, hypocalcemia if overcorrected

3. Targeted Therapies and Experimental Drugs

There are emerging treatments under investigation due to the rarity and limited response to conventional therapies:

* Surufatinib: A tyrosine kinase inhibitor shown in case reports to induce tumor regression in advanced cases. Side effects include hypertension, fatigue, diarrhea, and liver enzyme abnormalities.
* TKIs like sorafenib, cabozantinib, regorafenib: Used off-label in some metastatic cases with partial response. Side effects can include hypertension, hand-foot syndrome, fatigue, diarrhea, and skin changes.
* Immunotherapy: Agents like pembrolizumab (anti-PD1) have been tried in select patients with some benefit, especially if tumors have specific genetic mutations. Side effects are immune-related, such as skin rash, colitis, or thyroid dysfunction.
* mTOR inhibitors (everolimus) and other targeted combinations are experimental with potential benefits but side effects like mouth ulcers, infections, and fatigue can occur.

4. Radiation Therapy

* Not a drug, but sometimes used after surgery. Parathyroid cancer is usually radioresistant.

***Will my parathyroid cancer come back?***

More than half of people diagnosed with parathyroid cancer will experience recurrence (the cancer comes back).

Usually, the cancer comes back within a few years — anywhere from two to five years. In some cases, it can even happen decades later (20 years or more) after your original diagnosis and treatment.

***Can my parathyroid cancer spread to other parts of my body (metastasize)?***

Parathyroid cancer can spread (metastasize) to other parts of your body. Approximately 10% to 30% of people who are diagnosed with parathyroid cancer will have metastatic cancer at the time of diagnosis. In metastatic parathyroid cancer cases, the cancer most commonly spreads to your lung, bone or liver, but it could also spread to your lymph nodes or pancreas.

**PREVENTION TIPS**

Unfortunately, there isn’t a known way to prevent parathyroid cancer. You cannot reduce your risk of getting this condition either. If you have a family history of parathyroid conditions, talk to your healthcare provider.

**OUTLOOK / PROGNOSIS**

The prognosis for parathyroid cancer depends on the completeness of your parathyroid gland removal when you're first diagnosed, due to the limited secondary treatment options, like radiation and chemotherapy.

Survival rates for people who have parathyroid cancer and had a complete removal surgery (en bloc resection) with no cancer cells found in the tissue around their cancerous parathyroid gland (negative margins) are up to 90% at five years after the initial surgery and 67% at 10 years after the initial surgery.

More than half of people who’ve been diagnosed with parathyroid cancer and undergo surgery to remove their gland experience recurrence (the parathyroid cancer comes back). In these cases, the cancer usually comes back two to five years after the initial surgery, but it can come even decades later.

**WHEN TO SEE A DOCTOR / RED FLAG**

You should see a doctor for parathyroid carcinoma if you notice any signs or symptoms suggestive of this rare cancer or complications related to overproduction of parathyroid hormone and high calcium levels in your blood. Key situations to seek medical advice include:

* Symptoms related to hypercalcemia (high blood calcium), such as:
  + Persistent fatigue, weakness, or muscle aches
  + Nausea, vomiting, loss of appetite, or unexplained weight loss
  + Increased thirst and frequent urination
  + Constipation or abdominal pain
  + Confusion, trouble thinking clearly, or depression
  + Bone pain or fractures
  + Kidney problems, such as pain from kidney stones or kidney disease
* Presence of a lump or swelling in the neck
* Voice changes such as hoarseness or difficulty swallowing
* Any new or persistent neck pain or discomfort
* If you have a known parathyroid disorder and develop worsening symptoms or new findings

Early signs are often caused by hormone overproduction leading to high calcium rather than the tumor itself, so symptoms can be systemic and varied. Seeing a healthcare provider early allows for diagnosis through blood tests, imaging, and appropriate treatment, improving outcomes.

If you experience any of the above symptoms—especially in combination or persisting over weeks—it is important to consult your doctor promptly for evaluation and to rule out or confirm parathyroid carcinoma or other parathyroid disease

**DIFFERENTIAL DIAGNOSIS**

***Diagnostic Considerations***

The preoperative diagnosis of parathyroid carcinoma is generally considered to be impossible. However, certain clinical features should raise the possibility of parathyroid carcinoma. Rapidly progressive and severe hyperparathyroidism is characteristic of parathyroid carcinoma. Also, a hard or palpable parathyroid should be considered as carcinoma until proven otherwise.

Biopsy cannot reliably diagnose parathyroid carcinoma. Typically, surgical treatment should be performed presumptively based on clinical features or intraoperative findings.

"Atypical parathyroid tumor" was included in the 2022 World Health Organization (WHO) classification of parathyroid tumors. [20]  These tumors have some features of parathyroid carcinoma, such as fibrosis, but lack definitive hallmarks of carcinoma such as extracapsular invasion.

Typically these tumors have low malignant potential and are at low risk of recurrence, but long-term surveillance may be warranted.

***Differential Diagnoses***

* Hypercalcemia
* Hyperparathyroidism
* Paraneoplastic Syndromes
* Thyroid Nodule

**RECENT GUIDELINES OR UPDATES**

A definitive management of primary hyperparathyroidism (pHPT) include the following recommendations for diagnosis and treatment of parathyroid carcinoma:

* The diagnosis of parathyroid carcinoma should be considered in patients with pHPT, markedly elevated PTH levels and severe hypercalcemia
* Patients with pHPT who present with hypercalcemic crisis should be medically managed, followed by parathyroidectomy for suspected parathyroid carcinoma
* If parathyroid carcinoma is suspected, patients should be treated surgically because this is the only potentially curative treatment
* Parathyroidectomy is indicated when the clinical or biochemical evidence is consistent with parathyroid carcinoma
* Preoperative parathyroid fine-needle aspiration biopsy is not recommended
* Histologic diagnosis relies on identification of unequivocal angioinvasion and can be assisted by biomarkers
* When there is intraoperative suspicion of parathyroid carcinoma, complete resection avoiding capsular disruption offers potential cure; en bloc resection of adherent tissues may be required
* There is insufficient evidence to recommend prophylactic central or lateral neck dissection
* Adjuvant external beam radiotherapy is reserved for palliative care and should not be routinely performed after surgical resection

**EPIDEMIOLOGY**

Parathyroid cancer is a rare disease. Data from the National Cancer Institute’s Surveillance, Epidemiology, and End Results (SEER) program show an increasing annual incidence until approximately 2001.

More recently the annual incidence has stabilized to a rate of approximately 11 cases per 10,000,000 persons.Parathyroid cancer occurs in 0.5-1% of all cases of hyperparathyroidism.

A large European study also demonstrated the rarity of parathyroid carcinoma, with an estimated incidence of 2 cases per 10,000,000 persons/year.

A nationwide Korean study found that the age-standardized incidence rose from 3.8 cases per 10,000,000 persons/year in 2003 to 6.6 cases per 10,000,000 persons/year in 2016.

No known racial predilection exists. Parathyroid cancer occurs equally in males and females.Males fare slightly worse in prognosis.Parathyroid cancer usually occurs in patients older than 30 years. Beyond that, no predominant age association has been noted.

***Genetic Testing***

In patients with suspected or proven parathyroid cancer, genetic testing for germline *CDC73* mutation should be considered to rule out HPT-JT. For example, young patients with unexplained hyperparathyroidism should be considered for genetic screening.

Besides *CDC73* mutations that can be detected by conventional mutational analysis, intragenic deletion of *CDC73* has also been reported in patients with familial hyperparathyroidism and parathyroid carcinoma.

Therefore, intragenic and gross gene deletion should be assessed as a part of genetic analysis in patients with a high index of suspicion but negative *CDC73* mutational testing. Partial or complete gene deletion can be detected by methods such as exon array comparative genome hybridization.

***Staging***

Two staging systems have been developed and externally validated to classify risk in parathyroid carcinoma.

The first is called the differentiated system and is reminiscent of a typical tumor, node, metastasis (TNM) staging system. It divides patients into 4 classes of risk. The second system is simpler and divides patients simply into high and low risk.

***Differentiated system***

*T (tumor)*

* (Tx) - No information available
* T1 - Evidence of capsular invasion
* T2 - Invasion of surrounding soft tissues, excluding the vital organs of the trachea, larynx, and esophagus
* T3 - Evidence of vascular invasion
* T4 - Invasion of vital organs, such as the hypopharynx, trachea, esophagus, larynx, recurrent laryngeal nerve, carotid artery

*N (node)*

* (Nx) - Lymph node not assessed
* N0 - No regional lymph node metastases
* N1 - Regional lymph node metastases

*M (meastasis)*

* (Mx) - Distant metastases not assessed
* M0 - No evidence of distant metastases
* M1 - Evidence of distant metastases

*Classes*

* Class I - T1 or T2 N0 M0
* Class II - T3 N0 M0
* Class III - Any T, N1 M0, or T4
* Class IV - Any N, M1

High/low risk system

Criteria are as follows:

* Low risk - Capsular invasion combined with invasion of surrounding soft tissue
* High risk - Vascular invasion and/or lymph node metastases and/or invasion of vital organs and/or distant metastases

**PREDEFINED Q & A SETS**

***Question 1: “Has the cancer spread outside of my parathyroid gland?”  
Answer: “***Parathyroid cancer can spread (metastasize), most commonly to the lungs, bones, liver, lymph nodes, or pancreas. About 10% to 30% of patients have metastatic cancer at diagnosis. Your doctor will use imaging studies and tests to determine if the cancer has spread beyond the parathyroid gland.”

***Question 2: “What’s the best treatment for parathyroid cancer?’  
Answer: “***Surgery is the main and most effective treatment, especially complete removal of the tumor and affected tissue (en bloc resection). Chemotherapy and radiation are generally not very effective but may be used in certain cases, for example to reduce bone metastases or after surgery. Treatment also focuses on managing hypercalcemia (high blood calcium levels).”

***Question 3: “What are the treatment risks and side effects?”  
Answer: “***Surgery risks include injury to surrounding structures, hypocalcemia (low calcium), vocal cord paralysis, or recurrent laryngeal nerve damage. Radiation and chemotherapy side effects depend on the specific regimens used but are less commonly used in parathyroid cancer. Managing hypercalcemia is very important as severe hypercalcemia causes serious complications.”

***Question 4: “Is my family at risk for developing parathyroid cancer?”  
Answer: “***Parathyroid cancer is rare and usually sporadic, but some genetic syndromes (like hyperparathyroidism-jaw tumor syndrome) can increase risk. If you have a family history of parathyroid conditions or related syndromes, it is important to discuss genetic counseling and testing with your healthcare provider.”

***Question 5: “Can I get parathyroid cancer again?”  
Answer: “***Yes, recurrence is common. More than half of patients experience cancer recurrence, sometimes years or even decades after initial treatment. Lifelong monitoring is recommended to detect and treat recurrence early.”

***Question 6: “Am I at risk for other types of cancer?”  
Answer: “***Generally, parathyroid cancer does not directly increase risk for other cancers unless associated with a hereditary syndrome that predisposes to multiple tumors. Your healthcare provider can assess your overall cancer risk based on family history and genetics.”

***Question 7: “What type of follow-up care do I need after treatment?”  
Answer: “***Lifelong follow-up is needed with regular physical exams, blood tests to monitor calcium and parathyroid hormone levels, and imaging as needed to detect recurrence or metastases early. This follow-up helps manage complications like hypercalcemia and guides timely treatment if cancer returns.”

***Question 8: “Should I look out for signs of complications?”  
Answer: “***Yes. Signs to watch for include symptoms of high blood calcium such as nausea, vomiting, weakness, confusion, bone pain, kidney stones, or recurring fractures. Also report any new lumps, persistent neck pain, difficulty swallowing, or voice changes promptly to your doctor”

**DOCTOR-PATIENT CONVERSATIONS**

Doctor: Hello, I want to discuss the results of your tests. We have found that you have parathyroid cancer, which is a rare malignancy of the parathyroid gland. Do you have any questions so far?

Patient: Parathyroid cancer? I didn’t even know you could get cancer there. What does this mean?

Doctor: It means that some cells in your parathyroid gland have grown uncontrollably. The parathyroid glands help regulate your calcium levels. This cancer can cause high calcium in your blood, which may explain some of your symptoms like fatigue or weakness.

Patient: How serious is this? Can it be treated?

Doctor: Parathyroid cancer is rare, but surgery is usually the best and most effective treatment. We typically aim to remove the tumor along with any affected surrounding tissue. This offers the best chance for cure. Sometimes additional treatments like medication may be needed to manage calcium levels.

Patient: Will the cancer spread? What happens if it has spread?

Doctor: The cancer can sometimes spread to nearby lymph nodes or distant sites like the lungs or bones, but this is less common. We do imaging tests to check for spread. If it has spread, treatment may involve additional surgery or therapies, but we will tailor this to your specific situation.

Patient: What are the risks with surgery? Will there be side effects?

Doctor: Surgery carries some risks like injury to nearby nerves that control your voice or swallowing, and temporary or permanent low calcium levels if other parathyroid glands are affected. We monitor these carefully and provide supportive treatments if needed.

Patient: How will we know if the cancer comes back?

Doctor: We will do lifelong follow-up with regular blood tests to check your calcium and parathyroid hormone levels, and imaging if needed. Early detection of any recurrence gives us the best chance to treat it effectively.

Patient: Is parathyroid cancer hereditary? Should my family be checked?

Doctor: Most cases are sporadic, but some rare genetic syndromes can increase risk. We will review your family history and may refer you for genetic counseling if appropriate.

Patient: What symptoms should I watch for after treatment?

Doctor: Please report symptoms like new neck lumps, persistent hoarseness, difficulty swallowing, or signs of high calcium such as nausea, vomiting, confusion, or muscle weakness.

Patient: Thank you, Doctor. This helps me understand what to expect.

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**PERICHONDRITIS**

*ALTERNATIVE NAMES:* Perichondritis is also known by several alternative names, including auricular perichondritis and pinna perichondritis. These terms are often used interchangeably to describe the same condition, which involves an infection of the tissue surrounding the cartilage of the outer ear.

**DEFINITION / DESCRIPTION**

Perichondritis is an infection of the tissue covering the cartilage of your outer ear. This part of your ear is called the pinna or auricle. The infection is sometimes called auricular perichondritis or pinna perichondritis.

Perichondritis typically occurs due to some form of ear trauma. This can include injuries, burns and high ear piercings. It causes pain, swelling and redness of your outer ear. Sometimes a buildup of pus (abscess) may form between your ear tissue and cartilage.

Delayed diagnosis and treatment can cause serious complications. If left untreated, an abscess can cut off the blood supply to your cartilage. This can damage your cartilage and lead to tissue death (necrosis). Necrosis can cause a permanent ear deformity called cauliflower ear.

***Perichondritis and cellulitis***

Perichondritis and cellulitis are both skin infections that can cause inflammation. But they have different causes and symptoms. Perichondritis typically only affects the upper part of your outer ear. It doesn’t affect your earlobe. Cellulitis can affect your entire outer ear, including the upper part of your ear and your earlobe.

***How common is perichondritis?***

Perichondritis isn’t a very common skin condition, but researchers don’t know exact numbers. One study in England indicated cases doubled between 1990 and 1998. Researchers noted the increase was likely due to a rise in high ear piercings among adolescents.

Other studies have supported this theory. High ear piercings carry a greater risk of infection than earlobe piercings.

**CAUSES**

Perichondritis is a bacterial infection. Pseudomonas aeruginosa is the most common type of bacteria that causes the infection. Staphylococcus aureus and Escherichia coli can also cause perichondritis.

Perichondritis occurs when bacteria enter the cartilage of your outer ear. This most frequently occurs due to an injury or trauma.

Cartilage is the thick tissue that creates the shape of your outer ear. Cartilage has a thin layer of tissue surrounding it called perichondrium. The perichondrium provides nutrients to your cartilage.

The most common cause of perichondritis is high ear piercings through your cartilage. This type of piercing can damage your cartilage, which allows for the introduction of bacteria. Other causes of perichondritis may include:

* Trauma to the side of your head.
* Contact sports.
* Burns.
* Insect bites.
* Acupuncture.

Less common causes of perichondritis include:

* Ear surgery.
* Untreated ear infections that spread to your cartilage.
* Minor trauma, such as scratching your ear.
* Herpes zoster (shingles) infection.

Perichondritis may affect certain groups of people as well. This includes people who have:

* Inflammatory conditions, such as granulomatosis with polyangiitis.
* Weakened immune systems.
* Conditions like diabetes.

**SIGNS / SYMPTOMS**

The most common symptom of perichondritis is a painful, red and swollen outer ear. The redness typically surrounds a bite, cut, scrape or another injury. Perichondritis tends to affect the upper part of your outer ear but not your earlobe. If you have an abscess, you may experience fluid drainage. Other symptoms of perichondritis may include:

* Fever.
* Tenderness.
* Feeling of warmth in your ear.

**DIAGNOSIS METHODS**

Your healthcare provider will perform a physical examination to diagnose perichondritis. They’ll look at and feel your ear. They’ll also ask you about your symptoms.

Your healthcare provider will ask about your medical history, including any recent trauma to your ear. Trauma may include any piercings, burns, cuts or acupuncture procedures.

**TREATMENT OPTIONS**

Treatment guidelines for perichondritis typically include the use of antibiotics. The type of antibiotics depends on the severity of your condition and the kind of bacteria causing the infection.

A common antibiotic for the treatment of perichondritis is fluoroquinolone. You may take the antibiotic by mouth (orally) or apply it directly onto your skin (topically).

Perichondritis treatment for children may or may not include fluoroquinolone. Some studies have shown the antibiotic causes side effects like joint stiffness and tendon tears in children. But the American Academy of Pediatrics has said the antibiotic is safe for children.

Your healthcare provider will remove any foreign objects from your ear. This includes any high ear piercings and splinters. They may also recommend:

* A corticosteroid taken by mouth.
* Warm compresses.
* Over-the-counter pain relievers.

You may need surgery if you have a buildup of pus (abscess) cutting off the blood supply to your cartilage. Your healthcare provider will make a cut (incision) in the abscess. Then they’ll drain the pus and remove any dead cartilage and skin. The incision will allow blood to reach your cartilage again.

Your healthcare provider may leave a small drain in place for one to three days. To maintain blood supply to your cartilage, your healthcare provider may stitch your tissue to your cartilage.

**PREVENTION TIPS**

The best way to prevent perichondritis is to avoid piercing your high ear cartilage. Your risk of infection is much lower if you pierce your earlobe. Other ways to prevent perichondritis include:

* Don’t get acupuncture on your ears.
* Get ear infections treated promptly.
* Avoid contact sports.
* Try not to scratch your ears too much.

**OUTLOOK / PROGNOSIS**

With prompt diagnosis and antibiotic treatment, your symptoms should start to clear within two to three days. Some discomfort may remain for up to one month. But you can expect to make a full recovery.

If you have an abscess, your healthcare provider may have to perform surgery. They may leave in a drain that they’ll have to remove. After surgery, you’ll need to pack and dress your wound to let it heal properly.

If you don’t get treatment, an abscess can cut off the blood supply to your cartilage. This can destroy your cartilage and cause tissue death. This can lead to an ear deformity called cauliflower ear.

Perichondritis can also lead to an infection in your ear cartilage called chondritis. Chondritis can cause severe damage to your ear structure.

Part of your ear tissue may die and need to be surgically removed. You may need ear reshaping surgery to restore your ear to its normal shape.

**WHEN TO SEE A DOCTOR / RED FLAG**

* If you notice ear redness, swelling, or pain, especially after trauma or piercing
* If symptoms worsen or pus appears
* If fever or systemic symptoms develop
* Early treatment improves recovery and avoids complications

**DIFFERENTIAL DIAGNOSIS**

* Auricular cellulitis: Infection involving the skin and soft tissue including the earlobe, usually more diffuse swelling and erythema than perichondritis, which preferentially involves the cartilage and spares the earlobe.
* Auricular abscess: Localized collection of pus that may complicate perichondritis or other infections, requiring drainage.
* Otitis externa (ear canal infection): Inflammation/infection of the external auditory canal, causing pain and swelling, distinct from perichondritis which affects the pinna cartilage.
* Relapsing polychondritis: A rare autoimmune disorder causing recurrent inflammation of cartilage throughout the body, including the ear cartilage. It can mimic infectious perichondritis but is non-infectious and chronic, often bilateral, associated with systemic symptoms.
* Fungal infections: Can sometimes affect the ear and mimic perichondritis, especially in immunocompromised patients.
* Trauma or burns: Causes local inflammation that can present similarly.
* Other infections: Such as syphilis, leprosy (Hansen's disease), tuberculosis, or fungal infections may rarely involve auricular cartilage.
* Contact dermatitis or eczema: Skin conditions causing redness and swelling but primarily affecting the skin rather than cartilage.
* Auricular hematoma: Blood collection between cartilage and skin, usually following blunt trauma, can cause swelling and deformity resembling perichondritis but without infection.

**EPIDEMIOLOGY**

* Perichondritis, an infection or inflammation of the ear cartilage (pinna), is considered a rare condition, but exact incidence rates are not well established in the general population.
* One 10-year review in a rural hospital setting identified 30 cases of perichondritis, with a median patient age of about 28 years. In this study, 60% of cases were males, and about 23% had a known cause related to ear cartilage piercing. Other causes included otitis externa, trauma, insect bites, postoperative infections, and some cases had unknown causes.
* Perichondritis cases have been reported to rise in some regions, partly associated with the popularity of cartilage ear piercings, which predispose younger individuals and females more commonly to the condition.
* In another study, the incidence of perichondritis among ear injury patients was around 9.7%, indicating it is a recognized complication of auricular trauma.
* The condition can affect all ages but tends to be more common in younger individuals with cartilage piercings or ear trauma.
* Perichondritis is more common in males in some reports, but piercing-associated cases tend to occur more in females and younger patients.
* Indigenous populations in some rural settings appeared to have a higher likelihood of requiring surgical intervention and developing abscesses compared to non-Indigenous patients.
* Common pathogens include *Pseudomonas aeruginosa* and *Staphylococcus aureus*, with fungal cases noted especially in postoperative situations.
* The condition is not very common in general medical literature, and its precise population-based incidence remains largely unknown, but the number of cases has been observed to increase alongside increases in cartilage ear piercing practices

**PREDEFINED Q & A SETS**

Q1: “What is perichondritis?”

A: Perichondritis is an infection or inflammation of the tissue (perichondrium) covering the cartilage of the outer ear (pinna). It usually affects the ear's cartilage but spares the earlobe.

## Q2: “What causes perichondritis?”

A:

* Injury to the ear such as trauma, burns, or insect bites
* Ear cartilage piercings, especially in the upper ear (not the earlobe)
* Ear surgery or infections spreading from the skin
* Occasionally, autoimmune or inflammatory diseases in susceptible individuals

## Q3: “What are the main symptoms of perichondritis?”

A:

* Redness, swelling, and pain of the ear cartilage
* Warmth around the ear with possible pus formation
* Fever may be present in more severe cases
* The ear may become deformed (cauliflower ear) if untreated

## Q4: “How is perichondritis diagnosed?”

A

* Clinical examination based on symptoms and ear appearance
* Medical history including recent trauma or piercings
* Imaging (ultrasound or MRI) and lab tests if needed to assess severity or complications

## Q5: “What treatments are used for perichondritis?”

A:

* Prompt antibiotic therapy, often with fluoroquinolones, targeting common bacteria like *Pseudomonas aeruginosa* and *Staphylococcus aureus*
* Corticosteroids may be used to reduce inflammation in some cases
* Removal of any foreign bodies like earrings or splinters
* Incision and drainage if an abscess (pus collection) develops
* Pain management and warm compresses
* Surgery in severe or complicated cases to drain pus and restore blood flow to cartilage

## Q6: “What are the risks of complications with perichondritis?”

A:

* Cartilage necrosis due to interrupted blood supply
* Chronic recurrent infections
* Permanent deformity of the ear (cauliflower ear) if untreated or delayed treatment

## Q7: “How can perichondritis be prevented?”

A:

* Avoid upper ear cartilage piercings or practice meticulous care if pierced
* Promptly treat ear infections and injuries
* Avoid trauma or excessive ear scratching
* Avoid contact sports that risk ear injury

## Q8: “When should I see a doctor?’

A

* If you notice ear redness, swelling, or pain, especially after trauma or piercing
* If symptoms worsen or pus appears
* If fever or systemic symptoms develop
* Early treatment improves recovery and avoids complications

**DOCTOR-PATIENT CONVERSATIONS**

Doctor: Hello, I see you’re experiencing pain and swelling in your ear. Can you tell me when it started and what it feels like?

Patient: Yes, doctor. My ear has been red, swollen, and really sore, especially around the upper part. It’s been like this for a couple of days, and it hurts more when I touch it.

Doctor: From what you describe, it sounds like perichondritis, which is an infection or inflammation of the cartilage in the outer ear. It often causes redness, swelling, and pain, especially sparing the earlobe.

Patient: How did I get this? I haven’t had an injury or anything.

Doctor: Perichondritis often develops after trauma, like an ear piercing in the cartilage, insect bites, or skin infections. Sometimes it can happen without a clear cause. If you recently had a cartilage piercing or any injury, that might have introduced bacteria that cause this infection.

Patient: What kind of bacteria causes it? Is it serious?

Doctor: The most common bacteria are *Pseudomonas aeruginosa* and *Staphylococcus aureus*. If treated promptly with antibiotics, perichondritis usually resolves without problems. However, if left untreated, it can cause cartilage damage, deformity of the ear (sometimes called cauliflower ear), or abscess formation, which are more serious.

Patient: How do you treat this? Do I need surgery?

Doctor: We usually start with oral antibiotics, often a class called fluoroquinolones that targets the common bacteria effectively. Sometimes, if the swelling is severe or pus forms (an abscess), a minor surgical procedure to drain it might be necessary. We also recommend warm compresses and avoiding further irritation to the ear.

Patient: How soon will I feel better?

Doctor: Many patients start to feel improvement within a few days of starting antibiotics. It’s important to complete the full course of medication. If symptoms worsen or you notice increasing pain, pus, fever, or spreading redness, you should come back immediately.

Patient: Is there anything I can do to prevent this in the future?

Doctor: Yes, avoid unnecessary cartilage piercings or take good care of them with proper hygiene. Also, protect your ears from trauma, and promptly treat any outer ear infections or skin wounds.

Patient: Thank you, Doctor. I feel better knowing what it is and how to treat it.

Doctor: You’re welcome. We’ll keep an eye on it closely. Let me know if you have any concerns during your treatment.

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**PEDIATRIC GASTROESOPHAGEAL REFLUX DISEASE**

ALTERNATIVE NAMES:

**DEFINITION / DESCRIPTION**

GERD, or gastroesophageal reflux disease, is a long-term (chronic) digestive disorder. It happens when stomach contents flow back up (reflux) into the food pipe (esophagus).

GERD is a more serious and long-lasting form of gastroesophageal reflux (GER).

GER is common in babies under 2 years old. Most babies spit up a few times a day during their first 3 months. GER does not cause any problems in babies. In most cases, babies outgrow this by the time they are 12 to 14 months old.

It is also common for children and teens ages 2 to 19 to have GER from time to time. This doesn’t always mean they have GERD.

***When GER becomes GERD***

Your baby, child, or teen may have GERD if:

* Your baby’s symptoms prevent him or her from feeding. These symptoms may include: vomiting, gagging, coughing, and trouble breathing.
* Your baby has GER for more than 12 to 14 months
* Your child or teen has GER more than 2 times a week, for a few months

**CAUSES**

GERD is often caused by something that affects the LES, the lower esophageal sphincter. The LES is a muscle at the bottom of the food pipe (esophagus). The LES opens to let food into the stomach. It closes to keep food in the stomach. When the LES relaxes too often or for too long, stomach acid flows back into the esophagus. This causes vomiting or heartburn.

Everyone has reflux from time to time. If you have ever burped and had an acid taste in your mouth, you have had reflux. Sometimes the LES relaxes at the wrong times. Often your child will just have a bad taste in his or her mouth. Or your child may have a short, mild feeling of heartburn.

Babies are more likely to have a weak LES. This makes the LES relax when it should stay shut. As food or milk is digesting, the LES opens. It lets the stomach contents go back up to the esophagus.

Sometimes the stomach contents go all the way up the esophagus. Then the baby or child vomits. In other cases, the stomach contents only go part of the way up the esophagus. This causes heartburn or breathing problems. In some cases there are no symptoms at all.

Some foods seem to affect the muscle tone of the LES. They let the LES stay open longer than normal. These foods include:

* Chocolate
* Peppermint
* High-fat foods

Other foods cause the stomach to make more acid. These foods include:

* Citrus foods
* Tomatoes and tomato sauces

Other things that may lead to GERD include:

* Being obese
* Medicines, including some antihistamines, antidepressants, and pain medicines
* Being around secondhand smoke

**RISK FACTORS**

GERD is very common during a baby’s first year of life. It often goes away on its own. Your child is more at risk for GERD if he or she has:

* Down syndrome
* Neuromuscular disorders such as muscular dystrophy and cerebral palsy

**SIGNS / SYMPTOMS**

Heartburn, or acid indigestion, is the most common symptom of GERD. Heartburn is described as a burning chest pain. It begins behind the breastbone and moves up to the neck and throat. It can last as long as 2 hours. It is often worse after eating. Lying down or bending over after a meal can also lead to heartburn.

Children younger than age 12 will often have different GERD symptoms. They will have a dry cough, asthma symptoms, or trouble swallowing. They won’t have classic heartburn.

Each child may have different symptoms. Common symptoms of GERD include:

* Burping or belching
* Not eating
* Having stomach pain
* Being fussy around mealtimes
* Vomiting often
* Having hiccups
* Gagging
* Choking
* Coughing often
* Having coughing fits at night

Other symptoms may include:

* Wheezing
* Getting colds often
* Getting ear infections often
* Having a rattling in the chest
* Having a sore throat in the morning
* Having a sour taste in the mouth
* Having bad breath
* Loss or decay of tooth enamel

GERD symptoms may seem like other health problems. Make sure your child sees his or her healthcare provider for a diagnosis.

**DIAGNOSIS METHODS**

Your child's healthcare provider will do a physical exam and take a health history. Other tests may include:

* Chest X-ray. An X-ray can check for signs that stomach contents have moved into the lungs. This is called aspiration.
* Upper GI series or barium swallow. This test looks at the organs of the top part of your child’s digestive system. It checks the food pipe (esophagus), the stomach, and the first part of the small intestine (duodenum). Your child will swallow a metallic fluid called barium. Barium coats the organs so that they can be seen on an X-ray. Then X-rays are taken to check for signs of sores or ulcers, or abnormal blockages.
* Endoscopy. This test checks the inside of part of the digestive tract. It uses a small, flexible tube called an endoscope. It has a light and a camera lens at the end. Tissue samples from inside the digestive tract may also be taken for testing.
* Esophageal manometry. This test checks the strength of the esophagus muscles. It can see if your child has any problems with reflux or swallowing. A small tube is put into your child’s nostril, then down the throat and into the esophagus. Then it measures the pressure that the esophageal muscles make at rest.
* pH monitoring. This test checks the pH or acid level in the esophagus. A thin, plastic tube is placed into your child’s nostril, down the throat, and into the esophagus. The tube has a sensor that measures pH level. The other end of the tube outside your child’s body is attached to a small monitor. This records your child’s pH levels for 24 to 48 hours. During this time your child can go home and do his or her normal activities. You will need to keep a diary of any symptoms your child feels that may be linked to reflux. These include gagging or coughing. You should also keep a record of the time, type of food, and amount of food your child eats. Your child’s pH readings are checked. They are compared to your child’s activity for that time period.
* Gastric emptying study. This test is done to see if your child’s stomach sends its contents into the small intestine properly. Delayed gastric emptying can cause reflux into the esophagus

**TREATMENT OPTIONS**

In many cases, diet and lifestyle changes can help to ease GERD. Talk with your child’s healthcare provider about changes you can make. Here are some tips to better manage GERD symptoms.

For babies:

* After feeding, hold your baby in an upright position for 30 minutes.
* If bottle-feeding, keep the nipple filled with milk. This way your baby won’t swallow too much air while eating. Try different nipples. Find one that lets your baby's mouth make a good seal with the nipple during feeding.
* Adding rice cereal to feeding may be helpful for some babies.
* Burp your baby a few times during bottle-feeding or breastfeeding. Your child may reflux more often when burping with a full stomach.

For children:

* Watch your child's food intake. Limit fried and fatty foods, peppermint, chocolate, drinks with caffeine such as sodas and tea, citrus fruit and juices, and tomato products.
* Offer your child smaller portions at mealtimes. Add small snacks between meals if your child is hungry. Don’t let your child overeat. Let your child tell you when he or she is hungry or full.
* If your child is overweight, contact your child’s provider to set weight-loss goals.
* Serve the evening meal early, at least 3 hours before bedtime.

Other things to try:

* Ask your child's provider to review your child’s medicines. Some may irritate the lining of the stomach or esophagus.
* Don’t let your child lie down or go to bed right after a meal.
* Always check with your baby’s provider before raising the head of the crib if he or she has been diagnosed with gastroesophageal reflux. This is for safety reasons and to reduce the risk for SIDS and other sleep-related infant deaths.

***Medicines and other treatments***

Your child’s healthcare provider may also recommend other options.

Medicines. Your child's provider may prescribe medicines to help with reflux. There are medicines that help reduce the amount of acid the stomach makes. This reduces the heartburn linked to reflux. These medicines may include:

* H2-blockers. These reduce the amount of acid your stomach makes by blocking the hormone histamine. Histamine helps to make acid.
* Proton pump inhibitors. These help keep your stomach from making acid. They do this by stopping the stomach's acid pump from working.

The provider may prescribe another type of medicine that helps the stomach empty faster. If food doesn’t stay in the stomach as long as normal, reflex may be less likely to occur.

Calorie supplements. Some babies with reflux can’t gain weight because they vomit often. If this is the case, your child's healthcare provider may suggest:

* Adding rice cereal to baby formula
* Giving your baby more calories by adding a prescribed supplement
* Changing formula to milk- or soy-free formula if your baby may have an allergy

Tube feedings. In some cases tube feedings may be recommended. Some babies with reflux have other conditions that make them tired. These include congenital heart disease or being born too early (premature). These babies often get sleepy after they eat or drink a little. Other babies vomit after having a normal amount of formula. These babies do better if they are constantly fed a small amount of milk. In both of these cases, tube feedings may be suggested.

Formula or breastmilk is given through a tube that is placed in the nose. This is called a nasogastric tube. The tube is then put through the food pipe or esophagus, and into the stomach. Your baby can have a tube feeding in addition to a bottle feeding. Or a tube feeding may be done instead of a bottle feeding. There are also tubes that can be used to go around, or bypass, the stomach. These are called nasoduodenal tubes.

Surgery. In severe cases of reflux, surgery called fundoplication may be done. Your baby’s provider may recommend this option if your child is not gaining weight because of vomiting, has frequent breathing problems, or has severe irritation in the esophagus.

This is often done as a laparoscopic surgery. This method has less pain and a faster recovery time. Small cuts or incisions are made in your child’s belly. A small tube with a camera on the end is placed into one of the incisions to look inside. The surgical tools are put through the other incisions. The surgeon looks at a video screen to see the stomach and other organs. The top part of the stomach is wrapped around the esophagus. This creates a tight band. This strengthens the LES and greatly decreases reflux.

**POSSIBLE COMPLICATIONS**

Some babies and children who have GERD may not vomit. But their stomach contents may still move up the food pipe (esophagus) and spill over into the windpipe (trachea). This can cause asthma or pneumonia.

The vomiting that affects many babies and children with GERD can cause problems with weight gain and poor nutrition. Over time, when stomach acid backs up into the esophagus, it can also lead to:

* Inflammation of the esophagus, called esophagitis
* Sores or ulcers in the esophagus, which can be painful and may bleed
* A lack of red blood cells, from bleeding sores (anemia)

Adults may also have long-term problems from inflammation of the esophagus. These include:

* Narrowing, or stricture, of the esophagus
* Barrett’s esophagus, a condition where there are abnormal cells in the esophageal lining

**WHEN TO SEE A DOCTOR / RED FLAG**

Call you child's healthcare provider if your baby or child:

* Has reflux and is not gaining weight
* Has signs of asthma or pneumonia. These include coughing, wheezing, or trouble breathing.

**DIFFERENTIAL DIAGNOSIS**

***Diagnostic Considerations***

Vomiting is a symptom associated with many disorders. Accordingly, gastroesophageal reflux cannot be assumed to be the primary problem in infants and children who present with a history of emesis. Warning signals that herald the requirement for additional evaluation include the following:

* Bilious or forceful vomiting
* Hematemesis or hematochezia
* Vomiting with diarrhea
* Abdominal tenderness or distention
* Onset of vomiting after 6 months of life
* Fever, lethargy, hepatosplenomegaly
* Macrocephaly, microcephaly, seizures

The occurrence of any of these signs and symptoms indicates the need to consider a comprehensive metabolic, neurologic, and/or surgical evaluation, in addition to a gastroenterologic workup.

Conditions to consider in the differential diagnosis of gastroesophageal reflux include the following:

* Antral web
* Intestinal motility disorders, including achalasia, collagen-vascular disorders (e.g. systemic sclerosis)
* Eosinophilic esophagitis
* Helicobacter pylori associated gastritis
* Irritable bowel syndrome
* Peptic ulcer disease
* Tracheoesophageal fistula

***Differential Diagnoses***

* Acute Gastritis
* Chronic Gastritis
* Eosinophilic Esophagitis (EoE) Pathology
* Esophageal Motility Disorders
* Food Allergies
* Helicobacter Pylori Infection
* Hiatal Hernia
* Intestinal Malrotation
* Pediatric Duodenal Atresia and Stenosis Surgery
* Peptic Ulcer Disease

**RECENT GUIDELINES OR UPDATES**

Clinical practice guidelines on pediatric gastroesophageal reflux by the North American Society for Pediatric Gastroenterology, Hepatology, and Nutrition (NASPGHAN) and the European Society for Pediatric Gastroenterology, Hepatology, and Nutrition (ESPGHAN) recommend the following:

* Four to eight weeks of acid-suppression therapy is recommended to treat typical reflux symptoms. This should only occur after dietary modifications in infants are unsuccessful.
* Avoid acid suppression in infants and children with extraesophageal symptoms (eg, cough, wheezing, asthma).
* GERD complications should be assessed with esophagogastroduodenoscopy (EGD) with biopsies.
* pH/impedance studies are recommended to see the role of acid reflux in esophagitis and whether acid-suppression therapy was effective.
* Diagnosing GERD with salivary pepsin, extraesophageal biomarkers, and scintigraphy is not recommended.
* Only use manometry if there is suspicion of an underlying motility disorder.
* Positional therapy to treat reflux in infants is not recommended.
* Proton pump inhibitors can be used as treatment for erosive esophagitis.

**EPIDEMIOLOGY**

Gastroesophageal reflux is most commonly seen in infancy, with a peak at age 1-4 months. However, it can be seen in children of all ages, even healthy teenagers.

***United States statistics***

Approximately 85% of infants vomit during the first week of life, and 60-70% manifest clinical gastroesophageal reflux at age 3-4 months.

Symptoms abate without treatment in 60% of infants by age 6 months, when these infants begin to assume an upright position and eat solid foods. Resolution of symptoms occurs in approximately 90% of infants by age 8-10 months.

The estimated prevalence of gastroesophageal reflux among children aged older than 1 year and adolescents ranges from 0.9-18.8%.

**PREDEFINED Q & A SETS**

Q1: “What is a Pediatric GERD?”

A: GERD is a condition where stomach contents reflux back into the esophagus, causing troublesome symptoms or complications. It differs from simple reflux (GER), which is common in infants and usually resolves by 12 to 18 months without causing problems.

Q2: “What are the common symptoms of GERD in children?”

A:

* Frequent vomiting or regurgitation (more than normal spit-up)
* Irritability or feeding refusal, especially in infants
* Heartburn or chest pain (in older children)
* Persistent cough, wheezing, or recurrent respiratory infections
* Difficulty swallowing or painful swallowing
* Poor weight gain or growth problems
* Hoarseness, sore throat, or chronic throat clearing

Q3: “How is pediatric GERD diagnosed?”

A:

* Diagnosis mostly based on clinical history and physical exam
* Trials of feeding modifications or acid-suppressing medications to see if symptoms improve
* If symptoms persist or complications are suspected, tests may include:
  + Upper GI contrast x-ray
  + Esophageal pH or impedance monitoring
  + Endoscopy to assess esophageal inflammation or damage
  + Gastric emptying studies in select cases

Q4: “How is pediatric GERD treated?”

A:

* Lifestyle and feeding changes:
  + Smaller, more frequent meals
  + Thickening of feeds in infants
  + Upright positioning after feedings
  + Avoiding exposure to tobacco smoke
  + Avoiding foods that trigger symptoms (older children)
* Medications:
  + Acid suppressants like proton pump inhibitors (PPIs) or H2-receptor blockers for moderate to severe cases
  + PPIs are generally prescribed for 4 to 8 weeks but may be longer if needed
* Surgery:
  + Rarely needed, reserved for severe or complicated cases, often fundoplication procedure

Q5: “Are there any risks or complications of pediatric GERD?”

A:

* Esophagitis (inflammation of the esophagus)
* Strictures or narrowing of the esophagus
* Respiratory problems like asthma exacerbation or recurrent pneumonia
* Feeding difficulties and failure to thrive in severe cases

Q6: “When should I see a doctor for my child’s reflux?”

A:

* If reflux symptoms persist beyond infancy or worsen
* Feeding difficulties, poor weight gain, or growth problems
* Signs of respiratory issues like wheezing or chronic cough
* Vomiting blood or passing black stools
* Signs of pain, difficulty swallowing, or refusal to eat

**DOCTOR-PATIENT CONVERSATIONS**

Doctor: Hello, I understand you’re concerned about your child’s reflux symptoms. Can you tell me what you’ve observed?

Parent: Yes, my baby has been spitting up a lot and seems uncomfortable after feedings. Sometimes they arch their back and cry as if they’re in pain.

Doctor: That sounds like gastroesophageal reflux, which is quite common in infants. It means stomach contents are coming back up into the esophagus. Many babies “spit up” normally, but in some cases, it can cause discomfort or other problems, and we call that GERD.

Parent: Is this something serious? Will my baby outgrow it?

Doctor: Most infants with reflux get better as their digestive system matures, usually by 12 to 18 months of age. If your baby is growing well and not having breathing problems, we often recommend lifestyle changes first, such as feeding smaller amounts more frequently, keeping your baby upright after feeding, and burping them regularly.

Parent: What if the symptoms don’t improve or seem worse?

Doctor: If your baby isn’t gaining weight properly, has severe vomiting, or shows other concerning signs like coughing, wheezing, or refusal to eat, we may consider further evaluation. Sometimes medications like proton pump inhibitors can reduce stomach acid and help symptoms. But we usually try changes in feeding and positioning first.

Parent: Are there any risks with the medications?

Doctor: Medications generally are safe for children, but like all medicines, they can have side effects such as mild stomach upset or diarrhea. We use them only if lifestyle changes don’t help and monitor your child closely.

Parent: When would surgery be needed?

Doctor: Surgery is rare and reserved for severe cases where medicines don’t work or there are complications like aspiration pneumonia. The most common surgery is called fundoplication, which helps prevent reflux.

Parent: What can I do at home to help my child?

Doctor: Feeding your baby smaller amounts more frequently, keeping them upright for 20-30 minutes after feeding, avoiding tight diapers or clothing that puts pressure on the stomach, and avoiding exposure to tobacco smoke can all help.

Parent: What signs should make me call you?

Doctor: If your baby vomits blood or green bile, has trouble breathing, shows poor weight gain, develops persistent coughing or wheezing, or becomes very irritable or lethargic, please contact us promptly.

Parent: Thank you, Doctor. It helps to know what to expect and what I can do.

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**PEDIATRIC AIRWAY DISEASE**

ALTERNATIVE NAMES: “Reactive airway disease (RAD)” is an alternative name for pediatric airway disease, often used to describe a range of airway conditions in children that involve bronchial tubes overreacting to irritants.

It is sometimes referred to as “reactive airway disease (RAD)” and is often mislabeled as “asthma”, although they are not the same condition.

Other related terms include “pediatric airway disorders”, which encompass a variety of conditions affecting the airway, such as “subglottic stenosis”, “tracheomalacia”, and “laryngomalacia”.

Additionally, the term "narrow airway" is used to describe conditions where the airway is restricted or narrowed, leading to breathing difficulties.

**DEFINITION / DESCRIPTION**

Pediatric airway disease refers to a group of conditions that cause obstruction, inflammation, or functional problems in the airways of children, affecting their breathing. These disorders can be congenital (present at birth) or acquired and vary widely in severity and underlying cause.

Common Types and Examples

* Reactive Airway Disease (RAD): Often a provisional diagnosis used in young children with wheezing and coughing but without a firm asthma diagnosis; includes conditions with airway hyperreactivity.
* Asthma: A chronic inflammatory disease of the airways causing wheezing, coughing, chest tightness, and shortness of breath.
* Congenital airway anomalies: Such as laryngomalacia, tracheomalacia, subglottic stenosis.
* Infectious causes: Bronchiolitis, croup.
* Foreign body aspiration: Sudden onset airway obstruction symptoms.
* Other airway obstructions: Tumors, vascular rings.

**RISK FACTORS**

Pediatric airway disease has several risk factors that contribute to its development. These include both perinatal and postnatal factors. Preterm birth is a significant risk factor, as preterm infants are at higher risk of developing lung diseases such as bronchopulmonary dysplasia, recurrent wheezing, and asthma compared to term infants.

Additionally, perinatal factors such as chorioamnionitis, which is a common source of in utero inflammation, are associated with an increased risk of asthma in preterm infants.

Postnatal factors also play a crucial role. Exposure to environmental pollutants, such as diesel exhaust particles, has been shown to instigate asthma in mice and increase the risk of airway disease in children.

Children living near busy motorways or in high pollution areas have an increased risk of developing asthma. Furthermore, postnatal exposure to tobacco smoke, both prenatally and during the first year of life, predisposes children to reactive airway disease.

Other risk factors include viral infections during infancy, particularly respiratory syncytial virus (RSV) and human rhinovirus, which are predictors of subsequent asthma development.

Infections during the neonatal period, such as the need for continuous positive airway pressure or supplemental oxygen, also increase the risk of recurrent wheezing in late preterm infants.

Additionally, genetic factors, such as a family history of asthma, and environmental factors, such as exposure to cockroach allergens, contribute to the risk of developing pediatric airway disease. Obesity and early puberty are also associated with an increased risk of asthma.

In summary, the risk factors for pediatric airway disease include preterm birth, perinatal inflammation, postnatal environmental exposures, viral infections, genetic predisposition, and other factors such as obesity and early puberty.

**SIGNS / SYMPTOMS**

* Noisy breathing (stridor) often indicates upper airway obstruction.
* Wheezing: A high-pitched whistling sound during exhalation due to narrowed lower airways.
* Coughing, which may be persistent or worse during infections or exercise.
* Difficulty breathing or shortness of breath: Can be mild or severe.
* Chest tightness or congestion.
* Fatigue or poor exercise tolerance due to limited airflow.

**DIAGNOSIS METHODS**

* Based on clinical history and physical examination.
* Use of a stethoscope to detect wheezing or abnormal breath sounds.
* Sometimes imaging (chest X-ray, CT scan) or bronchoscopy.
* Pulmonary function tests in older children.
* Monitoring symptom patterns, triggers, and response to treatment.

**TREATMENT OPTIONS**

* Depends on the underlying cause.
* For asthma and RAD: inhaled corticosteroids, bronchodilators (e.g., albuterol), avoiding triggers.
* For congenital anomalies or foreign bodies: surgical or procedural interventions.
* Supportive care with oxygen, hydration, and infection control as needed.

**OUTLOOK / PROGNOSIS**

If a healthcare provider says you have RAD, it means you have breathing problems, but they aren’t sure of the cause. They’ll conduct a physical exam and tests to help determine the cause of your breathing issues so you can get proper treatment.

They may also refer you to a healthcare provider who specializes in conditions that affect your lungs (pulmonologist).

***What’s the outlook if I have reactive airway disease?***

If a healthcare provider describes your symptoms as RAD, your treatment, recovery and management depend on an official diagnosis. Some conditions are treatable, while others may progressively get worse.

**WHEN TO SEE A DOCTOR / RED FLAG**

Call your local emergency number or get to an emergency room right away if you’re using a lot of energy to breathe (severe respiratory distress), aren’t responding to breathing treatments, have low oxygen levels or notice anaphylaxis symptoms, including:

* Difficulty breathing.
* Drop in blood pressure.
* Swollen lips.
* Sudden weakness.
* Fainting (syncope).
* Confusion.

**DIFFERENTIAL DIAGNOSIS**

***Diagnostic Considerations***

The clinical diagnosis of asthma in children aged 5 years or younger is often based on symptoms during and between viral respiratory tract infections. The symptom pattern is as follows:

* Cough, wheeze, and heavy breathing for more than 10 days during upper respiratory tract infections
* More than three episodes per year or severe episodes and/or night worsening
* Cough, wheeze, or heavy breathing during play or when laughing, between episodes
* Allergic sensitization, atopic dermatitis, food allergy, or family history of asthma

***Differential Diagnoses***

* Anaphylaxis
* Aspiration Pneumonitis and Pneumonia
* Emergent Management of Croup (Laryngotracheobronchitis)
* Pediatric Acute Respiratory Distress Syndrome
* Pediatric Anaphylaxis
* Pediatric Epiglottitis
* Pediatric Foreign Body Ingestion
* Pediatric Pneumonia

***PROCEDURES***

Procedures include the following:

* Spirometry (decreased forced expiratory volume in one second [FEV1])
  + Bedside spirometry is the primary procedure for children with RAD who are older than 5 years.
  + Patients with decreased FEV1 require further evaluation and treatment.
* A barium swallow may be indicated to determine any esophageal, pulmonary, or vascular pathology, particularly a tracheoesophageal fistula.
* Bronchoscopy (rarely indicated) (see Table 1 below)
* Peak expiratory flow (PEF) is the most common form of pulmonary function test monitoring. Record the best of 3 attempts. Possible life-threatening asthma exacerbation with PEF predicted of less than 30%; severe exacerbation, with less than 50%; and moderate exacerbation, with less than 80%.

Peak flow rates are described in the table below.

***Table 1. Peak Flow Rates in Liters per Minute***

| Height in  Inches | Average  Rate | Range\* | Height in  Inches | Average  Rate | Range\* |
| --- | --- | --- | --- | --- | --- |
| 40 | 150 | 110-190 | 56 | 330 | 240-420 |
| 41 | 160 | 115-205 | 57 | 340 | 240-420 |
| 42 | 170 | 120-220 | 58 | 360 | 260-460 |
| 43 | 180 | 130-220 | 59 | 375 | 270-480 |
| 44 | 190 | 135-245 | 60 | 390 | 280-500 |
| 45 | 200 | 145-255 | 61 | 400 | 290-510 |
| 46 | 210 | 150-270 | 62 | 415 | 300-530 |
| 47 | 220 | 160-280 | 63 | 430 | 310-550 |
| 48 | 230 | 165-295 | 64 | 445 | 320-570 |
| 49 | 240 | 175-305 | 65 | 460 | 330-590 |
| 50 | 250 | 180-320 | 66 | 480 | 345-615 |
| 51 | 260 | 190-330 | 67 | 500 | 360-640 |
| 52 | 270 | 195-345 | 68 | 515 | 370-660 |
| 53 | 280 | 200-360 | 69 | 530 | 380-680 |
| 54 | 300 | 215-385 | 70 | 550 | 395-705 |
| 55 | 315 | 225-405 | 71 | 570 | 410-730 |

**RECENT GUIDELINES OR UPDATES**

Recommendations include the following:

* The ERS recommends spirometry as first-line diagnosis in children aged 5-16 years with suspected asthma.
* ERS recommends bronchodilator reversibility testing as first-line diagnosis in all children with FEV1 < LLN or < 80% predicted and/or FEV1/FVC < LLN or < 80% predicted.
* ERS recommends FeNO as first-line diagnosis in children aged 5-16 years with suspected asthma.
* ERS recommends against PEFR variability testing as the primary objective test on its own to diagnose asthma in children aged 5-16 years.
* ERS recommends against diagnosing asthma in children aged 5-16 years based on clinical history alone or following a single abnormal objective test.
* ERS recommends against using an improvement in symptoms after a trial of preventive medication alone to diagnose asthma in children aged 5-16 years.
* ERS recommends against using skin prick tests as diagnostic tests for asthma in children aged 5-16 years.
* ERS recommends a direct bronchial challenge test using methacholine in children aged 5-16 years with suspected asthma where asthma diagnosis could not be confirmed with first-line tests.
* ERS recommends an indirect bronchial challenge test using a treadmill or a bicycle in children aged 5-16 years with suspected asthma with exercise-related symptoms where asthma diagnosis could not be confirmed with first-line tests.

**EPIDEMIOLOGY**

***United States statistics***

Pediatric asthma is a chronic, multifactorial, lower airway disease that affects 5-15% of children (2.7 million children in the United States alone). In the United States, approximately one half of all emergency department (ED) and clinic visits for asthma are children younger than 18 years.

ED visits peak in the fall, whereas school holidays disrupt the spread of infections, resulting in a subsequent decrease in ED visits and hospitalizations. Status asthmaticus appears to be on the rise; several retrospective studies reflect an increase in hospital admissions, particularly in those younger than 4 years. Fewer hospital and ED visits occur in children using inhaled corticosteroid therapy.

Asthma prevalence appears to be increasing worldwide. Air pollutants may play a role in the prevalence increase. Higher prevalence occurs in poverty stricken urban areas where children are less likely to have routine doctor visits and readily available access to medications.

A correlation may exist between high levels of exposure to cockroach allergen and the frequency of asthma-related health problems in inner-city children. Homes in poverty areas were more likely to have high cockroach allergen levels.

Asthma may develop in children from early exposure to cockroach allergen.An association may exist between obesity and childhood asthma. Increased resistance, an adipokine produced by adipose tissue, may play a negative predictive role in asthma.

An algorithm has been developed to determine the risk factors for developing persistent asthma symptoms among children younger than 3 years of age who had 4 or more episodes of wheezing during the previous year.

The Asthma Predictive Index included either (1) one of the following: parental history of asthma, a physician diagnosis of atopic dermatitis, or evidence of sensitization to aeroallergens; or (2) two of the following: evidence of sensitization to foods, ≥4% peripheral blood eosinophilia, or wheezing apart from colds.

***International statistics***

Worldwide, the prevalence of asthma is increasing. Asthma is more common in Western countries than in developing countries. Asthma is more prevalent in English-speaking countries. Prevalence increases as a developing country becomes more Westernized and urbanized.

***Race-, sex-, and age-related demographics***

Reactive airway disease is more common in Black and Hispanic children; hospitalization rates in African Americans are 4 times greater than in the White population.

A correlation may exist between high levels of exposure to cockroach allergen and the frequency of asthma-related health problems in inner-city children. No correlation exists between education levels from a retrospective review.

The male-to-female ratio is 1.5:1. The peak prevalence of asthma is in those aged 6-11 years.

***Impact of COVID-19 on pediatric asthma exacerbation***

Pediatric asthma attacks decreased during the COVID-19 pandemic, probably because of reduced environmental allergen exposure and decreased risk of other viral respiratory tract infections. Asthma is not considered to be a risk factor for COVID-19 severity.

**PREDEFINED Q & A SETS**

Q1: “What is Pediatric Airway Disease?”

A: Pediatric airway disease refers to various conditions causing obstruction, inflammation, or dysfunction of the airways in children. It includes diseases like asthma, reactive airway disease (RAD), congenital airway anomalies, infections like bronchiolitis or croup, and foreign body aspiration.

Q2: “What are common symptoms of pediatric airway diseases?”

A:

* Wheezing (a high-pitched whistling sound when breathing out)
* Noisy breathing (stridor, especially indicating upper airway obstruction)
* Persistent cough
* Shortness of breath or difficulty breathing
* Chest tightness or congestion
* Fatigue due to poor oxygenation or sleep disruption
* In infants, symptoms may include grunting, fussiness, or difficulty feeding

Q3: “What causes pediatric airway diseases?”

A:

* Reactive airway inflammation often due to viral infections or allergens (as in asthma or RAD)
* Congenital abnormalities like laryngomalacia or tracheomalacia
* Respiratory infections such as bronchiolitis or croup
* Foreign body inhalation causing sudden airway blockage
* Other causes: reflux disease, allergies, or tumors (rare)

Q4: “How are pediatric airway diseases diagnosed?”

A:

* Primarily through clinical history and physical examination (listening to breathing sounds)
* In older children, pulmonary function tests such as spirometry can be used
* Imaging (chest X-ray, CT, MRI) or endoscopy like bronchoscopy may be done to assess airway structure or foreign bodies
* Allergy testing may be used if allergic triggers are suspected

Q5: “How are pediatric airway diseases treated?”

A:

* Treatment depends on the cause. For asthma and RAD: inhaled bronchodilators (e.g., albuterol) and corticosteroids to reduce inflammation
* Avoidance of triggers such as allergens, smoke, and infections
* Supportive care for infections (hydration, oxygen if needed)
* Surgical intervention for structural anomalies or removal of foreign bodies
* Education on asthma action plans and monitoring for early signs of exacerbation

Q6: “What are the complications or risks?”

A:

* Severe breathing difficulty or respiratory failure if untreated
* Recurrent infections or chronic lung damage in some cases
* Impact on growth, sleep, exercise tolerance, and quality of life

Q7: “When should I seek medical attention for my child?”

A:

* If your child shows persistent or worsening breathing difficulty
* If there is noisy or labored breathing, bluish discoloration (cyanosis), or fatigue
* If symptoms interfere with feeding, sleeping, or daily activity
* If a foreign body aspiration is suspected (sudden choking or inability to breathe properly)

**DOCTOR-PATIENT CONVERSATIONS**

Doctor: Hello, I understand you are concerned about your child’s breathing. Can you tell me what symptoms you have noticed?

Parent: Yes, my child has been wheezing a lot and sometimes has noisy breathing. They also cough frequently and sometimes seem short of breath.

Doctor: I see. These symptoms can be caused by various pediatric airway diseases, which means there is some obstruction or inflammation in your child’s airways making it harder to breathe.

Parent: What could be causing this? Is it serious?

Doctor: Common causes include asthma, viral infections like bronchiolitis, or sometimes structural issues in the airway such as laryngomalacia. The severity varies, but with proper treatment many children improve significantly.

Parent: How do you know what exactly it is?

Doctor: We start by examining your child, listening to their lungs, and reviewing their medical history. Sometimes we need tests like chest X-rays or breathing tests if your child is old enough. In some cases, further evaluation like bronchoscopy is considered.

Parent: What kinds of treatments are there?

Doctor: Treatment depends on the diagnosis. For asthma or reactive airway disease, we often use inhalers that open up the airways and steroids to reduce inflammation. For infections, supportive care with fluids and sometimes medications is given. Structural problems may require monitoring or surgery in rare cases.

Parent: Will my child get better? Is this something they will have for life?

Doctor: Many children outgrow these problems or learn to manage them well with medication and avoiding triggers like smoke or allergens. Early diagnosis and treatment are important to prevent complications.

Parent: What should I watch for that means I should call you or come back?

Doctor: If your child has increased difficulty breathing, persistent wheezing, blue lips, trouble feeding, lethargy, or if your child is not responding to usual medications, please contact us immediately.

Parent: Thank you, Doctor. That helps me understand what to expect and what to do.

*REFERENCES:*

<https://emedicine.medscape.com/article/800119-overview>

<https://www.boystownhospital.org/knowledge-center/reactive-airway-disease-children>

**PEDIATRIC HEAD MASSES**

*ALTERNATIVE NAMES /* **DEFINITION / DESCRIPTION**

Head and neck masses in children can have various alternative names depending on their origin and characteristics. Some common alternative names include:

* Thyroglossal duct cyst: A cyst that forms from remnants of the thyroglossal duct, which is a structure that develops during fetal growth and normally disappears after birth.
* Branchial cleft cyst: A cyst that develops from remnants of the branchial clefts, which are structures that form during fetal development and usually disappear after birth.
* Dermoid cyst: A cyst that contains skin structures such as hair, sweat glands, and sebaceous glands.
* Lymph node enlargement: Also known as lymphadenopathy, this refers to the swelling of lymph nodes, which can occur due to infections or other conditions.
* Neck mass: A general term for any lump or bump that develops on a child's neck.
* Congenital neck mass: A mass that is present at birth, which can be due to various developmental abnormalities.

**CAUSES**

A head or neck mass may be triggered by a variety of causes, including infection, tumors or cysts or masses formed while in the womb.

Conditions related to pediatric head and neck masses include:

* Branchial cleft anomaly. A branchial cleft anomaly is a cyst that develops in one or both sides of a child’s neck or below the collarbone. This condition is a birth defect and is also known as branchial cleft or pharyngeal arch remnant.
* Thyroglossal duct cyst. A thyroglossal duct cyst is a neck mass or lump that develops from cells and tissues left behind after the formation of the thyroid gland while a child is developing in the womb. Such cysts are most common in preschool-aged children or children in mid-adolescence, and they most often appear after an upper respiratory infection causes the cyst to become enlarged and painful.
* Enlarged lymph nodes. Enlarged (swollen) lymph nodes usually occur because of exposure to bacteria or viruses.
* Neck abscess. A neck abscess occurs when pus from an infection—such as a cold, tonsillitis, a sinus infection or an ear infection—gathers in the spaces between the structures of the neck. Neck abscesses are also known as cervical abscesses or deep neck infections.
* Head and neck cancer (tumors). Rare in children, head and neck cancer covers a group of cancers that can occur in the oral cavity, pharynx, larynx, paranasal sinuses and nasal cavity or the salivary glands.

**SIGNS / SYMPTOMS**

Symptoms of a branchial cleft anomaly include:

* Dimple, lump or skin tag on the neck, upper shoulder or just below the collarbone
* Fluid draining from the neck
* Swelling or tenderness in the neck, usually combined with an upper respiratory infection

The most common symptoms of a thyroglossal duct cyst are:

* Small, soft, round mass in the center of the front of the neck
* Tenderness, redness and swelling of the mass (if infected)
* Small opening in the skin near the mass with drainage
* Difficulty swallowing

The main symptoms of enlarged lymph nodes include:

* Tender and painful lymph nodes
* Swollen lymph nodes that may be the size of a pea or kidney bean or even larger

Depending on the cause of the enlarged lymph nodes, your child may also experience:

* Runny nose, sore throat or fever
* General swelling of the lymph nodes throughout the body
* Hardened, fixed and rapidly growing nodes
* Night sweats

Symptoms of a neck abscess can include:

* Fever
* Red, swollen, sore throat (sometimes on only one side) or bulge in the back of the throat
* Tongue pushed back against the throat
* Neck pain or stiffness
* Ear pain
* Body aches or chills
* Difficulty swallowing, talking or breathing

Symptoms of head and neck cancer include:

* Long-lasting sore throat or difficulty swallowing
* Change or hoarseness of the voice
* Mass in the neck or elsewhere in the head

Your child may exhibit additional symptoms based on where the tumor is located. Symptoms of cancer of the oral cavity include:

* White or red patching on the gums, tongue or lining of the mouth
* Swelling of the jaw
* Unusual bleeding or pain in the mouth

Symptoms of cancer of the pharynx include:

* Difficulty breathing or speaking
* Pain while swallowing or long-lasting pain in the neck or throat
* Frequent headaches
* Pain in the ears
* Difficulty hearing

Symptoms of cancer of the larynx include:

* Pain while swallowing
* Pain in the ears

Symptoms of cancer of the paranasal sinuses and nasal cavities include:

* Chronic sinus infections that do not respond to antibiotic treatment
* Nose bleeds
* Frequent headaches, swelling or other trouble with the eyes
* Pain in the upper teeth
* Blocked sinuses that will not clear

Symptoms of cancer of the salivary glands include:

* Swelling under the chin or around the jawbone
* Numbness or paralysis of the muscles in the face
* Persistent pain in the face, chin or neck

Many of the above symptoms occur with much more common, less severe conditions, so it is important for your child to be evaluated by a doctor in order to accurately diagnose head and neck cancer.

**DIAGNOSIS METHODS**

***Diagnosis of Head & Neck Masses***

To diagnose a head or neck mass, a pediatric ear, nose and throat specialist will perform a physical exam and review your child's medical history and current symptoms to narrow down the cause of symptoms.

A branchial cleft anomaly can be diagnosed using the following exams and tests:

* Physical exam. Your child's doctor will examine the neck and collarbone area to check for cysts.
* Imaging tests. The doctor may also order imaging tests such as magnetic resonance imaging (MRI), computed tomography (CT) or ultrasound to pinpoint the exact location of the anomaly.

A thyroglossal duct cyst can be diagnosed using the following exams and tests:

* Physical exam. Your child’s doctor will examine the throat and neck for signs of a cyst.
* Blood test. The doctor may order a blood test to assess your child’s thyroid function.
* Ultrasound. Ultrasound can be used to evaluate the mass and surrounding tissues.
* Thyroid scan. A thyroid scan is a type of nuclear medicine test that your child’s doctor may order to check the function of the thyroid.
* Fine needle aspiration. A fine needle aspiration is a type of biopsy that uses a thin needle to gather tissue that will be analyzed in a lab.

Enlarged lymph nodes can be diagnosed using the following exams and tests:

* Physical exam. The doctor will examine the lymph nodes near the surface of the skin for size, tenderness, warmth and texture.
* Blood test. Depending on what the doctor suspects is causing the enlarged lymph nodes, he or she may order a complete blood count (CBC) blood test to evaluate your child’s overall health and check for a range of conditions, including infections and leukemia.
* Imaging tests. Imaging tests such as an X-ray or CT scan can help the doctor determine potential sources of infection or to locate tumors.
* Biopsy. The doctor may remove a tiny sample of the enlarged lymph node for closer analysis in the lab.

A neck abscess can be diagnosed using the following exams and tests:

* Physical exam. The doctor will examine your child’s neck for the presence of an abscess.
* Throat culture. A throat culture involves collecting cells by swabbing the back of the throat. The tissue sample is sent to a laboratory to determine the type of organism causing the infection.
* Blood test. A blood test may be performed in order to measure the body’s response to infection.
* Biopsy. The doctor may remove a tiny sample of the abscess for closer analysis in the lab. Generally, if the doctor operates on a neck abscess to obtain the biopsy sample, he or she will also make an incision and drain the area.
* Imaging tests. The doctor may order an X-ray or CT scan in order to get a more detailed look at the abscess and surrounding tissues and bones.

Head and neck cancer can be diagnosed using the following exams and tests:

* Physical exam. A complete physical exam will be performed, including checking the location and size of the head or neck mass.
* Biopsy. The doctor will remove a sample of tissue from the mass to confirm if it is benign or malignant (cancerous).

**TREATMENT OPTIONS**

If your child has a branchial cleft anomaly accompanied with signs of infection, the doctor will likely prescribe antibiotic medicines to fight the infection. The doctor may also need to drain fluid from the cyst to reduce swelling. In some cases, surgery may be needed to prevent future infections.

This surgery is usually an outpatient procedure performed under general anesthesia. Recovery is relatively short, and your child can usually resume normal activity within a few days.

If your child has a thyroglossal duct cyst, the doctor may prescribe antibiotic medicine to treat the infection or recommend surgery to remove the cyst and the thyroglossal duct. This procedure is known as the Sistrunk procedure.

If your child has enlarged lymph nodes, treatment will depend on the underlying condition causing the condition. If the swollen lymph nodes are caused by a bacterial infection, the doctor may prescribe a course of antibiotic medicine.

If they are caused by certain rheumatologic conditions, such as lupus or arthritis, the doctor will treat that underlying condition. If the swollen lymph nodes are caused by cancer, the cancer will need to be treated with surgery, radiation therapy and/or chemotherapy.

If your child has a neck abscess, the doctor may prescribe antibiotic medicine to treat the infection. Depending on your child’s specific condition, the abscess may need to be drained through surgery using a needle. This procedure may require hospitalization.

The treatment of head and neck cancer is based on the location of the tumor, stage of the cancer and your child’s overall health. The doctor may recommend surgical removal of the tumor and chemotherapy or radiation therapy to shrink the tumor.

**PREVENTION TIPS**

Neck masses in children are usually benign and can be categorized into developmental, inflammatory, or neoplastic types. Preventive measures and early recognition are crucial for effective management. Here are some prevention tips and considerations:

* Avoiding exposure to infectious agents: Practicing good hygiene and avoiding contact with sick individuals can help prevent infections that may lead to inflammatory neck masses. For example, avoiding animal exposures like cat scratches can reduce the risk of infections such as cat-scratch disease.
* Regular health check-ups: Routine pediatric visits allow for early detection of any unusual masses or changes in the child's health. This is particularly important for monitoring any existing conditions or symptoms that may indicate a more serious issue.
* Maintaining a healthy lifestyle: Ensuring proper hydration and nutrition can support the immune system, potentially reducing the risk of infections that might contribute to the development of neck masses.
* Avoiding unnecessary medications: Some medications, such as phenytoin, can cause pseudolymphoma or lymphadenopathy. It is important to use medications only as prescribed and to discuss any concerns with a healthcare provider.
* Educating caregivers: Teaching caregivers about the signs and symptoms of neck masses, including changes in size, consistency, or associated symptoms, can facilitate early intervention.

**OUTLOOK / PROGNOSIS**

The outlook and prognosis for pediatric head masses vary depending on the type of mass, its location, and the treatment approach. For example, pilocytic astrocytoma (PA), a common brain tumor in children, generally has a favorable prognosis with surgical removal being the primary treatment.

However, the prognosis can be more complex for other types of brain tumors, such as medulloblastoma, which is more aggressive and has a poorer prognosis, especially in younger patients or when there is incomplete surgical resection or the presence of cerebrospinal fluid (CSF) metastases.

* Pilocytic Astrocytoma (PA): The prognosis is generally favorable with early detection and surgical removal. Regular follow-up is essential to monitor for recurrence.
* Medulloblastoma: This type of brain tumor is more aggressive and has a poorer prognosis, particularly in young children. Factors such as age at diagnosis, extent of surgical resection, and presence of CSF metastases significantly influence the prognosis.

**POSSIBLE COMPLICATIONS**

Ultrasound (US) is the first-line imaging modality for evaluating superficial pediatric head and neck masses, as it provides a quick, cost-effective, and non-invasive assessment of the mass's location, size, shape, internal content, and vascularity. However, in certain situations, further evaluation with cross-sectional imaging may be necessary.

For instance, if the mass is too large, deep, or hyperechoic to be fully assessed within the US field of view, or if malignancy or a high-flow vascular lesion is suspected, additional imaging such as MRI or CT may be required.

In terms of possible complications, pediatric head and neck masses can lead to various issues depending on their nature. For example, lymph nodes and their complications can include infections, abscesses, or even malignancies such as lymphoma.

Other potential complications include:

* Infection: Masses such as branchial cleft cysts or thyroglossal duct cysts can become infected, leading to inflammation, pain, and the formation of abscesses.
* Malignancy: While most pediatric head and neck masses are benign, some can be malignant, such as rhabdomyosarcoma or thyroid papillary carcinoma.
* Obstruction: Masses located in the airway or throat can cause breathing difficulties or swallowing problems.
* Neurological complications: Tumors near the brain or spinal cord can cause neurological symptoms, including headaches, seizures, or motor deficits.
* Cosmetic concerns: Some masses, such as hemangiomas or lymphangiomas, can cause visible deformities or affect the child's appearance.

In cases where a mass is suspected to be malignant, imaging techniques like PET/CT may be used for staging, and biopsy or excision may be necessary for a definitive diagnosis.

Additionally, in some cases, surgical intervention may be required to remove the mass, especially if it is causing symptoms or is suspected to be cancerous.

**WHEN TO SEE A DOCTOR / RED FLAG**

* The mass is rapidly growing, painful, or hard and fixed to underlying structures
* Presence of redness, tenderness, warmth, or pus, suggesting infection
* Associated systemic symptoms such as fever, weight loss, night sweats, or fatigue
* Neurologic symptoms like headaches, vomiting (especially projectile or recurrent), seizures, vision changes, weakness, difficulty walking or swallowing, or changes in behavior or personality
* Persistent or enlarging mass that does not resolve over weeks
* Difficulty breathing or swallowing related to the mass
* Any new onset of seizures or unprovoked neurological symptoms in a child with a head mass

**DIFFERENTIAL DIAGNOSIS**

1. Congenital Masses
   * Thyroglossal duct cyst: Midline neck mass that moves with swallowing or tongue protrusion; common in preschool or adolescent children.
   * Branchial cleft cysts: Usually located along the anterior border of the sternocleidomastoid muscle; second branchial cleft cyst is most common.
   * Dermoid cysts: Midline or lateral cystic masses often present since infancy.
   * Lymphatic malformations (cystic hygroma): Soft, compressible masses often in the posterior triangle of the neck.
   * Vascular malformations: Including hemangiomas (infantile), venous or capillary malformations.
   * Epidermoid cysts and others: Less common but possible.
2. Inflammatory / Infectious Masses
   * Reactive lymphadenopathy: The most common pediatric neck mass; often follows upper respiratory infections.
   * Suppurative lymphadenitis and abscesses: Tender, swollen lymph nodes with signs of infection; may require antibiotics or drainage.
   * Tuberculous lymphadenitis: Chronic, painless lymphadenopathy especially in endemic regions.
   * Cat scratch disease: Bartonella henselae infection causing regional lymphadenitis.
   * Inflammatory cysts/abscesses: Secondary to infections in the head and neck region.
3. Neoplastic Masses
   * Benign tumors: Such as hemangiomas, neurofibromas, or lipomas.
   * Malignant tumors: Less common but critical to consider — includes lymphoma (most common malignancy in head and neck), rhabdomyosarcoma, neuroblastoma metastases, thyroid carcinoma, sarcomas, salivary gland malignancies.
4. Other Causes
   * Salivary gland enlargement or tumors (including sialadenitis, cysts, or tumors)
   * Congenital cervical thymic cyst
   * Parathyroid or paragangliomas (rarely in pediatrics)

**EPIDEMIOLOGY**

* A large population-based study (1973–2008) using the SEER registry reported the most common pediatric head and neck tumors as salivary gland tumors, nasopharyngeal tumors, tumors of the nose/nasal cavity/middle ear, gum and other mouth tumors, and glossal (tongue) tumors. Salivary gland tumors were most frequent overall.
* In children under 12 years, most head and neck tumors are benign (about 68%), with lymphangiomas common benign lesions and non-Hodgkin lymphoma the most frequent malignant tumor identified.
* Among children aged 0 to 2 years, studies showed about 2.7% prevalence of head and neck masses in one hospital-based Iranian cohort, with congenital lesions (around 41%) and inflammatory/reactive lesions (about 35%) being the most common, followed by tumors (benign and malignant combined about 24%)—with malignancies about 3.6% of lesions.
* Males are generally slightly more affected than females with a male-to-female ratio ranging from about 1.2:1 to 1.7:1 in different studies.
* Locations vary, but the neck is the most common site for these masses, followed by oral cavity and salivary glands in many series.
* Pediatric head and neck malignancies are rare relative to benign masses but important. Lymphomas, rhabdomyosarcoma, neuroblastoma, and salivary gland malignancies comprise the majority of malignant cases with incidence varying by age group.
* Racial/ethnic disparities exist, e.g., nasopharyngeal cancers are more prevalent in Black and other non-White pediatric populations, while salivary gland tumors are more common in White pediatric patients

**PREDEFINED Q & A SETS**

Q1: “What is a pediatric head mass?”

A: A pediatric head mass is a lump or swelling in a child's head or neck area that can be caused by various benign or malignant conditions, including congenital cysts, infections, or tumors.

Q2: “What are the common causes of pediatric head masses?”

A:

* Congenital cysts or anomalies: such as thyroglossal duct cyst, branchial cleft cyst, dermoid cyst, or lymphatic malformations (cystic hygroma).
* Reactive or infectious lymphadenopathy: swollen lymph nodes often due to recent infections or inflammation.
* Benign tumors: such as hemangiomas or lipomas.
* Malignant tumors: including lymphomas, rhabdomyosarcoma, neuroblastoma, and thyroid cancer (less common).
* Other causes: salivary gland masses, abscesses, or thyroid masses.

Q3: “How can I tell if a head mass is serious?”

A:

* Rapid growth or hard, fixed swelling
* Pain, redness, warmth, or pus (suggesting infection)
* Associated systemic symptoms like fever, weight loss, night sweats, or fatigue
* Neurological symptoms such as headaches, vomiting, seizures, vision problems, or difficulty swallowing or breathing

Q4: “When should I see a doctor for my child’s head mass?”

A:

* If the mass is persistent for more than a few weeks or continues to grow
* If the mass is painful, red, warm, or draining pus
* If your child experiences systemic symptoms like fever or weight loss
* If there are signs of breathing difficulty, swallowing problems, or neurological symptoms

Q5: “How is a pediatric head mass diagnosed?”

A:

* Detailed medical history and physical examination
* Imaging tests such as ultrasound (common initial test), CT scan, or MRI to assess size, location, and characteristics
* Blood tests if infection or malignancy is suspected
* Fine needle aspiration biopsy or surgical biopsy in selected cases

Q6: “What treatments are used for pediatric head masses?”

A:

* Congenital cysts or benign masses: often surgical removal if symptomatic or cosmetically concerning
* Infectious causes: antibiotics and sometimes drainage of abscesses
* Malignant tumors: require specialized cancer treatment including surgery, chemotherapy, and/or radiation
* Supportive care: monitoring small, asymptomatic masses without immediate intervention

Q7: “Will my child fully recover from a pediatric head mass?”

A: Many pediatric head masses are benign and treatable with full recovery expected. Malignant masses require prompt treatment and careful follow-up. Early diagnosis and intervention improve outcomes.

**DOCTOR-PATIENT CONVERSATIONS**

Doctor: Hello, I see you’re here because of a lump or swelling on your child’s head or neck. Can you tell me when you first noticed it and if it has changed in size or caused any symptoms?

Parent: Yes, we found a small lump on the side of my child’s neck about a few weeks ago. It hasn’t been painful, but it seems to be growing slowly.

Doctor: Thank you for sharing that. Pediatric head masses are fairly common and can have many causes. Some lumps are congenital cysts that children are born with, others might be swollen lymph nodes due to infections, and less commonly some can be tumors.

Parent: What should I be worried about?

Doctor: Typically, if the mass is painless, soft, and has been stable or growing slowly without other symptoms, it’s usually benign, like a cyst or reactive lymph node. However, if the mass grows rapidly, becomes hard or fixed, causes pain, redness, or your child has symptoms like fever, night sweats, weight loss, or trouble swallowing or breathing, then it needs prompt evaluation.

Parent: What will you do to find out what it is?

Doctor: I will begin with a physical exam to check the lump’s size, consistency, and tenderness. Then, we often do an ultrasound, which helps us see if the mass is solid or cystic and its relation to nearby structures. Sometimes, blood tests or further imaging such as CT or MRI may be needed. If there is any concern about malignancy, a biopsy might be required.

Parent: Will my child need surgery?

Doctor: That depends on the cause. Many benign masses can be monitored or removed if they cause symptoms or cosmetic concerns. If it’s an infection, antibiotics or drainage might be needed. If it’s a tumor, your child will be referred to a specialist for further treatment, which may include surgery and other therapies.

Parent: What can I do at home in the meantime?

Doctor: Watch the mass closely. If it gets bigger quickly, becomes tender, red, or if your child develops systemic symptoms like fever or weight loss, bring them back immediately. Otherwise, try to keep the area clean and avoid any trauma.

Parent: Thank you, Doctor. That helps me feel more informed and less worried.

Doctor: You’re welcome. We will take good care of your child and keep you informed every step of the way.

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**PEDIATRIC NASAL MASSES**

**DEFINITION / DESCRIPTION**

Pediatric nasal masses are abnormal growths or lesions occurring in the nose, nasal cavity, or nasopharynx of children. They can be either congenital (present at birth) or acquired later, and may be benign (non-cancerous) or malignant (cancerous). These masses can arise from various nasal structures including the mucosa, cartilage, bones, and adjacent areas such as the nasopharynx.

Common types of pediatric nasal masses include:

* Congenital and developmental lesions such as nasolacrimal duct mucocele, dermoid cysts, encephaloceles, and nasal neuroglial heterotopia.
* Inflammatory and infectious masses like mucoceles, polyps, and pyogenic granulomas.
* Benign tumors including infantile hemangioma and juvenile nasopharyngeal angiofibroma.
* Malignant tumors such as rhabdomyosarcoma and nasopharyngeal carcinoma.
* Masses related to trauma (e.g., septal hematoma)

**CAUSES**

Common Types and Causes of Pediatric Nasal Masses

* Congenital/developmental masses: such as dermoid cysts, encephaloceles (brain tissue herniation), nasal gliomas (neuroglial heterotopia), and nasolacrimal duct mucoceles
* Inflammatory/infectious masses: mucoceles, nasal polyps (rare in young children), pyogenic granulomas
* Benign tumors: infantile hemangiomas, juvenile nasopharyngeal angiofibromas (vascular tumors usually in adolescent males)
* Malignant tumors: rhabdomyosarcoma (most common nasal malignancy in children), nasopharyngeal carcinoma, lymphoma
* Trauma-related masses: septal hematomas

**SIGNS / SYMPTOMS**

* Nasal obstruction or congestion (often one-sided/stuffy nose)
* Nasal discharge, which may be clear, thick, or bloody
* Epistaxis (nosebleeds)
* Postnasal drip (drainage down the back of the throat)
* Facial pain or headache (especially around eyes or nose)
* Snoring or sleep apnea symptoms
* Reduced or loss of smell
* Other possible symptoms: difficulty breathing through the nose, mouth breathing, swelling of the face, ear infections, or even eye symptoms like bulging or double vision if the mass is extensive
* Respiratory distress or obstructive sleep apnea may be seen in severe cases

**DIAGNOSIS METHODS**

1. Clinical Evaluation
   * Detailed medical history focusing on onset, duration, progression, associated symptoms (nasal obstruction, discharge, epistaxis, facial swelling, sleep issues).
   * Physical examination including anterior rhinoscopy and possibly nasal endoscopy to directly visualize the nasal cavity and identify the mass.
2. Imaging Studies
   * Magnetic Resonance Imaging (MRI): The preferred imaging modality in children due to its detailed soft tissue resolution without radiation exposure. MRI helps define the lesion’s extent, vascularity, and is critical to assess for any intracranial extension, especially in congenital midline lesions (e.g., encephaloceles).
   * Computed Tomography (CT) scan: Used as a complementary tool to assess bone involvement, calcifications, or when MRI is not feasible. Also useful for surgical planning.
   * Ultrasound may be used for superficial masses or associated neck masses but is less common for nasal cavity lesions.
3. Imaging is essential before biopsy or surgical intervention because some lesions have intracranial connections or are highly vascular (e.g., juvenile nasopharyngeal angiofibroma), where biopsy could cause serious complications like cerebrospinal fluid leak or severe bleeding.
4. Biopsy and Histopathological Analysis
   * Performed cautiously and usually after imaging rules out contraindications.
   * Necessary for definitive diagnosis of neoplastic or infectious masses.
   * Some congenital lesions may not require biopsy if imaging and clinical features are diagnostic.
5. Laboratory Tests
   * Blood tests to evaluate for inflammation or infection if suspected.
   * Allergy testing if nasal polyps or inflammatory processes are considered.
   * Additional tests (e.g., sweat chloride for cystic fibrosis) in specific contexts.

**TREATMENT OPTIONS**

1. Congenital and Benign Masses

* Surgical Removal: Most congenital masses such as dermoid cysts, encephaloceles, nasal gliomas, and nasolacrimal duct mucoceles are treated by surgical excision, often via endoscopic or external approaches depending on size and location.
* Infantile Nasal Hemangiomas: These benign vascular tumors usually undergo a phase of rapid growth in infancy followed by spontaneous involution.
  + First-line treatment: Propranolol (a beta-blocker) with or without corticosteroids to control growth during the proliferative phase.
  + Larger or complicated lesions may require surgery.
* Nasal Polyps:
  + Medical management with intranasal corticosteroids is first-line.
  + In severe or refractory cases, a short course of oral steroids may be given.
  + Surgical removal via endoscopic sinus surgery is reserved for persistent or causing obstruction.
  + Allergy testing and treatment of underlying causes (e.g., allergic rhinitis) are important adjuncts.

## 2. Malignant Tumors (e.g., nasopharyngeal carcinoma, rhabdomyosarcoma)

* Multimodal Therapy: Includes surgery where feasible, combined with chemotherapy and radiation therapy.
* Radiation therapy modalities include external beam and brachytherapy.
* Chemotherapy regimens are selected based on tumor type and stage.
* Immunotherapy and targeted therapies may be options in specific cases or trials.
* Treatment plans are individualized in specialized pediatric oncology centers.

## 3. Surgical Approaches

* Endoscopic Surgery: Preferred for many intranasal and benign lesions for minimal invasiveness and quicker recovery.
* External Approaches: Such as lateral rhinotomy or external rhinoplasty incisions are used for large, extensive, or externally visible lesions with or without intracranial extension.
* Combined Neurosurgical Approaches: Required for lesions with intracranial connections (e.g., encephaloceles).

## 4. Supportive and Adjunctive Care

* Antibiotics if associated infection is present.
* Management of airway obstruction and breathing difficulties if any.
* Close monitoring and follow-up for recurrence or complications.

**OUTLOOK / PROGNOSIS**

1. Benign and Congenital Masses

* Generally, these have an excellent prognosis following appropriate surgical removal or medical management.
* For example, dermoid cysts, nasal gliomas, and nasolacrimal duct mucoceles usually resolve well after surgery with minimal risk of recurrence.
* Infantile hemangiomas often involute spontaneously or respond well to propranolol therapy, resulting in a good long-term outlook.
* Nasal polyps managed medically or surgically tend to have good outcomes, though underlying allergy control is important for recurrence prevention.

1. Malignant Nasal Masses (Sinonasal Tumors)

* Pediatric sinonasal malignancies like rhabdomyosarcoma, lymphoma, nasopharyngeal carcinoma, and other soft tissue sarcomas carry a more guarded prognosis.
* The overall 5-year survival rates reported in various studies range from approximately 50% to 70%, depending heavily on tumor type and stage at diagnosis.
* Solid tumors (non-lymphomas) tend to show better survival compared to lymphomas in some series.
* Early-stage diagnosis and complete surgical resection tend to improve survival; incomplete resection or biopsy only is associated with worse outcomes.
* Metastatic disease at diagnosis carries a poor prognosis.
* Advances in multimodal therapy (surgery, chemotherapy, radiation) have improved outcomes over time.

**DIFFERENTIAL DIAGNOSIS**

* Acute Sinusitis
* Asthma
* Genetics of Neurofibromatosis Type 1 and Type 2
* Neuroblastoma Imaging
* Pediatric Rhabdomyosarcoma
* Sinonasal Manifestations of Cystic Fibrosis

**EPIDEMIOLOGY**

In the United States, the overall incidence of nasal polyps in children is 0.1%; the incidence in children with CF is 6-48%. Among adults, the incidence is 1-4% overall, with a range of 0.2-28%. Worldwide incidence is the same as the incidence in the United States.

Benign multiple nasal polyposis usually manifests in patients older than 20 years and is more common in patients older than 40 years. Nasal polyps are rare in children younger than 10 years.

Although the male-to-female ratio is 2-4:1 in adults, the ratio in children is unreported. A review of articles reporting on children whose nasal polyposis required surgery showed apparently equal prevalence in boys and girls, though the data are inconclusive.

The reported prevalence is equal in patients with asthma. Nasal polyps occur in all races and social classes.

**PREDEFINED Q & A SETS**

Q1: “What are pediatric nasal masses?”

A: Pediatric nasal masses are abnormal growths or swellings inside the nose or nasopharynx of children. They may be present at birth (congenital) or develop later and can be benign (non-cancerous) or malignant (cancerous). Nasal masses are uncommon but can cause nasal obstruction, discharge, bleeding, and affect breathing or facial development.

Q2: “What causes pediatric nasal masses?”

A:

* Congenital/developmental masses: dermoid cysts, encephaloceles, nasal gliomas (neuroglial heterotopia), nasolacrimal duct mucoceles
* Inflammatory/infectious lesions: nasal polyps (rare in young children), mucoceles, pyogenic granulomas
* Benign tumors: infantile hemangiomas, juvenile nasopharyngeal angiofibromas
* Malignant tumors: rhabdomyosarcoma (most common nasal malignancy in children), nasopharyngeal carcinoma, lymphoma
* Trauma-related masses: septal hematomas or post-traumatic swelling

Q3: “What symptoms do nasal masses cause?”

A:

* Unilateral or bilateral nasal obstruction or congestion
* Nasal discharge (which may be clear, thick, or bloody)
* Nosebleeds (epistaxis)
* Facial swelling or pain
* Snoring, sleep disturbances, or breathing difficulty
* Possible ear symptoms from eustachian tube blockage (ear infections, hearing issues)
* Rarely, eye symptoms such as bulging or double vision if the mass is large

Q4: “How are pediatric nasal masses diagnosed?”

A:

* Clinical evaluation with anterior rhinoscopy and sometimes nasal endoscopy
* Imaging studies, usually MRI to define soft tissue and exclude brain connection for congenital masses, and CT scan for bone involvement
* Biopsy may be performed safely after imaging if malignancy or uncertain diagnosis is suspected
* Additional blood tests or allergy evaluation as indicated

Q5: “How are pediatric nasal masses treated?”

A:

* Congenital/developmental masses: Generally surgical removal, often involving ENT and neurosurgery in lesions with intracranial extension
* Infantile hemangiomas: Medical therapy with propranolol and sometimes corticosteroids; surgery for complicated cases
* Nasal polyps: Medical therapy with corticosteroids; surgery if medical treatment fails
* Malignant tumors: Multimodal treatment including surgery, chemotherapy, and radiotherapy managed by pediatric oncology teams
* Infectious/inflammatory lesions: Antibiotics or drainage if abscess present

Q6: “What is the prognosis for pediatric nasal masses?”

A:

* Most benign and congenital masses have excellent prognosis after appropriate treatment
* Malignant tumors have variable prognosis depending on tumor type, stage at diagnosis, and response to treatment; early detection improves outcomes
* Infantile hemangiomas typically involute or respond well to medical therapy
* Ongoing follow-up is important to monitor for recurrence or complications

Q7: “When should I seek medical attention for my child’s nasal mass?”

A:

* If your child has persistent or worsening nasal blockage, especially if one-sided
* Recurrent nosebleeds or bloody nasal discharge
* Facial swelling, pain, or new neurological or visual symptoms
* Signs of breathing difficulty, sleep problems, or ear infections

**DOCTOR-PATIENT CONVERSATIONS**

Doctor: Hello, I understand you’re concerned about a lump or swelling inside your child’s nose. Can you tell me what symptoms you have noticed and when you first saw the mass?

Parent: Yes, I noticed my child has been having a blocked nose on one side for several weeks. Sometimes there’s some nosebleed and occasional discharge. They also snore at night, which seems worse lately.

Doctor: Thank you for telling me that. Nasal masses in children, although uncommon, can cause symptoms like nasal obstruction, discharge, or bleeding. These masses can be due to different causes, from harmless congenital cysts or benign growths to more serious tumors. It’s important we understand what type your child might have.

Parent: Is it something that is there from birth, or can it happen later?

Doctor: It can be both. Some nasal masses are congenital, meaning children are born with them, such as dermoid cysts or encephaloceles, which are rare. Others can develop later, like benign tumors such as infantile hemangiomas or inflammatory masses, and in rare cases, malignant tumors like rhabdomyosarcoma.

Parent: That sounds serious. How do you find out what it really is?

Doctor: We start with a physical exam and look inside the nose with a small camera called a nasal endoscopy.

We will also order imaging studies like an MRI, which gives us detailed pictures to see the size and exact location of the mass and whether it connects with any other structures, like the brain, especially for congenital masses.

Parent: Will you need to take a biopsy?

Doctor: If the imaging suggests the mass might be a tumor or infection that needs further evaluation, we may do a biopsy to get a small tissue sample. But we only proceed with that after careful imaging, since some masses are highly vascular or connected to the brain, and biopsy in those cases can be risky.

Parent: How do you treat these masses?

Doctor: Treatment depends on what the mass is. Many benign or congenital masses are treated with surgery to remove them. Some vascular tumors like hemangiomas respond to medications like propranolol. If it’s an infection, antibiotics may help. For malignant tumors, specialized cancer therapy including surgery, chemotherapy, and radiation is used.

Parent: Is surgery safe for my child?

Doctor: Yes, surgery is usually safe and planned carefully by a multidisciplinary team including ENT surgeons and sometimes neurosurgeons if the mass is close to the brain. Early treatment helps prevent complications and improve outcomes.

Parent: What should I watch for while waiting for tests or treatment?

Doctor: Watch for any worsening nasal blockage, increased bleeding, swelling of the face, changes in vision or behavior, breathing difficulty, or signs of infection like fever. If any of these happen, please bring your child in immediately.

Parent: Thank you, Doctor. It helps to know what to expect.

Doctor: You’re welcome. We will keep you informed at each step and work together to provide the best care for your child.

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**PEDIATRIC OBSTRUCTIVE SLEEP APNEA**

ALTERNATIVE NAMES: Pediatric obstructive sleep apnea is also known by several alternative names. These include "sleep apnea in children, obstructive" and "obstructive sleep apnea in children". Additionally, it is sometimes referred to as "sleep-disordered breathing (SDB)".

**DEFINITION / DESCRIPTION**

Pediatric obstructive sleep apnea is a condition in which a child's breathing is partly or completely blocked during sleep. Breathing can briefly stop and start again many times a night. The condition happens when the upper airway narrows or is blocked during sleep.

Obstructive sleep apnea can look different in children than it does in adults. Adults usually have daytime sleepiness. Children are more likely to have behavior issues, such as acting hyper or not paying attention.

Risk factors also differ. In adults, the key risk factors are obesity and age. Although obesity can play a role in children, the main risk factor in children is having tonsils and adenoids that are larger than usual. The adenoids are two small pads of tissue in the back of the nose. The tonsils are two oval-shaped pads in the back of the mouth.

It's important for healthcare professionals to find and treat pediatric obstructive sleep apnea as soon as possible. Early treatment helps prevent other health conditions called complications.

These can affect children's growth, learning, behavior and heart health. The first treatment may be surgery to remove enlarged tonsils and adenoids. But some children may get better using medical devices or taking medicines.

**CAUSES**

Pediatric obstructive sleep apnea is caused by muscles in the back of the throat relaxing and blocking the upper airway. In children, this leads to pauses in breathing that last about twice as long as the typical breath.

When breathing stops, this triggers the brain to wake up so that the airway can open again. This makes it hard to get enough rest.

Various conditions can raise the risk of the upper airway becoming blocked during sleep. Commonly, enlarged tonsils in the back of the mouth and enlarged adenoids in the back of the nose can cause a blockage. Other possible causes include being born with a birth defect related to the shape of the face or head and certain health conditions.

**RISK FACTORS**

The main risk factor for pediatric obstructive sleep apnea is enlarged tonsils and adenoids, especially in younger children. Obesity also is an important risk factor, mainly among teenagers.

Other risk factors for pediatric sleep apnea include having:

* A genetic condition such as Down syndrome or Prader-Willi syndrome.
* Birth defects in the skull or face.
* A group of conditions called cerebral palsy that affect movement and posture.
* A group of inherited blood disorders known as sickle cell disease.
* Conditions called neuromuscular disorders that affect the function of muscles due to problems with the nerves and muscles in the body.
* A history of low birth weight.
* A family history of obstructive sleep apnea.

**SIGNS / SYMPTOMS**

During sleep, symptoms of pediatric obstructive sleep apnea can include:

* Snoring.
* Pauses in breathing.
* Restless sleep.
* Snorting, gasping, coughing or choking.
* Mouth breathing.
* Nighttime sweating.
* Bed-wetting that starts after a long period of dry overnights.

Infants and young children with obstructive sleep apnea don't always snore. They might just have disturbed sleep.

During the day, children with sleep apnea might:

* Get headaches in the morning.
* Breathe through the mouth or have trouble breathing through the nose.
* Have trouble learning and paying attention.
* Do poorly in school.
* Have behavior issues such as acting hyper, impulsive or aggressive.
* Have poor weight gain.
* Talk about feeling sleepy, or fall asleep during school or during short car or bus rides.

**DIAGNOSIS METHODS**

Diagnosis involves the steps that a healthcare professional takes to find out if your child has pediatric obstructive sleep apnea.

A healthcare professional reviews your child's symptoms and health history and does a physical exam. Your child's healthcare professional likely will look at your child's head, neck, nose, mouth and tongue.

Other tests might be needed as well. The main test to check for sleep apnea in children is called a polysomnogram. This involves an overnight sleep test. Sensors are placed on your child's body.

The sensors record brain waves, breathing patterns, snoring, oxygen levels, heart rate and muscle activity while your child sleeps. This test may take place at a sleep center.

**TREATMENT OPTIONS**

Your child's healthcare professional works with you to find the right treatment for your child's pediatric obstructive sleep apnea. Most often, the first treatment for the condition is surgery to remove enlarged tonsils and adenoids. This is called adenotonsillectomy (ad-uh-no-ton-sil-EK-tuh-me). But some children get better with medicines or medical devices.

The right treatment plan for your child depends on your child's sleep apnea symptoms and risk factors. For most children, treatment includes adenotonsillectomy, but your child's healthcare professional may recommend other treatments if this surgery isn't right for your child. Other treatments also may be needed if the surgery doesn't fully treat your child's obstructive sleep apnea.

Some kids get better without sleep apnea treatments. It's possible for some children with mild to moderate obstructive sleep apnea to outgrow the condition.

A healthcare professional may recommend closely watching a child for up to six months to see if the symptoms get better. This is called watchful waiting. If the child also has allergies or other conditions that irritate the airway, watchful waiting can include treatment for those.

***MEDICATIONS***

Topical nasal steroids might ease sleep apnea symptoms for some children with mild obstructive sleep apnea. These medicines include fluticasone (Flovent HFA, Xhance, others) and budesonide (Rhinocort, Pulmicort Flexhaler, others). For kids with allergies, montelukast (Singulair) might help relieve symptoms when used alone or with nasal steroids.

***THERAPIES***

Your child's healthcare professional may recommend use of devices such as:

* Positive airway pressure therapy. Small machines gently blow air through a tube with treatments called continuous positive airway pressure (CPAP) and bilevel positive airway pressure (BPAP). The tube is attached to a mask that goes around your child's nose or nose and mouth during sleep. The machine sends air pressure into the back of your child's throat to keep your child's airway open. Often, positive airway pressure therapy is a treatment option if medicines or removal of adenoids and tonsils doesn't work.  
  Proper fitting of the mask and refitting as your child grows can make the mask more comfortable to wear.
* Oral appliances. These devices go in the mouth. They include dental devices and mouthpieces. Oral appliances help to expand the roof of the mouth and nasal passages. They also might move your child's bottom jaw and tongue forward to keep the upper airway open. Only some children benefit from these devices.

***SURGERY OR OTHER PROCEDURES***

Adenotonsillectomy to remove the tonsils and adenoids might improve obstructive sleep apnea by opening the airway. It's often a treatment option for children with moderate to severe obstructive sleep apnea.

Your child's primary healthcare professional might refer you to a pediatric ear, nose and throat specialist to talk about surgery. Other forms of upper airway surgery might be recommended based on your child's condition.

***Lifestyle and home remedies***

You can take the following steps at home to help your child with pediatric obstructive sleep apnea:

* Stay away from airway irritants and allergens. Allergens are things that cause allergies. Keep your child away from tobacco smoke and other indoor allergens or pollutants. This step is important for all children, but especially those with obstructive sleep apnea. Irritants and allergens can irritate the airway and cause congestion.
* Weight loss. If your child is obese, ask your child's healthcare professional about a weight-loss plan. The healthcare professional can give you and your child information on healthy changes in diet and physical activity. You also may be referred to specialists in managing obesity because it is a complex disease. Weight-loss surgery is a treatment option for some teenagers who have obstructive sleep apnea and severe obesity.
* Watchful waiting. Some children may outgrow their obstructive sleep apnea while their healthcare professionals track their health. This is especially true for kids with mild disease and no other risk factors.

**PREVENTION TIPS**

Most causes of childhood sleep apnea can’t be prevented.

You can help your child reduce their risk of obstructive sleep apnea by:

* Exercising regularly.
* Avoiding areas where there’s tobacco smoke.
* Managing seasonal allergies.

Your child’s provider can make personalized recommendations to help your child reduce their risk, especially if the condition runs in your biological family history.

**OUTLOOK / PROGNOSIS**

With treatment, childhood sleep apnea symptoms can go away and won’t have a long-term effect on your child as they grow. Untreated sleep apnea may be dangerous and can affect your child’s growth and development. Some children will have lingering symptoms as they age and may need lifelong management throughout adulthood.

***Does childhood sleep apnea go away?***

For certain causes, treatment is available to eliminate symptoms of obstructive sleep apnea in children. If your child has mild symptoms of sleep apnea, they may grow out of it. This can happen when tissue in the back of their throat shrinks as they get older, which opens their airways. Surgery to remove tonsils or adenoids effectively treats the condition. Symptoms of obstructive sleep apnea may return if your child develops a new blockage in their airway.

**POSSIBLE COMPLICATIONS**

Without treatment, pediatric obstructive sleep apnea can lead to other health conditions called complications. Rarely, pediatric obstructive sleep apnea can cause infants and young children not to grow as much as those who don't have the condition. Children who don't receive treatment also may have a higher risk of later complications such as:

* High blood pressure.
* High cholesterol.
* A higher than typical blood sugar level that raises the risk of diabetes.
* Other heart and blood vessel conditions.

Very rarely, children with certain genetic conditions can have serious symptoms of pediatric obstructive sleep apnea. These symptoms can lead to death. But in most children, treatment can help manage complications.

**WHEN TO SEE A DOCTOR / RED FLAG**

Visit a healthcare provider if you notice signs or symptoms of sleep apnea, especially if you notice your child waking up often in the middle of the night or they have interruptions to their breathing pattern as they sleep.

Visit the emergency room (ER), or call 911 or your local emergency services number, if:

* Your child has trouble breathing.
* Your child’s skin, lips and nails turn pale, blue or gray.

**DIFFERENTIAL DIAGNOSIS**

***Diagnostic Considerations***

Obstructive sleep apnea (OSA) must be differentiated from simple snoring, which is a vibratory inspiratory noise that is usually not accompanied by oxygen desaturation, hypercapnia, or sleep disruption. Overnight polysomnography can be performed to differentiate pronounced snoring from true obstructive sleep apnea in the pediatric age group.

***Daytime somnolence***

Daytime somnolence is a common complaint among individuals with obstructive sleep apnea. For teens and adults, this may be the presenting concern that brings them to medical attention.

However, keep in mind that not all children with excessive daytime somnolence have obstructive sleep apnea. Sleepiness during the day may be due to numerous factors in addition to sleep apnea. Many children are sleepy during the day simply because their parents do not have a clear idea as to how much sleep a child actually requires.

Chaotic sleep schedules with inconsistent bedtimes and rise times and with limited time allowed for sleep are major causes of daytime sleepiness and lassitude.

Any evaluation for suspected sleep apnea must include a careful history with inquiries about sleep times, bedtime routines, and a description of the sleeping environment. Parents should be asked to complete a sleep diary for 1-2 weeks to evaluate whether a child is sleeping enough.

***Narcolepsy***

Narcolepsy is a disease characterized by irresistible sleeping attacks that occur intermittently throughout the day. It is included in the differential diagnosis of excessive daytime sleepiness.

Patients with narcolepsy are tired throughout the day; thus, the disorder can be confused with obstructive sleep apnea syndrome. A history of episodic sleep-onset paralysis, hypnagogic (sleep-onset) hallucinations, or daytime memory lapses with automatic behaviors may help differentiate between narcolepsy and obstructive sleep apnea.

Sleep paralysis is a frightening experience that lasts from a few seconds to several minutes, during which an individual can breathe and move the eyes but otherwise cannot speak or move.

***Hypnagogic hallucinations***

Hypnagogic hallucinations are vivid lifelike dreams that occur just as one begins to fall asleep. These hallucinations often involve an awareness of another person or an animal in the room, bright colors, or unusual shapes.

Often, other senses are involved during the experience, including touch, smell, and hearing. Older patients with narcolepsy may experience cataplexy, or the sudden brief loss of muscular tone without loss of consciousness.

Multiple sleep latency testing (MSLT) following overnight polysomnography is necessary to confirm a diagnosis of narcolepsy and differentiate this from obstructive sleep apnea.

***Nocturnal gastroesophageal reflux***

Nocturnal gastroesophageal reflux may result in nocturnal restlessness, choking episodes during sleep, frequent awakenings, and labored breathing that resemble symptoms of obstructive sleep apnea syndrome.

***Other disorders***

Periodic limb movement disorder, nocturnal seizures, rhythmic movement disorder, and various parasomnias can be differentiated from obstructive sleep apnea on the basis of polysomnography.

***Differential Diagnoses***

* Childhood Sleep Apnea
* Chronic Fatigue Syndrome (Myalgic Encephalomyelitis)
* Congenital Stridor
* Nightmare Disorder
* Pediatric Gastroesophageal Reflux
* Pediatric Hypothyroidism
* Pediatric Obesity-Hypoventilation Syndrome
* Pediatric Sleep Disorders
* Sleep Terrors

**EPIDEMIOLOGY**

In nonobese and otherwise healthy children younger than 8 years, the prevalence of obstructive sleep apnea is estimated to be 1-3%. Habitual snoring is common during childhood and affects approximately 10% of children aged 2-8 years; the frequency decreases after age 9-10 years.

Obesity confers 4-fold to 5-fold added risk for sleep-disordered breathing. In children and adolescents with coexisting medical conditions such as trisomy 21, the prevalence of obstructive sleep apnea may be as high as 80%.

In the United Kingdom, approximately 1.75-2.25% of children aged 4-5 years are thought to have obstructive sleep apnea. Unfortunately, very few epidemiologic studies of childhood obstructive sleep apnea are available.

***Racial distribution***

Obstructive sleep apnea occurs more commonly among Black and Hispanic individuals than among White adults and children. In patients younger than 18 years, Blacks are 3.5 times more likely to develop obstructive sleep apnea than Whites.

The high frequency of obstructive sleep apnea in adult Asian populations indicates that the anthropometric characteristics of the craniofacial structures in this racial group also predispose to higher obstructive sleep apnea rates in children.

The frequency of obstructive sleep apnea in Hispanic children is equal to that of White children.

***Sex distribution***

The male-to-female ratio of obstructive sleep apnea in children is approximately 1:1. At puberty, the male-to-female ratio starts to increase. In older adolescents, a male preponderance emerges that essentially reflects the typical male predominance observed in the adult population. By adulthood, symptomatic men outnumber symptomatic women by 2:1 or more.

***Age distribution***

Obstructive sleep apnea is observed in children of all ages and may develop even in infancy. Retrospective studies note that a large number of parents with children in whom obstructive sleep apnea is diagnosed recall that their child's snoring began within the first months of life.

Preterm babies are at risk for more obstructive events while supine, but some have suggested that they are still at a lower risk of death from sudden infant death syndrome.

However, Moon et al, citing 3 studies, report that premature infants may be at 4 times increased risk for sudden infant death syndrome compared with term infants, with the risk increasing at lower gestational age and birthweight.

Most children with obstructive sleep apnea are aged 2-10 years (coinciding with adenotonsillar lymphatic tissue growth). Children with severe obstructive apnea are likely to present when aged 3-5 years. The mean age at diagnosis has been reported to be 14 months, plus or minus 12 months.

**PREDEFINED Q & A SETS**

Q1: “What is Pediatric Obstructive Sleep Apnea (OSA)?’

A: Pediatric OSA is a condition where a child’s airway becomes partially or completely blocked during sleep, causing disruptions in breathing and sleep quality. This blockage is often due to enlarged tonsils and adenoids but can result from other anatomical or medical factors.

Q2: “What are the common symptoms of Pediatric OSA?”

A:

* Loud snoring or noisy breathing while sleeping
* Pauses in breathing during sleep (apneas)
* Restless or disturbed sleep
* Mouth breathing or snorting/gasping at night
* Nighttime sweating
* Bedwetting after previously being dry
* Daytime symptoms including irritability, hyperactivity, difficulty paying attention, morning headaches, poor growth, and sleepiness during the day

Q3: “What causes Pediatric OSA?”

A:

* Enlarged tonsils and adenoids (most common cause)
* Obesity leading to excess soft tissue narrowing the airway
* Craniofacial abnormalities (e.g., small jaw, syndromes like Down syndrome)
* Neuromuscular disorders reducing airway muscle tone
* Allergies or nasal congestion
* Other airway abnormalities such as laryngomalacia or masses

Q4: “How is Pediatric OSA diagnosed?”

A:

* Clinical evaluation including history and physical exam
* Polysomnography (sleep study) is the gold standard test that records breathing, brain activity, oxygen levels, and other parameters overnight in a sleep lab to confirm diagnosis and severity

Q5: “How is Pediatric OSA treated?”

A:

* Surgery: Removal of enlarged tonsils and adenoids (adenotonsillectomy) is often the first and most effective treatment
* Positive airway pressure therapy (CPAP or BPAP): Machines that provide air pressure via a mask to keep the airway open during sleep, especially if surgery is not possible or fully effective
* Medications: Nasal steroids or allergy treatments may help mild cases or reduce inflammation
* Weight management: Important if the child is overweight or obese
* Other surgeries: May be needed for structural abnormalities if adenotonsillectomy is insufficient

What is the outlook for children with OSA?

With appropriate treatment, most children’s symptoms resolve completely, and their growth, behavior, and sleep quality improve significantly. Untreated OSA can lead to complications involving heart and lung function, cognitive and behavioral problems, and poor growth.

When should I see a doctor?

* If your child snores loudly or has noisy breathing during sleep regularly
* If you notice pauses in breathing or gasping during sleep
* If your child is unusually sleepy during the day, irritable, or having difficulty concentrating
* If you observe bedwetting, restless sleep, or mouth breathing
* For evaluation if your child is overweight or has craniofacial abnormalities

GENOMIC DATA

* Several genetic disorders predispose children to OSA because of anatomical and neuromuscular abnormalities, including Down syndrome, mucopolysaccharidoses, muscular dystrophies, and cerebral palsy. These conditions cause structural airway narrowing or muscle dysfunction contributing to OSA.
* Family history and ethnicity play important roles in OSA risk. Studies show African American children have a higher risk independent of obesity or other factors, suggesting genetic contributions to differences in airway anatomy or tissue characteristics.
* Recent genomic studies, such as a large genome-wide association study (GWAS) by researchers at Children's Hospital of Philadelphia (CHOP), have identified several chromosomal genetic markers associated with pediatric OSA risk. These include loci on chromosomes 1p36.22, 15q26.1, 18p11.32, among others, with some loci differing by ancestry group (European American vs African American) and sex. These findings suggest multiple genes contribute to OSA susceptibility through complex inheritance rather than a single-gene pattern

DOCTOR-PATIENT CONVERSATIONS

Doctor: Hello, I understand you're concerned about your child's sleep. Can you tell me what symptoms you've noticed during the night and day?

Parent: Yes, my child snores loudly almost every night and sometimes seems to stop breathing or gasp for air. They often seem very tired during the day, and lately, their teacher says they're having trouble concentrating.

Doctor: Thank you for sharing that. These symptoms suggest your child might have obstructive sleep apnea, which means their airway is partially blocked during sleep, causing breathing pauses and poor sleep quality.

Parent: What causes this to happen?

Doctor: The most common cause in children is enlargement of the tonsils and adenoids, which can narrow the airway. Other factors like allergies, nasal congestion, obesity, or facial structure can contribute as well.

Parent: How do you confirm that my child has sleep apnea?

Doctor: We typically recommend a sleep study, called polysomnography, that records your child's breathing, oxygen levels, and sleep patterns overnight. This helps us confirm the diagnosis and see how severe it is.

Parent: If the test shows sleep apnea, what treatments are available?

Doctor: The first-line treatment is usually surgical removal of the tonsils and adenoids, which often resolves the problem. In some cases or if surgery isn't enough, we may use a breathing machine at night, called CPAP, that helps keep the airway open. Addressing allergies or managing weight can also help.

Parent: Is surgery safe? Are there risks?

Doctor: Tonsil and adenoid removal is commonly done and generally safe in children, though like any surgery, there are risks like bleeding or infection, which we minimize with careful care and monitoring.

Parent: What if my child doesn't get better after surgery?

Doctor: Some children may need further evaluation to look for other causes of airway blockage or persistent symptoms. Additional treatments, including CPAP therapy or rarely further surgery, might be needed. We will follow your child closely after treatment.

Parent: What should I watch for as warning signs?

Doctor: If your child has increased breathing difficulty, choking episodes during sleep, pauses in breathing, or worsening daytime symptoms like extreme sleepiness or behavior changes, please seek medical attention promptly.

Parent: Thank you, Doctor. This helps me understand the condition and what to expect.

Doctor: You're very welcome. We will work together to ensure your child gets the best care and support.

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**Pediatric hearing loss**

**DEFINITION / DESCRIPTION**

Hearing loss in children can be present at birth (congenital) or develop later in childhood (acquired). Congenital hearing loss can be hereditary (genetic) or caused by infections during pregnancy, including infection with cytomegalovirus or rubella. Hearing loss is more common in babies who are in the neonatal intensive care unit (NICU). Hearing loss can be an isolated condition or a feature of a syndrome that causes additional symptoms. Genetic testing can help determine the cause of hearing loss in some cases. Acquired hearing loss can be caused by infectious diseases, such as meningitis or recurrent ear infections, as well as trauma and certain medications.

Depending on its cause and origin, the hearing loss can be:

* Sensorineural, a permanent type of hearing loss which occurs when the inner ear (cochlea) or the auditory nerve is damaged or malformed
* Conductive, which occurs when the sound can’t travel through the ear because of earwax build-up, a foreign body lodged somewhere in the ear, build-up of fluid or a punctured eardrum (Conductive hearing losses may be treated in some cases with medicine or surgery.)

Hearing loss is categorized as mild, moderate, severe or profound depending on its severity.

**CAUSES**

Hearing loss can happen any time during life—from before birth to adulthood.

The following are some of the things that can increase the chance a child will have hearing loss:

A genetic cause: About 1 out of 2 cases of hearing loss in babies is due to genetic causes. Some babies with a genetic cause for their hearing loss might have family members who also have a hearing loss. About 1 out of 3 babies with genetic hearing loss have a “syndrome.” This means they have other conditions in addition to the hearing loss, such as Down syndrome or Usher syndrome.

1 out of 4 cases of hearing loss in babies is due to maternal infections during pregnancy, complications after birth, and head trauma. For example, the child:

* Was exposed to infection, such as before birth.
* Spent 5 days or more in a hospital neonatal intensive care unit (NICU) or had complications while in the NICU.
* Needed a special procedure such as a blood transfusion to treat bad jaundice.
* Has head, face or ears shaped or formed in a different way than usual.
* Has a condition like a neurological disorder that may be associated with hearing loss.
* Had an infection around the brain and spinal cord called meningitis.
* Received a bad injury to the head that required a hospital stay.

**SIGNS / SYMPTOMS**

* Reduced hearing, such as inability to hear faint sounds
* Failure to respond to sound
* Delay of language and speech development in young children
* Unclear speech

The signs and symptoms of hearing loss are different for each child. If you think that your child might have hearing loss, ask the child's doctor for a hearing screening as soon as possible. Don't wait!

Even if a child has passed a hearing screening before, it is important to look for the following signs.

### Signs in babies

* Does not startle at loud noises.
* Does not turn to the source of a sound after 6 months of age.
* Does not say single words, such as "dada" or "mama" by 1 year of age.
* Turns head when he or she sees you but not if you only call out his or her name. This sometimes is mistaken for not paying attention or just ignoring, but could be the result of a partial or complete hearing loss.
* Seems to hear some sounds but not others.

### Signs in children

* Speech is delayed.
* Speech is not clear.
* Does not follow directions. This sometimes is mistaken for not paying attention or just ignoring, but could be the result of a partial or complete hearing loss.
* Often says, "Huh?"
* Turn the TV or radio volume up too high.

**DIAGNOSIS METHODS**

Hearing screening can tell if a child might have hearing loss. Hearing screening is easy and is not painful. In fact, babies are often asleep while being screened. It takes a very short time—usually only a few minutes.

### Babies

All babies should have a hearing screening no later than 1 month of age. Most babies have their hearing screened while still in the hospital after birth. If a baby does not pass a hearing screening, it’s very important to get a full hearing test as soon as possible, ideally no later than 3 months of age.

### Children

Children should have their hearing tested before they enter school or any time there is a concern about the child's hearing. Children who do not pass the hearing screening need to get a full hearing test as soon as possible.

**TREATMENT OPTIONS**

Treatment and intervention options for hearing loss in children include

* Working with a professional (or team) who can help a child and family learn to communicate.
* Getting a hearing device, such as a hearing aid.
* Joining support groups.
* Taking advantage of other resources available to children with hearing loss and their families.

#### Early Hearing Detection and Intervention (EHDI) Programs

Every state has an EHDI program. EHDI works to identify infants and children with hearing loss. EHDI also promotes timely follow-up testing and intervention services for any family whose child has a hearing loss.

#### Early Intervention (0-3 years)

Hearing loss can affect a child's ability to develop speech, language, and social skills. The earlier a child who is deaf or hard-of-hearing starts getting services, the more likely the child's communication (speech or sign language) and social skills will reach their full potential.

Early intervention program services help young children with hearing loss learn communication and other important skills. Research shows that early intervention services can greatly improve a child's development.

Babies who are diagnosed with hearing loss should begin to get intervention services as soon as possible, but **no later than 6 months of age**.

Services for children from birth through 36 months of age are called Early Intervention or Part C services. Even if your child has not been diagnosed with a hearing loss, he or she may be eligible for early intervention treatment services. The IDEA 2004 says that children under the age of 3 years (36 months) who are at risk of having developmental delays may be eligible for services. These services are provided through an early intervention system in your state. Through this system, you can ask for an evaluation.

#### Special Education (3-22 years)

Special education is instruction specifically designed to address the educational and related developmental needs of older children with disabilities, or those who are experiencing developmental delays. Services for these children are provided through the public school system. These services are available through the Individuals with Disabilities Education Improvement Act 2004 (IDEA 2004), Part B.

### Assistive Technology

Many people who are deaf or hard-of-hearing have some hearing. The amount of hearing a deaf or hard-of-hearing person has is called "residual hearing." Technology does not "cure" hearing loss but may help a child with hearing loss to make the most of their residual hearing. For those parents who choose to have their child use technology, there are many options, including

* Hearing aids
* Cochlear or brainstem implants
* Bone-anchored hearing aids
* Other assistive devices

#### Hearing Aids

Hearing aids

make sounds louder. They can be worn by people of any age, including infants. Babies with hearing loss may understand sounds better using hearing aids. This may give them the chance to learn speech skills at a young age.

There are many styles of hearing aids. They can help many types of hearing losses. A young child is usually fitted with behind-the-ear style hearing aids because they are better suited to growing ears.

#### Cochlear and Auditory Brainstem Implants

A cochlear implant may help many children with severe to profound hearing loss—even very young children. It gives that child a way to hear when a hearing aid is not enough. Unlike a hearing aid, cochlear implants do not make sounds louder. A cochlear implant sends sound signals directly to the hearing nerve.

Persons with severe to profound hearing loss due to an absent or very small hearing nerve or severely abnormal inner ear (cochlea), may not benefit from a hearing aid or cochlear implant. Instead an auditory brainstem implant may provide some hearing. An auditory brainstem implant directly stimulates the hearing pathways in the brainstem, bypassing the inner ear and hearing nerve.

Both cochlear and brainstem implants have two main parts. There are the parts that are placed inside the inner ear, the cochlea, or base of the brain, the brainstem during surgery; and the parts outside the ear that send sounds to the parts inside the ear.

#### Bone-Anchored Hearing Aids

This type of hearing aid can be considered when a child has either a conductive, mixed, or unilateral hearing loss and is specifically suitable for children who cannot otherwise wear ‘in the ear’ or ‘behind the ear’ hearing aids.

#### Other Assistive Devices

Besides hearing aids, there are other devices that help people with hearing loss. Examples of other assistive devices include

**Frequency Modulation (FM) System**. An FM system is a device that helps people with hearing loss hear in background noise. FM is the same type of signal used for radios. These systems send sound from a microphone used by someone speaking to a person wearing the receiver. FM is sometimes used with hearing aids. An extra piece is attached to the hearing aid that works with the FM system.

**Captioning.** Many television programs, videos, and DVDs are captioned. Television sets made after 1993 are made to show the captioning. You don't have to buy anything special. Captions show the conversation spoken in the soundtrack of a program on the bottom of the television screen.

**Other devices.** There are many other devices available for children with hearing loss. Some of these include

* Text messaging
* Telephone amplifiers
* Flashing and vibrating alarms
* Audio loop systems
* Infrared listening devices
* Portable sound amplifiers
* TTY (Text Telephone or teletypewriter)

### Medications and Surgery

Medications or surgery may also help make the most of a person’s hearing. This is especially true for a conductive hearing loss, or one that involves a part of the outer or middle ear that is not working in the usual way.

One type of conductive hearing loss can be caused by a chronic ear infection. A chronic ear infection is a build-up of fluid behind the eardrum in the middle ear space. Most ear infections are managed with medication or careful monitoring. Infections that don't go away with medication can be treated with a simple surgery that involves putting a tiny tube into the eardrum to drain the fluid out.

Another type of conductive hearing loss is caused by either the outer and or middle ear not forming correctly while the baby was growing in the mother's womb. Both the outer and middle ear need to work together in order for sound to be sent correctly to the inner ear. If any of these parts did not form correctly, there might be a hearing loss in that ear. This problem may be improved and perhaps even corrected with surgery. An ear, nose, and throat doctor (otolaryngologist) is the health care professional who usually takes care of this problem.

Placing a cochlear implant, auditory brainstem implant, or bone-anchored hearing aid will also require a surgery.

## Learning Language

## Without extra help, children with hearing loss have problems learning language. These children can then be at risk for other delays. Families who have children with hearing loss often need to change their communication habits or learn special skills (such as sign language) to help their children learn language. These skills can be used together with hearing aids, cochlear or auditory brainstem implants, and other devices that help children hear.

## 

## **PREVENTION TIPS**

The following are tips for parents to help prevent hearing loss in their children:

* Have a healthy pregnancy.
* Make sure your child gets all the childhood vaccines regularly.
* Keep your child away from high noise levels, such as from very loud toys. Visit the National Institutes of Health's website
* to learn more about preventing noise-induced hearing loss.

**OUTLOOK / PROGNOSIS**

The prognosis of hearing loss will vary considerably based on the underlying etiology. Congenital sensorineural hearing loss left untreated will invariably not improve or can progress, such as in the case of congenital CMV. On the other side of the spectrum, glue ear shows an excellent prognosis with the resolution of symptoms even without intervention

**WHEN TO SEE A DOCTOR / RED FLAG**

* If you think that your child might have hearing loss, ask the child's doctor for a **hearing screening** as soon as possible. Don't wait!
* If your child does not pass a hearing screening, ask the child's doctor for a **full hearing test** as soon as possible.
* If your child has hearing loss, talk to the child's doctor about **treatment and** **intervention services**.

**DIFFERENTIAL DIAGNOSIS**

* Acute otitis media
* Cholesteatoma
* Congenital stenosis
* Exostoses
* Foreign body
* Hemotympanum
* Impacted cerumen
* Keratosis obturans
* Middle ear tumour
* Otitis externa

**Categories of hearing loss are as follows:**

* Slight hearing loss: 16-25 dB
* Mild hearing loss: 26-40 dB
* Moderate hearing loss: 41-55 dB
* Severe hearing loss: 71-90 dB
* Profound hearing loss: 90 dB

**EPIDEMIOLOGY**

Hearing loss occurs in 1-3 newborns per 1000 births, with 1-2 per 1,000 suffering from permanent childhood hearing impairment.There is a slightly increased prevalence of hearing loss in boys compared to girls, with a ratio of 1.16:1.0.There are around 45,000 children with hearing loss in the UK, half of which are congenital in origin

**PREDEFINED Q & A SETS**

## What is pediatric hearing loss?

Pediatric hearing loss refers to a partial or complete inability to hear in one or both ears in children. It can affect speech, language development, learning, and social skills if not identified and managed promptly.

## What are the common types of hearing loss in children?

* Conductive hearing loss: Problems in the outer or middle ear (e.g., ear infections, fluid buildup, earwax blockage) that reduce sound conduction.
* Sensorineural hearing loss: Damage to the inner ear or auditory nerve, often permanent, caused by genetic factors, infections, or birth complications.
* Mixed hearing loss: Combination of conductive and sensorineural factors.

## What causes hearing loss in children?

* Genetic/hereditary causes: Many cases result from inherited conditions, some syndromic (with other features) or non-syndromic.
* Infections: Congenital infections like cytomegalovirus (CMV), rubella, meningitis, or others.
* Perinatal factors: Prematurity, birth asphyxia, ototoxic medications received in the neonatal intensive care unit.
* Middle ear problems: Otitis media with effusion (fluid), chronic ear infections.
* Noise exposure: Loud noises or injury to the ear.
* Trauma or structural abnormalities: Injuries to the ear/temporal bone or congenital malformations.

## What are the signs my child may have hearing loss?

* Delayed speech or language development
* Difficulty understanding speech or following instructions
* Frequently asking for repetition
* Turning one ear toward sound or favoring one side
* Speaking loudly or in a monotone voice
* Lack of response to loud noises or name calling
* Behavioral problems or attention difficulties possibly linked to hearing difficulties

## How is hearing loss diagnosed in children?

* Newborn hearing screening using otoacoustic emissions (OAE) or auditory brainstem response (ABR) tests.
* Age-appropriate hearing tests such as behavioral audiometry, visual reinforcement audiometry, or conditioned play audiometry.
* Physical exam including ear inspection and tympanometry.
* Imaging studies if structural abnormalities or neural involvement are suspected.
* Genetic testing if hereditary hearing loss is suspected.

## How is pediatric hearing loss treated?

* Conductive losses: Often treated by clearing infections, fluid drainage, or surgery for structural problems.
* Sensorineural losses: Hearing aids, cochlear implants for severe cases, or bone-anchored hearing devices.
* Early intervention with speech therapy and educational support is crucial.
* Management of any underlying medical conditions and monitoring for progressive loss.

## Can hearing loss in children be prevented?

* Prenatal care including vaccination against infections like rubella.
* Avoiding ototoxic medications unless necessary.
* Protecting children from excessive noise exposure.
* Prompt treatment of ear infections and regular health check-ups.

## What is the outlook for children with hearing loss?

* With early diagnosis and appropriate treatment, many children develop good speech, language, and social skills.
* Untreated hearing loss can lead to delays in communication and academic difficulties.
* Regular follow-up is important to monitor hearing status and device function.

**DOCTOR-PATIENT CONVERSATIONS**

Doctor: Hello, I understand you have some concerns about your child’s hearing. Can you tell me what you have noticed?

Parent: Yes, my child doesn’t seem to respond to sounds the way other kids do. Sometimes they don’t react when I call their name, and they speak less clearly than other children their age.

Doctor: Thank you for sharing that. Hearing loss in children can affect their speech, language, and learning if not addressed. It’s great that you noticed these signs early. There are different types of hearing loss—some temporary and some permanent. We’ll figure out what’s going on in your child’s case.

Parent: What causes hearing loss in kids?

Doctor: Causes vary. Some children have fluid in the middle ear from infections causing temporary hearing loss. Others might have problems with the inner ear or auditory nerve, which can be genetic or related to infections during pregnancy or around birth. Sometimes noise exposure or structural ear problems play a role.

Parent: How do you find out what type of hearing loss my child has?

Doctor: We start with a physical exam to look for infections or blockages. Then, we do hearing tests appropriate for your child’s age to check how well they hear different sounds. Newborn screening tests check for hearing problems early, but detailed testing is important as children grow. In some cases, imaging or genetic tests may be helpful.

Parent: If my child has hearing loss, what can be done?

Doctor: Treatment depends on the cause. If fluid is causing the problem, sometimes antibiotics or small tubes placed in the eardrum can help. For permanent hearing loss, hearing aids or cochlear implants can improve hearing. Also, early speech therapy and educational support are crucial for language development.

Parent: Is hearing loss permanent? Will my child ever hear normally again?

Doctor: Some causes, like fluid from infections, are temporary and improve with treatment. Sensorineural hearing loss, which is damage to the inner ear or auditory nerve, is usually permanent but can often be managed well with hearing devices and therapy. Early intervention makes a big difference in helping children communicate and learn.

Parent: What should I watch for at home?

Doctor: Watch how your child responds to sounds, how they speak and interact, and follow up regularly with your healthcare providers. If you notice worsening hearing or other concerns, let us know quickly.

Parent: Thank you, Doctor. This helps me understand what to do next.

Doctor: You’re welcome. We’re here to support your child’s hearing and development every step of the way.

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Pediatric sinusitis

**ALTERNATIVE NAMES**

Rhinosinusitis

**DEFINITION / DESCRIPTION**

Sinusitis (rhinosinusitis) in children can look different than sinusitis in adults. More often, children have a cough, bad breath, crankiness, low energy, and swelling around the eyes, along with a thick yellow-green nasal or post-nasal drip.

Most of the time, children are diagnosed with *viral* sinusitis (or a viral upper respiratory infection) that will improve by just being treated for its symptoms, but antibiotics can be considered in severe cases of *bacterial* sinusitis.1 In the rare child where medical therapy fails, surgery can be used as a safe and effective method of treating sinus disease in children.

Your child’s sinuses are not fully developed until late in the teen years. Although small, the maxillary (behind the cheek) and ethmoid (between the eyes) sinuses are present at birth. Like sinusitis in adults, pediatric sinusitis can be difficult to diagnose because the symptoms may be caused by other problems, such as a viral illness or allergy.

**CAUSES**

Young children are more prone to infections of the nose, sinus, and ears, especially in the first several years of life. Viruses, allergies, or bacteria usually cause sinusitis. Acute viral sinusitis is likely if your child has been sick for less than 10 days and is not getting worse. Acute bacterial sinusitis is likely when the sinusitis symptoms do not improve at all within 10 days of getting sick, or if your child gets worse within 10 days after beginning to get better.

Chronic sinusitis lasts 12 weeks or longer, and is usually caused by prolonged inflammation, instead of a long infection. Infection can be a part of chronic sinusitis, especially when it worsens from time to time, but is not usually the main cause.

**SYMPTOMS**

The following symptoms may indicate a sinus infection in your child:

* A cold lasting more than 10 to 14 days
* Low- or even high-grade fever
* Thick yellow-green nasal drainage for at least three days in a row
* Post-nasal drip, sometimes with sore throat, cough, bad breath, nausea and/or vomiting
* Headache, usually in children age six or older
* Irritability or fatigue
* Swelling around the eyes

**DIAGNOSIS METHODS**

Generally, your child's health care provider can diagnose sinusitis based on your child's symptoms and physical examination. In some cases additional tests may be performed to confirm the diagnosis. These may include:

* **Sinus X-rays.** Diagnostic test which uses invisible electromagnetic energy beams to produce images of internal tissues, bones, and organs onto film. (X-rays are not typically used, but may help assist in the diagnosis.)
* **Computed tomography (also called CT or CAT scan).** A diagnostic imaging procedure that uses a combination of X-rays and computer technology to produce horizontal, or axial, images (often called slices) of the body. A CT scan shows detailed images of any part of the body, including the bones, muscles, fat, and organs. CT scans are more detailed than general X-rays.  
  Your doctor may perform this procedure:
  + If treatment for sinusitis has been effective
  + If surgery is needed
  + If your child has experienced any complications due to sinusitis
* **Cultures from the sinuses.** Laboratory tests that involve the growing of bacteria or other microorganisms to aid in diagnosis.

**TREATMENT OPTIONS**

If you take your child to an ENT (ear, nose, and throat) specialist, or otolaryngologist, they will examine your child’s ears, nose, and throat. A thorough history and examination usually leads to the correct diagnosis. The doctor may also look for factors that make your child more likely to get a sinus infection, including structural changes, allergies, and problems with the immune system.

Occasionally, special instruments will be used to look into the nose during the office visit. Imaging (X-rays) of the sinuses, such as a CT scan, are not recommended in acute sinusitis unless there are complications from the infection. Radiation safety concerns may limit imaging scans, especially in children younger than six-years-old.2

*Acute sinusitis*—When bacterial sinusitis is present, most children respond very well to antibiotic therapy. Nasal steroid sprays or nasal saline (saltwater) drops or gentle sprays may also be prescribed for short-term relief of stuffiness. Over-the-counter decongestants and antihistamines are generally not effective for viral upper respiratory infections in children, and should not be given to children younger than two-years-old.

If your child has acute bacterial sinusitis, symptoms should improve within the first few days of treatment with antibiotics. Even if your child improves dramatically within the first week of treatment, it is important that you complete the antibiotic therapy. Your doctor may decide to treat your child with additional medicines if he/she has allergies or other conditions that make the sinus infection worse.

*Chronic sinusitis*—If your child suffers from two or more symptoms of sinusitis for at least 12 weeks and has signs of sinus pressure, he or she may have chronic sinusitis.3 Chronic sinusitis or more than four to six episodes of acute sinusitis per year indicates that you should see an ENT specialist, who can recommend appropriate medical or surgical treatment.

Surgery may be considered for a small percentage of children with severe or persistent sinusitis symptoms despite medical therapy. In children under 13-years-old, your doctor may advise removing adenoid tissue4 from behind the nose as part of the treatment for sinusitis. Although the adenoid tissue does not directly block the sinuses, infection of the adenoid tissue, called adenoiditis (infection of the back of the nose that can cause blockage), can cause many symptoms similar to sinusitis—runny nose, stuffy nose, post-nasal drip, bad breath, cough, and headache.

In older children and those for whom medical therapy has been unsuccessful, adenoidectomy or other surgical options may be recommended. An ENT surgeon can open the natural drainage pathways of your child’s sinuses and make the narrow passages wider. This also allows for culturing the infection so that antibiotics can be directed specifically against the bacteria causing your child’s sinus infection. Opening the sinuses allows nasal medications to be distributed more effectively, allowing air to circulate and usually reducing the number and severity of sinus infections.

## 1. Antibiotic Treatment

First-Line Antibiotics:

* Amoxicillin: Often the initial choice for uncomplicated acute bacterial sinusitis (ABS) in children .
  + Dosage: Standard dose is 45 mg/kg/day orally, divided into two doses. A higher dose of 90 mg/kg/day can be used for more severe cases or those at risk for antibiotic resistance .
  + Side Effects: Common side effects include diarrhea, nausea, vomiting, and rash. Allergic reactions, including severe ones, are possible .
* Amoxicillin-Clavulanate (Augmentin): Recommended as the first-line treatment for most cases of uncomplicated ABS where antibacterial resistance is not suspected, or for severe cases and those at risk for severe disease or antibiotic resistance . It combines amoxicillin with clavulanate, which helps overcome bacterial resistance.
  + Dosage: Standard dose is 45 mg/kg/day of the amoxicillin component, divided into two doses. High-dose is 90 mg/kg/day of the amoxicillin component, divided into two doses .
  + Side Effects: Similar to amoxicillin, but with a higher incidence of diarrhea, especially with higher clavulanate content . Other side effects include nausea, vomiting, and rash.

Alternative Antibiotics (for penicillin allergy or treatment failure):

* Cefpodoxime: 10 mg/kg/day orally divided into two doses .
* Cefdinir: 14 mg/kg/day orally in a single dose or divided into two doses .
* Levofloxacin: 10-20 mg/kg/day orally in a single dose or divided into two doses (typically reserved for severe cases or specific situations) .
* Macrolides (e.g., clarithromycin, azithromycin) or Erythromycin: May be used for children with severe penicillin allergies .
  + Side Effects: Macrolides can cause gastrointestinal upset, abdominal pain, and sometimes QT prolongation. Erythromycin can also cause GI upset .

Factors influencing antibiotic choice and dosage:

* Risk factors for bacterial resistance: Age under 2 years, recent antibiotic use, recent hospitalization, day-care attendance, incomplete pneumococcal vaccination, and immunodeficiency .
* Severity of illness: High fever and purulent nasal discharge are indicators of more severe disease .

Duration of Treatment: Antibiotic courses typically range from 7 to 10 days, or until symptoms resolve plus an additional 7 days .

## 2. Adjunctive Therapies

* Nasal Saline Sprays/Drops: Help clear purulent secretions and relieve stuffiness. Isotonic saline is generally preferred as hypertonic saline may decrease ciliary function .
  + Side Effects: Generally well-tolerated, minimal side effects.
* Nasal Steroid Sprays: May be prescribed for short-term relief of stuffiness, especially in children with allergic rhinitis .
  + Side Effects: Local irritation, nosebleeds, and rarely systemic effects with prolonged high-dose use.
* Analgesics/Antipyretics: For fever and pain relief (e.g., ibuprofen or acetaminophen) .
  + Side Effects: Vary depending on the medication (e.g., gastrointestinal upset with ibuprofen).
* Antihistamines and Decongestants: Generally not recommended for routine use in uncomplicated sinusitis unless there is a strong allergic component, as they may have anticholinergic drying effects

PREVENTION TIPS

OUTLOOK / PROGNOSIS

Acute Bacterial Sinusitis (ABS):

* The prognosis is generally *good* for uncomplicated acute sinusitis in children treated appropriately with antibiotics.
* Clinical improvement commonly occurs within 72 hours of starting antibiotics, with most children fully recovering without significant complications.
* Recurrence of acute sinusitis is uncommon in otherwise healthy children.
* Severe or complicated cases (e.g., with orbital or intracranial involvement) are rare but can lead to morbidity, requiring prompt specialized care.

Chronic Rhinosinusitis (CRS):

* Pediatric CRS is distinct and more challenging to treat than acute sinusitis.
* It often requires long-term management including antibiotics, nasal saline irrigation, intranasal steroids, and sometimes surgery (e.g., adenoidectomy, balloon sinuplasty, or functional endoscopic sinus surgery) if medical treatments fail.
* Though less dramatic, CRS can lead to complications such as bony changes (osteitis), mucoceles (sinus expansion), and impact conditions like asthma or cystic fibrosis.
* Surgical interventions generally show good outcomes, but recurrence is possible and long-term follow-up is important

## **Diagnostic Considerations**

These include the following:

* Adenoid hypertrophy
* Adenoiditis
* Benign tumors of the nasal cavity
* Benign tumors of the sinuses
* Ciliary dyskinesia
* Congenital malformations of the sinuses
* Immune deficiency
* Upper respiratory infection

## **Differential Diagnoses**

* Acute Frontal Sinusitis Surgery
* Allergic Fungal Sinusitis
* Chronic Sinusitis
* Fungal Sinusitis
* Malignant Tumors of the Nasal Cavity
* Malignant Tumors of the Sinuses
* Medical Treatment for Acute Sinusitis
* Sinonasal Manifestations of Cystic Fibrosis
* Sinonasal Papillomas
* Surgical Treatment of Acute Ethmoid Sinusitis
* Surgical Treatment of Acute Maxillary Sinusitis
* Surgical Treatment of Acute Sphenoid Sinusitis
* Surgical Treatment of Chronic Maxillary Sinusitis

**EPIDEMIOLOGY**

### Frequency

*United States*

Although the exact incidence of sinusitis in the pediatric population is unclear, it is diagnosed commonly, most often following a viral URTI. The number of URTIs that an individual has per year may be as high as 25 (children will have on average 6-8 per year); the number depends on several factors, including age, day care attendance, and number of siblings. Approximately 5-13% of URTIs are complicated by bacterial sinusitis. Many viral URTIs are mislabeled early in their course as acute sinusitis and are inappropriately treated with antibiotics.

A study by Gilani and Shin determined that in the United States between 2005 and 2012, in patients aged 0-20 years, there were 5.6 million ambulatory care visits per year for chronic rhinosinusitis. The condition was diagnosed in 2.1% of all pediatric ambulatory care visits, while acute rhinosinusitis was diagnosed in 0.6% of all such visits.

A retrospective study by Kalavacherla et al, using the Pediatric Health Information System, indicated that during and after the coronavirus disease 2019 (COVID-19) pandemic, the incidence of hospital admissions for pediatric acute sinusitis declined, while the rate of associated complications in these cases rose. The investigators looked at 2535 hospital admissions for pediatric acute sinusitis spread over three time periods, finding the incidence to be as follows:

* Pre-COVID-19 baseline - 31.5 cases/month
* Initial pandemic - 15.8 cases/month
* Post pandemic - 29 cases/month

In the meantime, the rate of complications during those periods was reported to be 27.5%, 37.4%, and 39%, respectively.

*International*

International incidence is similar to that in the United States.

### Mortality/Morbidity

Recent health-related quality of life measures showed a poor result in children with chronic rhinosinusitis. Because quantifying the morbidity caused by pediatric conditions is difficult, it must also be viewed in other terms. A child with an acute episode of sinusitis may lead the caregiver to experience emotional distress and lack of sleep and miss days from work. Chronic illness may have a negative impact on a child's quality of life in many ways, including complications of chronic antibiotic therapy, school absences, poor sleep patterns, impaired school performance, and irritability.

Children are also susceptible to more serious sequelae from a complication of sinusitis such as orbital cellulites (in about 9.3% of the cases) and intracranial complications (in 3.7-11% of patients). With close follow-up care, counseling of the family, and proper medical treatment, morbidity from this disease should be very low.

A study by Capra et al found a decrease between 2000 and 2009 in the estimated number of hospital admissions in the United States, from 5338 to 4511, for orbital complications of pediatric rhinosinusitis. The investigators suggested that the introduction of heptavalent pneumococcal vaccine was associated with the slight downward trend. The study also found that the mean patient age among children admitted for rhinosinusitis-related orbital complications rose from 4.77 years to 6.07 years and that the proportion of children who underwent surgery for these complications increased.

A study by Al-Madani et al of 616 patients indicated that in children, acute sinusitis most commonly involves the ethmoid sinus and that orbital complications are more common than they are in adults. The investigators also found that most patients in the study responded well to medical treatment.

### Race

No race predilection exists.

### Sex

No sex predilection exists.

### Age

The ethmoid and maxillary sinuses are present at birth. The sphenoid sinuses are pneumatized by age 5 years, and the frontal sinuses appear by age 7 years but are not completely developed until adolescence. Thus, children are predisposed to sinus infection at an early age. In young children, the most common sinuses involved are the ethmoid and maxillary sinuses. Acute sinusitis is much less common in young children than routine URTI or adenoiditis.

In an older child, the sphenoid and frontal sinuses are more likely to be involved with disease. Allergic rhinitis is also more common in older children. It affects only 1% of infants and 5% of children aged 5-9 years, while 15% of the adolescent population is affected. Allergic rhinitis is one of the most common predisposing factors for sinusitis, second only to viral URTIs.

## **Procedures**

See the list below:

* Rigid or flexible nasal endoscopy
  + Nasal endoscopy provides an excellent view of the OMC.
  + It is helpful for evaluation of the adenoid pad.
  + This procedure requires patient cooperation.
* Maxillary sinus puncture
  + This test is the criterion standard for obtaining maxillary sinus cultures.
  + Aerobic and anaerobic culture and sensitivity and Gram staining may enable pathogen-directed antibiotic therapy.
  + Indications for maxillary sinus puncture include the following:
    - Severe toxic illness
    - Acute illness unresponsive to antibiotics within 72 hours
    - Immunocompromised patients
    - Suppurative complications
    - Workup for fever of unknown origin
  + Contents of the maxillary sinus may be aspirated safely through the canine fossa or inferior meatus, but in the pediatric population this often requires a brief general anesthetic. In this instance, the physician may also consider obtaining a culture via the natural maxillary sinus ostia.
* Middle meatal swab
  + Cultures taken from the middle meatus or anterior middle turbinate have good (>80%) correlation with cultures taken from ipsilateral maxillary or ethmoid sinuses.
  + Having a carefully guided endoscopic sample of purulence from the middle meatus is important. Random nasal swabs show little correlation with maxillary cultures.
  + This procedure requires a cooperative child but is definitely less invasive than sinus puncture.

**PREDEFINED Q & A SETS**

## What is pediatric sinusitis?

Pediatric sinusitis is an infection or inflammation of the sinuses—air-filled spaces around the nose. It often occurs after a cold or with allergies. Children's sinuses are still developing, so sinus infections are common, especially in young children.

## What types of sinusitis are there?

* Acute sinusitis: Symptoms last less than 12 weeks and typically improve with treatment.
* Chronic sinusitis: Symptoms last longer than 12 weeks.
* Recurrent sinusitis: Multiple acute episodes, usually 3 or more per year.

## What causes sinusitis in children?

Sinusitis often follows a viral upper respiratory infection that causes swelling and blockage of sinus drainage pathways, allowing bacteria to grow. Common bacteria include *Streptococcus pneumoniae*, *Haemophilus influenzae*, and *Moraxella catarrhalis*. Allergies and other factors that cause nasal congestion also increase risk.

## What are the symptoms of pediatric sinusitis?

* Nasal congestion or stuffiness
* Nasal discharge, which may be yellow or green
* Cough, especially at night
* Facial pain or pressure (sometimes hard to localize in young children)
* Fever (often with acute bacterial sinusitis)
* Bad breath or fatigue
* Worsening or persistent cold symptoms lasting more than 10 days

## How is pediatric sinusitis diagnosed?

* Based on medical history and physical exam, including inspection of the nasal passages.
* Symptoms lasting more than 10 days without improvement or worsening after initial improvement suggest bacterial sinusitis.
* Imaging (CT scan) is reserved for complicated or chronic cases.
* Allergy testing or referral to a specialist may be done if recurrent or chronic sinusitis is suspected.

## How is pediatric sinusitis treated?

* Most cases, especially viral, improve without antibiotics. Supportive care includes nasal saline rinses and symptom relief.
* Antibiotics are used if symptoms are severe, prolonged (>10 days), or worsening.
* Treat underlying allergies if present.
* In chronic or recurrent sinusitis, referral to an ENT specialist may be necessary; surgery is rarely needed.

## When should I see a doctor?

* If your child has a fever higher than 100.4°F (38°C), persistent nasal discharge for more than 10 days, worsening symptoms after initial improvement, or severe facial pain/swelling.
* If your child has recurrent sinus infections (3 or more in a year).
* For symptoms not responding to usual care or concerns about complications.

## What is the outlook for children with sinusitis?

* Most children recover completely with appropriate treatment.
* Chronic or recurrent cases may require additional treatment and monitoring.
* Early care and management improve outcomes and prevent complications.

**DOCTOR-PATIENT CONVERSATIONS**

Doctor: Hello, I understand you’ve brought your child in because of a prolonged cold and nasal symptoms. Can you tell me what you’ve noticed and how long this has been going on?

Parent: Yes, my child has had a stuffy nose and a cough for about two weeks now. The nasal discharge is thick and yellow, and sometimes my child wakes up coughing more at night. They also had a mild fever a few days ago.

Doctor: Thank you. From what you describe, your child might have sinusitis, which is an inflammation or infection of the sinuses. It often happens after a cold and can cause symptoms like nasal congestion, thick nasal discharge, cough, and sometimes facial discomfort.

Parent: How do you know it’s sinusitis and not just a cold?

Doctor: Good question. Most colds improve within 7 to 10 days. When symptoms like nasal discharge and cough last longer than 10 days without improvement, or get worse after improving, it can suggest bacterial sinusitis. Your child’s thick yellow discharge and persistent symptoms fit this pattern.

Parent: What tests will you do?

Doctor: Usually, sinusitis is diagnosed based on the history and physical exam. I will check your child’s nose and face for tenderness and look for signs of inflammation. Imaging like a CT scan is only needed if complications are suspected or if the condition doesn’t improve with treatment.

Parent: What is the treatment?

Doctor: For bacterial sinusitis, antibiotics like amoxicillin or amoxicillin-clavulanate are typically used. We also recommend supportive care such as nasal saline sprays to help clear the nasal passages and using a humidifier. If your child has allergies, treating those can also help. Pain relievers can ease any discomfort or fever.

Parent: Are there any side effects or things I should watch out for with antibiotics?

Doctor: The common side effects are mild, like upset stomach or diarrhea. If you notice severe rash, persistent vomiting, or signs of worsening illness, please let us know right away.

Parent: How long until my child gets better?

Doctor: Many children start improving within 3 days of starting antibiotics, but make sure to complete the full course. If symptoms don’t improve after a week or worsen, please come back so we can reassess.

Parent: When should I seek emergency care?

Doctor: If you notice your child has severe facial swelling, intense pain, high fever unresponsive to medication, difficulty breathing, or vision changes, seek medical care immediately as these could indicate complications.

Parent: Thank you, Doctor. This helps me understand what to expect.

Doctor: You’re welcome. We’ll follow up to make sure your child improves and support you throughout the treatment.

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**Pediatric sleeping-disordered breathing**

**DEFINITION / DESCRIPTION**

Pediatric sleep-disordered breathing (SDB) is a general term for breathing difficulties during sleep. SDB can range from frequent loud snoring to obstructive sleep apnea (OSA), a condition where part, or all, of the airway is blocked repeatedly during sleep.

When a child’s breathing is disrupted during sleep, the body thinks the child is choking. The heart rate increases, blood pressure rises, the brain is aroused, and sleep is disrupted. Oxygen levels in the blood can also drop.

Approximately 10 percent of children snore regularly, and about two to four percent of children experience OSA. Recent studies indicate that mild SDB or snoring may cause many of the same problems as OSA in children.

**CAUSES**

A common physical cause of airway narrowing contributing to SDB is enlarged tonsils and adenoids. Overweight children are at increased risk for SDB because fat deposits around the neck and throat can also narrow the airway. Children with abnormalities involving the lower jaw or tongue, or neuromuscular deficits such as cerebral palsy, have a higher risk of developing SDB.

*How is Sleep Apnea Diagnosed?*

If you notice any of the symptoms described in this article, have your child checked by an ENT (ear, nose, and throat) specialist, or otolaryngologist. Sometimes physicians will make a diagnosis of SDB based on history and physical examination. In other cases, like children suspected of having severe OSA due to craniofacial syndromes, morbid obesity, neuromuscular disorders, or for children less than three-years-old, additional testing such as a sleep test may be recommended.

The sleep study, or polysomnography (PSG), is an objective test for SDB. Wires are attached to the head and body to monitor brain waves, muscle tension, eye movement, breathing, and the level of oxygen in the blood. The test is not painful and is generally performed in a sleep laboratory or hospital. Sleep tests occasionally produce inaccurate results, especially in children. Borderline or normal sleep test results may still result in a diagnosis of SDB based on parental observations and clinical evaluation.

**SIGNS / SYMPTOMS**

Potential symptoms and consequences of untreated pediatric SDB may include:

* *Snoring*—The most obvious symptom of SDB is loud snoring that is present on most nights. The snoring can be interrupted by complete blockage of breathing, with gasping and snorting noises associated with waking up from sleep. Loud snoring can also become a significant social problem if a child shares a room with siblings, or at sleepovers and summer camp.
* *Irritability*—A child with SDB may become irritable, sleepy during the day, or have difficulty concentrating in school. He or she may also display busy or hyperactive behavior.
* *Bedwetting*—SDB can cause increased urine production at night, which may lead to bedwetting (also called enuresis).
* *Learning difficulties*—Children with SDB may become moody and disruptive, or not pay attention, both at home and at school. SDB can also be a contributing factor to attention deficit disorders in some children.
* *Slow growth*—Children with SDB may not produce enough growth hormone, resulting in abnormally slow growth and development.
* *Cardiovascular difficulties*—OSA can be associated with an increased risk of high blood pressure, or other heart and lung problems.
* *Obesity*—SDB may cause the body to have increased resistance to insulin, and daytime fatigue can lead to decreased physical activity. These factors can contribute to obesity.

**DIAGNOSIS METHODS**

If you suspect your child may have a sleep disorder, you should see a doctor immediately. Sleep disorders can lead to **neurological** and cardiovascular problems later on.

Your pediatrician will ask if you have a family history of sleep apnea. He will then perform a physical exam to look for obstructions that might cause breathing difficulties. These might include things such as **enlarged tonsils** or narrow airways. If the doctor suspects sleep apnea, they may recommend a sleep study.

* **Sleep study** - A sleep study may occur at a lab that uses special technology to monitor your child’s behavior while sleeping. More often, your doctor will order a device such as an actigraph to track your child’s sleep patterns. The watch-like instrument is non-invasive and provides specialists with data they need to recommend appropriate treatments.

**TREATMENT OPTIONS**

Enlarged tonsils and adenoids are a common cause for SDB. Surgical removal of the tonsils and adenoids, called tonsillectomy and adenoidectomy (T&A), is generally considered the first line treatment for pediatric SDB if the symptoms are significant, and the tonsils and adenoids are enlarged. Of the more than 500,000 pediatric T&A procedures performed in the United States each year, the majority treat SDB. Many children with OSA show both short- and long-term improvement in their sleep and behavior after T&A.

Not every child with snoring needs to undergo T&A. If the SDB symptoms are mild or intermittent, academic performance and behavior is not an issue, the tonsils are small, or the child is near puberty (because tonsils and adenoids often shrink at puberty), it may be recommended that a child with SDB be watched conservatively and treated surgically only if symptoms worsen.

Recent studies have shown that some children have persistent SDB after T&A. A post-operative sleep study may be necessary, especially in children with persistent symptoms or increased risk factors for persistent apnea after T&A such as obesity, craniofacial anomalies or neuromuscular problems. Additional treatments such as weight loss, the use of continuous positive airway pressure (CPAP), or additional surgical procedures may sometimes be required.

## **Diagnostic Considerations**

A detailed sleep history, a thorough physical examination, and sleep logs provide the foundation for accurate diagnosis, treatment, and possible referral for polysomnography (PSG). Atypical presentations, snoring associated with daytime somnolence, behavioral-emotional problems, apneic or hypopneic episodes, suspicion of narcolepsy, abnormal and disruptive movements in sleep, unexplained or recalcitrant sleep difficulties, or daytime sleepiness indicate a need for sleep studies (see Workup).

## **Differential Diagnoses**

* Anorexia Nervosa
* Asthma
* Autism Spectrum Disorder
* Child Sexual Abuse
* Childhood Sleep Apnea
* Chronic Fatigue Syndrome (Myalgic Encephalomyelitis)
* Cognitive Deficits
* Down Syndrome
* Fibromyalgia
* Fragile X Syndrome
* Hyperthyroidism and Thyrotoxicosis
* Hypothyroidism
* Juvenile Idiopathic Arthritis
* Lactose Intolerance
* Nightmare Disorder
* Obesity
* Oppositional Defiant Disorder
* Pervasive Developmental Disorder
* Physical Child Abuse
* Pica
* Posterior Urethral Valves
* Prader-Willi Syndrome
* Rett Syndrome
* Rumination
* Sarcoidosis
* Schizophrenia
* Separation Anxiety and School Refusal
* Sleep Terrors
* Smith-Magenis Syndrome
* Williams Syndrome
* Wilson Disease

**EPIDEMIOLOGY**

* The overall prevalence of pediatric OSA in the general population is estimated to be 1% to 5%, with most studies reporting prevalence between 1% and 4% for moderate to severe OSA.
* Habitual snoring, which is part of the SDB spectrum, affects about 7.5% to 10% of children, with prevalence peaking between ages 2 and 8 years and decreasing after age 9-10.
* Boys appear to have a higher prevalence of SDB than girls in some studies, although data are not entirely consistent. Obesity is a strong risk factor, increasing risk 4- to 5-fold for sleep-disordered breathing.
* Racial and ethnic differences exist: Black children have a 3.5 times higher risk of developing OSA than White children. Hispanic children have similar prevalence to White children. Craniofacial differences may also influence prevalence in Asian populations.
* The highest risk for more severe OSA tends to be in children aged 1 to 5 years, likely due to anatomical factors such as adenotonsillar hypertrophy. In one referral population, about two-thirds of diagnosed children had moderate-to-severe OSA, with many children from this group being very young preschoolers.
* Children with certain medical conditions (e.g., trisomy 21 / Down syndrome) have much higher prevalence of OSA, up to 80% in some reports

**PREDEFINED Q & A SETS**

## What is pediatric sleep-disordered breathing (SDB)?

Pediatric SDB refers to a spectrum of breathing problems during sleep, ranging from habitual snoring to obstructive sleep apnea, where the airway is partially or completely blocked, causing breathing disruptions that affect sleep quality and overall health.

## What are the common symptoms of pediatric SDB?

* Loud, frequent snoring most nights
* Breathing pauses, gasping, or choking sounds during sleep
* Mouth breathing or nasal voice
* Restless, disrupted sleep with tossing and turning
* Daytime sleepiness or fatigue
* Behavioral problems like irritability, hyperactivity, or learning difficulties
* Morning headaches
* Night sweats, bedwetting, or sleepwalking in some children.

## What causes pediatric sleep-disordered breathing?

* Enlarged tonsils and adenoids (most common cause) narrowing the airway
* Obesity, causing excess fat deposits around the neck and throat
* Craniofacial abnormalities affecting airway size or shape
* Neuromuscular disorders impairing airway muscle control (e.g., cerebral palsy)
* Nasal congestion from allergies or chronic rhinitis
* Structural abnormalities like a small lower jaw or tongue position.

## How is pediatric SDB diagnosed?

* By clinical history and physical exam, including observation of sleep behaviors
* Polysomnography (sleep study) is the gold standard to confirm diagnosis, measuring breathing patterns, oxygen levels, and sleep stages overnight
* Sleep questionnaires and screening tools may be used to assess risk but are not definitive.

## How is pediatric sleep-disordered breathing treated?

* Adenotonsillectomy: Surgical removal of enlarged tonsils and adenoids is the primary and most effective treatment in many children
* Continuous Positive Airway Pressure (CPAP): Non-surgical option for children who are not surgical candidates or have persistent symptoms after surgery
* Medical management of allergies, nasal obstruction, or obesity
* Close follow-up to monitor improvement or persistent symptoms.

## What is the outlook for children with pediatric SDB?

With timely diagnosis and treatment, most children experience significant improvement in sleep quality, behavior, growth, and cognitive function. Untreated SDB can lead to cardiovascular problems, poor growth, learning difficulties, and behavioral issues.

## When should I see a doctor for my child?

* If your child snores loudly or frequently
* If you notice pauses in breathing or choking during sleep
* If your child shows daytime sleepiness, hyperactivity, learning or behavioral problems
* If your child breathes primarily through the mouth at night
* If your child has difficulty waking up, morning headaches, or bedwetting

**DOCTOR-PATIENT CONVERSATIONS**

Doctor: Hello, I understand you're concerned about your child's sleep. Can you tell me what symptoms you have noticed during the night and day?

Parent: Yes, my child snores loudly almost every night and sometimes seems to stop breathing or gasp for air. They often seem very tired during the day, and their teacher says they have trouble concentrating.

Doctor: Thank you for sharing that. These symptoms are common in children with sleep-disordered breathing, which means the airway may be partially blocked during sleep. This can cause pauses in breathing and disrupt their sleep quality.

Parent: What causes this to happen in children?

Doctor: The most common cause is enlarged tonsils and adenoids, which can narrow the airway. Other factors like allergies, nasal congestion, obesity, or certain facial structures can also contribute.

Parent: How do you confirm whether my child has this condition?

Doctor: The best way to confirm is with a sleep study called a polysomnography. It records your child's breathing patterns, oxygen levels, and sleep stages overnight to evaluate if obstructive events occur and how severe they are.

Parent: What treatments are available?

Doctor: In many cases, surgery to remove the enlarged tonsils and adenoids is very effective. Some children benefit from a breathing support machine called CPAP if surgery isn’t possible or doesn’t fully help. Managing allergies, nasal congestion, and maintaining a healthy weight can also improve symptoms.

Parent: Is surgery safe for children?

Doctor: Tonsil and adenoid removal is a common procedure in children and generally safe. Like any surgery, it carries some risks like bleeding or infection, but these are uncommon with careful care.

Parent: How soon can we expect improvement?

Doctor: Many children start to sleep and breathe better soon after surgery or with treatment, leading to better daytime focus and energy. We will follow up closely to ensure your child's symptoms improve and to adjust treatment if needed.

Parent: What warning signs should I watch for?

Doctor: If your child has difficulty breathing while awake, choking episodes at night, very loud and frequent apneas, extreme daytime sleepiness, or behavioral changes, please bring them in immediately.

Parent: Thank you, Doctor. This helps me understand what to expect and how to help my child.

Doctor: You're welcome. We’ll work together to take the best care of your child's sleep and health.

REFERENCES:

<https://pm.amegroups.org/article/view/5050/html>

[Pediatric Sleep-disordered Breathing - ENT Health](https://www.enthealth.org/conditions/pediatric-sleep-disordered-breathing/)

**POST-NASAL DRIP**

**ALTERNATIVE NAMES:** Post-nasal drip is also known by several alternative names, including upper airway cough syndrome (UACS), postnasal drainage, and post nasal drip syndrome. It is also referred to as posterior rhinorrhea, which includes synonyms such as discharge from the back of the nose, discharge from the nasopharynx, and postnasal catarrh. Additionally, it has been previously called postnasal drip syndrome (PNDS).

**DEFINITION / DESCRIPTION**

Postnasal drip is when more mucus than normal gathers and drips down the back of your throat. You may feel like you have a tickle in the back of your throat. Postnasal drip can be a bothersome condition that can lead to a chronic cough.

The glands in your nose and throat are constantly making mucus. This is normal. These glands produce one to two quarts of mucus per day. Mucus has many important functions, such as:

* Moistens and cleans your nasal lining.
* Moistens the air you breathe.
* Traps and clears whatever you inhale.
* Helps fight infections.

You normally swallow mucus unconsciously. You don't notice it because it mixes with your saliva and drips harmlessly down the back of your throat. But when you feel like mucus is gathering in your throat or dripping from the back of your nose, it becomes more obvious.

**CAUSES**

You can get postnasal drip for many different reasons. One of the most frequent causes of postnasal drip is allergies, which is often called allergic postnasal drip.

Another cause is a deviated septum. If you have a deviated septum, it means the wall of cartilage between your nostrils (septum) is crooked. The misplaced structure of your nose makes one of your nasal passages smaller than the other. This can prevent mucus from draining properly and can lead to postnasal drip. Other postnasal drip causes may include:

* Colds and flu.
* Bacterial infections.
* Sinus infections (sinusitis).
* Cold temperatures, changing weather and dryness in the air.
* Bright lights.
* Spicy foods.
* Pregnancy.
* Certain medicines, such as birth control pills and high blood pressure (hypertension) medications.
* Age.
* Chronic acid reflux (GERD).

***Is postnasal drip contagious?***

Postnasal drip itself isn’t contagious. But the cause of it may be contagious. For example, if you develop postnasal drip because of a viral infection such as a cold, you could pass the virus on to someone else.

**SIGNS / SYMPTOMS**

Postnasal drip can cause an irritated sore throat. Your tonsils and other tissues in your throat may swell up, leading to discomfort. You may feel like there’s a lump in the back of your throat. Other symptoms of postnasal drip may include:

* A feeling of mucus draining into your throat.
* Frequent swallowing.
* Gurgling or hoarseness.
* Urge to clear your throat.
* Bad breath (halitosis).
* Cough that bothers you more at night.
* Nausea and vomiting from excess mucus draining to your stomach.

Postnasal drip can also cause painful ear infections if mucus clogs up your Eustachian tubes. Your Eustachian tubes are what connect your nose and throat to your middle ears.

**DIAGNOSIS METHODS**

Your healthcare provider may diagnose postnasal drip by performing a physical exam of your ears, nose and throat. They may use a special camera called an endoscope to look inside of your nose and throat. This procedure is called a nasal endoscopy. They may also order X-rays

**TREATMENT OPTIONS**

Postnasal drip can be hard to cure. Treatment depends on the cause of the condition. For common colds and flu, you can try drinking warm liquids like soup or tea to help thin out the excess mucus. Along with drinking plenty of water, these home remedies will also keep you hydrated. Other treatment options may include:

#### **Allergies**

To fix postnasal drip due to allergies, you should avoid things you’re allergic to. Symptom relief may include medicines like:

* Antihistamines, such as loratadine-pseudoephedrine (Claritin®).
* Decongestants.
* Cromolyn and steroid nasal sprays.
* Oral steroids.

In addition, immunotherapy with allergy shots or drops under your tongue may be a good remedy for the condition.

#### **Deviated septum**

If you have a deviated septum, you may need a surgery called a septoplasty to permanently treat postnasal drip. Septoplasty straightens your septum and provides better airflow.

#### **Bacterial infections**

To get rid of postnasal drip due to a bacterial infection, your healthcare provider may recommend certain medications, including:

* Antibiotics.
* Nasal sprays.
* Decongestants, such as pseudoephedrine (Sudafed®).
* Nasal saline irrigations, such as neti pots.

If postnasal drip is due to chronic sinusitis, your healthcare provider may recommend sinus surgery. Sinus surgery can open your blocked sinuses.

#### **Chronic acid reflux**

Treatment for postnasal drip caused by GERD (gastroesophageal reflux disease) may include:

* Avoiding foods and drinks for at least three hours before bedtime.
* Keeping your head elevated six inches to eight inches above your body at bedtime.
* Losing any excess weight.
* Cutting out caffeine and alcohol.
* Taking antacids such as TUMS® or acid blockers such as Pepcid AC®.

**PREVENTION TIPS**

One way to prevent postnasal drip is by reducing your exposure to things you’re allergic to as much as possible. Ways to prevent the condition may include:

* Taking a daily allergy pill.
* Keeping your house dust-free and clean.
* Using pillow covers and mattress covers to prevent dust mites.
* Changing the air filters on your HVAC system frequently.
* Showering before you go to bed if you’ve spent time outdoors.

***How do I take care of myself if I have a postnasal drip?***

There are many things you can do at home to help clear up your postnasal drip. You may need more fluids to thin out your secretions. Home remedies may include:

* Drink more water.
* Cut out caffeine.
* Avoid diuretics, if possible.

You can also try a mucus-thinning medication (expectorant) such as guaifenesin (Mucinex®). These may make your secretions thinner. Saline nasal irrigations lessen thickened secretions. Saline nasal sprays can help moisten your nose.

**OUTLOOK / PROGNOSIS**

Postnasal drip is very common and has many different causes. While the condition isn’t usually serious, it can be annoying. With some over-the-counter medications and other home remedies, it should clear up on its own.

If you have repeated cases of postnasal drip or other symptoms along with it, reach out to your healthcare provider. You may have a bacterial infection or other condition that requires medical care.

**WHEN TO SEE A DOCTOR / RED FLAG**

If you’ve taken steps to clear up your postnasal drip and they’re not working, call your healthcare provider. You may have a bacterial infection that needs an antibiotic. Symptoms of a bacterial infection may include:

* Fever.
* Wheezing.
* Odor-causing mucus

**DIFFERENTIAL DIAGNOSIS**

* Allergic Rhinitis: One of the most common causes. Allergic inflammation causes increased mucus production and congestion leading to postnasal drainage.
* Infectious Rhinitis / Viral Upper Respiratory Infections: The common cold or other viral infections cause mucosal irritation and increased mucus production, typically transient.
* Sinusitis (Acute or Chronic): Bacterial or viral sinus infections result in inflamed and blocked sinuses producing thick mucus that drains posteriorly.
* Nonallergic (Vasomotor) Rhinitis: A hypersensitive nasal mucosa reacting to irritants like cold air, smoke, perfumes, which can cause increased mucus production without infection or allergy.
* Reflux (Gastroesophageal Reflux Disease - GERD): Acid reflux can irritate the throat and upper airway, worsening cough and mucus sensation.
* Nasal Polyps or Structural Abnormalities: Benign nasal growths or anatomical issues can obstruct drainage, causing mucus buildup and PND.
* Foreign Body in the Nose (especially in children): Can cause unilateral postnasal drip, foul odor, and mucopurulent discharge.
* Medications: Some drugs can thicken nasal secretions or cause rebound congestion (e.g., overuse of nasal decongestants, certain antihypertensives).
* Other Causes: Rarely, tumors or other masses can cause obstruction and mucus retention leading to symptoms similar to postnasal drip.

**EPIDEMIOLOGY**

* Prevalence: PND commonly occurs as a symptom in conditions like allergic and nonallergic rhinitis, which affect about 30-40% of the general population. Chronic idiopathic PND patients have been reported in studies with an average age in the mid-50s, but PND symptoms can affect all ages depending on the cause.
* Associated conditions: Allergic rhinitis is a leading cause of PND and affects about 10-30% of adults globally and up to 40% of children. Nonallergic rhinitis is also common, more so in females. Sinusitis is another frequent contributor to PND.
* Symptom characteristics: In patients with chronic idiopathic PND, throat discomfort occurs in about 74%, and cough is present in about 30%. Symptoms can be persistent or recurrent, lasting months to years.
* Clinical burden: PND is a frequent cause of chronic cough, particularly when associated with upper respiratory conditions. Studies show PND symptoms significantly prolong cough duration in patients with asthma.
* Response to treatment: Over 70% of patients with chronic idiopathic PND respond to antihistamine-decongestant therapy, though recurrence rates can be around 26%.

**PREDEFINED Q & A SETS**

Q1: “What is a postnasal drip?”

A: Postnasal drip is when extra mucus builds up inside the nose or sinuses and drips down the back of the throat. This can cause throat irritation, frequent throat clearing, coughing, or a feeling of mucus in the throat.

Q2: “What causes postnasal drip?”

A: Postnasal drip is caused by increased mucus production, often due to:

* Allergies (allergic rhinitis)
* Viral or bacterial sinus infections (sinusitis)
* Common cold or upper respiratory infections
* Nonallergic irritants like smoke, dry air, or strong smells
* Gastroesophageal reflux (acid reflux)
* Nasal structural problems or nasal polyps
* Foreign bodies in the nose (especially in children)

Q3: “What are the symptoms?”

A:

* Feeling of mucus dripping down the throat
* Frequent throat clearing or coughing
* Sore or scratchy throat
* Hoarseness or voice changes
* Nasal congestion
* Difficulty swallowing or feeling like something is stuck in the throat
* Bad breath (in some cases)

Q4: “How is postnasal drip diagnosed?”

A: Diagnosis is usually made by your healthcare provider based on your symptoms and a physical exam, including examination of your nose and throat. In some cases, they may use:

* Nasal endoscopy (using a small camera to look inside the nose)
* Imaging tests (like sinus X-rays or CT scans) if sinus disease or structural issues are suspected
* Allergy testing if allergies are thought to be a cause

Q5: “How is postnasal drip treated?”

A: Treatment depends on the underlying cause:

* For allergies: Antihistamines, nasal steroid sprays, and avoiding triggers
* For sinus infections: Antibiotics if bacterial, nasal saline rinses, and possibly nasal steroids
* For acid reflux: Lifestyle changes and medications to reduce stomach acid
* Home remedies: Staying well-hydrated, using saline nasal sprays, gargling with salt water, and using a humidifier
* Avoid irritants like smoke, strong odors, alcohol, and caffeine which can worsen symptoms

Q6: “When should I see a doctor?”

A: You should consult a healthcare provider if:

* Postnasal drip persists despite treatment
* You have symptoms lasting longer than 10 days or worsen
* You notice blood in your mucus
* You develop fever, wheezing, difficulty breathing, or severe throat pain
* You have foul-smelling nasal discharge or bad breath that doesn’t improve

DOCTOR-PATIENT CONVERSATIONS

Doctor: Hello, what brings you in today?

Patient/Parent: My child has been complaining about a constant feeling of mucus dripping down the back of their throat. They keep clearing their throat and sometimes cough, especially at night.

Doctor: It sounds like your child might be experiencing postnasal drip. This means extra mucus from the nose or sinuses is running down the back of their throat, which can cause that throat irritation and coughing.

Patient/Parent: What causes postnasal drip?

Doctor: There are several common causes. Allergies are a frequent reason—it makes the nose produce more mucus. It can also be due to colds, sinus infections, irritants like smoke or strong smells, or even acid reflux from the stomach irritating the throat. Sometimes structural issues like nasal polyps or a foreign body can cause it, especially in children.

Patient/Parent: How do you find out what is causing it?

Doctor: First, I’ll ask about other symptoms—any history of allergies, recent colds, or signs of sinus infection like facial pain or fever. I’ll also examine the nose and throat. If needed, we might do allergy testing or imaging if sinus disease or structural problems are suspected.

Patient/Parent: How is postnasal drip treated?

Doctor: Treatment targets the underlying cause. For allergies, nasal steroid sprays and antihistamines help. If there’s a sinus infection, sometimes antibiotics are needed. For acid reflux, lifestyle changes and medications can reduce stomach acid. Also, saline nasal sprays and staying well hydrated can relieve symptoms.

Patient/Parent: Are there any things I should avoid or watch for?

Doctor: Avoid exposure to irritants like smoke or strong perfumes. If symptoms last more than 10 days, get worse, or if your child develops fever, breathing difficulties, or blood in mucus, you should come back for further evaluation.

Patient/Parent: Thank you, Doctor. This helps me understand what is going on and what to do.

Doctor: You’re welcome. Let’s work together to manage your child's symptoms and improve their comfort.

*REFERENCES:*

<https://my.clevelandclinic.org/health/diseases/23082-postnasal-drip>

**PETROSITIS**

**ALTERNATIVE NAMES:** Alternative names for petrositis include infection/osteomyelitis of the petrous temporal bone, petrous apicitis, and Gradenigo's syndrome (a particular syndromal presentation of petrositis). Other synonyms mentioned are inflammation of the petrous bone , and it is also referred to as petrositides. Additionally, it is known as petrous apicitis , and in some contexts, it is called Gradenigo's syndrome.

**DEFINITION / DESCRIPTION**

Petrositis (also called petrous apicitis) is an infection and inflammation of the petrous portion of the temporal bone in the skull, which is the area surrounding the inner ear. It typically results from the spread of a middle ear or mastoid infection (such as otitis media or mastoiditis) into this part of the bone.

This condition is relatively rare but serious because the infection can involve nearby cranial nerves and structures. The classic presentation, known as Gradenigo's syndrome, includes a triad of symptoms:

* Retro-orbital (behind the eye) pain due to trigeminal nerve (cranial nerve V) involvement
* Sixth cranial nerve (abducens nerve) palsy causing lateral rectus muscle paralysis and resulting in double vision (diplopia)
* Otorrhea (ear discharge) from the middle ear infection

**CAUSES**

Primary causes of petrositis (or petrous apicitis) are infections that spread from the middle ear or mastoid air cells to the petrous portion of the temporal bone. This spread happens mainly through vascular canals or bone resorption and via pneumatized air cell tracts in the bone. The infections typically arise as complications of acute or chronic otitis media (middle ear infection) or mastoiditis.

The most common bacteria implicated include *Pseudomonas aeruginosa* and other pathogens involved in middle ear infections. The petrous apex may contain air cells that when blocked by inflammation or mechanical obstruction allow bacteria to become trapped, leading to infection in this area.

In summary, petrositis usually results from:

* Extension of infection from the middle ear or mastoid to the petrous apex
* Blockage and subsequent infection of the pneumatized air cell system within the petrous bone
* Often associated with acute or chronic otitis media or mastoiditis

**SIGNS / SYMPTOMS**

symptoms can include headaches, hearing loss, vertigo (due to inner ear involvement), facial nerve paralysis, and signs of deep infection or inflammation in the temporal bone

**DIAGNOSIS METHODS**

* Imaging Studies:
  + Computed Tomography (CT) scan is the standard diagnostic tool that shows opacification (clouding) of the mastoid air cells including the petrous apex, bone erosion, and can reveal extension of infection. CT is useful for delineating bone destruction and surgical planning.
  + Magnetic Resonance Imaging (MRI) provides additional details not visible on CT, such as better visualization of soft tissue inflammation and involvement of adjacent structures like the cavernous sinus. Contrast-enhanced MRI can detect enhancement due to inflammation or abscess.
  + Single-photon emission computed tomography (SPECT) may be used if CT or MRI findings are inconclusive.
* Laboratory tests:
  + Culturing middle ear effusions if present, to identify bacteria and guide antibiotic therapy.
  + Routine blood tests including complete blood count (CBC) and inflammatory markers supported diagnosis of infection.
* Clinical Examination:
  + Assessment of symptoms (pain, cranial nerve palsies, ear discharge).
  + Otoscopic exam for evidence of middle ear disease or mastoid infection contributing to petrositis.
* Additional imaging/tests:
  + Conventional X-rays may show bone erosion but are largely replaced by CT and MRI.
  + Audiometric tests may be performed to assess hearing loss associated with petrositis

**TREATMENT OPTIONS**

* Antibiotics:  
  High-dose, broad-spectrum intravenous (IV) antibiotics are the mainstay of treatment, targeting the typical bacterial pathogens from middle ear infections. Early and adequate antibiotic therapy can lead to resolution without surgery, especially if diagnosed before serious complications develop. Patients often require prolonged courses of IV antibiotics, sometimes several weeks.
* Surgical intervention:  
  Surgery, such as mastoidectomy or approaches to drain the petrous apex, is indicated if there is failure to respond to antibiotics, disease progression, or development of complications (like abscess formation or cranial nerve deficits). Techniques include transmastoid drainage and more extensive procedures like circumferential petrosectomy for maximal infected bone removal while preserving facial nerve and hearing function. Surgical treatment is reserved for severe or refractory cases.
* Conservative management examples:  
  Cases have shown complete symptom resolution with IV antibiotics alone, and some reports describe avoiding even myringotomy if early diagnosis is made.
* Treatment goals:  
  Prevention of complications such as cranial nerve palsies or life-threatening intracranial spread.

**OUTLOOK / PROGNOSIS**

* Improved outcomes with antibiotics: Early diagnosis and treatment with high-dose intravenous broad-spectrum antibiotics often lead to full resolution without the need for surgery. Several recent case reports document complete clinical and radiological recovery within weeks of medical treatment alone.
* Surgical intervention reserved: Surgery is usually reserved for cases that fail medical management or have complications such as abscesses or cranial nerve palsies. When surgery is performed timely, it also contributes to good outcomes.
* Potential complications: If untreated or inadequately treated, petrositis can cause serious complications including facial nerve paralysis, intracranial infections (meningitis, abscess), venous sinus thrombosis, labyrinthine fistula, and even death. Thus, delayed treatment worsens prognosis.
* Mortality rates: Mortality is rare with modern antibiotic therapy but was more common in the pre-antibiotic era. Recent systematic reviews report low death rates (~5%) mainly related to severe or complicated infections.
* Long-term recovery: Patients typically recover fully if managed early. Some neurological symptoms like cranial nerve palsies improve gradually with treatment

**DIFFERENTIAL DIAGNOSIS**

* Cholesterol granuloma of the petrous temporal bone: a cystic lesion caused by chronic inflammation and bleeding, often showing characteristic imaging features.
* Congenital cholesteatoma: a benign growth of keratinizing squamous epithelium in the petrous bone that can cause bone erosion.
* Mucocoele of the petrous apex: a cystic expansion due to trapped mucus, less common but can present similarly.
* Base of skull tumors: including meningioma or metastatic lesions that can involve the petrous bone.
* Jugular paraganglioma (glomus tumor): a vascular tumor near the skull base.
* Acoustic neuroma (vestibular schwannoma): which can extend toward the petrous apex and affect cranial nerves.
* Otitis externa: infection of the ear canal skin, distinguished clinically but included in the broader differential when assessing ear symptoms.
* Sigmoid sinus thrombosis: can present with headache and ear symptoms and mimic petrositis complications.
* Meningitis and labyrinthitis: infections of the meninges and inner ear, respectively, may have overlapping symptoms such as cranial nerve palsies and ear pain.
* Neoplasia: other neoplastic lesions affecting the temporal bone or skull base.

**EPIDEMIOLOGY**

Petrous apicitis was commonly encountered before the introduction of antibiotics. It now appears quite rarely. Most reports on petrous apicitis in the literature are single clinical cases, and determining its frequency is difficult.

In a retrospective review of petrous apicitis cases occurring over a 40-year period, Gadre and Chole found that six out of 44 patients with the condition (13.6%) had Gradenigo syndrome.

**PREDEFINED Q & A SETS**

Q1: “What is petrositis?”  
A: Petrositis is an infection and inflammation of the petrous portion of the temporal bone, the part of the skull surrounding the inner ear. It often occurs as a complication of middle ear infections (otitis media) or mastoiditis, and may involve serious spread to nearby cranial nerves and structures.

Q2: “What causes petrositis?”  
A: Petrositis is typically caused by the spread of bacterial infection from the middle ear or mastoid air cells into the petrous apex via air cell tracts or bone erosion. Common bacteria include those involved in chronic or acute otitis media, such as *Pseudomonas aeruginosa* and *Streptococcus pneumoniae*.

Q3: “Who is affected by petrositis?”  
A: Petrositis is rare today, largely due to effective antibiotic use. It most often affects patients with prolonged or complicated middle ear infections or mastoiditis. The petrous apex is pneumatized (contains air cells) in about 30% of people, which can predispose to infection in this region.

Q4: “What are the typical symptoms?”

A:

* Retro-orbital pain (behind the eye) due to trigeminal nerve involvement
* Sixth cranial nerve palsy (causing double vision)
* Otorrhea (ear discharge)
* Headaches, hearing loss, vertigo, facial nerve paralysis (in severe cases)
* Fever and other signs of infection

The classic symptom triad is known as Gradenigo's syndrome: deep facial pain, sixth nerve palsy, and otorrhea, but not all cases present fully.

Q5: “How is petrositis diagnosed?”

A:

* Clinical suspicion based on symptoms and history of middle ear infection
* Imaging with CT scan (shows bone erosion and air cell opacification) and MRI (soft tissue details and abscesses)
* Microbiologic cultures from ear discharge
* Blood tests indicating infection/inflammation
* Otoscopic examination for middle ear disease.

Q6: “How is petrositis treated?”

A:

* High-dose intravenous antibiotics targeting typical ear pathogens are the main treatment and can resolve many cases without surgery
* Surgical drainage (mastoidectomy or approaches to drain petrous apex) is reserved for refractory or complicated cases
* Additional support with pain management and sometimes corticosteroids
* Myringotomy with tympanostomy tubes may be used to ventilate the middle ear and aid drainage.

Q7: “What is the prognosis?”  
A: With early diagnosis and adequate antibiotic treatment, the prognosis is generally favorable, with full recovery common. Untreated petrositis can lead to serious complications including intracranial infection, cranial nerve paralysis, and death. Surgical intervention reduces risks in complicated cases.

Q8: “Can it be prevented?”  
A: Prompt and effective treatment of otitis media and mastoiditis reduces the chance of petrositis developing.

DOCTOR-PATIENT CONVERSATIONS

Doctor: Hello, your child has a condition called petrositis, which is an infection and inflammation of part of the temporal bone near the ear called the petrous apex.

Parent: What does that mean? How serious is it?

Doctor: It is a rare but serious complication that can happen after an ear infection or mastoiditis. The infection affects a deep bone in the skull, causing symptoms like severe ear pain, pain behind or around the eye, and sometimes even double vision because it can affect certain cranial nerves.

Parent: What symptoms should I watch for?

Doctor: Key symptoms include persistent ear discharge, deep aching pain behind the eye or face, fever, and difficulty moving the eye outward which can cause double vision. If your child complains of severe facial or ear pain along with eye problems, it’s important to contact us promptly.

Parent: How do you treat petrositis?

Doctor: We usually start with intravenous antibiotics to fight the infection. Most children respond well to this medical treatment without needing surgery. However, if symptoms do not improve or if complications arise, surgery may be required to drain the infected area.

Parent: Will my child need surgery right away?

Doctor: Not always. Thanks to modern imaging and effective antibiotics, many cases can be managed conservatively. Surgery is reserved for children whose symptoms persist or worsen despite antibiotics, or if complications such as abscess formation develop.

Parent: How long will recovery take?

Doctor: Recovery varies, but with proper treatment, symptoms often improve within days to weeks. Follow-up imaging will help us ensure the infection is resolving. We monitor closely for any signs of complications.

Parent: What are the risks if untreated?

Doctor: If left untreated, the infection can spread to other parts of the brain and nerves, causing serious complications like meningitis, brain abscess, or permanent cranial nerve damage. That’s why timely treatment is crucial.

*REFERENCES:*

<https://emedicine.medscape.com/article/883256-overview#a7>

**PHARYNGITIS (VIRAL AND BACTERIAL)**

**ALTERNATIVE NAMES:** Pharyngitis is commonly referred to as "sore throat" and can be caused by viral or bacterial infections. Alternative names for pharyngitis include "acute sore throat". Viral pharyngitis is often associated with symptoms similar to the common cold, while bacterial pharyngitis, particularly when caused by group A Streptococcus, is commonly known as "strep throat".

**DEFINITION / DESCRIPTION**

A sore throat, or “pharyngitis,” is a scratchy, painful feeling in the back of your throat (pharynx). It happens when the tissue lining your throat (mucosa) becomes inflamed. If you have a sore throat, it may hurt to swallow or talk.

Many things cause pharyngitis, from viral and bacterial infections to allergies and sleeping with your mouth open. Most sore throat symptoms go away with home care within a few days.

But you should contact a healthcare provider if your sore throat lasts longer than a week, gets worse or you develop symptoms like a fever or swollen lymph nodes.

***Types of pharyngitis***

There are two main types of pharyngitis. Healthcare providers categorize them based on how long symptoms last:

* Acute pharyngitis: A sore throat that lasts from about three to 10 days. Most sore throats are acute pharyngitis.
* Chronic pharyngitis: A sore throat that lasts for more than 10 days (usually several weeks) or that keeps returning after you get better.

**CAUSES**

Most sore throats happen because you have a viral infection, like the common cold or flu. Less often, a sore throat (pharyngitis) may be a sign of the following conditions or issues:

* Bacterial infection: Conditions like strep throat and bacterial sinus infections may cause a sore throat.
* Tonsillitis: Tonsillitis occurs when your tonsils become infected and inflamed. Bacteria and viruses can both cause tonsillitis.
* Allergies: Allergic reactions from pollen, dust mites, pets or mold can cause mucus from your nose to drip down the back of your throat (postnasal drip). This can lead to a sore throat.
* Acid reflux: People with gastroesophageal reflux disease (GERD) feel burning and pain in their throats. This pain, called heartburn, happens when acid from your stomach backs up into your food tube (esophagus).
* Overuse or irritants: You can strain your throat by yelling or screaming. You may also develop a sore throat if you eat spicy food, smoke or drink very hot liquids.
* Mouth breathing: You may have a sore throat if you breathe through your mouth instead of your nose when you’re sleeping.
* Tumors: Although a sore throat rarely means cancer, it’s one potential symptom of throat cancer or benign (noncancerous) growths.

**SIGNS / SYMPTOMS**

Pharyngitis may start with a raspy feeling in your throat, as if your throat is dry. If your sore throat gets worse, you may feel a sharp pain in your throat when you swallow or talk. You may feel the pain in your ears or down the side of your neck.

If an infection is causing your sore throat, additional symptoms may include:

* Fever.
* Headache.
* Upset stomach.
* Swollen lymph nodes.
* Nasal congestion (stuffy nose).
* Runny nose.
* Cough.
* Fatigue.
* Hoarseness.
* Redness or swelling in your throat and tonsils.
* White patches, spots or streaks in your throat and tonsils.

**DIAGNOSIS METHODS**

Your healthcare provider will ask about your symptoms. They’ll perform a physical exam that involves looking at your throat, tongue and possibly your ears. They may do a strep test to check for the bacteria that causes strep throat.

Testing is important in case your provider can’t tell from your symptoms alone if your sore throat is related to a virus or bacteria. The results determine what treatments you’ll need.

**TREATMENT OPTIONS**

Treatment depends on what’s causing your pharyngitis. Viral infections usually clear up on their own within a week. In the meantime, your healthcare provider may recommend over-the-counter (OTC) medicines to ease your sore throat.

Other treatments for a sore throat may include:

* Antibiotics: You may need antibiotics to treat a bacterial infection. Most people need to take them for 10 days.
* Over-the-counter antihistamine medications: Antihistamines may dry postnasal drip related to allergies.
* Over-the-counter antacids: Antacids may help with acid reflux that causes heartburn and sore throats. Other steps, like avoiding big meals right before bedtime, may help.
* Prescription mouthwash: For a severe sore throat related to conditions like cancer, your provider may prescribe a special mouthwash (sometimes called “magic mouthwash”) that contains a mix of a numbing agent (like lidocaine), Benadryl® and Maalox®.

**PREVENTION TIPS**

Viral infections like colds and the flu often cause pharyngitis. You can reduce your chances of getting a sore throat by protecting yourself against these common infections. You can:

* Stay up to date on vaccinations for the flu and COVID-19.
* Wash your hands often, using soap and water or alcohol-based sanitizers.
* Avoid sharing food, drinks or utensils.
* Avoid people with colds or other contagious respiratory infections.
* Avoid being around others when you’re sick to reduce the risk of spreading infectious diseases.

**OUTLOOK / PROGNOSIS**

Usually, a sore throat isn’t a serious medical issue. Instead, it’s typically an unpleasant sign that you’re sick with a cold or the flu.

Most sore throats related to a viral infection go away within three to 10 days. If your sore throat lasts longer than this, it may be a symptom of a condition that requires prescription medications or other treatments your healthcare provider can recommend.

**HOME REMEDIES**

It may take some time to cure what’s causing your pharyngitis. In the meantime, there are at-home remedies you can try to relieve your sore throat. You can:

* Take over-the-counter pain relievers. Acetaminophen (Tylenol®), naproxen sodium (Aleve®) and ibuprofen (Advil®) are all options that may help with a sore throat.
* Suck on throat lozenges or hard candy. This increases spit (saliva) production, which adds soothing moisture to your throat. But never try this with children under 5, who may choke.
* Try ice or cold foods. You can also try sucking on ice chips or popsicles to ease sore throat pain.
* Drink soothing liquids. Drinking tea with lemon and honey, broth or bouillon may help dry, scratchy throats. If coolness feels better on the back of your throat, try sipping ice water.
* Gargle saltwater. Gargling a mixture that’s ½ teaspoon of salt per 1 cup of water can ease inflammation and irritation in your throat.
* Use a humidifier or vaporizer. Adding moisture to your environment, especially to your bedroom when you’re sleeping, helps ease dry throats.
* Rest your voice. If your throat is sore from shouting, screaming, singing or even talking a lot, resting your voice may help.
* Avoid irritants. Avoid exposure to secondhand smoke, and steer clear of spicy foods and very hot liquids that may irritate your sore throat. If you smoke, work with your healthcare provider to quit.

**WHEN TO SEE A DOCTOR / RED FLAG**

Contact your healthcare provider if your sore throat lasts longer than a week. Reach out if you have any of the following symptoms:

* Severe throat pain.
* Trouble breathing or swallowing.
* A fever, especially if it’s over 100.4 degrees Fahrenheit (38 degrees Celsius).
* A visible bulge in the back of your throat.
* Blood in your saliva or phlegm.
* A rash anywhere on your body.

**DIFFERENTIAL DIAGNOSIS**

***Diagnostic Considerations***

These include the following:

* Allergic rhinitis with postnasal drip
* Airway obstruction
* Head and neck neoplasias
* Gastroesophageal reflux disease (GERD)
* Peritonsillar cellulitis

Differential Diagnoses

* Candidiasis in Emergency Medicine
* Diphtheria
* Emergent Management of Croup (Laryngotracheobronchitis)
* Emergent Management of Gonorrhea
* Emergent Management of Pediatric Epiglottitis
* Epiglottitis
* Hand-Foot-and-Mouth Disease in Emergency Medicine
* Herpes Simplex Virus (HSV) in Emergency Medicine
* Infectious Mononucleosis (IM) in Emergency Medicine
* Pediatric Pharyngitis
* Peritonsillar Abscess in Emergency Medicine
* Pharyngitis
* Pneumonia, Mycoplasma
* Retropharyngeal Abscess
* Rheumatic Fever in Emergency Medicine
* Scarlet Fever

**RECENT GUIDELINES OR UPDATES**

The Choosing Wisely medical initiative picked the top guidelines from the Italian Panel of the National Guidelines for the Management of Acute Pharyngitis in Children. The chosen recommendations are as follows:

* Blood exams should not be performed
* Antibiotics should not be administered unless microbiologic confirmation of streptococcal infection has been carried out
* If a throat culture is performed, susceptibility tests on isolates should not be executed
* The antibiotic course should not be shortened
* Because penicillin V is not available in Italy, amoxicillin (50 mg/kg/d in 2-3 doses orally) for 10 days is the first choice treatment
* Steroids should not be administered, to avoid masking a possible underlying severe condition

A literature review by the Italian Panel for the Management of Acute Pharyngitis in Children indicated that guidelines for the treatment of acute pharyngitis vary in their treatment recommendations, with one group of guidelines recommending that GABHS be treated with antibiotics to avoid the development of acute rheumatic fever, another group of guidelines pointing to acute pharyngitis as self resolving and advising that antibiotic therapy be used only in complicated cases, and a third group of guidelines supporting the use of differing strategies tailored to each patient’s acute rheumatic fever risk. The investigators state that “[s]ince GABHS pharyngitis could affect the global burden of GABHS disease, it is advisable to define a shared strategy worldwide.”

**EPIDEMIOLOGY**

### ***United States***

Children experience more than 5 upper respiratory infections (URIs) per year and an average of one streptococcal infection every 4 years. The occurrence in adults is about one half that rate. The most significant bacterial agent causing pharyngitis in both adults and children is GAS infection (*Streptococcus pyogenes*), and the most common viruses are rhinovirus and adenovirus. GAS is most prevalent in late fall through early spring.

### ***International***

The incidence of pharyngitis is higher internationally. Antibiotic resistance may be more prevalent in some countries because of overprescription of antibiotics. Note, however, that despite this, there has never been a documented case of GAS resistant to penicillin anywhere in the world.

A study by Banigo et al reported that the reduction in the number of tonsillectomies performed in England (28,309 in 1990/1991 vs 6327 in 2013/2014) correlates with an increase in the number of hospital admissions in that country for acute tonsillitis and pharyngitis and with an increase in invasive group A beta-hemolytic streptococcal (GABHS) infections. Indeed, over the course of the 1990/1991 to 2013/2014 period, the number of invasive GABHS infections rose more than two-fold in children aged 14 years or younger.

**PREDEFINED Q & A SETS**

Q1: “What is pharyngitis?”  
Pharyngitis is inflammation of the pharynx, the back of the throat, resulting in a sore throat. It commonly causes discomfort, pain, scratchiness, and difficulty swallowing or speaking.

Q2: “What causes pharyngitis?”  
Pharyngitis is most often caused by viral infections such as the common cold viruses (rhinovirus, coronavirus), influenza, adenovirus, Epstein-Barr virus (mononucleosis), and others. Less commonly, it is caused by bacterial infections, primarily group A streptococcus (strep throat). Other rare bacterial causes include gonorrhea and chlamydia. Non-infectious causes include allergies, acid reflux, irritants like smoke, and overuse of the throat muscles.

Q3: “Who commonly gets pharyngitis?”  
Pharyngitis can affect people of all ages but is especially common during colder months and among people frequently exposed to respiratory infections, such as healthcare workers and children.

Q4: “What are the typical symptoms?”

Answer:

* Sore or scratchy throat
* Pain or difficulty swallowing and talking
* Red, swollen throat and tonsils, sometimes with white patches or pus
* Fever
* Headache, body aches, fatigue
* Swollen lymph nodes in the neck
* Cough, runny nose (especially in viral cases)  
  Symptoms can vary depending on the cause.

Q5: “How is pharyngitis diagnosed?”  
Diagnosis is based on clinical history and physical examination of the throat. Throat swabs or rapid antigen tests for group A streptococcus may be done to differentiate bacterial infections from viral ones. In some cases, blood tests or other cultures may be needed.

Q6: “How is pharyngitis treated?”

Answer:

* Viral pharyngitis typically resolves on its own, with symptomatic relief using pain relievers, hydration, and throat lozenges.
* Bacterial pharyngitis (e.g., strep throat) requires antibiotics to prevent complications and speed recovery.
* Avoidance of irritants and treatment of underlying causes like acid reflux or allergies is important for non-infectious pharyngitis.

Q7: “What are possible complications?”

Untreated streptococcal pharyngitis can lead to complications such as rheumatic fever, kidney inflammation, or peritonsillar abscess. Viral pharyngitis rarely leads to serious complications.

Q8: “Can pharyngitis be prevented?”  
Good hygiene practices such as handwashing, avoiding close contact with infected individuals, and not sharing utensils can reduce the risk. Prompt treatment of bacterial infections also helps prevent spread and complications

**DOCTOR-PATIENT CONVERSATIONS**

Doctor: Hello, what brings you in today?

Patient: Hi, doctor. I’ve had a sore throat for the last couple of days, and it’s been painful to swallow.

Doctor: I see. Do you have any other symptoms? Fever, cough, runny nose, or swollen glands?

Patient: Yes, I have a mild fever and some headache. No cough or runny nose though.

Doctor: Have you noticed any white patches or pus on your tonsils?

Patient: Actually, yes, there are some white spots on my tonsils when I look in the mirror.

Doctor: Okay, that can suggest a bacterial infection. Have you been around anyone who is sick recently?

Patient: Not that I know of.

Doctor: Let me examine your throat. Please open your mouth wide and say “ahh.” [Examines the throat] Your tonsils look red and swollen with some white exudate. You also have tender lymph nodes under your jaw. Based on this, I think you likely have strep throat.

Patient: Is that serious?

Doctor: It’s quite common and treatable. I will order a rapid strep test to confirm the diagnosis, but I’ll also start you on an antibiotic to clear the infection. It’s important to complete the full course.

Patient: How long will it take to get better?

Doctor: With antibiotics, symptoms usually improve in a few days. You should rest, drink plenty of fluids, and avoid irritants like smoking. If your symptoms worsen or you develop difficulty breathing, swelling, or rash, come back immediately.

Patient: Should I stay home from work or school?

Doctor: Yes, it’s best to stay home while you have a fever and for at least 24 hours after starting antibiotics to avoid spreading the infection.

Patient: Thank you, doctor.

Doctor: You’re welcome. Let me know if you have any questions or concerns.

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**PITUITARY TUMORS**

**ALTERNATIVE NAMES:** Alternative names for pituitary tumors include pituitary adenoma (benign pituitary tumor), pituitary neuroendocrine tumors (PitNETs), microadenoma (benign tumor smaller than 10 mm), macroadenoma (benign tumor larger than 10 mm), prolactinoma (a type of functioning pituitary adenoma that secretes excess prolactin), and corticotroph adenoma (a type of functioning pituitary tumor that secretes adrenocorticotropic hormone). Other names include pituitary carcinomas and metastatic pituitary neuroendocrine tumors (PitNETs) for cancerous types.

**DEFINITION / DESCRIPTION**

Pituitary tumors are unusual growths that develop in the pituitary gland. This gland is an organ about the size of a pea. It's located behind the nose at the base of the brain. Some of these tumors cause the pituitary gland to make too much of certain hormones that control important body functions. Others can cause the pituitary gland to make too little of those hormones.

Most pituitary tumors are benign. That means they are not cancer. Another name for these noncancerous tumors is pituitary adenomas. Most adenomas stay in the pituitary gland or in the tissue around it, and they grow slowly. They typically don't spread to other parts of the body.

Pituitary tumors can be treated in several ways. The tumor may be removed with surgery. Or its growth may be controlled with medications or radiation therapy. Sometimes, hormone levels are managed with medicine. Your health care provider may suggest a combination of these treatments. In some cases, observation — also called a ''wait-and-see'' approach — may be the right choice.

***Types***

Types of pituitary adenomas include:

* **Functioning.** These adenomas make hormones. They cause different symptoms depending on the kind of hormones they make. Functioning pituitary adenomas fall into several categories, including those that make:
  + **Adrenocorticotropic hormone.** This hormone also is known as ACTH. These tumors are sometimes called corticotroph adenomas.
  + **Growth hormone.** These tumors are called somatotroph adenomas.
  + **Luteinizing hormone and follicle-stimulating hormone.** These hormones also are known as gonadotropins. Pituitary tumors that make these hormones are called gonadotroph adenomas.
  + **Prolactin.** These tumors are called prolactinomas or lactotroph adenomas.
  + **Thyroid-stimulating hormone.** These tumors are called thyrotroph adenomas.
* **Nonfunctioning.** These adenomas don't make hormones. The symptoms they cause are related to the pressure their growth puts on the pituitary gland, nearby nerves and the brain.
* **Macroadenomas.** These are larger adenomas. They measure about 1 centimeter or more. That's slightly less than a half-inch. They can be functioning or nonfunctioning.
* **Microadenomas.** These adenomas are smaller. They measure less than 1 centimeter. That's slightly less than a half-inch. They can be functioning or nonfunctioning.

Pituitary tumors are different from pituitary cysts. A cyst is a sac that may be filled with air, fluid or other material. A tumor is an unusual mass of cells that may grow over time. Cysts may form on or near the pituitary gland, but they are not tumors or adenomas.

**CAUSES**

The pituitary gland is a small organ about the size of a pea. It's located behind the nose at the base of the brain. Despite its small size, the pituitary gland has an effect on nearly every part of the body. The hormones it makes control important body functions, such as growth, blood pressure and reproduction.

The cause of uncontrolled cell growth in the pituitary gland, which creates a tumor, remains unknown. In rare cases, pituitary tumors can be caused by genes you've inherited. But most have no clear hereditary cause. Still, scientists think that changes in genes may play an important role in how pituitary tumors develop.

**RISK FACTORS**

Most people who get pituitary tumors don't have any factors that put them at higher risk of developing these tumors. Environment and lifestyle choices don't seem to have an effect on a person's risk of pituitary tumors.

Although genetics seems to play a role, most people who have pituitary tumors don't have a family history of them.

The only known risk factors are several rare hereditary conditions that raise the risk of many health problems, including pituitary tumors. These conditions include:

* Multiple endocrine neoplasia, type 1, also called MEN 1.
* Multiple endocrine neoplasia, type 4, also called MEN 4.
* Carney complex.
* McCune-Albright syndrome.

**SYMPTOMS**

Not all pituitary tumors cause symptoms. Sometimes these tumors are found during an imaging test, such as an MRI or a CT scan, that is done for another reason. If they don't cause symptoms, pituitary tumors usually don't need treatment.

Pituitary tumor symptoms may be caused by a tumor putting pressure on the brain or on other parts of the body nearby. Symptoms also can be caused by a hormone imbalance. Hormone levels can rise when a pituitary tumor makes too much of one or more hormones. Or a large tumor that disrupts the way the pituitary gland works may cause hormone levels to fall.

### **Symptoms from tumor pressure**

Macroadenomas can put pressure on the pituitary gland, on nerves, on the brain and on other parts of the body nearby. That can cause symptoms such as:

* Headache.
* Eye problems due to pressure on the optic nerve, especially loss of side vision, also called peripheral vision, and double vision.
* Pain in the face, sometimes including sinus pain or ear pain.
* Drooping eyelids.
* Seizures.
* Nausea and vomiting.

***Symptoms from hormone changes***

***Low amounts of hormones***

Macroadenomas can limit the pituitary gland's ability to make hormones. When that happens, symptoms may include:

* Tiredness or weakness.
* Lack of energy.
* Sexual problems, such as problems with erections and less interest in sex.
* Changes in menstrual cycles.
* Nausea.
* Feeling cold.
* Losing or gaining weight without trying.

#### **High amounts of hormones**

Functioning pituitary adenomas typically make a large amount of one hormone. That exposes the body to high levels of that hormone. Rarely, a pituitary adenoma may make more than one hormone. The following types of functioning pituitary adenomas cause different symptoms depending on the hormones they make.

#### **Pituitary tumors that make adrenocorticotropic hormone**

Pituitary tumors that make adrenocorticotropic hormone are called corticotroph adenomas. Adrenocorticotropic hormone, also called ACTH, causes the adrenal glands to make the hormone cortisol. ACTH tumors trigger the adrenal glands to make too much cortisol. This causes a condition called Cushing disease. Cushing disease is one cause of Cushing syndrome.

Cushing disease symptoms include:

* Weight gain and fatty tissue deposits around the midsection and upper back.
* Rounded face.
* Stretch marks.
* Thin skin that bruises easily.
* Thinning of the arms and legs with muscle weakness.
* Thicker or more visible body hair.
* Slow healing of cuts, insect bites and infections.
* Areas of darkened skin.
* Acne.
* Changes in menstrual cycles.
* Sexual problems, including problems with erections and less interest in sex.

#### **Pituitary tumors that make growth hormone**

Pituitary tumors that make growth hormone also are called growth hormone-secreting tumors or somatotroph adenomas. Too much growth hormone leads to a condition known as acromegaly. Acromegaly can cause:

* Changes in facial features, including larger lips, nose and tongue; longer lower jaw; and wide spaces between teeth.
* Growth of hands and feet.
* Thicker skin.
* More sweating and body odor.
* Joint pain.
* A deeper voice.

Children and teens who have too much growth hormone might also grow faster or taller than usual. This condition is called gigantism.

#### **Pituitary tumors that make luteinizing hormone and follicle-stimulating hormone**

Luteinizing hormone (LH) and follicle-stimulating hormone (FSH) are also known as gonadotropins. Pituitary tumors that make these hormones are called gonadotroph adenomas.

It's uncommon for these adenomas to make too many hormones that then trigger symptoms. Instead, symptoms from these adenomas usually are due to tumor pressure. If symptoms do happen because of too much LH and FSH, they affect women and men differently.

Symptoms in women may include:

* Change in menstrual cycles.
* Fertility problems.
* Enlargement of and pain in the ovaries caused by a condition called ovarian hyperstimulation syndrome.

Symptoms in men may include:

* Enlarged testicles.
* Higher levels of testosterone.

#### **Pituitary tumors that make prolactin**

These adenomas are called prolactinomas. Too much of the hormone prolactin can lead to a decrease in the body's levels of sex hormones — estrogen and testosterone. Too much prolactin affects men and women differently.

In women, too much prolactin might cause:

* Irregular menstrual cycles.
* Lack of menstrual cycles.
* Milky discharge from the breasts.
* Breast tenderness.
* Problems with fertility.
* Less interest in sex.

In men, too much prolactin can cause a condition called male hypogonadism. Symptoms might include:

* Problems with erections.
* Less interest in sex.
* Breast growth.
* Problems with fertility.
* Less body and facial hair.

#### **Pituitary tumors that make thyroid-stimulating hormone**

Pituitary tumors that make thyroid-stimulating hormone are called thyrotroph adenomas. They also may be referred to as thyroid-stimulating hormone-secreting tumors. They cause the thyroid gland to make too much of the hormone thyroxine, also called T-4. That leads to a condition called hyperthyroidism, also known as overactive thyroid disease. Hyperthyroidism can speed up the body's metabolism causing many symptoms. Some of the most common include:

* Weight loss.
* Rapid or irregular heartbeat.
* Nervousness, anxiety or irritability.
* Frequent bowel movements.
* Sweating.
* Tremor.
* Sleep problems.

**DIAGNOSIS METHODS**

Pituitary tumors often aren't noticed or aren't detected. In many cases, that's because the symptoms caused by pituitary tumors that make hormones, called functioning adenomas, and large tumors, called macroadenomas, are similar to those of other medical conditions. It's also because they grow very slowly over time. Small pituitary tumors that don't make hormones, called nonfunctioning microadenomas, often don't cause symptoms. If they are detected, it's typically because of an imaging exam, such as an MRI or a CT scan, that's done for another reason.

To detect and diagnose a pituitary tumor, your health care provider will likely talk with you about your personal and family medical history and do a physical exam. Testing to detect a pituitary tumor also may include:

* **Blood tests.** Blood tests can show whether your body has too much or too little of certain hormones. For some hormones, blood test results that show too much of the hormone may be all that's needed for your health care provider to diagnose a pituitary adenoma.  
  For other hormones, such as cortisol, a blood test result that shows too much of the hormone may need to be followed by other tests. Those tests can show if the earlier result was caused by a pituitary adenoma or by another health concern.  
  Results that show hormone levels are too low need to be followed with other tests, usually imaging exams, to see if a pituitary adenoma may be the cause of those test results.
* **Urine tests.** A urine test may be used to help diagnose a pituitary adenoma that's making too much of the hormone ACTH. Too much ACTH leads to too much cortisol in the body and causes Cushing disease.
* **MRI scan.** A magnetic resonance imaging scan, also called an MRI scan, is a test that uses a magnetic field and computer-generated radio waves to create detailed images of the body's organs and tissues. An MRI of the brain can help detect a pituitary tumor and show its location and size.
* **CT scan.** A computed tomography scan, also called a CT scan, is a type of imaging test that combines a series of X-rays to create cross-sectional images. MRI scans are used more often than CT scans to detect and diagnose pituitary tumors. But a CT scan may be helpful in planning surgery if your health care provider tells you that a pituitary tumor must be removed.
* **Vision testing.** A pituitary tumor can affect eyesight, especially side vision, also called peripheral vision. Testing your eyes to check how well you can see may help your health care provider decide if other tests may be needed to detect a pituitary tumor.

Your health care provider may refer you to a specialist in hormone disorders, called an endocrinologist, for more testing.

**TREATMENT OPTIONS**

Many pituitary adenomas don't need treatment. They are not cancer, so if they don't cause symptoms, simply watching them over time may be a good approach. If treatment is needed, the specific treatment depends on the tumor type, size, location and growth over time. If a tumor is causing too much or too little of certain hormones in the body, that also affects the treatment. Your age and overall health play a role in treatment planning too.

The goal of treatment is to:

* Return hormone levels to a healthy range.
* Avoid more damage to the pituitary gland and restore its regular function.
* Reverse symptoms caused by tumor pressure or prevent them from getting worse.

If a pituitary adenoma needs treatment, it may include surgery to remove the tumor. Medication or radiation therapy also might be used to treat a pituitary adenoma. Treatment involves a team of medical experts. The team may include a:

* Brain surgeon, also called a neurosurgeon.
* Nose and sinus surgeon, also called an ENT surgeon.
* Hormone disorder specialist, also called an endocrinologist.
* Radiation therapy specialist, also called a radiation oncologist.

### **Surgery**

Surgery to treat a pituitary tumor involves removing the tumor. This is sometimes called a tumor resection. A surgeon may suggest surgery if a pituitary adenoma:

* Presses on the optic nerves and limits eyesight.
* Causes other symptoms, such as headache or facial pain.
* Lowers hormone levels in the body due to pressure on the pituitary gland.
* Causes the body to make too much of some hormones.

Results after surgery typically depend on the adenoma type, its size and location, and whether the tumor has grown into tissues around it.

Surgeries to remove a pituitary tumor include endoscopic transnasal transsphenoidal surgery and craniotomy.

### **Endoscopic transnasal transsphenoidal surgery**

This surgery also is called adenomectomy. It's the most common surgery used to remove a pituitary adenoma.

During the surgery, a surgeon — typically a neurosurgeon partnering with a nose and sinus surgeon — removes the adenoma through the nose and sinuses. The surgery doesn't require an external cut, also called an incision. It does not affect other parts of the brain. The surgery doesn't cause a scar that you can see.

Large macroadenomas may be hard to remove with this surgery. That's particularly true if a macroadenoma has spread to nearby nerves, blood vessels or other parts of the brain.

### **Transcranial surgery**

This surgery also is called a craniotomy. It's used less often than endoscopic transnasal transsphenoidal surgery for pituitary tumors. This surgery makes it easier to reach and remove large macroadenomas or pituitary tumors that have spread to nearby nerves or brain tissue. It also makes it easier for the surgeon to see the extent of the tumor, as well as the parts of the brain around it. During transcranial surgery, the surgeon removes the tumor through the upper part of the skull through a cut in the scalp.

Endoscopic transnasal transsphenoidal surgery and transcranial surgery are generally safe procedures. Complications are uncommon. But as with any surgery, there are risks. Complications after pituitary tumor surgery can include:

* Bleeding.
* Infection.
* Reaction to the medicine that puts you in a sleep-like state during surgery This sleep-like state is called anesthesia.
* Temporary headache and nasal congestion.
* Brain injury.
* Double vision or loss of vision.
* Damage to the pituitary gland.

### **Diabetes insipidus**

Surgery to remove a pituitary tumor might damage the pituitary gland. That can limit its ability to make hormones, leading to other medical problems including diabetes insipidus. This condition happens when the pituitary gland cannot make enough of the hormone vasopressin. That hormone is made in the back of the gland, an area called the posterior pituitary. Diabetes insipidus causes the body's fluids to go out of balance, which then causes the body to make large amounts of urine. That can cause extreme thirst and raise the risk of dehydration. Diabetes insipidus after surgery to remove a pituitary tumor usually is short term. It typically goes away without treatment within a few days. If diabetes insipidus lasts longer than that, treatment with a manufactured form of vasopressin may be used. The condition often goes away after several weeks or months.

If your health care provider suggests surgery to treat a pituitary tumor, ask about which surgery is right for you. Talk about the possible complications, risks and side effects. Ask what you can expect during recovery.

### **Radiation therapy**

Radiation therapy uses high-energy sources of radiation to treat pituitary tumors. Radiation therapy can be used after surgery. Or it can be used alone if surgery isn't an option.

Radiation therapy can be helpful if a pituitary tumor:

* Isn't completely removed with surgery.
* Comes back after surgery.
* Causes symptoms that medications don't relieve.

The goal of radiation therapy for pituitary adenomas is to control adenoma growth or to stop the adenoma from making hormones.

Methods of radiation therapy that can be used to treat pituitary tumors include:

* **Stereotactic radiosurgery.** Often delivered as a single high dose, this type of radiation therapy precisely focuses radiation beams on the tumor. Although the word "surgery" is in its name, no cut into the skin is needed. It delivers radiation beams the size and shape of the tumor into the tumor with the aid of brain-imaging techniques. This requires attaching a head frame to the skull. The frame is removed right after treatment. Little radiation comes in contact with healthy tissue near the tumor. That lowers the risk of damage to the healthy tissue.
* **External beam radiation.** This method also is called fractionated radiation therapy. It delivers radiation in small amounts over time. A series of treatments usually is done five times a week for 4 to 6 weeks.
* **Intensity modulated radiation therapy.** This type of radiation therapy, also called IMRT, uses a computer that allows the beams to be shaped to surround the tumor from many angles. The strength of the beams can be limited. That lowers the risk of side effects on healthy tissue.
* **Proton beam therapy.** Another radiation option, proton beam therapy uses positively charged ions, called protons, to target tumors. Proton beams stop after releasing their energy within the tumor. This means the beams can be controlled to target a pituitary adenoma with less risk of side effects in healthy tissue. This type of radiation therapy requires special equipment. It isn't widely available.

Potential side effects and complications of radiation therapy for pituitary adenomas can include:

* Damage to the pituitary gland that limits its ability to make hormones.
* Damage to healthy tissue near the pituitary gland.
* Vision changes due to damage to the optic nerves.
* Damage to other nerves close to the pituitary gland.
* Slightly increased risk of developing a brain tumor.

A radiation oncologist evaluates your condition and talks with you about the benefits and risks of radiation therapy for your situation. It usually takes months to years to see the maximum benefit of radiation therapy for pituitary adenomas. Side effects and complications from radiation therapy usually don't show up right away either. It is important to have regular long-term follow-up care to detect any hormone problems that may happen due to radiation therapy.

### **Medications**

Treatment with medications can be useful for the management of pituitary adenomas. They can help lower the amount of hormones the body makes due to a tumor. Some medications also can shrink certain types of pituitary tumors.

### **Pituitary tumors that make prolactin**

The following medications are used to lower the amount of prolactin a pituitary adenoma makes. They also can often shrink the tumor.

* Cabergoline.
* Bromocriptine (Parlodel, Cycloset).

Possible side effects include:

* Dizziness.
* Mood disorders, including depression.
* Headache.
* Weakness.

Some people develop compulsive behaviors, such as problems with gambling, while taking these medications. Those behaviors also are called impulse control disorders.

### **Pituitary tumors that make adrenocorticotropic hormone**

Tumors that make adrenocorticotropic hormone, also called ACTH, cause the body to make too much cortisol. That condition is known as Cushing disease. Medications that can lower the amount of cortisol the body makes include:

* Ketoconazole.
* Metyrapone (Metopirone).
* Osilodrostat (Isturisa).

Possible side effects of these medications include a heart problem that can lead to a serious heartbeat irregularity.

Another medication called mifepristone (Korlym, Mifeprex) can be used for people with Cushing disease who have type 2 diabetes or glucose intolerance. Mifepristone doesn't lower the amount of cortisol the body makes. Instead, it blocks the effects of cortisol on the body's tissues.

Side effects of mifepristone include:

* Tiredness.
* Weakness.
* Nausea.
* Heavy vaginal bleeding.

The medication pasireotide (Signifor) works by lowering the amount of ACTH a pituitary adenoma makes. It's taken as a shot twice a day. Providers typically suggest pasireotide when surgery to remove an adenoma doesn't work. It also may be used when an adenoma can't be removed with surgery.

Side effects of pasireotide include:

* Diarrhea.
* Nausea.
* High blood sugar.
* Headache.
* Stomach pain.
* Tiredness.

### **Pituitary tumors that make growth hormone**

Two kinds of medication can treat pituitary tumors that make growth hormone. Providers often prescribe these medications when surgery to remove a pituitary adenoma hasn't worked to return the amount of growth hormone in the body to a healthy level.

* **Somatostatin analogs.** This type of medication lowers the amount of growth hormone that the body makes. It also may partially shrink a pituitary adenoma. Somatostatin analogs include:
  + Octreotide (Sandostatin).
  + Lanreotide (Somatuline Depot).
* These medications are given as a shot, usually every four weeks. A form of octreotide that can be taken in a pill, called Mycapssa, also is available. It works like the forms that are given as a shot and has similar side effects.  
  Side effects of somatostatin analogs include:
  + Nausea and vomiting.
  + Diarrhea.
  + Stomach pain.
  + Dizziness.
  + Headache.
  + Pain at the site of the shot.
  + Gallstones.
  + Worsening diabetes.
* Many of these side effects improve over time.
* **Pegvisomant (Somavert).** This medication blocks the effect of too much growth hormone on the body. It's taken as a shot every day. This medication may cause the side effect of liver damage in some people.

### **Pituitary hormone replacement**

The pituitary gland controls growth, thyroid function, adrenal function, reproductive function and the balance of water in the body. One or all of those can be harmed by a pituitary adenoma or by its treatment with surgery or radiation. This is because of the hormone changes they can cause. If your hormones fall to unhealthy levels, you may need to take hormone replacement therapy. This can restore hormones to a healthy level.

### **Watchful waiting**

In watchful waiting — also known as observation, expectant therapy or deferred therapy — you might need regular follow-up tests to see if a tumor grows or if hormone levels change. Watchful waiting may be a choice for you if an adenoma isn't causing any symptoms or triggering other health problems. Talk to your health care provider about the benefits and risks of watchful waiting versus treatment in your situation.

**OUTLOOK / PROGNOSIS**

### ***Visual prognosis***

Visual field improvement has been reported in 79–95% of patients undergoing pituitary adenoma resection, and visual acuity improvement in 45–86%.

The pattern of visual function recovery depends on the severity of the anterior visual pathway neuronal dysfunction:

* Visual impairment due to focal conduction block: This is the first stage; patients will experience normalization of the visual field defect within a week after optic chiasm decompression.
* Visual impairment secondary to demyelination or decreased axonal transport without axonal loss: This is the second stage; patients will experience a slow progressive visual function recovery between 1 to 4 months after optic chiasm decompression.
* Visual impairment due to irreversible neuronal damage: Patients will have minimal visual improvement through visual pathway remodeling and remyelination.

Optical coherence tomography (OCT) can be used as a tool for predicting visual function recovery after pituitary tumor treatment. Normal macular ganglion cell-inner plexiform layer (mGCIPL) and peripapillary retinal nerve fiber layer (pRNFL) thickness have been associated with a better postoperative visual outcome.

OCT cutoff values are difficult to determine given the variation in values depending on the machine manufacturer used. Average pRNFL thickness below 75 to 81 um, and average mGCIPL thickness below 67 um have been associated with a worse visual outcome.

***Tumor recurrence***

A high mitotic index, a Ki67 index greater than 3%, and histological subtypes such as sparsely granulated somatotroph adenomas, silent corticotrophin adenomas, Crooke cell adenomas, plurihormonal PIT1-positive tumors, and lactotroph macroadenomas in men have been suggested as potential prognostic pathological markers for aggressiveness. [7]  Additionally, an evidence-based score, PANOMEN 3, has been proposed to assess prognosis in patients with resected and unresected adenomas.

Approximately 30% of resected adenomas have persistent progressive growth in the next 4 years after surgery.

A meta-analysis that included 17,509 patients with pituitary adenomas treated surgically showed a 5-year postoperative recurrence rate of 4% in somatotroph adenomas, 11% in corticotroph adenomas, 12% in nonfunctioning adenomas, and 18% in prolactinomas.

### ***Pituitary carcinoma***

The presence of metastasis secondary to pituitary carcinomas can occur. However, its prevalence is extremely low. Pituitary carcinomas constitute less than 0.1% of all anterior pituitary tumors.

**POSSIBLE COMPLICATIONS**

Pituitary tumors usually don't spread to other parts of the body. They can affect a person's health, though. Pituitary tumors may cause:

* Problems with eyesight, including vision loss.
* High blood pressure.
* High blood sugar.
* Bone loss.
* Heart problems.
* Problems with thinking and memory.

### **Seizures**

If a pituitary tumor presses on part of the brain called the medial temporal lobe, it may lead to a seizure. This type of seizure is known as a focal seizure with impaired awareness.

These seizures involve a change in or loss of consciousness or awareness. If you have one of these seizures, you may seem to be awake. But you stare into space and don't respond as you typically would to the environment around you. You may not remember the seizure after it happens.

### **Permanent low hormone levels**

Having a pituitary tumor or having one removed with surgery may permanently change your body's hormone supply. As a result, you may need hormone replacement therapy for the rest of your life.

### **Pituitary apoplexy**

A rare but potentially serious complication of a pituitary tumor is pituitary apoplexy. This happens when there is sudden bleeding into the tumor. Symptoms include:

* A severe headache, likely worse than you've ever had before.
* Problems with your eyesight, including double vision or loss of vision in one or both eyes.
* Nausea and vomiting.
* Confusion or other reduced mental function.

Pituitary apoplexy requires emergency treatment. The treatment usually includes taking corticosteroid medicine to ease swelling around the tumor. You also may need surgery to remove the tumor.

**WHEN TO SEE A DOCTOR / RED FLAG**

Call your healthcare provider if your symptoms return or you have new symptoms.

**DIFFERENTIAL DIAGNOSIS**

***Diagnostic Considerations***

Several other intracranial neoplasms can present as intrasellar tumors. These include craniopharyngiomas, meningiomas, neurofibromas, ectopic germinomas, and, rarely, metastatic tumors.

Granulomatous and infectious disorders can localize to the sellar region or the hypothalamus (eg, sarcoid, tuberculomas, and granulomatous polyangiitis vasculitis).

Carotid artery aneurysm can occur in the intrasellar region.

Lesions in the sphenoid sinus, such as a mucocele, can mimic the clinical picture of a pituitary adenoma.

An unusual postpartum lymphocytic inflammatory pituitary lesion can mimic a pituitary tumor. This is known as lymphocytic hypophysitis.

Acromegaly can result from a nonpituitary source of increased growth hormone.

Differentiating between Cushing disease and Cushing syndrome related to adrenal hyperplasia or tumor is important.

Hypothalamus compression can cause increased prolactin levels because of a decrease in the prolactin inhibitory factor. Thus, hyperprolactinemia may be seen with nonprolactin-secreting pituitary adenomas and other sellar lesions with hypothalamic compression.

Other causes of hyperprolactinemia that are unrelated to mass lesions in the pituitary or the hypothalamus include the following:

* Intracranial - Empty sella syndrome, status post cranial irradiation
* Pharmacological - Antipsychotics (and other dopamine receptor antagonists), methyldopa, reserpine, verapamil, estrogen, opiates, cimetidine, sulpiride
* Endocrine - Primary hypothyroidism
* Metabolic - Chronic renal failure, cirrhosis
* Other unusual causes - Breast manipulation, chest wall lesions, spinal cord lesions, stress

In some cases, a specific cause cannot be established.

***Differential Diagnoses***

* Basilar Artery Thrombosis
* Brainstem Gliomas
* Cavernous Sinus Syndromes
* Cerebral Venous Thrombosis
* Glioblastoma
* Intracranial Hemorrhage
* Leptomeningeal Carcinomatosis (Metastasis) Imaging
* Low-Grade Astrocytoma
* Meningioma
* Pediatric Craniopharyngioma
* Pediatric Ependymoma
* Primary CNS Lymphoma
* Thoracic Aneurysm
* Tuberculous Meningitis

**EPIDEMIOLOGY**

***Frequency***

Pituitary tumors represent 10%–15% of all intracranial tumors.Whereas incidental pituitary tumors are found in approximately 10% of autopsies and neuroimaging studies of healthy subjects, clinically evident pituitary adenomas are present in 1 of 11,000 subjects in the general population.

The relative frequency of clinically evident pituitary adenomas is approximately 41%–62% in prolactinomas, 15–48% in clinically nonfunctioning adenomas, 6%–14% in somatotroph adenomas, 2–6% in corticotrophin adenomas, and approximately 1% in thyrotroph adenomas.

The incidence of clinically evident pituitary adenomas has been estimated to be 40 per million individuals per year. The annual incidence of pituitary adenomas per 1 million individuals is estimated to be in the range of 16 to 26 for prolactinomas, approximately 10 for somatotroph adenomas, and 1.6 for corticotrophin adenomas. However, it is important to note that these rates can vary based on population demographics and geographic location.

***Demographics***

Among patients with a clinically evident pituitary adenoma, 62%–77% are women. However, sex distribution is influenced by the type of pituitary adenoma and age.

Microprolactinomas have a female to male ratio of 20:1. Likewise, annual incidence of prolactinomas is higher in women than in men: 24–37 vs 7.6–9 per million, respectively.

Nevertheless, after menopause, prolactinoma incidence is similar in both sexes. Cushing disease is also more frequent in women with a female-to-male ratio of 3:1. On the contrary, nonfunctioning adenoma and acromegaly have similar frequency among males and females

Pituitary adenoma diagnosis predominates in middle-aged and elderly adults. However, its diagnosis can occur at any age. Peak acromegaly diagnosis occurs at age 40 to 60 years, prolactinoma in women usually predominates at age 25 to 40 years, and Cushing disease disproportionately affects young women.

***Mortality/Morbidity***

The mortality rate related to pituitary tumors is low. Advances in medical and surgical management of these lesions and the availability of hormonal replacement therapies have contributed to successful management. Nevertheless, pituitary apoplexy is a life-threatening complication.

Because of cardiometabolic and respiratory comorbidities, acromegaly standardized mortality ranges from 1.41 to 1.45. Standardized mortality ratio in patients with active Cushing disease is 4 to 16 times higher compared to the general population, secondary to stroke and coronary disease Prolactinomas have not reported an increase in standardized mortality ratio.

Morbidity associated with macroadenomas may include permanent visual loss, ophthalmoplegia, and other neurological complications. Tumor recurrence after surgical removal may also occur.

Central nerve system metastases and, rarely, distant metastases can occur with pituitary tumors.Endocrine abnormalities are amenable to correction.

However, damage in many organ systems as a result of longstanding uncorrected deficiencies may be irreversible. Additionally, overtreatment with glucocorticoids or thyroid hormones in patients with hypopituitarism may lead to cardiovascular and bone comorbidities

**PREDEFINED Q & A SETS**

Q1: “What is a pituitary tumor?”  
A pituitary tumor is an abnormal growth in the pituitary gland, a small pea-sized gland at the base of the brain that controls hormone production and regulates many bodily functions.

Most pituitary tumors are benign (non-cancerous) and are also called pituitary adenomas. Some may secrete excess hormones, causing symptoms, while others do not produce hormones but can cause symptoms by pressing on adjacent structures.

Q2: “Are pituitary tumors cancerous?”  
In over 99% of cases, pituitary tumors are benign and not cancer. Malignant pituitary tumors (pituitary carcinomas) are extremely rare.

Q3: “What symptoms do pituitary tumors cause?”  
Symptoms depend on tumor size and hormone activity:

* Hormone-secreting tumors can cause symptoms related to hormone excess (e.g., acromegaly from excess growth hormone, Cushing's disease from excess cortisol)
* Large tumors can cause headaches, vision problems (due to pressure on the optic nerves), and sometimes pituitary hormone deficiencies by compressing normal pituitary tissue.

Q4: “How are pituitary tumors diagnosed?”  
Diagnosis is made through:

* Clinical evaluation of symptoms and hormone levels via blood tests
* Imaging studies, especially MRI of the pituitary region
* Visual field testing if vision symptoms are present.

Q5: “Who treats pituitary tumors?”  
Treatment often involves a multidisciplinary team including endocrinologists (for hormone management), neurosurgeons (for surgery if needed), and sometimes otolaryngologists (ENT surgeons) especially if surgery involves the transsphenoidal approach through the nasal passages.

Q6: “What are the treatment options?”

* Observation: Many small, non-secreting tumors without symptoms are simply monitored over time.
* Medications: Some hormone-secreting tumors can be treated with drugs to normalize hormone levels.
* Surgery: Often done via a minimally invasive transsphenoidal approach to remove the tumor.
* Radiation therapy: Used in certain cases where surgery and medications are insufficient.

Q7: “What is the prognosis?”  
With appropriate treatment, patients with pituitary tumors generally have a good outlook and can lead normal lives. Hormone deficiencies after treatment may require lifelong hormone replacement therapy. Regular follow-up is important to monitor for tumor recurrence or hormonal changes.

Q8: “What questions should I ask my doctor?”

* What type of pituitary tumor do I have?
* Is it hormone-secreting?
* What treatment do you recommend and why?
* What are the risks and benefits of treatment?
* What symptoms should prompt me to seek urgent care?
* How often will I need follow-up and monitoring?

**DOCTOR-PATIENT CONVERSATIONS**

Doctor: Hello, what brings you in today?

Patient: Hi doctor, I’ve been having headaches and some vision problems lately. Also, I feel unusually tired and have noticed some changes in my body.

Doctor: I see. Have you noticed any changes like unexplained weight gain or loss, changes in your menstrual cycle or libido, or any unusual swelling?

Patient: Yes, I’ve gained some weight and my periods have become irregular. My hands and feet also feel larger than before.

Doctor: These symptoms can sometimes be related to pituitary gland issues, including tumors that might affect hormone production and nearby structures like your optic nerves. We will need to do some tests to find out more.

Patient: What kind of tests will I need?

Doctor: We will start with blood tests to check your hormone levels and an MRI scan of your brain to look at the pituitary gland. The MRI will help us see if there is a tumor and its size and location.

Patient: If you find a tumor, what are the treatment options?

Doctor: Treatment depends on the type and size of the tumor and whether it's causing hormone problems. Options include careful observation with regular monitoring if the tumor is small and not causing symptoms, medications to control hormone secretion if it’s hormone-secreting, and surgery to remove the tumor if necessary. Sometimes radiation therapy is used as well.

Patient: What does the surgery involve?

Doctor: Most pituitary tumors can be removed through a minimally invasive procedure called transsphenoidal surgery. It’s done through your nasal passages, so there’s no need for an external incision. The surgery aims to remove the tumor while preserving normal pituitary function.

Patient: What are the risks and recovery like?

Doctor: Like any surgery, there are risks such as bleeding or infection, but serious complications are rare. Recovery usually involves monitoring hormone levels and may require hormone replacement if the normal pituitary tissue is affected. Most patients recover well and can return to normal activities with follow-up.

Patient: How often will I need follow-up?

Doctor: You’ll need regular visits with hormone tests and imaging to monitor for any recurrence or changes. These follow-ups are important for long-term management.

Patient: Thank you. I feel better knowing there are options.

Doctor: You’re welcome. We will work together to find the best treatment for you. Please ask any questions you have at any time.

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**PRESBYCUSIS**

**ALTERNATIVE NAMES:** Alternative names for presbycusis include age-related hearing loss. Presbycusis is also referred to as hearing loss – age related.

### 

**DEFINITION / DESCRIPTION**

Presbycusis (prez-buh-KYOO-sis) is hearing loss that develops gradually as you get older. It’s the most common cause of hearing loss worldwide and affects 2 out of 3 people in the U.S. age 70 or older. The medical term comes from the Greek words for elderly (*presby*-) and hearing (*akousis*). Right now, there’s no cure for age-related hearing loss, but using hearing aids may help to improve hearing.

**CAUSES**

Presbycusis is a type of sensorineural hearing loss, or hearing loss that happens when something damages your inner ear. Risk factors for presbycusis include:

* A family history of hearing loss: Age-related hearing loss can run in families. If some of your family members developed hearing loss as they grew older, chances are you’ll have the condition, as well.
* Ototoxicity: This is inner ear damage that may be a side effect of certain medications.
* Noise exposure: Research shows people with noise-induced hearing loss often develop more severe age-related hearing loss.

Research suggests there’s a relationship between some medical conditions and age-related hearing loss, including:

* Coronary artery disease (CAD).
* Diabetes.
* Hypertension (high blood .
* Stroke.

**SIGNS / SYMPTOMS**

The most obvious symptom is not being able to hear as well as you used to. Presbycusis symptoms develop gradually. You may be developing age-related hearing loss if you:

* Can’t hear certain high-pitched sounds like birds singing or alarms.
* Always need to increase the volume on your cell phone, television, computer or tablet.
* Have tinnitus.
* Have trouble understanding what people are saying when you’re in a crowded room or a noisy place like a restaurant.

**DIAGNOSIS METHODS**

Diagnosis may include:

* Physical examination by a primary care provider. They’ll examine your ears for signs of eardrum damage, inflammation, swelling or an infection. They may use an otoscope to examine your ears. Otoscopes are small magnifying instruments with lights that let providers look at your eardrum and outer ear canal.
* Hearing tests done by an audiologist. The pure tone test is a common test to diagnose presbycusis. This test finds the quietest volume you can hear at each pitch. You’ll wear headphones or earplugs to hear the sounds and speech.

#### **Types of presbycusis**

Your audiologist’s diagnosis may include details about the type of presbycusis that you have. The type you have depends on specific damage to your inner ear. To understand presbycusis types, it may help to know more about your inner ear structure and how hearing works:

1. Your inner ear contains your cochlea. It contains many tiny hair cells (stereocilia).
2. When sound vibrations from your middle ear reach these cells, they send a signal (electrical impulse) to your auditory nerve. This nerve connects your ears to your brain.
3. Your auditory nerve transmits the signal from your stereocilia to your brain, which receives the signals and translates them into sound.

The types of presbycusis include:

* Sensory presbycusis: This hearing loss happens when you lose outer hair cells in the organ of Corti at the base of your cochlea.
* Neural presbycusis: Your ability to hear relies in part on nerve cells in your cochlea and the auditory pathway to your brain. You lose nerve cells over time as part of growing older.

**TREATMENT OPTIONS**

The most common presbycusis treatment is to use hearing aids. Hearing aids amplify sounds and deliver them to your ear. However, hearing aids can’t cure presbycusis or restore lost hearing.

#### **Does presbycusis type affect treatment?**

No, the treatment is the same for all types of presbycusis. But diagnosing presbycusis type is one way your provider can predict the impact that age-related hearing loss will have on your life.

For example, neural presbycusis affects your ability to understand what people are saying (speech discrimination).

**PREVENTION TIPS**

Presbycusis is a common kind of hearing loss that happens as you age. You can’t prevent it, but there are things you can do that may slow down hearing loss or reduce your risk of developing severe presbycusis:

* Wear earplugs or other ear protection in situations or places where there will be loud sounds, like at a concert or a construction site.
* Turn down the volume of your television or music, particularly if you use headphones to listen to music, watch movies or listen to audiobooks.
* Move away from loud noise if you can’t protect yourself from it. For example, if you’re at a concert but don’t have earplugs, move away from the speakers.
* Stop smoking. Smoking can affect your circulation and your hearing, including increasing the chance you’ll have age-related hearing loss at an earlier age than usual. Talk to your provider about helpful resources for quitting smoking.
* Take care of your overall health. Experts think there may be a link between hearing loss, including presbycusis, and conditions like high blood pressure, stroke, diabetes and coronary artery disease. Reducing your risk for these conditions may help slow hearing loss.

**OUTLOOK / PROGNOSIS**

Presbycusis is a progressive condition, meaning it gets worse over time. But you can manage hearing loss with hearing aids. Most people with age-related hearing loss can manage their condition

.

**POSSIBLE COMPLICATIONS**

Presbycusis does more than take away your ability to hear. If you have trouble hearing, you may feel anxious about talking to others because you strain to understand what they’re saying. Research shows having presbycusis increases your risk of developing:

* Social isolation.
* Mental health issues like anxiety disorder or depression.

***HOME REMEDIES***

How do I take care of myself?

Hearing aids will help you to hear better, but your hearing won’t come back. Living with hearing loss can be challenging even if you’re using hearing aids. Here are some suggestions for managing presbycusis:

* Share your situation: Tell family, friends and colleagues that you have hearing loss. They’ll want to know how they can help, like being sure to face you when they speak.
* Protect your hearing: Loud noise can make presbycusis worse. Remember to wear ear protection anytime you’re going to be around loud noise, whether that’s at work or attending events like concerts.
* Wear your hearing aids: Sometimes, people stop using hearing aids because the devices make their ears hurt. If you have presbycusis, wearing your hearing aids every day may slow down presbycusis. Talk to your audiologist if wearing your hearing aids is uncomfortable. They’ll be glad to adjust them.

**WHEN TO SEE A DOCTOR / RED FLAG**

You’ll probably need annual checkups so your audiologist can do hearing tests to see if your hearing loss is getting worse. Contact your audiologist if you notice you’re having issues hearing even when you wear your hearing aids.

**DIFFERENTIAL DIAGNOSIS**

Presbycusis is a diagnosis of exclusion. If the pattern is consistent with sensorineural hearing loss, the patient should be referred for formal audiometry testing. The differential diagnosis for sensorineural hearing loss includes:

* Noise exposure
* Infection
* Ménière disease
* Trauma
* Autoimmune disease
* Perilymph fistula
* Genetically-inherited hearing loss
* Otosclerosis
* Tumor
* Exposure to ototoxic agents
* Metabolic dysfunction

In addition to audiometry, further testing such as imaging or metabolic assessment can be considered if hearing loss does not follow classic presbycusis characteristics.

Other conditions associated with presbycusis, such as diabetes, hypertension, renal impairment, and hyperlipidemia, should be evaluated.

If the pattern of hearing loss is conductive, then an alternative diagnosis to presbycusis should be considered. These include:

* Cerumen impaction
* Foreign body
* Tumor obstruction
* Infection
* Perforation
* Otosclerosis
* Cholesteatoma

Presbycusis should be considered in older adults presenting with changes in mood and cognition as these may be due to underlying hearing impairment.

**RECENT GUIDELINES OR UPDATES**

Recommendations regarding management include, but are not limited to, the following:

* It is recommended that patients with presbycusis be followed up at least yearly by the hearing aid prescriber
* If both ears have an intelligibility threshold under 60 dB in the free field without lip-reading with a dissyllabic list and a hearing aid, referral to a reference center, where cochlear implant indications can be discussed, is recommended
* Screening for presbycusis-associated comorbidities (cognitive deficit, anxiety and depression, visual or eating disorders, loss of autonomy), with patient referral to the appropriate practitioners, is recommended
* Ensuring management of the patient’s cardiovascular risk factors is recommended
* It is recommended that patients undergoing audiologic rehabilitation with hearing aids be offered a stereophonic fitting so that these individuals can benefit from binaural hearing
* In vivo measurement is recommended for accurate adjustment of hearing aid amplification
* With regard to speech and language therapy for auditory cognitive training, the patient should be offered combined auditory and cognitive therapy no matter how much time has elapsed since the hearing aid fitting
* To optimize management of presbycusis, evaluation of cognitive, verbal, non-verbal, emotional, and lip-reading skills is recommended as part of the speech-language pathology assessment

**EPIDEMIOLOGY**

***Frequency***

*United States*

No accurate account of the incidence of presbycusis in the United States is available. Approximately 25-30% of people aged 65-74 years are estimated to have impaired hearing. For people aged 75 years and older, this incidence is thought to rise to 40-50%.

According to the 2014 National Health Interview Survey, 43.2% of American adults above age 70 years reported having hearing trouble, compared with 5.5% of adults aged 18-39 years and 19.0% of adults aged 40-69 years. It is estimated that by 2025, over 500 million individuals worldwide will suffer from clinically significant hearing impairment.

***International***

The international incidence of presbycusis varies widely among societies. Westernized foreign countries and primitive civilizations have very different patterns of hearing loss.

A 1962 study by Rosen and colleagues of a remote tribe of the Sudan called the Mabaans revealed significantly less hearing loss in the elderly population than in similarly aged people of urban societies.

Whether this is because of the lack of chronic noise exposure or the paucity of other systemic ailments that are common in industrialized societies (eg, atherosclerosis, diabetes, reactive airway disease) is not known. In general, most of the world's population experiences some degree of decline in hearing with advancing age.

*Race:* No known difference exists in prevalence of presbycusis based on race.

*Sex:* No difference in the prevalence of presbycusis between the sexes is found.

*Age:* By definition, prevalence of presbycusis increases with advancing age.

***GENOMIC DATA***

* Genetic Contribution: Studies estimate that about 35–55% of presbycusis cases have a genetic background. Family studies like the Framingham study show heritability around 40%, and some families present higher incidence rates.
* Candidate Genes: Multiple genes have been implicated, including *MYO6* (myosin VI), *GRM7*, *GRHL2*, *KCNQ4*, and others involved in cochlear structure and function. Mutation in these genes can affect hair cell integrity or auditory pathways.
* Mitochondrial DNA (mtDNA) Mutations: Damage and deletions in mitochondrial DNA related to oxidative stress accumulate with age and contribute to cochlear cell dysfunction, especially in the energy-demanding inner ear sensory cells, leading to hearing loss.
* Genome-wide Association Studies (GWAS): These have identified dozens of risk variants influencing presbycusis, supporting its polygenic nature. Variants often have small individual effects but a cumulative impact. No single gene fully explains most cases.
* Ultra-rare Variants: Some cases, particularly those with earlier onset (around age 50), carry rare mutations in genes known to cause congenital or childhood hearing loss, contributing to familial presbycusis

**PREDEFINED Q & A SETS**

Q1: “What is presbycusis?”  
Presbycusis is the gradual loss of hearing that occurs as people age, usually affecting both ears. It is one of the most common causes of hearing loss in older adults and primarily affects the ability to hear higher-pitched sounds.

Q2: “What causes presbycusis?”  
Presbycusis results from progressive changes in the inner ear, especially the loss of sensory hair cells in the cochlea. Contributing factors include aging itself, genetic predisposition, long-term noise exposure, certain medications (ototoxicity), and health conditions such as diabetes or cardiovascular disease.

Q3: “Who is most commonly affected?”  
Presbycusis commonly affects adults over the age of 65. Approximately 1 in 3 adults aged 65 to 74 experience some degree of hearing loss, increasing to nearly half of those 75 and older.

Q4: “What are the typical symptoms?”

Answer:

* Difficulty hearing high-pitched sounds like a phone ringing or birds singing
* Trouble understanding speech, especially in noisy environments
* Sounds may seem muffled or unclear
* Needing to increase volume on phones, TVs, or radios
* Occasional tinnitus (ringing in the ears)
* Difficulty following conversations, especially with background noise

Q5: “How is presbycusis diagnosed?”

A: Diagnosis involves:

* Medical history and symptom review
* Physical examination including otoscopic inspection of the ear canal and eardrum
* Audiometric tests conducted by audiologists to measure hearing thresholds and speech discrimination ability
* Sometimes imaging or blood tests rule out other causes of hearing loss.

Q6: “How is presbycusis treated?”

A:

* Hearing aids are the most common treatment and help amplify sounds to improve hearing clarity
* Assistive listening devices and cochlear implants might be options for some patients with severe hearing loss
* Counseling and communication strategies (like lip reading) may also be helpful
* No current cure exists to reverse presbycusis, but these interventions can significantly improve quality of life.

Q7: “Can presbycusis be prevented?”

A:

* Avoiding excessive noise exposure (using hearing protection)
* Managing cardiovascular health and other chronic conditions that affect hearing
* Avoiding ototoxic medications when possible
* There is no guaranteed prevention, but these strategies may reduce risk or slow progression.

Q8: “What complications are associated with presbycusis?”

A:

* Social isolation and communication difficulties
* Increased risk of depression and cognitive decline due to hearing impairment
* Safety issues from not hearing alarms or warnings properly

**DOCTOR-PATIENT CONVERSATIONS**

Doctor: Hello, what brings you in today?

Patient: Hi doctor, I’ve been having trouble hearing lately, especially in noisy places. I keep asking people to repeat themselves, and it’s getting frustrating.

Doctor: I understand. Have you noticed if it’s harder to hear high-pitched sounds, like the phone ringing or birds?

Patient: Yes, exactly. And sometimes when people talk, their voices sound muffled or unclear, particularly in crowds.

Doctor: That sounds like age-related hearing loss, also called presbycusis. It’s a common condition affecting many adults as they get older. It usually affects both ears and makes it hard to hear higher-frequency sounds.

Patient: Is this something that will get worse?

Doctor: Presbycusis tends to progress slowly over time. While there’s no cure to reverse it, hearing loss can often be managed effectively with hearing aids or other assistive devices.

Patient: What causes it?

Doctor: It results mainly from gradual changes in the inner ear’s sensory cells as we age. Other factors like noise exposure, certain medications, and health conditions can contribute as well.

Patient: What can I do to help manage it?

Doctor: Using hearing aids can significantly improve your ability to hear and understand speech. Also, protecting your hearing from loud noises and managing any health conditions can help slow progression. Counseling about communication strategies is useful too.

Patient: I sometimes find it hard to follow conversations, especially when people speak quickly or don’t face me.

Doctor: That’s very common. When speaking with others, it helps if they face you directly in good lighting so you can see their lips and facial expressions. You can also ask them to speak slowly and clearly.

Patient: Should I bring someone with me to appointments to help?

Doctor: That can be very helpful, especially for remembering information and asking questions. Also, I will make sure to speak clearly and check that you understand everything. If needed, I can provide written instructions too.

Patient: What tests will you do?

Doctor: We’ll do a hearing test, called audiometry, to measure what you can hear and how well you understand speech. Sometimes, additional tests help rule out other causes.

Patient: Thank you, doctor. I feel better knowing there are options.

Doctor: You’re welcome. We’ll work together to find the best way to help you hear better and maintain your quality of life. Please let me know if you have questions anytime.

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**PHARYNGOMAXILLARY SPACE ABSCESS**

**ALTERNATIVE NAMES**

* Parapharyngeal space abscess
* Lateral pharyngeal space abscess
* Pharyngomaxillary (or pharyngo-maxillary) abscess (variation in spelling)

**DEFINITION / DESCRIPTION**

A pharyngomaxillary space abscess is an infection with pus accumulation in the pharyngomaxillary space, which is part of the parapharyngeal space — the area lateral to the pharynx and medial to the pterygoid muscles near the upper jawbone. This deep neck space infection commonly originates from infections in the tonsils, pharynx, or nearby odontogenic (dental) sources.

**CAUSES**

* Infections spreading from adjacent sites: Most abscesses in the pharyngomaxillary or adjacent deep neck spaces arise from infections in nearby areas. These include:
  + *Upper respiratory infections* (e.g., nasopharyngitis, tonsillitis, adenoiditis)
  + *Rhinosinusitis*
  + *Otitis media* (middle ear infection)
  + *Dental infections or dental procedures*
  + *Peritonsillar abscess* or other oropharyngeal infections
* Suppuration of lymph nodes: In children, lymph nodes in and around the pharyngomaxillary region drain the nasopharynx, oropharynx, and adjacent areas. Infections often cause lymphadenitis that can progress to cellulitis and abscess formation.
* Trauma or penetrating injury: Direct inoculation via foreign bodies, trauma, or medical instrumentation (e.g., endoscopy, intubation) can introduce infection.
* Spread from nearby deep neck infections: Infections from the parapharyngeal or retropharyngeal spaces can extend to the pharyngomaxillary space.

**RISK FACTORS**

* Young age/immature immune system: Younger children are more susceptible due to the presence of more prominent lymphoid tissue that can become infected, and less mature immune responses.
* Recent or ongoing upper respiratory infections: Conditions like viral or bacterial nasopharyngitis, tonsillitis increase risk.
* Poor dental hygiene or dental infections
* Trauma or instrumentation to the oral-pharyngeal region
* Chronic ear infections or mastoiditis which can spread infection into deep neck spaces
* Anatomical factors: Proximity of lymphatic drainage and fascial planes facilitates spread

**SIGNS / SYMPTOMS**

* Fever
* Sore throat and odynophagia (painful swallowing)
* Neck swelling extending down to the hyoid bone
* Trismus (difficulty opening the mouth), especially with anterior compartment abscesses
* Medial bulging of the tonsil and lateral pharyngeal wall on examination
* In severe or posterior infections, patients may have high fever, rigors, bacteremia, neurological signs, or signs of vascular involvement like carotid artery erosion or internal jugular vein thrombosis

**DIAGNOSIS METHODS**

* Clinical Evaluation:  
  Based on symptoms such as fever, sore throat, painful swallowing (odynophagia), neck swelling (especially down to the hyoid bone), and sometimes difficulty opening the mouth (trismus). Physical exam may show medial bulging of the tonsil and lateral pharyngeal wall in anterior abscesses or posterior pharyngeal wall swelling in posterior abscesses.
* Imaging:
  + Contrast-enhanced CT scan is the diagnostic gold standard. It clearly shows the size, location, extent of the abscess, involvement of adjacent structures, and guides surgical planning.
  + Ultrasound can be used as an adjunct to assess abscess volume but is less definitive than CT.
  + MRI may be used in complex cases or where further soft tissue detail is needed.
* Laboratory Tests:
  + Complete blood count (CBC) typically shows elevated white blood cells indicating infection.
  + Inflammatory markers (such as C-reactive protein) may be elevated.
  + Culture and sensitivity testing of throat swabs or pus (if drainage is performed) to identify causative bacteria and guide antibiotic therapy.
* Other Investigations:
  + Endoscopy may be used to visualize the oropharynx and pharyngeal walls, especially when differentiation from other deep neck processes is necessary.

**TREATMENT OPTIONS**

* Airway Control: This is critical due to the risk of airway obstruction from swelling. Patients require close monitoring and possible airway intervention if compromised.
* Antibiotics:  
  Parenteral (intravenous) broad-spectrum antibiotics are started to cover the likely pathogens. Common regimens include drugs such as ceftriaxone, ampicillin/sulbactam, or clindamycin. Antibiotic therapy often continues for several days after drainage, followed by 10 to 14 days of oral antibiotics based on culture results.
* Surgical Drainage:
  + Anterior abscesses can often be drained via an intra-oral incision inside the mouth.
  + Posterior abscesses or larger collections usually require an external surgical approach through the submaxillary (below the jaw) area to safely access and drain the pus.
  + Drainage is essential to remove the collection and prevent spread or complications.
* Supportive Care: Includes pain management, hydration, and monitoring for complications.
* Occasionally, small abscesses without airway compromise may respond to IV antibiotics alone, avoiding surgery.

**OUTLOOK / PROGNOSIS**

* Most children improve clinically within 48 to 72 hours after proper surgical drainage combined with antibiotics, with very low rates of complications reported.
* In uncomplicated cases and relatively healthy children, complete recovery without sequelae is excellent.
* Early intervention reduces the risk of serious complications such as airway obstruction, spread of infection to surrounding structures, and systemic illness.
* Conservative treatment with intravenous antibiotics may be successful, especially in younger children with smaller abscesses, but surgical drainage is often required for larger or refractory abscesses.
* The recurrence rate of deep neck abscesses is low (estimated 1-5%) but must be addressed promptly if it occurs.
* Hospital stays typically range from a few days to about a week depending on severity and treatment modality.

**WHEN TO SEE A DOCTOR / RED FLAG**

* Fever and sore throat that worsen or do not improve
* Difficulty swallowing, painful swallowing, or refusal to eat or drink
* Swelling in the neck or throat area, especially near the jaw or tonsils
* Trismus (lockjaw or difficulty opening the mouth)
* Muffled or "hot potato" voice
* Drooling or excessive saliva due to difficulty swallowing
* Difficulty breathing, noisy breathing, or changes in voice indicating possible airway obstruction
* Severe neck pain, restricted or painful neck movement, or torticollis (neck stiffness/tilting)
* Facial swelling or noticeable lumps near the angle of the jaw
* Signs of systemic illness such as irritability, malaise, or appearing very unwell

**DIFFERENTIAL DIAGNOSIS**

* Other Deep Neck Space Infections:
  + Peritonsillar abscess: Usually more superficial with prominent tonsillar swelling and muffled "hot potato" voice.
  + Retropharyngeal abscess: Located more posteriorly with bulging of the posterior pharyngeal wall and possible airway obstruction.
  + Submandibular (Ludwig’s) angina/abscess: Involves the floor of mouth and submandibular space, causing tongue elevation and dental origin.
  + Parotid gland abscess or sialadenitis
* Infectious Conditions:
  + Bacterial pharyngitis or tonsillitis: Acute sore throat without deep space involvement.
  + Epiglottitis: Acute inflammation of epiglottis with drooling and airway distress.
  + Lemierre’s syndrome: Internal jugular vein thrombophlebitis following oropharyngeal infection.
* Neoplastic lesions:
  + Primary tumors of the parapharyngeal space such as:
    - Deep lobe parotid tumors
    - Carotid body tumors (paragangliomas)
    - Lymphoma
    - Local spread of tonsillar carcinoma
* Vascular Lesions:
  + Carotid artery aneurysm or pseudoaneurysm
  + Venous thrombosis
* Non-infectious causes:
  + Reactive lymphadenopathy
  + Tuberculous lymphadenitis or spinal tuberculosis involving cervical region
  + Foreign body or trauma-related swelling

**EPIDEMIOLOGY**

* The incidence ranges from about 0.3 to 2 cases per 1000 people annually in the general population, according to various studies.
* It most commonly affects young adults between 20 and 40 years of age.
* In children, the infection is less common but can have a more severe course due to anatomical and physiological differences.
* Risk factors include dental infections, upper respiratory tract infections, trauma to soft tissues of the face or neck, immune system disorders, and poor hygiene.
* The incidence of parapharyngeal abscesses broadly (including pharyngomaxillary) is estimated around 1 to 2 cases per 100,000 population per year in pediatric epidemiological data, with a higher predisposition in young children under 5 years and males.
* Deep neck infections, including pharyngomaxillary abscesses, historically comprised a significant proportion of deep neck infections prior to the antibiotic era, and though incidence has decreased, cases still occur especially with underlying infections or immune compromise

**PREDEFINED Q & A SETS**

Q1: “What is a pharyngomaxillary space abscess?”  
It is a localized collection of pus (abscess) in the pharyngomaxillary space, which is part of the parapharyngeal space near the upper jawbone and lateral to the throat. It usually arises from bacterial infections originating in the tonsils, pharynx, or dental sources.

Q2: “What causes it?”  
Bacterial infections from upper respiratory tract infections (such as tonsillitis or rhinitis), dental infections (pulpitis, periodontitis), or trauma to the area can lead to abscess formation.

Q3: “What are the common symptoms?”

Answer:

* Fever and general malaise
* Sore throat and painful swallowing (odynophagia)
* Neck swelling, often extending down to the level of the hyoid bone
* Trismus (difficulty opening the mouth) especially in anterior abscesses
* Swelling and bulging of the tonsil and lateral pharyngeal wall
* In severe cases, airway compromise, drooling, or neurological symptoms if vital structures are involved

Q4: “How is it diagnosed?”

Answer:

* Clinical evaluation based on symptoms and physical examination
* Laboratory tests including complete blood count (CBC) to assess infection and bacterial cultures if drainage is done
* Imaging, primarily contrast-enhanced CT scan, to confirm and define the abscess size, location, and extent
* Ultrasound or MRI may be used additionally in some cases
* Endoscopy for visual assessment if needed

Q5: “What is the treatment?”

Answer:

* Prompt airway management if compromised
* Broad-spectrum intravenous antibiotics covering aerobic and anaerobic bacteria (e.g., ceftriaxone, ampicillin/sulbactam, clindamycin)
* Surgical drainage:
  + Anterior abscesses are often drained via an intra-oral approach
  + Posterior or extensive abscesses may require external surgical drainage through the submandibular region
* Supportive care including pain control and hydration
* In select small or early abscesses without airway threat, intravenous antibiotics alone may suffice

Q6: “What are possible complications?”

Answer:

* Spread of infection to adjacent spaces or blood vessels
* Internal jugular vein thrombophlebitis (Lemierre's syndrome)
* Carotid artery rupture or erosion
* Sepsis and systemic infection
* Airway obstruction requiring emergency intervention

Q7: “What is the prognosis?”  
With early recognition, appropriate antibiotics, and timely surgical drainage when needed, the prognosis is generally good. Delayed treatment increases risk of severe complications.

**DOCTOR-PATIENT CONVERSATIONS**

Doctor: Hello, what brings you in today?

Patient: Doctor, I’ve had a severe sore throat and neck swelling for a few days. It’s painful to swallow, and my mouth is hard to open.

Doctor: I see. Have you noticed any fever or chills?

Patient: Yes, I have a high fever and feel quite unwell. My neck is also swollen on one side.

Doctor: These symptoms suggest a deep neck infection called a pharyngomaxillary space abscess. It is a collection of pus in the space near your throat and upper jawbone. It often happens as a complication from infections like tonsillitis or dental infections.

Patient: That sounds serious. How do you confirm this?

Doctor: We will examine you carefully and order a contrast-enhanced CT scan of your neck. This will show us the exact location and size of the abscess. Blood tests will check for infection, and we might also take a sample if we drain the abscess.

Patient: What is the treatment?

Doctor: Treatment includes intravenous antibiotics to fight the infection and usually surgical drainage to remove the pus. For abscesses near the front, we often drain through the mouth. If it involves deeper or more posterior areas, surgery through a small neck incision might be necessary. We will also monitor your airway closely since swelling can make breathing difficult.

Patient: Will I need to stay in the hospital?

Doctor: Yes, typically you will need hospital care for a few days to receive IV antibiotics and after surgery to recover safely. In some very early or small abscesses without airway risk, antibiotics alone might be enough, but most cases require drainage.

Patient: What are the risks if not treated properly?

Doctor: Without prompt treatment, the infection can spread to other neck spaces and vital structures, causing serious complications like airway obstruction, blood clots in neck veins, or even life-threatening spread of infection.

Patient: How soon can I get better?

Doctor: If treated early, most patients recover well within a week or two, though you might need follow-ups to ensure complete healing.

Patient: Thank you, doctor. I’m glad I came in early.

Doctor: You did the right thing. We will take good care of you and keep you informed every step of the way. Please let us know if your symptoms worsen or if you experience difficulty breathing.

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**PARADOXICAL VOCAL FOLD MOTION DISORDER**

**ALTERNATIVE NAME**S: Alternative names for paradoxical vocal fold motion disorder include vocal cord dysfunction (VCD), paradoxical vocal fold motion (PVFM), paradoxical vocal cord movement (PVCM), inducible laryngeal obstruction, functional upper airway obstruction, functional dysphonia, psychogenic upper airway obstruction, and spasmodic dysphonia.

**DEFINITION / DESCRIPTION**

Paradoxical vocal fold motion disorder (PVFM), also known as vocal cord dysfunction (VCD), is an acquired disorder that can affect both breathing and voicing.

It causes episodes of shortness of breath (SOB) that may be triggered by a variety of stimuli, such as exercise (sometimes only one particular sport), strong smells (perfume, smoke, household cleaners) and/or intense emotions (anxiety/stress). Sometimes people lose their voice or the quality changes during episodes.

During normal breathing, the vocal folds open during inhalation (breathing in). However, during an episode of PVFM, the vocal folds partially or almost fully close with inhalation, thus temporarily narrowing the airway.

This behavior is felt to be due to the vocal folds over-protecting the trachea (airway) during exposure to certain stimuli. Over time, this behavioral response can become automatic.

It does not improve with inhalers, as the condition is due to muscle movement in the throat rather than a lung disorder. It is possible to have both PVFM and underlying lung conditions, requiring joint management with pulmonology.

**CAUSES**

Problems associated with vocal cord dysfunction (VCD) include the following:

* Gastroesophageal reflux disease, laryngopharyngeal reflux
* Upper airway inflammation due to allergies (rhinitis), sinusitis, or recurrent viral infections
* Strenuous exercise
* Occupational or other exposure to irritant fumes
* Environmental allergens and/or pollutants
* Psychogenic causes (eg, severe emotional stress)

Bodily sensations such as throat tightening, wheeze, and cough can feed into a cycle of panic that exacerbates symptoms.

**SIGNS / SYMPTOMS**

***Paradoxical Vocal Fold Motion Disorder Symptoms, Signs***

Patients with PVFM often report symptoms of increased difficulty with inhalation, feeling as if the throat is tightening or closing off, and a sound (stridor) with inhalation.

**DIAGNOSIS METHODS**

***Diagnosis and Tests***

The condition can be challenging for healthcare providers to diagnose. For example, studies suggest providers often mistakenly diagnose vocal cord dysfunction as asthma or croup.

To diagnose the condition, a healthcare provider may do a physical examination, ask about your medical history and ask about activities or exposures that could result in VCD. They may order the following tests:

* Laryngoscopy. Your provider will use a camera mounted on a flexible tube that they insert through your nose or throat to examine your vocal cords. They’ll ask you to breathe and speak while they watch to see when your vocal cords open and close. They may do a provocation test to trigger an episode of VCD while looking in your throat so they can understand better what is causing the shortness of breath.
* Pulmonary function tests and spirometry. In these tests, you breathe into a spirometer, a device that measures how much air you inhale and exhale and how quickly you exhale. Your provider may do this test to rule out asthma. Sometimes, the test will show changes in the flow-volume loop that can be a sign of VCD. It’s important to know that these tests can’t diagnose ILO

Diagnosis is made through the process of elimination by first ruling out other conditions that can cause shortness of breath.

Before your referral to the Voice and Swallowing Center, you will typically have seen a pulmonologist (lung doctor) and/or been given inhalers by your PCP and may have had a cardiac (heart) work-up.

If these tests are normal, or the shortness of breath is not fully managed by inhalers, you are then sent to us. We review prior test results and identify any behavioral patterns (chronic throat clearing, hydration, speaking patterns, reflux symptoms) that could be contributing factors.

The gold standard of diagnosing PVFM is by visualizing vocal fold movement during an episode of shortness of breath via a transnasal flexible laryngoscopy or video laryngostroboscopy.

If you have specific triggers (e.g. strong odors/perfumes, physical exertion) we may expose you to those triggers or have you briskly walk or run in an attempt to trigger complaints.

If we see abnormal vocal fold movement, we immediately coach you through breathing exercises to resolve the episode of shortness of breath. The video of the examination is recorded and we review it together.

**TREATMENT OPTIONS**

***Treatment for Paradoxical Vocal Fold Motion Disorder***

Patients with PVFM are often referred for behavioral treatment also known as laryngeal control therapy with one of our specialized voice pathologists.

In laryngeal control therapy, you will be instructed on a set of breathing (respiratory retraining) exercises designed to keep the vocal folds open and the throat relaxed. The goal of therapy is to reduce the effort it takes to breathe and to retrain the breath cycle in an attempt to prevent shortness of breath.

**PREVENTION TIPS**

Avoiding situations and managing conditions that cause your episodes may keep them from happening. For example, if you know that exposure to certain workplace chemicals often leads to ILO symptoms, protecting yourself from exposure by wearing protective breathing gear may reduce the chance you’ll have an episode.

**OUTLOOK / PROGNOSIS**

Prognosis for vocal cord dysfunction (VCD) is good with effective response to speech therapy, which allows patients to take control of their disorder.

VCD can feel life-threatening, but it actually isn’t.

Don’t hesitate to go to the emergency department if you’re having trouble breathing. But tests will likely show you’re getting enough oxygen even though it doesn’t feel like it.

**WHEN TO SEE A DOCTOR / RED FLAG**

Contact your provider if you still have symptoms even though you’re doing your breathing exercises, managing underlying conditions and avoiding situations that lead to episodes. Call 911 or go to the emergency room if you have shortness of breath that doesn’t go away.

**DIFFERENTIAL DIAGNOSIS**

***Diagnostic Considerations***

Laryngomalacia, vocal cord paresis, and CNS causes must be differentiated from vocal cord dysfunction (VCD).Additionally, VCD is often misdiagnosed as asthma.

Antenatal prolapse must also be considered.

***Table 1. Differential diagnosis of laryngeal movement disorders***

| VCD |  |
| --- | --- |
| Psychogenic | Somatoform disorder, conversion disorder, abuse, anxiety disorder, depression, Munchausen syndrome, malingering |
| Exercise | Exercise |
| Irritant | Extrinsic (chemical irritants, olfactory stimuli)  Intrinsic (GERD, laryngopharyngeal reflux rhinits/post nasal drip, sinusitis) |
| Laryngospasm | Intubation, airway manipulation, IgE mediated, nocturnal aspiration |
| Vocal Cord Paresis/Paralysis | Prolonged intubation, recurrent laryngeal or vagus nerve damage during chest or thyroid surgery, idiopathic |
| Infectious | Epiglottis, bronchiolitis, laryngotracheobronchitis (croup), laryngitis, pharyngeal abscess, diphtheria, pertussis, laryngeal papillomatosis |
| Neoplastic | Head and neck malignancy, cystic hygroma, hemangioma, rhabdomyosarcoma, teratoma, lymphoma, papilloma |
| Endocrine | Thyroid goiter |

***Differential Diagnoses***

* Allergic and Environmental Asthma
* Anaphylaxis
* Asthma
* Bilateral Vocal Fold Paralysis
* Epiglottitis
* Exercise-Induced Asthma
* Foreign body obstruction
* Laryngeal abnormalities (eg, neoplasm, polyps, cyst)
* Laryngeal edema from C1INH deficiency or ACE inhibitor use
* Laryngeal spasm
* Unilateral Vocal Fold Paralysis
* Upper Respiratory Tract Infection
* Vocal Polyps and Nodules

**EPIDEMIOLOGY**

***Frequency***

Vocal cord dysfunction (VCD) is observed in up to 10% of patients at referral centers seeking evaluation of asthma that is unresponsive to aggressive therapy.

The literature reveals a high incidence of VCD in persons with psychiatric conditions (eg, depression, obsessive-compulsive disorder, borderline personality disorder, neuroses induced by childhood sexual abuse),persons with an increased body mass index,and medical personnel. VCD may complicate true asthma in a small number of patients.

***Mortality/Morbidity***

Mortality rates are unknown, but morbidity is often significant from years of corticosteroid use, resulting in iatrogenic Cushing-like syndrome, bone density loss, and growth suppression in the pediatric population.

Misdiagnosis of VCD as asthma may lead to significant morbidity and increased costs, and misuse of measures of asthma control may be contributing to these findings.

***Demographics***

This condition is predominantly observed in females.The authors' review of the published literature indicates a female-to-male ratio of approximately 3:1.

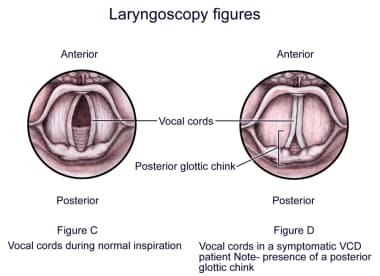
This condition predominates in people aged 20–40 years, but it can occur in people aged 6–83 years. Recent literature suggests an increase of this condition in children and adolescents.

***Procedures***

***Laryngoscopy***

The criterion standard for the diagnosis of vocal cord dysfunction (VCD) is direct visualization of the paradoxical adduction of the true vocal cords during inspiration.

The classic textbook picture is the adduction of the anterior two thirds of the vocal cords with a posterior diamond-shaped chink through which air flows during the inspiratory phase.



Laryngoscopic views of the vocal cords.

If the patient is not symptomatic at the time of laryngoscopy or rhinoscopy, typical vocal cord changes may often be induced by exercise, hyperventilation, or a maximal forced expiratory effort followed by rapid inspiration. These maneuvers may increase the sensitivity of the test.

**PREDEFINED Q & A SETS**

Q1: “What is a paradoxical vocal fold motion disorder?”  
Paradoxical vocal fold motion disorder is a condition where the vocal folds (vocal cords) close when they should open during breathing. Normally, vocal folds open to let air pass when breathing in and close only for speaking, swallowing, or coughing. In PVFM, the vocal folds are inappropriately narrow or closed during inhalation, causing airway obstruction and difficulty breathing.

Q2: “What causes PVFM?”  
The exact cause is unknown but may be triggered by factors including acid reflux, exercise, allergies or airborne irritants, strong emotions, voice overuse, cough, or exposure to fumes. It is often seen in people with a sensitive or reactive airway. Psychological factors may also play a role.

Q3: “Who is affected by PVFM?”  
PVFM can affect people of all ages and is often misdiagnosed, especially in individuals thought to have asthma. It can occur alone or coexist with asthma.

Q4: “What are the symptoms of PVFM?”

Answer:

* Throat or chest tightness
* Noisy breathing or noisy inhalation (stridor)
* Difficulty getting air in (inspiratory dyspnea)
* Feeling of throat closing or choking sensation
* Intermittent shortness of breath or difficulty breathing
* Voice changes or inability to speak during episodes

Q5: “How is PVFM diagnosed?”  
Diagnosis includes clinical history focusing on symptoms that do not improve with asthma treatment, pulmonary function tests showing inspiratory flow limitation, and direct visualization of the vocal folds via laryngoscopy during an episode showing paradoxical vocal fold adduction.

Q6: “How is PVFM treated?”

Answer:

* Respiratory retraining and breathing exercises led by a speech-language pathologist to teach proper vocal fold control
* Identification and avoidance of triggers such as acid reflux or allergens
* Treatment of underlying medical conditions such as GERD or allergies if present
* Psychological counseling if emotional stress is a significant trigger
* Education to differentiate PVFM from asthma, as typical asthma medications may not help

Q7: “What is the prognosis?”  
With proper diagnosis and treatment, most people improve significantly. Treatment focuses on symptom management and preventing episodes.

**DOCTOR-PATIENT CONVERSATIONS**

Doctor: Hello, what brings you in today?

Patient: Hi doctor, I’ve been having trouble breathing, especially when I exercise or get stressed. Sometimes I feel like my throat is closing and I have noisy breathing. It feels really scary.

Doctor: I understand. Have you been diagnosed with asthma or had breathing treatments before?

Patient: Yes, I was told I might have asthma, but the inhalers don’t seem to help much.

Doctor: That’s important information. Your symptoms could be due to a condition called paradoxical vocal fold motion disorder. In this, the vocal cords close when they should open during breathing, especially on inhalation, causing airway narrowing and difficulty breathing.

Patient: So it’s not asthma? How do you diagnose it?

Doctor: We diagnose it mainly by looking at your vocal cords during an episode using a flexible laryngoscope inserted through your nose or mouth. You may also have pulmonary function tests that show difficulty breathing in. We consider your history of symptoms, triggers like reflux or irritants, and how asthma treatments are working.

Patient: What causes this condition?

Doctor: It can be triggered by many things, including acid reflux, allergies, exercise, stress, or irritants like smoke or strong odors. Sometimes emotional stress can play a role too.

Patient: How is it treated? Will I need medication?

Doctor: The main treatment is speech therapy with a specialist who will teach you breathing techniques to control your vocal cords and keep them open during breathing. We also try to manage any triggers like reflux or allergies. Unlike asthma, inhalers usually don’t help with PVFM itself. Psychological support might help if stress is a big factor.

Patient: Can I expect to get better?

Doctor: Yes, most people improve significantly with proper diagnosis and therapy. Learning to recognize and manage your triggers along with these breathing exercises often reduces or stops episodes.

Patient: What should I do if I have a sudden breathing problem?

Doctor: If you feel severe breathing difficulty, seek emergency care. Meanwhile, try to stay calm and use the breathing techniques you learn. Avoid panicking as that can make symptoms worse.

Patient: Thank you, doctor. This helps me understand what’s going on.

Doctor: You’re welcome. We’ll work together to get you the right care. Please let me know if you have new or worsening symptoms.

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**PARAGANGLIOMA**

**ALTERNATIVE NAMES**

Extra-adrenal pheochromocytoma

**DEFINITION / DESCRIPTION**

A paraganglioma (also known as an extra-adrenal pheochromocytoma) is a rare neuroendocrine tumor (NET) that forms near your carotid artery (the major blood vessels in your neck), along nerve pathways in your head and neck and in other parts of your body.

The tumor is made of a certain type of cell called chromaffin cells, which produce and release certain hormones known as catecholamines.

Your adrenal glands, the two glands that are on top of each kidney, make several hormones. Among these are hormones called catecholamines that help control the following important bodily functions:

* Heart rate.
* Blood pressure.
* Blood sugar (blood glucose).
* The way your body responds to stress.

The primary catecholamines include:

* Dopamine.
* Epinephrine (adrenaline).
* Norepinephrine (noradrenaline).

Even though paragangliomas don’t form in your adrenal glands, they’re made of tissue that’s found in your adrenal glands. Paragangliomas may release extra catecholamines into your blood, causing certain signs and symptoms.

### **Paraganglioma and pheochromocytoma**

Paraganglioma and pheochromocytoma are both rare tumors that form from the same type of cells known as chromaffin cells. The difference is where they form in your body.

Pheochromocytomas form in the center of your adrenal gland (adrenal medulla), and paragangliomas form outside your adrenal gland, usually along the arteries or nerves in your neck. Paragangliomas are also called extra-adrenal pheochromocytomas.

### **Is paraganglioma cancer?**

Paragangliomas can be benign (not cancer) or malignant (cancer). Approximately 20% of paragangliomas are malignant.

It can be very challenging for healthcare providers to tell if a paraganglioma is cancerous or not — even after they’ve looked at the tumor tissue under a microscope after it’s been removed. Because of this, a paraganglioma is often considered cancer if it has:

* Spread to nearby tissues (regional spread of paraganglioma).
* Spread to distant areas like your lungs or bone (metastasized).
* Come back after initial treatment (recurred).

There’s no standard staging system for paraganglioma if it’s cancerous. Instead, it’s described as the following:

* Localized paraganglioma: The tumor is in one area only.
* Regional paraganglioma: Cancer has spread to lymph nodes or other tissues near its original location.
* Metastatic paraganglioma: Cancer has spread to other parts of your body, such as your liver, lungs, bone or distant lymph nodes. Approximately 35% to 50% of malignant paragangliomas may spread to other parts of your body (metastasize).
* Recurrent paraganglioma: Cancer has recurred (come back) after it has been treated. It may come back in the same place or in another part of your body.

### **How fast does a paraganglioma grow?**

Paragangliomas usually grow very slowly. But this could vary from case to case.

### **Who does paraganglioma affect?**

Anyone at any age can get a paraganglioma, but they occur most often in people between 30 and 50 years of age. Approximately 10% of cases occur in children.

### **How common is paraganglioma?**

Paraganglioma is a rare tumor. It’s estimated that only 2 out of every 1 million people have paraganglioma.

**CAUSES**

In most cases of paraganglioma, the exact cause is unknown, and it occurs randomly. Approximately 25% to 35% of people who have paraganglioma have a hereditary condition (passed through the family) that’s linked to paraganglioma, including:

* Multiple endocrine neoplasia 2 syndrome, types A and B (MEN2A and MEN2B).
* Von Hippel-Lindau (VHL) disease.
* Neurofibromatosis type 1 (NF1).
* Hereditary paraganglioma syndrome.
* Carney-Stratakis dyad (paraganglioma and gastrointestinal stromal tumor [GIST]).
* Carney triad (paraganglioma, GIST, and pulmonary chondroma).

**SIGNS / SYMPTOMS**

Signs and symptoms of paraganglioma happen when the tumor releases too much adrenaline or noradrenaline into your blood. However, some paraganglioma tumors don’t make extra adrenaline or noradrenaline and don’t cause symptoms (are asymptomatic). Common symptoms of paraganglioma include episodes of:

* High blood pressure (hypertension).
* Headaches.
* Excessive sweating for no known reason.
* A pounding, fast or irregular heartbeat.
* Feeling shaky.

Less common symptoms of pheochromocytoma include:

* Being much paler than you usually are.
* Nausea and/or vomiting.
* Diarrhea.
* Constipation.
* Elevated blood glucose levels (hyperglycemia).
* An extreme drop in blood pressure upon standing suddenly (orthostatic hypotension).
* Unexplained weight loss.

Some people who have a paraganglioma may experience symptoms infrequently or in bursts.

**DIAGNOSIS METHODS**

Since paraganglioma is a rare tumor and is sometimes asymptomatic, it can be difficult to diagnose. Healthcare providers sometimes find paragangliomas when they order a test or procedure for another reason.

A provider may suspect a diagnosis of paraganglioma after reviewing the following factors:

* A detailed medical history, including previous pheochromocytoma or paraganglioma cases in your family.
* A thorough physical and medical evaluation.
* Certain symptom characteristics, such as high blood pressure that’s unresponsive to standard treatment.

### **What tests are used to diagnose paraganglioma?**

Your healthcare provider may use the following tests and procedures to diagnose paraganglioma:

* Physical exam: Your provider will perform a physical exam of your body to check general signs of health, such as your blood pressure. They will also ask you about your medical history, including your family’s medical history as it relates to endocrine issues.
* 24-hour urine test: This type of urine (pee) test involves collecting samples of your urine for 24 hours to measure the amounts of adrenal hormones called catecholamines in your urine. Substances that result from the breakdown of these hormones are also measured. Higher-than-normal amounts of certain catecholamines in your urine may be a sign of paraganglioma.
* Blood catecholamine tests: These tests measure the level of catecholamines in your blood. Substances that result from the breakdown of these hormones are also measured. Higher-than-normal amounts of certain catecholamines in your blood may be a sign of paraganglioma.
* PET scan (positron emission tomography scan): A positron emission tomography (PET) scan uses a safe injectable radioactive chemical called a radiotracer and a device called a PET scanner to produce images of your organs and tissues. The scanner detects metabolically active cells and tumors that absorb large amounts of the radiotracer, which indicates a potential health problem. This imaging test is particularly good for determining the location of paragangliomas.
* CT scan (computer tomography scan): A CT scan is an imaging procedure that takes a series of X-ray images from different angles to provide detailed pictures of areas inside your body. Your provider may recommend a CT scan so that they can determine where the tumor is (most commonly in the neck area).
* MRI (magnetic resonance imaging): An MRI is an imaging procedure that uses a magnet, radio waves and a computer to make a series of detailed pictures of areas inside your body. Your provider may recommend an MRI so that they can look at the region of your body where the tumor is located.

After your provider has diagnosed you with paraganglioma, they’ll likely perform additional tests to see if it has spread to other parts of your body.

### **Is there genetic testing for paraganglioma?**

If you’re diagnosed with paraganglioma, your provider will likely recommend genetic counseling to find out your risk for having an inherited syndrome and associated cancers.

Your healthcare provider may recommend genetic testing if any of the following situations apply to you:

* You have a personal or family history of traits linked with inherited pheochromocytoma or paraganglioma syndrome.
* You have signs or symptoms of higher-than-normal catecholamine levels in your blood or cancerous paraganglioma.
* You’ve been diagnosed with paraganglioma before age 40.

If your genetic counselor finds certain gene changes in your testing results, they will likely recommend that your family members who are at risk but don't have signs or symptoms be tested as well.

**TREATMENT OPTIONS**

Treatment options for paraganglioma depend on several factors, including:

* The size of the tumor.
* If the tumor is benign (not cancer) or malignant (cancer).
* If you have symptoms caused by higher-than-normal levels of catecholamines.
* If the tumor is in one area only or has spread to other places in your body (metastasized).
* If the tumor has been diagnosed for the first time or has come back (recurred).

If you have a paraganglioma that causes symptoms due to excess adrenal hormones, your healthcare provider will likely recommend medication to manage the symptoms. Medications may include:

* Medication that keeps your blood pressure regulated, such as alpha-blockers.
* Medication that keeps your heart rate normal, such as beta-blockers.
* Medication that blocks the effect of the excess hormones released by your adrenal gland(s).

Treatment options for paraganglioma include:

* Surgery and tumor removal.
* Radiation therapy.
* Chemotherapy.
* Ablation therapy.
* Targeted therapy.

Together, you and your healthcare team will determine a treatment plan that works best for you and your situation.

#### **Surgery and tumor removal**

Surgery is the main form of treatment for paragangliomas. During the surgery to remove the tumor, your surgeon will check the surrounding tissue and lymph nodes to see if the tumor has spread. If it has, your surgeon will remove the affected tissue(s) as well, if possible.

Most paragangliomas can be removed using minimally invasive techniques such as laparoscopic surgery, which involves making a few small incisions in your skin and removing the tumor with special instruments. However, traditional open surgery may be needed for large tumors.

After surgery, your provider will check your catecholamine levels in your blood or urine. Normal catecholamine levels are a sign that all the paraganglioma cells were removed.

#### **Radiation therapy**

Radiation therapy is a cancer treatment that focuses strong beams of energy to destroy cancer cells or keep them from growing while sparing as much surrounding healthy tissue as possible.

There are two types of radiation therapy:

* External radiation therapy: This therapy uses a machine outside your body to send radiation toward cancer.
* Internal radiation therapy: This therapy uses a radioactive substance sealed in needles, seeds, wires or catheters that a healthcare provider places directly into or near cancer.

The type of radiation therapy your provider may recommend depends on whether your cancer is localized, regional, metastatic or recurrent. Providers most often use external radiation therapy and/or 131I-MIBG therapy to treat malignant paraganglioma. The treatment 131I-MIBG is a radioactive substance infusion that collects in certain kinds of tumor cells, killing them with the radiation that it gives off.

#### **Chemotherapy**

Chemotherapy is the standard therapy for treating metastatic paraganglioma. It’s a cancer treatment that uses drugs to stop the growth of cancer cells by killing the cells or by preventing them from dividing and multiplying. Chemotherapy is usually given through a vein (intravenously). It’s usually an effective treatment, but it can cause side effects.

#### **Ablation therapy**

Ablation therapy is a minimally invasive treatment option that uses very high or very low temperatures to destroy tumors. Ablation therapies that can help kill cancer cells and abnormal cells include:

* Radiofrequency ablation: This is a therapy that uses radio waves to heat and destroy cancer cells and abnormal cells. The radio waves travel through electrodes (small devices that carry electricity).
* Cryoablation: This therapy uses liquid nitrogen or liquid carbon dioxide to freeze and destroy cancer cells and abnormal cells.

#### **Targeted therapy**

Targeted therapy is a treatment option that uses medications or other substances to attack specific cancer cells without harming healthy cells. Healthcare providers use targeted therapies to treat metastatic and recurrent paraganglioma.

Researchers are currently studying sunitinib, a type of tyrosine kinase inhibitor, for treatment for metastatic paraganglioma. Tyrosine kinase inhibitor therapy is a type of targeted therapy that prevents tumors from growing.

**PREVENTION TIPS**

Unfortunately, you can’t prevent developing a paraganglioma. However, if you’re at risk for developing a paraganglioma due to certain inherited syndromes and genes, genetic counseling can help screen for paraganglioma and potentially catch it in its early phases.

Talk to your healthcare provider if you have any first-degree relatives (siblings and parents) that have been diagnosed with paraganglioma or pheochromocytoma and/or any of the following genetic conditions:

* Multiple endocrine neoplasia 2 syndrome.
* Von Hippel-Lindau (VHL) disease.
* Neurofibromatosis type 1.
* Hereditary paraganglioma syndrome.
* Carney-Stratakis dyad.
* Carney triad.

**OUTLOOK / PROGNOSIS**

The prognosis (outlook) for paraganglioma varies depending on certain factors, including:

* Where the tumor is in your body and how big it is.
* If it’s cancer and has spread to other parts of your body.
* If the tumor was able to be surgically removed and if so, how much of the tumor was removed during surgery.

People who have a small paraganglioma that has not spread to other parts of their body (has not metastasized) have a five-year survival rate of about 95%. People who have paraganglioma that has come back after initial treatment (recurred) or spread to other parts of their body (metastasized) have a five-year survival rate between 34% and 60%.

There are also cases of aggressive paraganglioma tumors that haven’t metastasized but have invaded local tissue to the point where surgery can’t fully remove it. In these cases, the excess release of adrenaline and noradrenaline can be dangerous and difficult to treat.

If paragangliomas are left untreated, whether benign or malignant, they can potentially cause serious, life-threatening complications due to the excess amounts of adrenaline and noradrenaline they can secrete. Complications can include:

* Heart muscle disease (cardiomyopathy).
* Inflammation of your heart muscle (myocarditis).
* Uncontrolled bleeding in your brain (cerebral hemorrhaging).
* Accumulation of fluid in your lungs (pulmonary edema).
* Heart attack (myocardial infarction).
* Stroke.
* Coma.
* Death.

**WHEN TO SEE A DOCTOR / RED FLAG**

If you’ve been diagnosed with paraganglioma and experience concerning symptoms, contact your healthcare provider.

If you’re experiencing symptoms of paraganglioma, such as high blood pressure and headaches, talk to your provider. Even though paraganglioma is rare and the likelihood of having it is low, it’s important to treat high blood pressure regardless.

If you’ve recently found out that one of your first-degree relatives (siblings and parents) has a genetic syndrome, such as multiple endocrine neoplasia 2 syndrome or von Hippel-Lindau (VHL) disease, that puts you at a higher risk of developing a paraganglioma, ask your provider about genetic testing.

**DIFFERENTIAL DIAGNOSIS**

The following list encompasses possible differential diagnoses based on symptom characteristics:

* Endocrine
  + Pheochromocytoma
  + Thyrotoxicosis
  + Insulinoma
  + Hypoglycemia
  + Medullary thyroid carcinoma
* Cardiovascular
  + Labile essential hypertension
  + Positional orthostatic hypotension
  + Carcinoid syndrome
  + Pulmonary edema
  + Syncope
  + Orthostatic hypotension
  + Cardiac arrhythmia
  + Angina
  + Renovascular disease
* Psychological
  + Hyperventilation
  + Vancomycin infusion reaction
  + Anxiety and panic attacks
  + Factitious disorder
  + Somatization disorder
* Pharmacologic
  + Sympathomimetic drug ingestion
  + Recreational drug ingestion (cocaine, phencyclidine [PCP], lysergic acid diethylamide [LSD])
  + Chlorpropamide-alcohol flush
  + Withdrawal of adrenergic inhibitor
  + Monoamine oxidase inhibitor treatment combined with a decongestant
* Neurologic
  + Autonomic neuropathy
  + Migraine
  + Cerebrovascular accident
  + Autonomic seizures
* Other
  + Mast cell disease
  + Recurrent idiopathic anaphylaxis

**EPIDEMIOLOGY**

Paragangliomas are related to pheochromocytomas and are often grouped together, contributing to some uncertainty regarding their precise incidence. In the US, medical professionals diagnose approximately 500 to 1000 paragangliomas annually. The combined incidence of paragangliomas and pheochromocytomas is 0.7 to 1.0 per 100,000 person-years.

Most patients with sporadic paraganglioma present between their 30s and 50s, with the mean age of diagnosis being 47. Tumors located in the skull base and neck tend to occur in older patients.

In contrast, abdominal tumors are more likely to present in patients at a younger age, around 36, compared to 43 for those with tumors in the skull base and neck.

Patients with hereditary paragangliomas usually present about 10 years earlier than those with sporadic disease, often in their early 30s.

Most paragangliomas are benign. The incidence of malignant paragangliomas is around 90 to 95 cases per 400 million person-years.

Among patients with hereditary paraganglioma, the male-to-female ratio is approximately equal. In contrast, sporadic tumors are much more prevalent in females, with a female-to-male ratio of 3 to 1

***Genetic Tests***

All patients with a paraganglioma should undergo genetic testing.[7] Standard testing evaluates for pathologic variants in *RET, VHL, NF-1, SDHD, SDHC, SDHB, SDHA, SDHAF2, TMEM127,* and *MAX*.

Targeted testing can be considered if the patient presents with an apparent paraganglioma-related syndrome. Genetic testing plays a crucial role in preventing recurrence and guiding long-term observation.

In cases where a genetic variant is identified, it is essential to notify and recommend testing for all first-degree relatives. Patients who are known carriers should undergo the following testing:

* All *SDHx* pathogenic variant carriers
  + Annual screening with plasma or 24-hour urine collection for fractionated metanephrines
* All *SDHB, SDHC,* and at-risk *SDHD* or *SDHAF2* carriers
  + CT or MRI of the skull base, neck, chest, abdomen, and pelvis every 2 to 3 years
  + I-123 MIBG scintigraphy or 68Ga-dotatate–labeled PET-CT every 5 years.

**PREDEFINED Q & A SETS**

Q1: “What caused my paraganglioma?”  
Paragangliomas are rare neuroendocrine tumors caused by abnormal growth of cells in paraganglia, clusters of neuroendocrine cells associated with nerves and blood vessels. Causes include:

* Genetic mutations: About 30-40% of paragangliomas are linked to inherited genetic changes passed down in families. Common syndromes include Multiple Endocrine Neoplasia type 2 (MEN 2), Von Hippel-Lindau disease, Neurofibromatosis type 1, hereditary paraganglioma syndromes, and Carney-Stratakis dyad. These DNA changes predispose individuals to develop these tumors.
* Other factors: Although less common, environmental factors like radiation exposure or low oxygen conditions (hypoxia) may contribute. However, these are not well established causes.
* Many paragangliomas arise sporadically with no clear cause identified.

Q2: “Could my children and/or relatives develop a paraganglioma?”  
Yes, if your paraganglioma is linked to an inherited genetic mutation or syndrome, there is a possibility that your children or close relatives could develop paragangliomas.

Genetic counseling and testing are recommended to evaluate family risk. Some hereditary paraganglioma syndromes show autosomal dominant inheritance, meaning a 50% chance of passing the mutation to offspring.  
If no hereditary syndrome is found, the risk to relatives is much lower but still worth discussing with your healthcare provider.

Q3: “What are my treatment options?”  
Treatment depends on tumor size, location, symptoms, growth rate, and overall health:

* Active surveillance: Small, asymptomatic, or slow-growing tumors may be monitored regularly with imaging and exams without immediate intervention.
* Surgical removal: Often recommended for accessible tumors causing symptoms or risk of complications. Surgery may require specialized approaches depending on tumor location. Sometimes embolization (cutting blood supply) precedes surgery to reduce bleeding risk.
* Radiation therapy: Precision radiotherapy techniques such as stereotactic radiosurgery (e.g., CyberKnife) can control tumor growth and are used as primary treatment or post-surgery.
* Combination therapies: Some patients require a combination of surgery and radiation.
* Systemic therapies: For metastatic or unresectable tumors, targeted therapies or chemotherapy may be considered. Clinical trials may offer additional options.

Q4: “What are the side effects of different treatment therapies?”

Answer:

* Surgery: Risks include bleeding, nerve injury causing hearing loss, facial weakness, or swallowing difficulties depending on tumor site. Recovery time varies with surgical extent.
* Radiation therapy: May cause fatigue, skin irritation, temporary swelling, or rarely damage to nearby tissues. Long-term risks include radiation-induced tissue changes.
* Medications/systemic therapy: Potential side effects depend on the drugs used but can include nausea, fatigue, or immune suppression.
* Many side effects can be managed with supportive care and rehabilitation.

Q5: “How can I manage my symptoms?”

Answer:

* Treatment of symptoms depends on tumor effects. If the tumor secretes excess hormones (catecholamines), medications to manage high blood pressure and related symptoms are essential.
* Supportive therapies like physical therapy, speech therapy, or audiology may help with nerve-related symptoms after treatment.
* Regular follow-up with your care team is important to monitor tumor status and symptoms.
* Lifestyle measures such as stress reduction, healthy diet, and avoiding stimulants (e.g., caffeine) may help with symptom control but do not treat the tumor itself

**DOCTOR-PATIENT CONVERSATIONS**

Doctor: Hello, what brings you in today?

Patient: Doctor, I was recently told I have a paraganglioma. I’m a bit worried and would like to understand more about it.

Doctor: I can definitely help explain. A paraganglioma is a rare type of tumor that arises from specialized nerve cells called paraganglia. These cells are found near nerves and blood vessels, often in the head and neck, abdomen, or pelvis.

Patient: What causes it? Is it cancer?

Doctor: Paragangliomas are usually benign, meaning they are not cancerous. However, some can be locally aggressive or rarely spread. The exact cause is not always clear. In about 30-40% of cases, genetic mutations inherited in families play a role. Sometimes, it happens sporadically.

Patient: Could my family members also get this tumor?

Doctor: If your tumor is linked to a hereditary genetic mutation, there could be a risk for your children or relatives. That’s why we recommend genetic counseling and testing to assess your family risk and guide screening if necessary.

Patient: What symptoms can this tumor cause?

Doctor: Symptoms depend on the tumor’s size and location:

* In the head and neck, you might notice a painless lump, hearing changes, or nerve-related symptoms like facial weakness.
* If the tumor produces hormones, it can cause high blood pressure, headaches, sweating, or palpitations.
* Sometimes, there are no symptoms, and tumors are found incidentally.

Patient: How do you diagnose a paraganglioma?

Doctor: Diagnosis involves imaging studies like MRI or CT scans, and sometimes specialized scans that detect hormone activity. Blood and urine tests can check for hormone secretion. A biopsy is often not done before surgery because of bleeding risk.

Patient: What treatment options are available?

Doctor: Treatment depends on several factors, including tumor size, location, symptoms, and whether it is hormone-secreting. Options include:

* Surgical removal, which is often the primary treatment.
* Radiation therapy, especially if surgery isn’t feasible or as an adjunct.
* Active surveillance if the tumor is small and not causing problems.

Patient: Are there risks or side effects from treatment?

Doctor: Surgery carries typical risks like bleeding and nerve injury, which may affect functions depending on tumor location. Radiation can cause fatigue or local tissue effects. We will discuss these in detail tailored to your situation.

Patient: What can I do to manage symptoms?

Doctor: If your tumor secretes hormones, medications might be needed to control blood pressure or other symptoms. Supportive therapies and regular follow-up are important. Lifestyle measures like stress reduction can help but don’t replace formal treatment.

Patient: What’s my outlook?

Doctor: Many patients do well with proper treatment and follow-up. Some tumors grow slowly and don’t cause problems for years. We will develop a personalized plan and monitor closely.

Patient: Thank you, doctor. This helps me understand much better.

Doctor: You’re welcome. Please feel free to ask any questions anytime. We are here to support you throughout your care journey.

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**PAROTID TUMORS**

**ALTERNATIVE NAMES:** Alternative names for parotid tumors include salivary duct tumor, and in some cases, they may be referred to as benign mixed tumors, particularly for pleomorphic adenomas. Other specific types, such as Warthin's tumor, are also known as cystadenolymphoma.

**DEFINITION / DESCRIPTION**

Parotid tumors are growths of cells that start in the parotid glands. The parotid glands are two salivary glands that sit just in front of the ears. There is one on each side of the face. Salivary glands make saliva to help with chewing and digesting food.

There are many salivary glands in the lips, cheeks, mouth and throat. Growths of cells, which are called tumors, can happen in any of these glands. The parotid glands are the most common place that salivary gland tumors happen.

Most parotid tumors aren't cancerous. These are called noncancerous or benign parotid tumors. Sometimes the tumors are cancers. These are called malignant parotid tumors or parotid gland cancers.

Parotid tumors often cause swelling in the face or jaw. Other symptoms include problems swallowing or a loss of facial movement.

Diagnosis and treatment for parotid tumors is often done by doctors who specialize in problems that affect the ear, nose and throat. These doctors are called ENT specialists or otolaryngologists.

**CAUSES**

It's not clear what causes parotid tumors. These tumors start as a growth of cells in the parotid glands. The parotid glands are two salivary glands that sit just in front of the ears. There is one on each side of the face. Salivary glands make saliva to help with chewing and digesting food.

Parotid tumors happen when cells in the parotid glands develop changes in their DNA. A cell's DNA holds the instructions that tell the cell what to do. In healthy cells, the DNA gives instructions to grow and multiply at a set rate. The instructions tell the cells to die at a set time. In tumor cells, the DNA changes give different instructions. The changes tell the tumor cells to grow and multiply quickly. Tumor cells can keep living when healthy cells die. This causes too many cells.

Sometimes the changes in the DNA turn the cells into cancer cells. Cancer cells can invade and destroy healthy body tissue. In time, cancer cells can break away and spread to other parts of the body. When cancer spreads, it's called metastatic cancer.

**RISK FACTORS**

Factors that increase the risk of parotid tumors include:

* **Increasing age.** Parotid tumors can happen at any age. However, they are more common in older adults.
* **Previous radiation therapy treatments.** People who have had radiation therapy to the head and neck area in the past are at a higher risk of parotid tumors.
* **Exposure to harmful substances.** People who work with certain substances may have an increased risk of salivary gland tumors, including parotid tumors. Examples of industries associated with an increased risk include those that involve rubber manufacturing and nickel.

**SIGNS / SYMPTOMS**

Signs and symptoms of parotid tumors include:

* A lump or swelling on one side of the face that might appear near the ear or cheek.
* Trouble swallowing.
* Trouble opening the mouth widely.
* Numbness in part of the face.
* Muscle weakness in part of the face.
* Pain in the face.

**DIAGNOSIS METHODS**

Tests and procedures used to diagnose a parotid tumor may include:

* **A physical exam.** A healthcare professional feels the jaw, neck and throat for lumps or swelling.
* **Collecting a sample of tissue for testing.** A biopsy is a procedure to collect a sample of tissue for testing. It typically involves using a needle to collect fluid or tissue from the parotid gland. The needle may be inserted through the skin on the face and into the parotid gland.  
  In the lab, tests can show what types of cells are involved and tell if they're cancerous. This information helps your healthcare team understand your prognosis and which treatments are best for you.  
  Results from a needle biopsy aren't always correct. Sometimes the results say a tumor isn't cancerous when it is. For this reason, some healthcare professionals don't do a biopsy before surgery. Instead, they may take a sample of tissue for testing during surgery.
* **Imaging tests.** Imaging tests help your healthcare team understand the size and location of your tumor. If your parotid tumor is cancerous, imaging tests help look for signs that the cancer has spread. Tests may include ultrasound, MRI and CT.

**TREATMENT OPTIONS**

Parotid tumor treatment often involves surgery to remove the tumor. If the tumor is cancerous, you might need more treatment. This could be with radiation therapy and chemotherapy.

### **Surgery**

Operations used to remove parotid tumors include:

* **Removing part of the parotid gland.** For most parotid tumors, surgeons may cut away the tumor and some of the healthy parotid gland tissue around it. The part of the parotid gland that's left continues working as before.
* **Removing all of the parotid gland.** Surgery to remove all of the parotid gland is called parotidectomy. It might be needed for larger tumors, tumors that are cancerous and those that affect the deeper parts of the parotid gland.
* **Removing more tissue to get all of the cancer.** If parotid gland cancer has grown into nearby bone and muscles, some of these may be taken out with the parotid gland. Surgeons try to remove all of the cancer and a small amount of the healthy tissue that surrounds it. Then they work to repair the area so that you can continue to chew, swallow, speak, breathe and move your face. This may involve moving skin, tissue, bone or nerves from other parts of your body to make repairs. This type of surgery isn't needed for parotid tumors that aren't cancerous.

To get to the parotid gland, surgeons make a cut in the skin near the ear. The cut is often hidden in a crease of skin or behind the ear.

Sometimes a sample of tumor tissue is tested during surgery to see if it's cancer. A doctor who uses blood and body tissue to diagnose diseases, who is called a pathologist, looks at the sample right away. The pathologist tells the surgeon if the tumor is cancerous. This helps the surgeon decide how much of the parotid gland to remove. The pathologist also might test nearby lymph nodes and other tissue for signs of cancer.

The parotid gland surrounds the nerve that moves the muscles of the face. This nerve is called the facial nerve. Surgeons take special care to avoid hurting it. They might use electrical devices to check on the nerve and make sure it works as expected after surgery.

Sometimes the facial nerve gets stretched during surgery. This can cause loss of movement in the face muscles. Muscle movement often gets better over time. Rarely, the facial nerve must be cut in order to get all of the tumor. Surgeons can repair the facial nerve using nerves from other areas of the body or from artificial nerves.

Parotid tumor surgery can be complex. It requires well-trained surgeons and specialists for the best outcome. If you're facing surgery for a parotid tumor, meet with your surgeon before your operation to ask questions. Learning more about the procedure can help you feel more comfortable about your treatment plan. You might consider asking:

* Where will you cut into the skin to reach the parotid gland? Will I have a scar?
* How much of the parotid gland do you plan to remove?
* How likely is it that the facial nerve will be hurt? How will you manage this?
* How will you be sure that you've removed all of the tumor?
* Will you remove any lymph nodes?
* Will I need reconstructive surgery? What will that involve?
* What should I expect during recovery? How long will it take to heal?

### **Radiation therapy**

Radiation therapy uses powerful energy beams to kill cancer cells. The energy can come from sources such as X-rays and protons.

Radiation therapy is used to treat parotid gland cancers. Radiation therapy might be recommended after surgery. The radiation can kill any cancer cells that remain. If surgery isn't possible, radiation therapy might be the first treatment for parotid cancers.

### **Chemotherapy**

Chemotherapy uses strong medicines to kill cancer cells. Chemotherapy is sometimes used to treat parotid gland cancers. It might be needed if there's a risk that the cancer might spread or if surgery isn't an option. In these situations, chemotherapy might be done at the same time as radiation therapy.

Chemotherapy is sometimes used on its own for advanced cancer, such as cancer that has spread to other parts of the body. Chemotherapy may help relieve pain and other symptoms caused by the cancer.

### **Targeted therapy**

Targeted therapy uses medicines that attack specific chemicals in the cancer cells. By blocking these chemicals, targeted treatments can cause cancer cells to die.

Targeted therapy might be an option for treating parotid gland cancers when other treatments haven't helped.

**PREVENTION TIPS**

You may not be able to prevent these tumors. Avoiding known risk factors like using tobacco may lower your risk.

**OUTLOOK / PROGNOSIS**

That depends on whether you have a cancerous or noncancerous tumor. Surgery typically cures noncancerous parotid tumors. It may cure cancerous tumors that healthcare providers detect and treat before they can spread. But these cancerous tumors can come back (recur) years after treatment.

If you had treatment for a cancerous parotid gland tumor, your healthcare provider will schedule regular checkups, including imaging tests, for up to 20 years after treatment. Your checkup schedule may look like the following:

| **Post-treatment timeline** | **Checkup schedule** |
| --- | --- |
| Year 1 | Every one to three months. |
| Year 2 | Every two to six months. |
| Year 3 to 5 | Every four to eight months. |
| Year 5 and beyond | Every 12 months. |

#### **What are the survival rates for people with cancerous parotid gland tumors?**

Survival rates are estimates of the percentage of people with this condition who were alive five years after diagnosis. Rates vary widely depending on factors like:

* Tumor subtype: There are more than 20 tumor subtypes.
* Tumor stage: The higher the tumor stage, the lower the survival rate.
* Tumor grade: People with low-grade tumors are more likely to survive longer than people with high-grade tumors.

Mucoepidermoid carcinoma (MEC) is the most common subtype of cancerous parotid gland tumor. The survival rate ranges from 86% for low-grade tumors to 22% for high-grade tumors.

Here are some things to keep in mind when you think about survival rates for cancerous parotid gland tumors:

* Survival rates vary depending on the tumor subtype, stage and grade.
* Five-year survival rates are estimates based on other people’s experiences.
* Survival rates are updated every five years and may be different now.

If you’re receiving treatment for a cancerous parotid gland tumor, ask your healthcare provider what you can expect.

### **How do I take care of myself?**

Cancerous parotid gland tumors can come back. Here are some suggestions for taking care of yourself after treatment:

* Attend your follow-up appointments: Tests to look for recurring parotid gland cancer may detect cancerous tumors before they spread.
* Avoid tobacco: Experts suspect tobacco use increases your risk of developing parotid gland cancer. If you want to stop using tobacco, ask your healthcare provider about programs to help you do that.
* Consider cancer survivorship: Cancerous parotid gland tumors can come back. Knowing that could make you feel anxious. Cancer survivorship programs may help.

**WHEN TO SEE A DOCTOR / RED FLAG**

You’ll have frequent follow-up appointments if you had treatment for a cancerous parotid gland tumor. But you should contact your healthcare provider if you notice changes like a new lump developing in front of one of your ears.

**DIFFERENTIAL DIAGNOSIS**

Benign Parotid Tumors

* Pleomorphic adenoma (most common benign tumor)
* Warthin tumor (papillary cystadenoma lymphomatosum; often in older men and smokers)
* Basal cell adenoma
* Oncocytoma
* Lipoma (soft tissue benign tumor)
* Schwannoma (nerve sheath tumor)

## Malignant Parotid Tumors

* Mucoepidermoid carcinoma (most common malignant tumor)
* Adenoid cystic carcinoma (noted for perineural invasion)
* Acinic cell carcinoma
* Carcinoma ex pleomorphic adenoma (malignant transformation of pleomorphic adenoma)
* Salivary duct carcinoma
* Squamous cell carcinoma (often metastatic from skin)
* Basal cell adenocarcinoma

## Non-Neoplastic Conditions Mimicking Tumors

* Chronic sialadenitis (inflammation of the gland)
* Necrotizing sialometaplasia (benign inflammatory lesion mimicking malignancy)
* Benign lymphoepithelial cysts (especially in HIV-positive patients)
* Lymphoma (primary or involvement secondary to systemic disease)

## Metastatic Tumors

* Metastases from cutaneous squamous cell carcinoma or melanoma, especially from the head and neck region

**EPIDEMIOLOGY**

* Parotid tumors represent the majority of salivary gland tumors, with about 59-70% of all salivary gland tumors located in the parotid gland.
* Most parotid tumors are benign, accounting for approximately 65-80% of cases, with the most common benign tumors being pleomorphic adenoma (~70%) and Warthin tumor (~17%).
* About 20-35% of parotid tumors are malignant, with mucoepidermoid carcinoma and adenoid cystic carcinoma being the most frequent malignant types.
* Parotid tumors show a slight female predilection overall (around 54%), though some recent data indicate increasing male predominance in certain regions.
* The peak incidence occurs in adults aged between their 4th and 7th decades (30s to 60s), with about 70% of tumors presenting in this age range.
* The overall incidence of salivary gland tumors (including parotid) is relatively low, estimated around 1 to 3 cases per 100,000 population per year.
* The data on trends suggest stable to slightly increasing incidence rates in some areas, with some controversies about possible associations (e.g., mobile phone use); however, no definitive causal link has been established.

**PREDEFINED Q & A SETS**

Q1: “Do I have a parotid tumor?”  
Diagnosis is based on clinical examination, imaging (usually ultrasound, MRI, or CT), and often a biopsy or fine needle aspiration (FNA) to determine if a lump in the parotid region is a tumor and its nature.

Q2: “Is my parotid tumor cancerous?”  
Only pathological examination after biopsy or surgery can confirm if the tumor is benign or malignant (cancerous). Most parotid tumors are benign, but about 15-25% can be malignant.

Q3: “What is the stage of my parotid tumor?”  
Staging is done using imaging studies (CT, MRI, PET) and sometimes surgery, to determine tumor size, invasion of surrounding tissues, and lymph node involvement. Staging helps guide treatment choice.

Q4: “Has my parotid tumor spread to other parts of my body?”  
Spread (metastasis) is assessed using imaging and physical exams. Malignant tumors can spread to lymph nodes in the neck and distant organs, but this varies by tumor type and stage.

Q5: “Will I need more tests?”  
Yes, your doctor might order more imaging tests, biopsies, blood tests, or referral to specialists depending on initial findings and staging. Tests help confirm diagnosis, plan treatment, and check for spread.

Q6: “What are the treatment options?”

Answer:

* Surgery (parotidectomy): Surgical removal of the tumor is the main treatment. Types include superficial (part of gland) or total parotidectomy, depending on tumor size and location. Surgeons aim to preserve the facial nerve if possible.
* Radiation therapy: Often used after surgery for malignant tumors to kill residual cancer cells or when surgery isn't feasible.
* Chemotherapy: Less common but may be used for advanced or metastatic cancer, or combined with radiation.
* Targeted therapy: Some cancers may respond to drugs targeting specific tumor markers.
* Observation: Sometimes small benign tumors may be monitored without immediate surgery.

Q7: “How much does each treatment increase my chances of a cure or prolong my life?”

Answer:

* Surgery offers the best chance for cure for most benign and many malignant tumors detected early.
* Radiation improves local control and survival in malignant cases post-surgery or when surgery isn't possible.
* Chemotherapy has limited effectiveness as a primary treatment but may help with advanced disease.
* Outcomes depend heavily on tumor type, grade, stage, and patient factors. Your specialist will discuss your prognosis specifically.

Q8: “What are the potential side effects of each treatment?”

Answer:

* Surgery: Risks include facial nerve injury causing weakness or paralysis, numbness, scarring, and cosmetic changes.
* Radiation: May cause skin reactions, dryness, swelling, and rarely damage to surrounding tissues.
* Chemotherapy: Can cause nausea, fatigue, hair loss, and immune suppression, depending on drugs used.

Q9: “How will each treatment affect my daily life?”

Answer:

* Surgery usually requires recovery time and may temporarily or permanently affect facial movement and sensation.
* Radiation therapy involves multiple sessions and side effects that can impact comfort and eating.
* Chemotherapy causes systemic effects that may reduce energy and immunity.
* Rehabilitation and support services help manage these impacts.

Q10: “Is there one treatment option you believe is the best?”  
Treatment is individualized based on your tumor’s type, size, spread, and your overall health. Surgery is often the cornerstone, but your doctor will tailor the plan to maximize cure while preserving function and quality of life.

Q11: “What would you recommend to a friend or family member in my situation?”  
Most doctors would recommend timely evaluation and treatment by an experienced head and neck surgeon or oncologist, with a multidisciplinary approach including pathology, radiology, and supportive care.

Q12: “Should I see a specialist?”  
Yes, parotid tumors require evaluation by specialists such as head and neck surgeons, otolaryngologists (ENT doctors), or oncologists experienced in salivary gland tumors.

**DOCTOR-PATIENT CONVERSATIONS**

Doctor: Hello, what brings you in today?

Patient: Doctor, I found a lump near my jaw and I'm worried it might be a tumor. Can you tell me what it could be?

Doctor: I understand your concern. A lump in the area near your jaw could be a swelling of the parotid gland, which is one of your salivary glands. This swelling might be due to many causes, including a parotid tumor, which can be benign or malignant.

Patient: How do you find out if it's cancer?

Doctor: We start with a physical examination and often use imaging tests like an ultrasound, CT scan, or MRI to see the size and characteristics of the lump. A biopsy, usually done with a fine needle, may be taken to examine the cells and tell if the tumor is benign or cancerous. Sometimes, the tissue is checked during surgery as well.

Patient: What treatments are available?

Doctor: Treatment depends on the tumor type. Most benign tumors can be cured by surgically removing part or all of the affected parotid gland. Surgery is carefully done to avoid damaging the facial nerve, which controls your facial muscles. If the tumor is cancerous, you might need additional treatments like radiation therapy. We will tailor the treatment to your specific diagnosis.

Patient: Will surgery leave a scar or cause facial weakness?

Doctor: Surgery will usually leave a scar near the jawline, but surgeons try to place it where it’s less visible. There is a risk to the facial nerve because it runs through the gland. Usually, the nerve is preserved, but occasionally it may be stretched or affected temporarily, leading to weakness that often improves over weeks to months. Rarely, if the tumor invades the nerve, part of it might need to be removed and repaired.

Patient: How will the treatment affect my daily life?

Doctor: After surgery, you will need some recovery time. You might have some swelling or numbness around the ear and face. Most people return to normal activities within a few weeks. If radiation is needed, it usually involves daily sessions over several weeks and can cause fatigue or skin changes. We support you through all these steps.

Patient: What is the outlook?

Doctor: The prognosis depends on whether the tumor is benign or malignant, its type, and stage. Most benign tumors have an excellent outcome with surgery alone. Early-stage cancers that are treated appropriately can also have good outcomes. We will monitor you closely with regular follow-up appointments to catch any recurrence early.

Patient: Should I see a specialist?

Doctor: Yes, a head and neck surgeon or an ENT specialist with experience in salivary gland tumors will manage your care. They work in a multidisciplinary team including radiologists, pathologists, and oncologists to give you the best care.

Patient: Are there any materials or websites I can read?

Doctor: Absolutely, I can provide printed brochures and recommend trustworthy websites such as the Mayo Clinic and Cleveland Clinic's patient resources on parotid tumors for more detailed information.

Patient: Thank you, doctor. That helps me understand what to expect.

Doctor: You're welcome. Feel free to ask me any more questions anytime. We’re here to support you through your treatment.

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**PETROUS APEX LESION**

**ALTERNATIVE NAMES:** An alternative name for a petrous apex lesion is a petrous apex cyst. These lesions are typically fluid-filled and can take the form of a cholesterol granuloma, cholesteatoma (epidermoid), or other types of cysts. Cholesterol granuloma is the most common type of lesion found within the petrous apex , while cholesteatomas can be either congenital or acquired. Other types of lesions include mucoceles and fibrous dysplasia.

**DEFINITION / DESCRIPTION**

A petrous apex lesion refers to an abnormal growth, cyst, tumor, or infection occurring in the petrous apex, which is a pyramidal-shaped, medial projection of the petrous portion of the temporal bone at the skull base.

This region is anatomically complex and situated near crucial structures such as cranial nerves, the cavernous sinus, the internal carotid artery, Dorello canal, and Meckel cave, making lesions here potentially serious due to their proximity to these important anatomic features.

The petrous apex itself can vary in its degree of pneumatization (air cell-filled spaces) or may contain marrow fat. Lesions can arise from the bone, pneumatized air cells, or adjacent vascular structures, and can be intrinsic or result from extension of infections or tumors from nearby areas like the nasopharynx or sinonasal regions.

***Types of petrous apex lesions include:***

* Developmental and benign cystic lesions: such as cholesterol granulomas (common, benign cysts filled with thick material), cholesteatomas, mucoceles, and epidermoid cysts.
* Benign tumors: including meningiomas, schwannomas, and paragangliomas (glomus tumors).
* Inflammatory/infectious lesions: such as petrous apicitis (infection of petrous apex air cells).
* Malignant tumors: like chordomas, chondrosarcomas, nasopharyngeal carcinoma, and metastatic lesions.
* Anatomic variants and pseudolesions: asymmetrical pneumatization or marrow signal variations that mimic pathology.

**CAUSES**

The exact cause of some petrous apex lesions is not clear. It is believed that cholesterol granulomas may develop as the result of air infiltrating the bone marrow and creating pockets that then fill with blood and trigger the formation of a cyst.

Cholesterol granulomas may also be linked to dysfunctions involving the Eustachian tube. The purpose of the Eustachian tube, which connects the tympanic cavity to the back of the nose, is to allow air to enter behind the eardrum while also allowing fluid and mucus to drain.

Cholesteatomas involving the petrous apex may be congenital, meaning the condition is present at birth, or it may be acquired during the patient’s lifetime. Congenital cholesteatomas are typically caused by skin cells left behind during embryonic development.

The cells form a skin cyst known as a cholesteatoma. Acquired cholesteatomas are normally caused by Eustachian tube dysfunction, or rarely from trauma to or surgery in the ear. Both congenital and acquired cholesteatomas can become quite large and impinge on nerves and other nearby inner ear structures. Some congenital cholesteatomas occur outside the petrous apex between the dura and the temporal bone.

**SIGNS / SYMPTOMS**

Small petrous apex lesions may not cause any issues or symptoms. Problems may arise when the lesions grow and impinge on other nerves such as the facial movement or sensory nerves, the hearing or balance nerves, or nerves that move the eyes. A lesion impinging on the various nerves may affect sensations or movement in the face or impact eye movement. Other symptoms may include a feeling that the ear is plugged up, foul smelling drainage coming from the ear, decreased hearing, balance issues, and rarely pain behind the ear

**DIAGNOSIS METHODS**

A doctor may arrive at a preliminary diagnosis of a petrous apex lesion based on the patient’s symptoms and the results of an initial evaluation. Computed tomography or magnetic resonance imaging scans are normally used to confirm the diagnosis and to determine the best treatment approach. Most petrous apex lesions are diagnosed incidentally on MRI’s obtained for other reasons such as headaches.

**TREATMENT OPTIONS**

The treatment of petrous apex lesions depends on the type of lesion, the severity of the patient’s symptoms, and the amount of damage that the growth has caused to the surrounding structures of the ear.

Cholesterol granulomas affecting the petrous apex are fluid-filled cysts, so the primary treatment involves draining the cyst. There are a number of different approaches that the surgeon may use to access the cyst; however, the most frequently used technique is the infracochlear approach. This technique involves accessing the granuloma from a position just below the cochlea of the inner ear. More recently, we have started observing some cholesterol granulomas since some of these have been found to not grow.

Petrous apex cholesteatomas, on the other hand, are a little more complicated since they must be completely removed. Although it is possible to access the cholesteatoma through the inner ear, it is sometimes also necessary to access it through the middle fossa located below the brain. In some cases, the cholesteatoma can be approached via a transcochlear or transotic approach when the hearing is not salvageable.

**PREVENTION TIPS**

In many cases, such as with congenital cholesteatomas, there is nothing that a person can do to prevent a petrous apex lesion. The key is to seek medical attention as soon as possible after symptoms develop. Individuals should consult their doctor if they experience:

* Recurrent or chronic ear infections
* Changes in their hearing
* Issues maintaining balance
* Pain in or around their ear
* Drainage from the ears
* Trauma to the ears
* Changes in sensation of the face
* Facial paralysis that lasts more than 6 months warrants further work up

**OUTLOOK / PROGNOSIS**

* Common lesion types and prognosis: The most common petrous apex lesion is the cholesterol granuloma, a benign cyst-like lesion that can be effectively managed with drainage. Surgical drainage procedures typically yield good outcomes with symptom relief in the majority of patients.
* Symptom improvement: Post-treatment, many patients experience significant improvement or resolution of symptoms such as headache, dizziness, hearing loss, diplopia (double vision), and paresthesia. For example, symptom relief was reported in about 77% of patients following surgery for cholesterol granulomas. Facial nerve and cranial nerve deficits can improve or resolve after treatment depending on lesion extent and duration of symptoms.
* Surgical risks and complications: Surgery is generally safe but not without risks. Complications such as hearing loss, facial nerve weakness, carotid injury, and CSF leaks have been reported, though these are relatively rare. More aggressive surgical resections are usually reserved for solid tumors or recurrences, while cystic lesions are often treated with drainage to minimize morbidity.
* Recurrence and follow-up: Recurrence rates vary; for cholesterol granulomas, rates around 10-17% have been reported, often due to drainage tract occlusion. Long-term radiographic follow-up is important as recurrences can happen years after treatment. Use of drainage tubes or stents may reduce recurrence risk.
* Incidental or asymptomatic lesions: Small, asymptomatic petrous apex lesions (such as cholesterol granulomas) are sometimes discovered incidentally. In such cases, a "wait-and-scan" approach with regular imaging follow-up is a valid strategy, reserving intervention for symptomatic or enlarging lesions.
* Malignant or infectious lesions: Prognosis worsens significantly if the lesion is malignant (e.g., chondrosarcoma) or infectious (e.g., petrous apicitis). Prompt diagnosis and aggressive treatment including surgery and antibiotics are critical to avoid serious complications such as cranial nerve damage, vascular injury, meningitis, or brain abscess

**WHEN TO SEE A DOCTOR / RED FLAG**

* Severe or persistent ear pain (otalgia) or ear discharge (otorrhea)
* Deep facial, retroorbital (behind the eye), or head pain
* Hearing loss or changes in hearing
* Balance problems, dizziness, or vertigo
* Facial weakness, spasms, or numbness indicating cranial nerve involvement
* Double vision or difficulty moving the eyes (due to cranial nerve palsies such as the sixth nerve)
* Headache that is severe or progressively worsening
* Fever, which may indicate infection such as petrous apicitis

**DIFFERENTIAL DIAGNOSIS**

## Congenital / Anatomic Variants (Pseudolesions)

* Asymmetrical pneumatization of petrous apex air cells
* Marrow signal variation (normal fatty marrow with characteristic MRI signals)
* Cerebrospinal fluid (CSF) intensity lesions/pockets
* Petrous apex cephalocele (meningeal herniation into the petrous apex)

## 2. Inflammatory / Infectious Lesions

* Petrous apicitis (infection of petrous apex, often from otitis media)
* Osteomyelitis of the temporal bone
* Petrous apex effusion (fluid accumulation, non-infectious or infectious)

## 3. Cystic Lesions

* Cholesterol granuloma (most common cystic lesion; T1 and T2 hyperintense on MRI)
* Mucocele (mucous-filled cyst causing smooth bone remodeling)
* Cholesteatoma (keratin debris-filled cyst with bone erosion; diffusion-weighted imaging positive)
* Epidermoid cyst

## 4. Benign Tumors

* Schwannoma (usually cranial nerve schwannomas such as from CN VII or VIII)
* Meningioma (may involve skull base and petrous apex)
* Chondroma
* Fibrous dysplasia (osseous dysplasia presenting as expansile bone lesion)

## 5. Malignant Tumors

* Chordoma (midline skull base tumor, can extend to petrous apex)
* Metastatic disease (from various primary tumors)
* Primary bone malignancies (rare)
* Malignant peripheral nerve sheath tumors (rare)

## 6. Vascular Lesions

* Petrous internal carotid artery (ICA) aneurysm or dolichoectasia (can mimic mass lesion)
* Carotid artery pseudoaneurysm

**EPIDEMIOLOGY**

* Petrous apicitis, an infectious inflammation of the petrous apex often secondary to middle ear or mastoid infection, was once more common but became rare after antibiotics introduction. It now occurs infrequently, estimated at approximately 2 cases per 100,000 with acute otitis media in some studies. Most contemporary literature consists of small case series or single reports due to its rarity.
* The petrous apex is pneumatized in about 9 to 30% of individuals, which predisposes to infection spreading into this region from the middle ear.
* Among petrous apex lesions overall, cholesterol granulomas are the most common cystic lesion, accounting for about 60% of petrous apex lesions in surgical series. These are rare with an incidence estimated around 0.6 cases per million population, but represent the majority of benign lesions encountered at this site.
* Other lesions such as meningiomas constitute about 25% of petrous apex pathologies, while petrous internal carotid artery aneurysms and petrous apicitis comprise smaller proportions (for example, apicitis around 2.6% in some series).
* Symptoms from petrous apex lesions vary and may remain silent for long periods; when symptomatic, headaches, visual symptoms, hearing loss, or cranial neuropathies are common presentations.
* Because of the diversity and rarity of petrous apex lesions, epidemiologic data mainly come from retrospective case series and institutional reviews rather than large population-based studies

**PREDEFINED Q & A SETS**

Q1: “What are petrous apex lesions?”  
Petrous apex lesions are cysts or tumors that form in the petrous apex, a part of the temporal bone near the middle of the skull base and ear. They can be cystic or solid and are often discovered incidentally or during evaluation of symptoms such as ear pain or hearing loss.

Q2: “What causes petrous apex lesions?”  
They can arise from a variety of causes, including:

* Benign cystic lesions: cholesterol granuloma (most common), mucocele, cholesteatoma, epidermoid cyst
* Benign tumors: meningioma, schwannoma, paraganglioma (glomus tumor)
* Malignant tumors or extending cancers: chordoma, chondrosarcoma, nasopharyngeal carcinoma, metastases
* Inflammatory or infectious: petrous apicitis (infection of petrous apex, often from middle ear infection)
* Congenital or anatomical variants: pneumatization variations, marrow fat, petrous apex cephalocele (meningeal herniation).

Q3: “What symptoms do petrous apex lesions cause?”  
Symptoms vary widely depending on the lesion size and involvement of nearby nerves, and may include:

* Headache (temporal or retro-orbital)
* Ear pain, ear discharge or pus
* Hearing loss or tinnitus
* Facial weakness or numbness
* Double vision due to cranial nerve involvement.

Some small lesions may be asymptomatic and found incidentally.

Q4: “How are petrous apex lesions diagnosed?”  
Diagnosis relies heavily on imaging studies:

* CT scans evaluate bone anatomy, erosion, and pneumatization.
* MRI characterizes lesion content (solid vs cystic), signal intensity, and contrast enhancement patterns. Cholesterol granulomas typically show high signal on T1 and T2 MRI sequences, whereas cholesteatomas show restricted diffusion on diffusion-weighted imaging.
* Clinical correlation and sometimes biopsy or surgical exploration may be needed for definitive diagnosis.

Q5: “What are treatment options?”  
Treatment depends on lesion type, symptoms, growth, and risk of complications:

* Observation: Many small or asymptomatic lesions, such as stable cholesterol granulomas or air cell fluid, can be monitored with periodic imaging.
* Surgical treatment: Indicated for symptomatic lesions causing nerve compression, infection, or growth. Surgical options include drainage (especially for cystic lesions) or complete removal, though the location can make surgery challenging.
* Antibiotics: Required for infectious lesions like petrous apicitis.
* Individualized treatment plans are created by specialists, often involving ENT surgeons and neurosurgeons.

Q6: “What is the prognosis?”  
Most benign lesions have a good prognosis when treated appropriately. Risks include recurrence after drainage (for cystic lesions) and potential nerve injury from surgery. Malignant lesions require more aggressive treatment and have a variable prognosis depending on type and stage

**DOCTOR-PATIENT CONVERSATIONS**

Doctor: Hello, what brings you in today?

Patient: Doctor, I recently had some imaging done for headaches and ear pain, and they mentioned a lesion in my petrous apex. I’m not sure what that means. Can you explain?

Doctor: Of course. The petrous apex is a part of the temporal bone near your inner ear deep in your skull. A lesion there means there is an abnormal finding like a cyst, growth, or sometimes an infection in that area.

Patient: What kinds of lesions are there, and are they dangerous?

Doctor: There are several types. Common benign lesions include cholesterol granulomas, which are cyst-like and usually slow growing. Other possibilities include cysts like mucoceles or cholesteatomas, benign tumors like schwannomas or meningiomas, infections such as petrous apicitis, and rarely malignant tumors.

Many petrous apex lesions grow slowly and may not cause serious problems, but some can affect nearby nerves and cause symptoms that need treatment.

Patient: What symptoms should I watch for?

Doctor: Symptoms can include headache, ear pain or discharge, hearing loss, dizziness, facial numbness or weakness, or double vision. Sometimes small lesions cause no symptoms and are found incidentally.

Patient: How do you diagnose what type of lesion it is?

Doctor: Imaging studies are key. CT scans show the bone structure and whether there is bone erosion or remodeling. MRI helps us see the tissue characteristics inside the lesion, helping distinguish cystic from solid lesions and identifying likely lesion types.

Because of the deep location, biopsy is usually not done before surgery, so we rely mostly on imaging and clinical findings.

Patient: What treatment options are available?

Doctor: Treatment depends on symptoms, the type of lesion, and growth. Many small or asymptomatic lesions can just be monitored with periodic imaging.

If the lesion causes symptoms, grows, or risks damaging nerves, surgery or drainage may be needed. Surgery can be complex because of the location near critical nerves and vessels, and requires careful planning with imaging. Infection like petrous apicitis requires antibiotics and sometimes surgical drainage.

Patient: What are the risks of treatment?

Doctor: Surgery risks include injury to nearby nerves, such as the facial nerve, hearing loss, dizziness, or bleeding. That’s why surgery is only done when clearly necessary and by an experienced skull base surgeon.

Patient: What is the outlook for someone with a petrous apex lesion?

Doctor: Many patients do very well, especially with benign or infectious causes treated early. Slow-growing lesions monitored carefully have a low risk of complications. The key is close follow-up to catch any changes early.

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**RAMSAY HUNT SYNDROME**

**ALTERNATIVE NAMES:** Ramsay Hunt syndrome is also known by several alternative names, including herpes zoster oticus, Hunt syndrome, geniculate ganglionitis, and herpetic geniculate ganglionitis. It is sometimes referred to as Ramsay Hunt syndrome type 2. Other names include Ramsay Hunt disease and geniculate herpes.

Herpes zoster oticus

**DEFINITION / DESCRIPTION**

Ramsay Hunt syndrome (herpes zoster oticus) occurs when a shingles outbreak affects the facial nerve near one of your ears. In addition to the painful shingles rash, Ramsay Hunt syndrome can cause facial paralysis and hearing loss in the affected ear.

Ramsay Hunt syndrome is caused by the same virus that causes chickenpox. After chickenpox clears up, the virus still lives in your nerves. Years later, it may reactivate. When it does, it can affect your facial nerves.

Prompt treatment of Ramsay Hunt syndrome can reduce the risk of complications, which can include permanent facial muscle weakness and deafness.

**CAUSES**

Ramsay Hunt syndrome occurs in people who've had chickenpox. Once you recover from chickenpox, the virus stays in your body — sometimes reactivating in later years to cause shingles, a painful rash with fluid-filled blisters.

Ramsay Hunt syndrome is a shingles outbreak that affects the facial nerve near one of your ears. It can also cause one-sided facial paralysis and hearing loss.

**RISK FACTORS**

Ramsay Hunt syndrome can occur in anyone who has had chickenpox. It's more common in older adults, typically affecting people older than 60. Ramsay Hunt syndrome is rare in children.

Ramsay Hunt syndrome isn't contagious. However, reactivation of the varicella-zoster virus can cause chickenpox in people who haven't previously had chickenpox or been vaccinated for it. The infection can be serious for people who have immune system problems.

Until the rash blisters scab over, avoid physical contact with:

* Anyone who's never had chickenpox or who's never had the chickenpox vaccine.
* Anyone who has a weak immune system.
* Newborns.
* Pregnant women.

**SIGNS / SYMPTOMS**

The two main signs and symptoms of Ramsay Hunt syndrome are:

* A painful red rash with fluid-filled blisters on, in and around one ear.
* Facial weakness or paralysis on the same side as the affected ear.

Usually, the rash and the facial paralysis occur at the same time. Sometimes one can happen before the other. Other times, the rash never occurs.

If you have Ramsay Hunt syndrome, you might also experience:

* Ear pain.
* Hearing loss.
* Ringing in your ears (tinnitus).
* Difficulty closing one eye.
* A sensation of spinning or moving (vertigo).
* A change in taste perception or loss of taste.
* Dry mouth and eyes.

**DIAGNOSIS METHODS**

Doctors often can identify Ramsay Hunt syndrome based on medical history, a physical exam, and the disorder's distinctive signs and symptoms. To confirm the diagnosis, your doctor might take a sample of fluid from one of the rash blisters in your ear for testing.

**TREATMENT OPTIONS**

Prompt treatment of Ramsay Hunt syndrome can ease pain and decrease the risk of long-term complications. Medications may include:

* Antiviral drugs. Medications such as acyclovir (Zovirax), famciclovir (Famvir) or valacyclovir (Valtrex) often help combat the chickenpox virus.
* Corticosteroids. A short regimen of high-dose prednisone appears to boost the effect of antiviral drugs in Ramsay Hunt syndrome.
* Anti-anxiety medications. Drugs such as diazepam (Valium) can help relieve vertigo.
* Pain relievers. The pain associated with Ramsay Hunt syndrome can be severe. Prescription pain medications may be needed.

***Self care***

The following can help reduce the discomfort of Ramsay Hunt syndrome:

* Keep areas affected by the rash clean.
* Apply cool, wet compresses to the rash to ease pain.
* Take an over-the-counter pain reliever or anti-inflammatory drug, such as ibuprofen (Advil, Motrin IB, others).

If facial weakness makes it difficult for you to close one of your eyes, take the following steps to protect your vision:

* Use moisturizing eyedrops throughout the day if your eye becomes dry.
* At night, apply ointment to the eye and tape your eyelid shut or wear an eye patch.

**PREVENTION TIPS**

Children are now routinely vaccinated against chickenpox, which greatly reduces the chances of becoming infected with the chickenpox virus. A shingles vaccine for people age 50 or older also is recommended.

**OUTLOOK / PROGNOSIS**

Ramsay Hunt syndrome recovery time depends on the severity of your facial paralysis. If you don’t have much damage to your nerve and you’re otherwise healthy, your symptoms may improve within a few weeks to a few months.

However, it may take up to a year to achieve a full recovery. If the damage to your facial nerve is more severe, your chances of a full recovery are less likely.

Antiviral medications can help, but your chances of a full recovery are better if you start treatment within three days of symptoms appearing.

Early diagnosis and prompt treatment with antiviral therapy seem to improve long-term outcomes. About 70% of people with Ramsay Hunt syndrome will return to complete or almost complete facial function. However, even with prompt treatment, some cases result in permanent facial paralysis or hearing loss.

***Can Ramsay Hunt syndrome recur?***

While very rare, Ramsay Hunt syndrome can come back just as a shingles infection can. That’s because the varicella-zoster virus remains present within your nerves forever

**POSSIBLE COMPLICATIONS**

Complications of Ramsay Hunt syndrome may include:

* Permanent hearing loss and facial weakness. For most people, the hearing loss and facial paralysis associated with Ramsay Hunt syndrome is temporary. However, it can become permanent.
* Eye damage. The facial weakness caused by Ramsay Hunt syndrome may make it difficult for you to close your eyelid. When this happens, the cornea, which protects your eye, can become damaged. This damage can cause eye pain and blurred vision.
* Postherpetic neuralgia. This painful condition occurs when a shingles infection damages nerve fibers. The messages sent by these nerve fibers become confused and exaggerated, causing pain that may last long after other signs and symptoms of Ramsay Hunt syndrome have faded.

**WHEN TO SEE A DOCTOR / RED FLAG**

Call your doctor if you experience facial paralysis or a shingles rash on your face. Treatment that starts within three days of the start of signs and symptoms may help prevent long-term complications.

**DIFFERENTIAL DIAGNOSIS**

***Diagnostic Considerations***

In the differential diagnosis of Ramsay Hunt syndrome, several other conditions with overlapping symptoms must be considered.Shingles, caused by the reactivation of the varicella zoster virus, results in a painful rash and can affect nerves throughout the body, not just in the facial region.

This condition shares the viral etiology with Ramsay Hunt syndrome but typically lacks the specific facial nerve involvement seen in Ramsay Hunt.

Bell palsy is another key differential diagnosis, characterized by sudden onset facial paralysis often preceded by symptoms such as slight fever, pain behind the ear, and neck stiffness.Unlike Ramsay Hunt syndrome, Bell palsy does not present with a rash and the facial paralysis is generally less severe at onset.

The paralysis in Bell palsy is believed to be due to non-infective inflammation and swelling of the facial nerve, with potential viral and immune factors involved.

Distinguishing features such as ear pain, dizziness, and hearing loss in Ramsay Hunt syndrome can help differentiate it from Bell palsy.

Acoustic neuroma and trigeminal neuralgia are also considered in the differential diagnosis.Acoustic neuroma, a benign tumor of the vestibulocochlear nerve, can cause symptoms like tinnitus, hearing loss, and muscle weakness due to nerve compression.

It may also lead to coordination issues, facial numbness, and speech difficulties as the tumor grows. Trigeminal neuralgia involves the trigeminal nerve and is marked by intense, stabbing facial pain.

The exact cause of trigeminal neuralgia remains unclear, but it is characterized by episodes of severe facial pain that can mimic some symptoms of Ramsay Hunt syndrome. Each of these conditions has distinct features that, upon careful evaluation, help in distinguishing them from Ramsay Hunt syndrome.

***Differential Diagnoses***

* Acoustic Neuroma
* Bell Palsy
* Persistent Idiopathic Facial Pain
* Postherpetic Neuralgia
* Temporomandibular Disorders
* Trigeminal Neuralgia

**EPIDEMIOLOGY**

***Frequency***

Ramsay Hunt syndrome is a rare neurologic disorder that arises as a complication of latent varicella-zoster virus (VZV) infection. It is estimated to strike approximately 5 out of every 100,000 people annually in the United States and affects both males and females equally.

The syndrome can occur in anyone who has previously had chickenpox, though it predominantly affects adults older than 60 years and is extremely rare in children younger than 6 years. Ramsay Hunt syndrome accounts for about 16% of all causes of unilateral facial palsies in children and 18% in adults. It is also thought to be responsible for as many as 20% of clinically diagnosed cases of Bell palsy.

The condition may present without a cutaneous rash, known as zoster sine herpete. Interestingly, VZV has been detected by polymerase chain reaction (PCR) in the tear fluid of patients diagnosed with Bell palsy,indicating a possible link. Due to the potential for Ramsay Hunt syndrome to go undiagnosed or be misdiagnosed, accurately determining its true prevalence in the general population is challenging.

The incidence of Ramsay Hunt syndrome among patients with HIV infection is not well-documented but may occur at a higher rate than in the general population due to the increased risk of VZV infection in individuals with HIV.

***Mortality/morbidity***

Ramsay Hunt syndrome is not usually associated with mortality. It is a self-limiting disease; the primary morbidity results from facial weakness. Unlike Bell palsy, this syndrome has a complete recovery rate of less than 50%.

***Procedures***

In the setting of a peripheral facial palsy, cerebrospinal fluid (CSF) rarely is analyzed. Although lumbar puncture is not recommended in the diagnosis of this disease, CSF findings can be helpful in confirming the diagnosis.

In one study, CSF findings were abnormal in 11% of 230 patients with idiopathic peripheral facial palsy, in 60% of 17 patients with Ramsay Hunt syndrome (abnormal finding was pleocytosis), in 25% of 8 patients with Lyme disease, and in all 8 patients with HIV infection.Thus, if the CSF is abnormal, a specific cause should be sought.

Temporary relief of otalgia in geniculate neuralgia may be achieved by applying a local anesthetic or cocaine to the trigger point, if in the external auditory canal.

***Staging***

Several scales have been developed to quantify the degree of facial muscle weakness. Of those, the House-Brackmann scale is most commonly used. However, the Yanagihara facial nerve grading scale is also clinically helpful.

The House-Brackmann facial neuropathy scale is as follows:

* 1- Normal
* 2 - Mild dysfunction (slight weakness only noticeable on close inspection)
* 3 - Moderate dysfunction (obvious weakness, but not disfiguring differences between both sides)
* 4 - Moderately severe dysfunction (obvious weakness and disfigurement)
* 5 - Only barely perceptive motor function
* 6 - Complete paralysis

The Yanagihara facial nerve grading scale is as follows:

* At rest 0 - 2 - 4
* Wrinkle forehead 0 - 2 - 4
* Close eyes normally 0 - 2 - 4
* Close eyes forcefully 0 - 2 - 4
* Close eye on involved side 0 - 2 - 4
* Wrinkle nose 0 - 2 - 4
* Blow out cheek 0 - 2 - 4
* Wristle 0 - 2 - 4
* Grin 0 - 2 - 4
* Depress lower lip 0 - 2 - 4

Each function is scored 0 (complete palsy), 2 (partial palsy), or 4 (nearly normal).

***TREATMENT DRUG AND THEIR SIDE EFFECTS***

1. Antiviral Medications

Common antivirals prescribed to combat the varicella-zoster virus causing RHS include:

* Acyclovir (Zovirax)
* Valacyclovir (Valtrex)
* Famciclovir (Famvir)

Purpose: These medications help reduce viral replication, ease pain, speed healing of the rash, and reduce risk of long-term complications such as facial paralysis. Early treatment (within 3 days of symptom onset) improves outcomes.

Common Side Effects:

* Nausea
* Headache
* Diarrhea
* Fatigue
* Rarely, kidney toxicity (especially if dehydrated), requiring dose adjustment or hydration.

2. Corticosteroids

* The most commonly used steroid is Prednisone (oral corticosteroid).
* Sometimes given in high doses for a short period (e.g., 5 to 10 days) with gradual tapering.

Purpose: Corticosteroids reduce nerve inflammation and swelling, helping to preserve nerve function and improve recovery of facial movement.

Common Side Effects:

* Increased blood sugar
* Mood changes (irritability, anxiety)
* Insomnia
* Increased appetite and weight gain
* Stomach upset or ulcers
* Elevated blood pressure
* Long-term use risks include immune suppression, osteoporosis, and adrenal suppression, but these are uncommon with short courses typically used in RHS.

3. Other Medications to Manage Symptoms

* Pain relievers: Prescription analgesics or over-the-counter NSAIDs (e.g., ibuprofen) to control pain.
* Vestibular suppressants or anti-anxiety drugs: Medications like diazepam (Valium) may be used to control vertigo or dizziness.
* Neuropathic pain agents: If patients develop nerve pain (postherpetic neuralgia), drugs such as gabapentin (Neurontin) may be prescribed.

**PREDEFINED Q & A SETS**

Q1: “What is Ramsay Hunt syndrome?”  
Ramsay Hunt syndrome is a neurological condition caused by reactivation of the varicella-zoster virus (the same virus that causes chickenpox and shingles) in the facial nerve near the ear. It results in a painful red rash with fluid-filled blisters on or around one ear, and sudden weakness or paralysis of the facial muscles on the same side.

Q2: “What causes Ramsay Hunt syndrome?”  
After a chickenpox infection, varicella-zoster virus remains dormant in nerve cells. Reactivation—often triggered by stress, aging, or weakened immunity—causes inflammation of the facial nerve, leading to the symptoms of RHS.

Q3: “Who is affected by Ramsay Hunt syndrome?”  
RHS most commonly occurs in adults over 50 years old and is rare in children. Immunosuppressed individuals are at higher risk. It can happen to anyone who had chickenpox earlier in life.

Q4: “What are the main symptoms?”

Answer:

* Sudden-onset facial paralysis or weakness on one side of the face
* Painful red rash with blisters on ear, ear canal, or mouth (though rash may sometimes be absent or delayed)
* Severe ear or facial pain
* Hearing loss, tinnitus (ringing in the ear), and vertigo (dizziness or spinning sensation)
* Difficulty closing one eye
* Changes in taste or dry mouth/eyes

Q5: “How is Ramsay Hunt syndrome diagnosed?”  
Diagnosis is usually clinical, based on symptoms and visible rash. If diagnosis is uncertain, testing of fluid from blisters can confirm varicella-zoster virus by PCR. Imaging may be used to rule out other causes of facial paralysis.

Q6: “What treatments are available?”

Answer:

* Antiviral medications: such as acyclovir, valacyclovir, or famciclovir to reduce virus replication
* Corticosteroids: typically prednisone to reduce inflammation and nerve swelling
* Pain relief: including NSAIDs or prescription pain medication for severe pain
* Supportive care: protecting the eye if facial paralysis causes incomplete closure (lubricating drops, ointment, taping eyelid)
* Other medications: anti-vertigo drugs (e.g., diazepam) or neuropathic pain agents if needed

Early treatment, ideally within 72 hours of symptom onset, improves recovery outcomes.

Q7: “What is the prognosis?”  
Most patients begin to recover facial movement within weeks to months. Recovery may take up to a year. Early initiation of treatment improves chances of full or near-full recovery. Delayed treatment and severe nerve damage may cause permanent facial weakness or hearing loss.

Q8: “What should I do to manage symptoms at home?”

Answer:

* Keep the rash clean and use cool compresses to relieve pain
* Take prescribed medications as directed
* Protect your affected eye to prevent dryness and injury
* Report any new or worsening symptoms such as severe headache, vision changes, or increased facial weakness promptly

**DOCTOR-PATIENT CONVERSATIONS**

Doctor: Hello, I want to talk to you about a condition called Ramsay Hunt syndrome. It happens when a virus called herpes zoster, which also causes shingles, infects a nerve near your ear. This can cause a painful rash and weakness or paralysis on one side of your face.

Patient/Parent: What kind of symptoms should I look out for?

Doctor: The main symptoms include a painful blistering rash around the ear, sometimes inside the ear canal, and facial weakness or paralysis on the same side. You might also have ear pain, difficulty closing one eye, or trouble with your facial expressions. Some people experience changes in hearing, dizziness, or a dry mouth or eye because the nerves involved control those functions.

Patient/Parent: How serious is this? Is it like Bell’s palsy?

Doctor: It’s a bit different and often more severe than Bell’s palsy because it's caused by a viral infection in the nerves and also comes with that rash. The facial weakness can be more pronounced, and recovery can take longer—sometimes weeks to months. There’s also a higher chance that some symptoms might not fully go away, compared to Bell’s palsy.

Patient/Parent: How is it treated?

Doctor: We usually start treatment right away with high-dose antiviral medications and corticosteroids to reduce viral activity and inflammation. Pain control is also important because it can be quite intense. Early treatment improves the chances of recovery. In some cases, physical therapy for the facial muscles may be recommended to help with healing and prevent complications like abnormal facial movements.

Patient/Parent: Will my facial movement come back to normal?

Doctor: Many patients improve significantly, but about 70% recover completely. Some people might have some lingering weakness or have abnormal muscle movements called synkinesis, where different parts of your face move together unintentionally. If that happens, physical therapy or sometimes injections can help.

Patient/Parent: Should we worry about complications?

Doctor: Yes, uncommonly Ramsay Hunt syndrome can cause permanent nerve damage, hearing loss, or persistent pain called postherpetic neuralgia. But with prompt medical care, these complications are less likely.

Patient/Parent: When should we seek help if symptoms start?

Doctor: If you or your child suddenly develops facial weakness, ear pain, or a rash near the ear, seek medical care right away. Early treatment is key to better outcomes.

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**RECURRENT RESPIRATORY PAPILLOMATOSIS (RRP)**

**ALTERNATIVE NAMES:** Recurrent respiratory papillomatosis (RRP) is also known by several alternative names, including laryngeal papilloma , juvenile papillomatosis , adult papillomatosis , and squamous cell papillomatosis. Additionally, it is sometimes referred to as recurrent respiratory papillomatosis (RRP).

**DEFINITION / DESCRIPTION**

Recurrent respiratory papillomatosis (RRP) is a condition caused by the human papillomavirus (HPV). Another name for RRP is laryngeal papilloma.

People with RRP develop wart-like tumors (papillomas) in their respiratory tract, the airways traveling from their mouth and nose to their lungs. Tumors usually grow on and around vocal cords located in your voice box (larynx).

Tumors can also occasionally spread downward to the other structures of your respiratory system, such as:

* Trachea.
* Bronchi.
* Lungs.

Though RRP is noncancerous (benign), the tumors can grow quickly and often require surgical removal. RRP treatment can be challenging because the tumors frequently come back (recur). Some people with RRP may require multiple surgeries per year.

### **Who does recurrent respiratory papillomatosis affect?**

Recurrent respiratory papillomatosis affects both children and adults.

* Juvenile-onset RRP is usually diagnosed before age 5. Compared with adults, RRP in children is often more aggressive and more likely to recur after treatment.
* Adult-onset RRP can occur at any age — usually after age 14 — and is less likely to recur.

### **How common is recurrent respiratory papillomatosis?**

Recurrent respiratory papillomatosis is a rare disease. Estimates of how often it occurs range from 1 in 100,000 people to 4 in 100,000 people.

HPV vaccination, which began in the U.S. in 2006, is effective against RRP. HPV vaccination is for teens starting at age 11 and up to age 26. Currently, more than 50% of teens in the U.S. are vaccinated. As vaccinated teens become adults, public health experts predict a decline in HPV-related cancers and conditions (including RRP).

***Types of HPV cause recurrent respiratory papillomatosis***

HPV is a group of more than 100 types of related viruses. HPVs cause warts on your skin and moist tissues (mucous membranes) found in your mouth, throat, vagina and anus. Because HPV spreads easily through vaginal, anal and oral sex, it’s the most common sexually transmitted disease in the U.S.

Two types of HPV (HPV 6 and HPV 11) cause RRP. These are the same types that cause most cases of genital warts. HPV also causes several types of cancer, including cervical cancer and cancer of the larynx (laryngeal). On rare occasions, it can also cause lung cancer.

HPV infection is very common. Usually, the infection resolves on its own without causing any negative health effects. Healthcare providers aren’t sure why HPV causes disease in some people but not others.

### **How is HPV transmitted in recurrent respiratory papillomatosis?**

Healthcare providers don’t fully understand HPV transmission in RRP. In children, HPV can be passed on during childbirth if the mother has active genital warts. But only a small percentage of babies exposed to genital warts during delivery develop RRP.

RRP transmission can also occur when your baby is born via cesarean section. This suggests that HPV can be transmitted before birth, while the baby is still in their mother’s uterus.

In adults, HPV may be transmitted through oral sex. Another theory is that adults with RRP were exposed to HPV during birth. The HPV then hid in healthy cells. Something activated this “hidden” HPV, which led to RRP during adulthood.

**CAUSES**

* HPV infection: RRP results from persistent infection of the respiratory epithelium by HPV, mostly types 6 and 11. In a smaller number of cases, other subtypes like HPV 16 and 18 may be involved.
* Transmission from mother to child: In children (juvenile onset RRP), the most common route is vertical transmission of HPV during passage through the birth canal in vaginal delivery from an infected mother. This is the primary cause of juvenile onset RRP.
* Adult onset RRP: The cause is less clear but may include reactivation of latent HPV acquired in childhood or new infection, potentially through oral sexual contact.

**RISK FACTORS**

* For juvenile onset RRP (JoRRP):
  + Being a first-born child
  + Vaginal delivery (rather than cesarean)
  + Prolonged labor
  + Maternal age under 20 years
  + Maternal presence of genital or anal warts (condylomas) during pregnancy, indicating active HPV infection
* For adult onset RRP (AoRRP):
  + Increased number of lifetime sexual partners
  + Frequency of oral sex, which may be a mode of HPV transmission
  + Possible immunodeficiency (e.g., HIV infection) can worsen disease severity
  + Smoking and gastroesophageal reflux disease (GERD) have been implicated in worsening adult RRP
* HPV subtype: HPV 11 is generally associated with a more aggressive disease course and airway obstruction.
* Age of onset: Earlier onset in childhood often correlates with a more aggressive disease.
* Co-existing infections: Other viral infections may influence disease progression.

Vaccination against HPV can prevent RRP by protecting against HPV 6 and 11 infection.

**SIGNS / SYMPTOMS**

As the tumors grow on the vocal cords and in your respiratory tract, they can cause speaking, breathing and swallowing problems. The most common symptoms of RRP include:

* Hoarse voice or loss of voice.
* A noisy or high-pitched sound when breathing (stridor).
* Rapid or difficult breathing.
* Chronic cough.
* Difficulty swallowing.

Your healthcare provider will evaluate you or your child carefully to distinguish RRP from other similar conditions, including asthma and allergies.

**DIAGNOSIS METHODS**

Your healthcare provider assesses your or your child’s symptoms and does a physical examination. They may place a mirror and bright light at the back of your throat to look at your vocal cords. If your symptoms and exam suggest RRP, your healthcare provider will refer you to an ear, nose and throat (ENT) specialist.

### **What tests are used to diagnose recurrent respiratory papillomatosis?**

Your ENT specialist may perform a laryngoscopy. This procedure uses an endoscope (a thin, flexible tube with a small camera and light) to view your throat and larynx. The ENT specialist:

1. Numbs the inside of your or your child’s nose and throat.
2. Thread the scope into your nose and down the back of your throat to your voice box.
3. Examine your respiratory tract for tumors that look like bumps or tiny bunches of grapes.

To confirm an RRP diagnosis, an ENT specialist may need to do a biopsy to collect a small tissue sample for HPV testing. This procedure is similar to a laryngoscopy but happens in an operating room under general anesthesia.

The ENT specialist uses an endoscope with special tools to collect a tissue sample. A lab analyzes the tissue to confirm the presence and type of HPV.

On rare occasions, a lung specialist may need to look inside of your windpipe in a similar fashion to collect a sample.

**TREATMENT OPTIONS**

RRP has no cure. The goal of treatment is to remove the growths and keep them from coming back. You or your child may need a combination of surgery and medication.

### **How is surgery used to treat recurrent respiratory papillomatosis?**

The primary treatment for recurrent respiratory papillomatosis is surgery. Your healthcare provider removes as many of the growths as possible without harming healthy tissue. Techniques for surgical removal include:

* Laser surgery: This treatment uses beams of light to shrink or destroy tumors.
* Microdebrider:This advanced tool uses gentle suction to pull the tumor toward a specialized, precise blade, which quickly and efficiently removes it.
* Cryoablation: During cryoablation, healthcare providers apply freezing temperatures to kill tumor cells.

Most RRP surgeries, especially in children, occur under general anesthesia in the operating room. You’re asleep during the surgery.

Office-based laser surgery is an option for some adults. In-office procedures are faster and eliminate the need for general anesthesia. You receive local anesthesia to keep you comfortable, but are awake during the procedure.

### **How often will my child or I need surgery for RRP?**

After surgery, RRP tumors often return. Some people need retreatment every few weeks. Others may go a year or more between treatments. Recurrence is more common in children.

### **How do medications treat recurrent respiratory papillomatosis?**

Medications can treat RRP in people who have:

* More than four RRP surgeries per year.
* Rapid tumor regrowth.
* Difficulty breathing due to tumors blocking their airway.
* Tumors that have spread to multiple locations.

These medications are called adjuvant therapies because they’re used after the primary treatment to help prevent the return of tumors. About 20% of people with RRP (1 in 5) require adjuvant therapy.

RRP adjuvant medications use different processes to reduce tumor growth:

* Cidofovir, an antiviral, prevents replication of the HPV virus.
* Immunotherapy agents, such as pembrolizumab, use your body’s immune system to destroy tumor cells.

### **Can the HPV vaccine help treat recurrent respiratory papillomatosis?**

Researchers have found that the HPV vaccine may also be an effective adjuvant treatment option. Early studies show that HPV vaccination slows tumor growth and decreases the frequency of surgeries.

**PREVENTION TIPS**

The HPV vaccine is effective at preventing RRP. The Centers for Disease Control and Prevention (CDC) recommends HPV vaccination for all children at age 11 or 12. People who weren’t vaccinated as teens can receive the vaccine up to age 26.

Condoms can also prevent RRP by lowering your risk of getting HPV through sex. But condoms don’t protect against HPV if the genital warts are on areas where condoms can’t cover.

If you have genital warts and are pregnant or thinking about becoming pregnant, talk to your healthcare provider. Overall, the risk of transmission from you to your baby is low. If you and your healthcare provider decide to treat genital warts during pregnancy, several safe treatment options are available.

**OUTLOOK / PROGNOSIS**

The outlook for most children and adults with RRP is good. Surgery and medication therapy can effectively manage tumors, so they don’t interfere with daily activities. Rarely, RRP will require a tracheostomy or become cancerous.

**POSSIBLE COMPLICATIONS**

In severe RRP, your airway can become blocked. To provide air to the lungs, you or your child may need a tracheostomy, which is a hole through your neck into your trachea. A tracheostomy affects your ability to speak. Your healthcare team will help you care for the tracheostomy and learn how to use your voice.

Cancer is another complication that can happen. Though rare, RRP can transform from a benign disease to (malignant) cancer. Transformation can occur many years after diagnosis and is more likely if the tumors have spread to lower parts of your respiratory tract. Healthcare providers aren’t sure why RRP becomes cancerous.

**DIFFERENTIAL DIAGNOSIS**

***Diagnostic Considerations***

Due to its nonspecific clinical manifestations, recurrent respiratory papillomatosis (RRP) is easily mistaken for asthma, acute laryngitis, upper respiratory infection, or bronchitis.

Other conditions to consider in the diagnosis of RRP include the following:

* Benign lung, laryngeal, or tracheal tumors
* Laryngeal infection
* Malignant laryngeal or tracheal tumors
* Squamous cell lung cancer

***Differential Diagnoses***

* Bilateral Vocal Fold Paralysis
* Foreign Body Aspiration
* Gastroesophageal Reflux Disease
* Malingering
* Relapsing Polychondritis
* Relapsing Polychondritis
* Secondary Lung Tumors
* Subglottic Stenosis in Adults
* Subglottic Stenosis in Children
* Tracheomalacia
* Vocal Cord Dysfunction

**EPIDEMIOLOGY**

***United States data***

Prior to the introduction of a quadrivalent vaccine for prevention of genital HPV in 2006, the estimated incidence of recurrent respiratory papillomatosis (RRP)in the United States was 4 per 100,000 in juvenile‐onset RRP (JORRP) and 2 per 100,000 in adult–onset RRP (AORRP).

RRP is declining in incidence; however it remains the most common benign laryngeal neoplasm in children.Roughly 15,000 surgical procedures for the condition are performed each year, at an estimated cost of $100 million.

***International data***

A study by Donne et al estimated the prevalence of patients with recurrent respiratory papillomatosis in the United Kingdom to be 1.42 per 100,000.

Australia introduced a nationwide immunization effort in 2007 which provided the HPV vaccine to at least half of all females aged 12-26.

The Australian National Immunisation Program began providing immunizations to all students aged 12-13 years (girls in 2007 and boys in 2013) through a school-based initiative.

As a result, the incidence of HPV in women 18 to 24 dropped from 22.7% in 2005 to 1.1% in 2015.The incidence of JORRP declined to 0.022 per 100,000 in 2016.

***Race-, sex-, and age-related demographics.***

The National Registry for Juvenile-Onset Recurrent Respiratory Papillomatosis,a registry including 603 children from around the United States, reports that 63% of the children were white, 28.4% were black, 0.8% were Asian, 0.8% were Native American, and 7% were unknown.

Ethnicity was reported for 367 (60.9%) of the patients; 57 (15.5%) were identified as Hispanic and 310 (84.5%) as white or black non-Hispanic.

JORRP affects males and females in equal numbers, whereas AORRP is more common in males.

The mean age at diagnosis of JORRP is 3.1 years.The adult form usually manifests in the third or fourth decade of life but may rarely manifest in patients older than 60 years

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***Staging***

A staging system for recurrent respiratory papillomatosis was published in 1998 and assigns a numerical score based on clinical and anatomic parameters.The clinical parameters include the urgency of intervention, voice quality, and the presence and severity of stridor and respiratory distress. The anatomic score is based on the number of sites of disease and whether the lesion is surface, raised, or bulky. Elements of the staging system have been found to help predict the frequency of needed surgical intervention

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***Procedures***

Laryngoscopy or bronchoscopy findings provide confirmation of the diagnosis of recurrent respiratory papillomatosis (RRP). Confirmation in patients presenting with hoarseness or voice change requires visualization of the vocal cords.

Because laryngeal involvement occurs in more than 95% of patients, direct laryngoscopy reveals the characteristic warty growths. Rarely, the trachea is involved without laryngeal lesions, and bronchoscopy may be required for diagnosis. These procedures reveal the characteristic cauliflowerlike warty growths

**PREDEFINED Q & A SETS**

## Q1: “What is Recurrent Respiratory Papillomatosis (RRP)?”

Recurrent respiratory papillomatosis is a rare disease caused by human papillomavirus (HPV), primarily types 6 and 11. It leads to the growth of benign wart-like tumors (papillomas) in the respiratory tract, most commonly on the vocal cords and larynx but can also affect the nose, throat, trachea, and lungs.

## Q2: “What causes RRP?”

RRP is caused by infection with HPV, often transmitted from mother to child during childbirth or acquired later in life. HPV types 6 and 11 are most commonly involved, with type 11 associated with more aggressive disease.

## Q3: “Who is affected by RRP?”

RRP can affect both children and adults. It often presents in children before age 5 but can appear at any age. The disease course varies; some have mild symptoms with infrequent growths, while others require frequent surgeries and have aggressive airway involvement.

## Q4: “What are the symptoms of RRP?”

Answer:

* Hoarseness or changes in voice quality (most common early symptom)
* Chronic cough
* Noisy breathing or stridor (high-pitched wheezing sound) due to airway obstruction
* Difficulty breathing or respiratory distress, especially in severe cases
* Difficulty swallowing
* Choking episodes
* Recurrent pneumonia if papillomas spread to the lungs
* Failure to thrive in infants with severe airway compromise

## Q5: “How is RRP diagnosed?”

Diagnosis is usually made by visualization of papillomas through laryngoscopy. A flexible scope passed through the nose and throat can detect papillomas on vocal cords and upper airway. In some cases, direct microscopic laryngoscopy and bronchoscopy under anesthesia are needed to examine deeper airway segments.

## Q6: “What is the treatment for RRP?”

Answer:

* Surgical removal: The main treatment is to remove papillomas to maintain a patent airway and improve voice. This often requires repeated surgeries because papillomas tend to recur. Techniques include microdebriders, lasers, and surgical instruments.
* Adjuvant medications: For aggressive or recurrent cases, antiviral agents like cidofovir or interferon alpha may be used, though responses vary.
* Prevention: HPV vaccination reduces the risk of HPV infection and thus the development or recurrence of RRP.

## Q7: “What is the prognosis?”

The course of RRP is highly variable. Some patients experience spontaneous remission, especially as children approach puberty, while others have recurrent growths requiring lifelong management. Rarely (less than 1%), papillomas may undergo malignant transformation into squamous cell carcinoma if untreated.

## Q8: “What should patients and parents watch for?”

Seek medical advice if hoarseness persists, breathing noises (stridor) appear, or there is difficulty breathing or swallowing. Early intervention can prevent airway obstruction and serious complications.

**DOCTOR-PATIENT CONVERSATIONS**

Doctor: Hello, what brings you in today?

Patient: Doctor, my child has been hoarse for several weeks, and I heard you might have found some growths in their throat. What exactly is this condition?

Doctor: Your child has a condition called recurrent respiratory papillomatosis, or RRP for short. It’s a rare disease caused by a virus called human papillomavirus, or HPV. This virus causes benign, wart-like growths called papillomas to develop in the respiratory tract, mainly on the vocal cords.

Patient: Is it cancer? Will it hurt my child?

Doctor: No, these growths are not cancerous, but they can cause problems by growing and blocking airflow or affecting the voice. The main symptoms are hoarseness and sometimes breathing difficulties if the growths become large or widespread.

Patient: How do you know my child has RRP?

Doctor: We use a procedure called laryngoscopy, which involves passing a small camera through the nose or mouth to look at the vocal cords and throat. The papillomas have a characteristic warty appearance. Sometimes we take a small biopsy to confirm the diagnosis and test for HPV.

Patient: How is it treated? Can it be cured?

Doctor: While there is no cure yet for RRP, the main treatment is surgical removal of the papillomas to keep the airway open and improve the voice. These growths often recur, so surgeries may be needed repeatedly over time. Newer treatments and vaccines help reduce the frequency of surgery and severity.

Patient: Are the surgeries safe? Will my child have problems afterward?

Doctor: Surgery is generally safe and usually done under general anesthesia. We try to remove as much abnormal tissue as possible without hurting normal vocal cord tissue. Recovery is usually quick. Some children need multiple procedures, but with expert care, most do well.

Patient: What about the HPV vaccine? Can it help?

Doctor: Yes, HPV vaccination can help prevent RRP if given before infection, and there’s some evidence it may reduce recurrence in those already affected. We recommend it as part of overall management if appropriate.

Patient: What can I do to help my child at home?

Doctor: You should watch for any worsening hoarseness, noisy breathing, or difficulty swallowing and bring your child in for check-ups as scheduled. Keeping all appointments and following treatment recommendations is important.

Patient: Will my child outgrow this?

Doctor: Some children experience fewer recurrences as they get older, especially around puberty, but the disease course is unpredictable. We monitor closely and adjust treatment as needed.

Patient: Thank you, doctor. This has been very helpful.

Doctor: You’re welcome. We’re here to support you and your child every step of the way. Please don’t hesitate to ask any questions.

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**RHABDOMYOSARCOMA**

**ALTERNATIVE NAMES:** Rhabdomyosarcoma is also known by several alternative names, including soft tissue cancer - rhabdomyosarcoma, soft tissue sarcoma, alveolar rhabdomyosarcoma, embryonal rhabdomyosarcoma, and sarcoma botryoides.

**DEFINITION / DESCRIPTION**

Rhabdomyosarcoma is a rare type of cancer known as soft tissue sarcoma. It develops in your skeletal muscles. Rhabdomyosarcoma mostly affects children and teenagers but can affect adults as well. Each year, between 400 and 500 people in the U.S. receive a diagnosis of rhabdomyosarcoma.

There are different types of rhabdomyosarcomas, some of which are aggressive and more difficult to treat. In some cases, treatment often puts the condition into remission, but rhabdomyosarcoma can come back (cancer recurrence).

***Types of this condition***

There are several types of rhabdomyosarcomas:

* Embryonal rhabdomyosarcoma: This is the most common type of rhabdomyosarcoma. It affects more children than adults. It typically develops in the head and neck, including the membrane that covers your child’s brain, eye socket and other parts of their head and neck. Subtypes of embryonal rhabdomyosarcoma may also develop in hollow organs like your child’s bladder and vagina (botryoid rhabdomyosarcoma) or in the area around your child’s testicles (spindle cell rhabdomyosarcoma).
* Alveolar rhabdomyosarcoma: This condition affects older children, teenagers and young adults between the ages of 20 and 35. It typically develops in your arms, legs or trunk (torso). Alveolar rhabdomyosarcoma is aggressive, meaning it spreads quickly soon after it develops.
* Pleomorphic rhabdomyosarcoma: This type typically affects adults ages 50 and older. It can develop anywhere in your body but mostly affects your legs. It also appears in your arms, chest, belly (abdomen), and parts of your head and neck.

**CAUSES**

Rhabdomyosarcoma happens when immature muscle cells mutate, becoming cancerous cells that multiply and create tumors. Certain genetic mutations, including a change that creates the fusion gene *PAX/FOX01*, may cause a type of rhabdomyosarcoma. People with certain inherited disorders have an increased risk of developing the condition:

* Li Fraumeni syndrome.
* Beckwith Wiedemann syndrome.
* Neurofibromatosis.
* Costello syndrome.
* Cardiofasciocutaneous syndrome.

**RISK FACTORS**

Factors that may increase the risk of rhabdomyosarcoma include:

* **Younger age.** Rhabdomyosarcoma most often happens to children younger than 10.
* **Inherited syndromes.** Rarely, rhabdomyosarcoma has been linked to genetic syndromes that are passed from parents to children. These include neurofibromatosis 1, Noonan syndrome, Li-Fraumeni syndrome, Beckwith-Wiedemann syndrome and Costello syndrome.

There is no way to prevent rhabdomyosarcoma.

**SIGNS / SYMPTOMS**

Symptoms vary depending on the tumor’s location. For example, a tumor in your child’s ear may cause earache or discharge from their ear. A tumor behind your eye may make it swell or bulge out of the socket. Other symptoms by tumor location include:

* Arm or leg muscle: Mass, lump or swelling that may be painful.
* Belly (abdomen): Belly pain, constipation or vomiting.
* Bladder and urinary tract: Blood in urine (hematuria) or difficulty peeing.
* Nasal cavity: Nosebleed (epistaxis) or sinus infection symptoms.
* Vagina: Mass or lump growing from your child’s vagina.
* Testicles: Fast-growing mass or lump around your child’s testicles.

Rhabdomyosarcoma symptoms may resemble less serious conditions. Many health issues can cause symptoms like nosebleeds, vomiting or lumps and bumps and may not be symptoms of rhabdomyosarcoma. That being said, you should contact a healthcare provider if you or your child have symptoms or changes that don’t go away or seem to be getting worse.

**DIAGNOSIS METHODS**

A healthcare provider will ask about your or your child’s symptoms. They’ll also ask about your family medical history to learn if anyone has an inherited disorder that increases the risk of rhabdomyosarcoma.

They’ll do a physical examination to look for symptoms like lumps or growths. Providers may do the following tests to diagnose rhabdomyosarcoma:

* Computed tomography (CT) scan or magnetic resonance imaging (MRI) scan.
* Positron emission tomography (PET) scan.
* Bone scan.
* Lumbar puncture.
* Bone marrow biopsy.
* Biopsy to obtain tissue samples.
* Immunohistochemistry.
* Cytology tests.

#### **Does rhabdomyosarcoma show up in bloodwork?**

No, it doesn’t. Your or your child’s oncologist may order blood tests after they diagnose rhabdomyosarcoma. For example, they may do a complete blood count (CBC) test to see if the condition has spread to your bone marrow. They may do blood tests to monitor rhabdomyosarcoma treatment.

### **Rhabdomyosarcoma risk groups**

Oncologists who care for children with rhabdomyosarcoma classify the condition by risk group. They establish risk group classification based on three factors:

* Tumor stage: Medical pathologists use the TNM staging system to establish the rhabdomyosarcoma stage. T stands for tumor size and location, N stands for signs of cancerous cells in lymph node location and M stands for metastasis, or if a tumor spreads from where it started.
* Clinical group: This classification reflects the outcome of tissue biopsies or initial surgeries. For example, if a biopsy or surgery completely removes a tumor, the case receives Group I classification. Group classifications are 1 to 4.
* Gene changes: The presence of the fusion gene *PAX/FOX01*.

Risk group classifications are low risk, intermediate risk and high risk. Your child’s oncology team uses risk group classification to plan treatment, assess the chance the tumor will come back after treatment and establish a prognosis, or what you can expect to happen after treatment.

The risk group classification process relies on very specific health information. You may not understand all the factors that go into establishing the risk group classification process.

Don’t hesitate to ask your child’s care team to explain all the information they use to place your child’s case in a specific risk group. They’ll be glad to explain the process and what it means for your child’s treatment and prognosis.

**TREATMENT OPTIONS**

Treatment varies depending on the condition type. Rhabdomyosarcoma is a rare disease; if you or your child has it, ask the cancer care team about participating in a clinical trial. In general, oncologists use the following treatments:

* Surgery.
* Radiation therapy.
* Chemotherapy.

**OUTLOOK / PROGNOSIS**

Sometimes, treatment can cure rhabdomyosarcoma. This is called remission, which means that you don’t have symptoms and tests don’t detect signs of cancer. In many cases, remission is permanent, but rhabdomyosarcoma can come back. In general, adults are less likely to be cured than children.

### **What is the life expectancy for someone with rhabdomyosarcoma?**

There’s no data on how long someone with rhabdomyosarcoma can expect to live. Researchers do track the percentage of people who were alive five years after receiving a diagnosis of rhabdomyosarcoma.

Survival rates vary widely depending on factors like the type of rhabdomyosarcoma, risk group classification and whether the condition comes back after treatment. Overall, 70% of children with this condition were alive five years after diagnosis. The five-year survival rate for adults is 20%.

Regardless of your situation, it’s important to remember that survival rates are estimates based on the experiences of other people who receive treatment for rhabdomyosarcoma. If you or your child have this condition, it’s understandable that you may want to know what to expect. If that’s your situation, your oncology team is your best resource for information.

### **How do I take care of my child or myself?**

Cancer disrupts your daily life, and rhabdomyosarcoma is no exception. If you or your child have this condition, you may feel overwhelmed and under stress. Here are some suggestions that may help:

* Consider palliative care: Cancer symptoms and cancer treatment can be tough to handle. Palliative care is a type of treatment that focuses on quality of life, from easing symptoms to finding mental health support.
* Talk to a child life specialist: Cancer upends children’s lives, taking them away from their friends and activities. Having cancer can be lonely for a child who’s going through something their friends may not understand. Child life specialists are specially trained healthcare providers who help children cope with medical experiences.
* Get some rest: Cancer treatment — and caring for a child with cancer — can be exhausting. If you’re receiving treatment, try to rest whenever you need to, not just when you have time. If you’re caring for a child with rhabdomyosarcoma, talk to your healthcare provider about programs and services that provide respite care.
* Consider cancer survivorship: Rhabdomyosarcoma can come back after treatment. If you or your child are worried cancer will come back, ask your provider about cancer survivorship support.

**POSSIBLE COMPLICATIONS**

Complications of rhabdomyosarcoma and its treatment include:

* **Cancer that spreads.** Rhabdomyosarcoma can spread from where it started to other parts of the body. When cancer spreads, it might require more-intense treatments. This can make recovery harder. Rhabdomyosarcoma most often spreads to the lungs, lymph nodes and bones.
* **Long-term side effects.** Rhabdomyosarcoma and its treatments can cause many side effects, both short and long term. Your healthcare team can help you manage the side effects that happen during treatment. And the team can give you a list of side effects to watch for in the years after treatment.

**WHEN TO SEE A DOCTOR / RED FLAG**

If you or your child are receiving treatment, contact your oncologist if treatment side effects are stronger than you expect. Depending on your situation, your oncologist may have specific guidance on symptoms that could mean rhabdomyosarcoma is spreading or coming back. Don’t hesitate to contact them with your concerns.

**DIFFERENTIAL DIAGNOSIS**

Other musculoskeletal neoplasms that should be excluded include:

* Ewing sarcoma
* Li-Fraumeni syndrome
* Lipomas
* Liposarcoma
* Lymphadenopathy
* Lymphoproliferative disorders
* Neurofibromatosis type 1
* Osteosarcoma
* Wilms tumor

**EPIDEMIOLOGY**

Rhabdomyosarcoma (RMS) is a rare condition making up 3% of all pediatric cancers. However, RMS is the most common childhood and adolescent soft tissue sarcoma, comprising 50% of soft tissue sarcomas in individuals younger than 20. In the US, there are approximately 350 newly diagnosed patients each year. All histological subtypes of RMS have also been shown to be significantly more prevalent in males. RMS has a much lower incidence in adults, accounting for approximately 1% of solid cancers.

Although the various subtypes of RMS can arise anywhere in the body, the embryonal histological type is the most common, and the head and neck is the most frequently involved area. RMS involving the extremities is more frequently observed with the alveolar subtype.

The pleomorphic and alveolar subtypes have the highest rates of metastases and, consequently, the poorest prognosis compared to other histologic types. The most common metastatic sites include the lungs, bone marrow, and lymph nodes

**PREDEFINED Q & A SETS**

Q1: “What is rhabdomyosarcoma?”

Rhabdomyosarcoma is a rare, malignant cancer that arises from immature muscle cells called rhabdomyoblasts. It is the most common soft tissue sarcoma in children but can occur at any age.

The tumor can develop in various parts of the body, including the head and neck, genitourinary tract, limbs, and trunk.

Q2: “What causes rhabdomyosarcoma?”

Rhabdomyosarcoma develops when muscle progenitor cells undergo genetic mutations that cause them to grow uncontrollably.

Some cases are associated with genetic syndromes like Li-Fraumeni syndrome or neurofibromatosis. Specific genetic alterations, such as fusion genes PAX3-FOXO1 or PAX7-FOXO1, are linked with the alveolar subtype.

Q3: “Who is affected by rhabdomyosarcoma?”

Rhabdomyosarcoma primarily affects children and adolescents, with peak incidence between ages 2 and 6 and again in adolescence. It is less common in adults. Both males and females can be affected.

Q4: “What are the symptoms of rhabdomyosarcoma?”

Symptoms vary depending on tumor location but often include:

* A rapidly growing, painless mass or swelling
* Symptoms related to local compression, such as difficulty breathing if in the airway, or vision problems if near the eye
* Pain if the tumor presses on nerves
* Bleeding or discharge if involving mucosal surfaces (e.g., nose, throat, vagina)
* Obstruction symptoms such as urinary difficulty if in the genitourinary tract

Q5: “How is rhabdomyosarcoma diagnosed?”

Answer:

* Physical Examination: Initial assessment of the mass or symptoms.
* Imaging Tests: MRI and CT scans to define tumor size, location, and spread. PET scans and bone scans may be used to check for metastases.
* Biopsy: Tissue sampling (needle or surgical) to confirm diagnosis via histopathology and immunohistochemistry, with markers like desmin, myogenin.
* Genetic Testing: May be done to identify characteristic gene fusions, especially in alveolar subtype.

Q6: “What are the treatment options for rhabdomyosarcoma?”

Treatment usually involves a combination of:

* Surgery: To remove the tumor if feasible, aiming for complete excision.
* Chemotherapy: Systemic treatment to kill cancer cells and shrink tumors before or after surgery.
* Radiation Therapy: Used when surgery is incomplete or not possible, or to treat residual disease.

The treatment plan depends on tumor location, size, subtype, and whether it has spread. Multidisciplinary care in specialized centers is essential.

Q7: “What is the prognosis for rhabdomyosarcoma?”

Prognosis depends on factors such as tumor subtype, location, size, patient age, and presence of metastasis. Localized embryonal RMS has a good prognosis with 5-year survival rates exceeding 70-80%, whereas metastatic or alveolar RMS has a poorer outlook. Early diagnosis and multimodal treatment improve outcomes.

Q8: “What are common side effects of treatment?”

Answer:

* Surgery may cause functional or cosmetic effects depending on location.
* Chemotherapy can cause nausea, fatigue, hair loss, immunosuppression, and increased infection risk.
* Radiation may cause skin irritation, fatigue, and potential late effects on growth or organ function.

If you want, I can provide tailored Q&A for patients, caregivers, or clinicians with more specific diagnostic criteria, subtype details, or emerging therapies.

***Genomic Data on Rhabdomyosarcoma Subtypes:***

1. Embryonal Rhabdomyosarcoma (ERMS)

* Most common subtype in young children (0-5 years).
* Genetically characterized by aneuploidy with multiple chromosomal gains and losses (e.g., gains in chromosomes 2, 7, 8, 11, 12, 13, 17-20; losses including 1p, 9, 10, 14-16).
* Frequent mutations in the RAS pathway genes: NRAS, KRAS, and HRAS (up to ~50% of cases). Some HRAS mutations link to Costello syndrome and other RASopathies.
* Other recurrent mutations include TP53 (up to 15%), associated with poorer prognosis, BCOR, DICER1 (notably in tumors of the female genital tract), FGFR4, NF1, and FBXW7.
* No specific gene fusions characterize ERMS.

1. Alveolar Rhabdomyosarcoma (ARMS)

* More common in adolescents (10-20 years) and extremity locations.
* Defined by characteristic and pathognomonic gene fusions involving the PAX3-FOXO1 or PAX7-FOXO1 genes, resulting from chromosomal translocations t(2;13)(q35;q14) and t(1;13)(p36;q14), respectively.
* These fusion genes encode aberrant transcription factors that drive tumor development.
* Fusion-positive ARMS is associated with poorer prognosis compared to fusion-negative tumors.

1. Spindle Cell/Sclerosing RMS

* Occurs across a wide age range including infants and adults.
* Infantile forms involve recurrent gene fusions such as VGLL2-CITED2, VGLL2-NCOA2, TEAD1-NCOA2, and SRF-NCOA2.
* Many adult and pediatric cases show MYOD1 p.L122R point mutations, often linked to aggressive behavior.
* Some MYOD1-mutated tumors also harbor PIK3CA mutations and deletions in CDKN2A.

1. Pleomorphic RMS

* Seen mainly in adults.
* Genetically complex with multiple chromosomal abnormalities but no specific recurrent fusion genes identified.

Additional Genomic Insights:

* Germline mutations in cancer predisposition genes such as TP53 (Li-Fraumeni syndrome) and NF1 are found more frequently in embryonal RMS.
* Recent large cohort studies identified recurring mutations above 5% incidence in genes like NRAS, BCOR, NF1, TP53, FGFR4, KRAS, HRAS, and CTNNB1.

**DOCTOR-PATIENT CONVERSATIONS**

Doctor: Hello, I understand you have some questions about the diagnosis of rhabdomyosarcoma. How are you feeling today?

Patient (or parent): Thank you, doctor. I’m quite worried. What exactly is rhabdomyosarcoma?

Doctor: Rhabdomyosarcoma, or RMS, is a rare cancer that starts in immature muscle cells. It most commonly affects children, but can occur at any age. It can appear in different parts of the body, such as the head and neck, arms, legs, or urinary tract.

Patient: How do you know if someone has this cancer?

Doctor: Diagnosis usually begins with a physical exam if a lump or swelling is noticed. We then use imaging tests like MRI or CT scans to understand the size and location of the tumor. A biopsy is essential to confirm the diagnosis by examining the tumor cells under a microscope. Sometimes genetic tests help classify the tumor subtype, which is important for planning treatment.

Patient: What kind of treatments are available?

Doctor: Treatment typically involves a combination of surgery to remove the tumor if possible, chemotherapy to target cancer cells throughout the body, and sometimes radiation therapy to the tumor site. The exact plan depends on the tumor’s size, location, and whether it has spread.

Patient: What are the side effects of these treatments? Are they dangerous?

Doctor: Treatments can have side effects. Surgery may cause some functional or cosmetic issues depending on where the tumor is. Chemotherapy can cause nausea, hair loss, fatigue, and a lower immune response, which means more risk of infections. Radiation might cause skin changes or affect growth if given to growing bones or organs. We monitor and manage these side effects carefully to keep you as comfortable as possible.

Patient: What is the outlook or prognosis?

Doctor: Prognosis depends on several factors such as the tumor subtype, size, and whether it has spread. Some types and early-stage tumors respond very well to treatment, with a good chance of cure. More advanced cases require intensive therapy but many children still do well with expert care.

Patient: Will we need long-term follow-up?

Doctor: Yes. After treatment, regular follow-up visits with physical exams and imaging are important to monitor for any recurrence or long-term side effects. We also assess growth, organ function, and provide rehabilitation if needed.

Patient: What can we do to support recovery?

Doctor: Besides medical treatment, support such as physical therapy, counseling, and nutritional help can be very beneficial. It’s also important to attend all follow-up appointments and to communicate any new symptoms promptly. We are here to support both you and your family throughout this journey.

Patient: Thank you, doctor. It helps to understand what to expect.

Doctor: You're welcome. Please feel free to ask any questions at any time. We’ll create a care plan tailored for you and provide the best treatment and support available.

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**RUPTURED EARDRUM (PERFORATED TYMPANIC MEMBRANE)**

**ALTERNATIVE NAMES:** Alternative names for a ruptured eardrum (perforated tympanic membrane) include tympanic membrane perforation, perforated eardrum, and eardrum - ruptured or perforated.

**DEFINITION / DESCRIPTION**

A ruptured eardrum (also known as a perforated eardrum) is a hole or tear in the thin tissue that separates your outer ear from your middle ear. It can happen because of a bad ear infection or an injury to your ear.

A ruptured eardrum can affect your hearing or sense of balance. But in many cases, it heals on its own. Sometimes, you may need surgery to fix it. The best way to prevent it is to protect your ears from injury and infection.

If you think you’ve ruptured your eardrum, don’t panic — but don’t ignore it, either. Most tears heal with time, but some may need treatment.

**CAUSES**

The most common cause of a perforated eardrum is a middle ear infection. When fluid builds up behind your eardrum, it creates pressure that can make it tear or pop.

Other common causes include:

* Ear injuries or trauma: A hard hit to your ear or head, like from a fall or a slap, can rupture your eardrum.
* Loud, explosive sounds: Your eardrums may rupture if you’re close by when there’s an explosion or gunfire.
* Pressure changes (barotrauma): If air gets trapped in your middle ear during flying or diving, a sudden change in pressure can make your eardrum burst.
* Sticking objects in your ears: Using cotton swabs or other small objects to clean or scratch your ear can puncture your eardrum. Even accidents, like bumping into a twig or getting hit by a thrown object, can cause damage.

***How to lower your risk***

Protecting your ears is the best way to prevent a ruptured eardrum. Here are a few simple tips:

* Clean with care: Skip cotton swabs. Use a clean cloth to gently wipe the outside of your ear.
* Prevent airplane ear: Flying can cause pressure changes that affect your ears. Chew gum, yawn, or wear special earplugs during takeoff and landing.
* Protect your ears from loud blasts: Gunfire or explosions can rupture your eardrum. Use earplugs or hearing protection if you’ll be near loud or explosive noise.
* Treat ear infections early: Signs of a middle ear infection include ear pain, stuffy nose, fever or trouble hearing. See a provider if symptoms last more than a few days.

**SIGNS / SYMPTOMS**

You might not know your eardrum is ruptured right away, especially if nothing hits your ear. Many people notice changes in hearing or drainage from their ear as the first signs.

Common symptoms include:

* Ear pain: This pain can come on suddenly and may go away quickly.
* Fluid draining from your ear: It may look like pus or have blood in it.
* Sudden hearing loss: Sounds may seem muffled or harder to hear.
* Tinnitus: This is a ringing, buzzing or humming sound in your ear that doesn’t come from outside.

If you notice any of these symptoms, it’s a good idea to get checked out. A ruptured eardrum usually heals on its own, but it’s best to let a provider confirm what’s going on.

**DIAGNOSIS METHODS**

To check for a ruptured eardrum, your provider will look inside your ear with a tool called an otoscope. It shines a light into your ear so they can see your eardrum clearly.

You might also have hearing tests. These help measure how well your eardrum is working and how well you hear.

Common tests include:

* Audiometry (audiogram): Checks how well you hear soft sounds and different tones
* Tympanometry: Shows how well your eardrum moves

These tests help your provider figure out if you have an eardrum rupture or another ear issue

**TREATMENT OPTIONS**

In many cases, a ruptured eardrum heals on its own without treatment. But if it doesn’t, you may need to see an ear, nose and throat specialist (ENT) for help.

Treatment options may include:

* Myringoplasty: Your ENT places a small paper patch over the hole. The patch helps your eardrum grow back and close the tear.
* Tympanoplasty: This surgery uses tissue from another part of your body (like skin or cartilage) to repair the eardrum.

***Recovery time***

Most ruptured eardrums heal within a few weeks, but some may take a few months. Contact your healthcare provider if you still have pain, drainage or hearing problems. You might need more treatment.

**OUTLOOK / PROGNOSIS**

Most ruptured eardrums heal on their own, but how you care for your ear can make a big difference. Here’s how to protect your ear while it heals:

* Avoid blowing your nose: If possible, don’t blow your nose while your eardrum is healing. If you have allergies, ask your healthcare provider about ways to prevent a stuffy nose.
* Don’t clean your ears: Even gentle cleaning may keep your eardrum from healing.
* Keep your ear dry: Use waterproof earplugs or cotton balls coated with petroleum jelly when you shower or take a bath. If you swim for exercise, please find another way to keep moving. Moisture in your ear from swimming may affect your eardrum.

***What side should I sleep on with a ruptured eardrum?***

If you have a ruptured eardrum, try sleeping on the side that doesn’t hurt or doesn’t have fluid coming out. This keeps pressure off the damaged ear.

**POSSIBLE COMPLICATIONS**

A ruptured eardrum is sometimes a serious issue, especially if it doesn’t heal on its own. Possible long-term complications include:

* A hole that doesn’t close
* Dizziness or balance problems
* Ongoing hearing loss
* Repeat ear infections
* Skin growth that can damage your middle ear (cholesteatoma)

Your eardrum also acts as a barrier. When it’s torn, germs, water and debris can enter more easily and raise your risk of infection

**WHEN TO SEE A DOCTOR / RED FLAG**

Call your provider if your symptoms don’t improve within a few weeks, or if they get worse.

Go to the emergency room if:

* Something is stuck in your ear
* You have sudden, severe ear pain or hearing loss
* You think a sharp object damaged your ear

**DIFFERENTIAL DIAGNOSIS**

Tympanic membrane rupture is generally easy to identify based on history and physical exam alone. However, there is other pathology that needs to be considered when assessing for TM perforation.

Differential diagnosis includes AOM, otitis externa, traumatic otorrhea (CSF otorrhea), brain or inner ear neoplasm, posterior stroke, Ramsay Hunt syndrome, ear FB, auricular hematoma, bullous myringitis, labyrinthitis, cholesteatoma, and perilymphatic fistula, to name a few. A thorough history and physical exam can help narrow down the diagnosis.

**EPIDEMIOLOGY**

While TM perforation incidence is unknown overall, given that many heal spontaneously, it is not uncommon to see a ruptured tympanic membrane in clinical practice.

One study of nearly 1,000 patients in the United States showed that men more commonly had traumatic rupture compared to women in a ratio of 1.49:1. A study out of Nigeria looking at 529 patients found similar statistics to the United States with a male to female ratio of 2:1.

Another study with 80 participants showed that the average age of patients who experience TM perforation was 26.7 +/- 14.6 years, with children making up 25% of the sample size

**PREDEFINED Q & A SETS**

Q1: “What is a ruptured eardrum?”

A ruptured eardrum is a tear or hole in the thin membrane (tympanic membrane) that separates the ear canal from the middle ear. This membrane helps transmit sound and protects the middle ear from bacteria and water.

Q2: “What causes a ruptured eardrum?”

Answer:

* Middle ear infections (most common cause) that increase pressure behind the eardrum.
* Trauma to the ear such as a direct blow, foreign objects, loud noises, or sudden pressure changes (barotrauma during flying or diving).
* Severe head injuries.
* Sudden changes in air pressure or loud blasts.

Q3: “What are the symptoms of a ruptured eardrum?”

Answer:

* Sudden ear pain that may quickly subside.
* Drainage from the ear that may be clear, pus-filled, or bloody.
* Hearing loss, usually partial and conductive (sound is softer).
* Ringing in the ear (tinnitus).
* Vertigo or dizziness in some cases.

Q4: “How is a ruptured eardrum diagnosed?”

Answer:

* Physical exam using an otoscope (a lighted instrument to look inside the ear).
* Hearing tests to assess the degree of hearing loss.
* Sometimes imaging if complications are suspected.

Q5: “How is a ruptured eardrum treated?”

Answer:

* Most ruptures heal on their own within a few weeks to 2 months.
* Pain relievers such as acetaminophen or ibuprofen can ease discomfort.
* Keep the ear dry—avoid getting water in the ear when bathing or swimming. Use cotton wool with petroleum jelly when washing hair.
* Avoid putting anything inside the ear (no cotton swabs or eardrops unless prescribed).
* Avoid blowing your nose hard to prevent pressure on the healing eardrum.
* Antibiotics may be prescribed if infection is present or to prevent an infection.
* If the perforation does not heal on its own, surgical options such as myringoplasty (patching the eardrum) or tympanoplasty (using a graft to repair) may be needed.

Q6: “What can I do to protect my ear while it heals?”

Answer:

* Sleep on the side opposite to the affected ear to reduce pressure.
* Avoid swimming or exposing the ear to moisture.
* Use protection like earplugs during showers or hair washing (covered with petroleum jelly).
* Follow your doctor's recommendations regarding medication and follow-up appointments.

Q7: “When should I see a doctor?”

Answer:

* If symptoms such as pain, drainage, or hearing loss persist beyond a few weeks.
* If symptoms worsen or new symptoms develop (increased pain, fever, dizziness, facial weakness).
* If the ruptured eardrum does not seem to be healing.

Q8: “What is the outlook?”

Answer:

* Most ruptured eardrums heal completely without treatment and hearing returns to normal.
* If healing is delayed or complications occur, medical or surgical treatments can effectively repair the eardrum and restore hearing.

**DOCTOR-PATIENT CONVERSATIONS**

Doctor: Hello, what brings you in today?

Patient: Doctor, I suddenly felt sharp pain in my ear, and now I think my hearing is worse. I also noticed some fluid coming from my ear. Could it be a ruptured eardrum?

Doctor: It sounds like you might have a perforated or ruptured eardrum, which means there is a tear or hole in the thin membrane inside your ear that separates your ear canal from the middle ear. The symptoms you describe, like sudden pain, hearing loss, and discharge, are common with this condition.

Patient: How does this happen? Can the eardrum heal on its own?

Doctor: A ruptured eardrum can happen from an ear infection, sudden changes in pressure like flying or diving, loud noises, or direct injury to the ear. The good news is that most ruptured eardrums heal by themselves within a few weeks to a couple of months without surgery.

Patient: How do you diagnose it?

Doctor: I will look into your ear with a special lighted instrument called an otoscope to see if I can spot the tear. Sometimes, if there’s fluid or discharge, it can make the view harder, but often we can see the perforation directly. We might also do a hearing test to assess how much the hearing is affected.

Patient: Is there anything I should do to help it heal and avoid infection?

Doctor: Yes, it’s very important to keep your ear dry while it heals. Avoid getting water in your ear when bathing or swimming. You can use a waterproof earplug or place a cotton ball coated with petroleum jelly gently in your ear during showers. Also, don’t insert anything into your ear canal, like cotton swabs. If you experience pain, over-the-counter pain relievers can help.

Patient: What if it doesn’t heal or if I get an infection?

Doctor: If the tear doesn’t heal on its own or if you develop persistent infections, we can place a small patch over the hole to help it close. This might need to be done a few times for full healing. In rare cases, surgery called tympanoplasty is needed to repair the eardrum.

Patient: How long will it take to recover my hearing?

Doctor: Hearing often improves as the eardrum heals, but it may take a few weeks. If the hearing loss is significant or doesn’t improve, we can perform further tests or treatments.

Patient: When should I come back or seek emergency care?

Doctor: If you have severe symptoms like sudden total hearing loss, continuous bleeding, severe pain that doesn’t improve, dizziness that makes you vomit, or facial weakness, seek emergency care immediately. Otherwise, we’ll schedule follow-up visits to monitor your healing.

Patient: Thank you, doctor. That makes me feel better about what to expect.

Doctor: You’re welcome. Feel free to call or visit if you have any questions as you recover.

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**RETROPHARYNGEAL ABSCESS**

*ALTERNATIVE NAMES:* Retropharyngeal abscess: A collection of pus in the back of the throat, often resulting from a bacterial infection affecting the throat's lymph nodes.

* Deep neck infections: A term used to describe infections involving the retropharyngeal space or the parapharyngeal (lateral pharyngeal) space.
* Parapharyngeal abscesses: A type of deep neck infection that involves the parapharyngeal space.
* Lateral pharyngeal abscess: Another term for a parapharyngeal abscess.
* Pharyngomaxillary abscess: A term used to describe a retropharyngeal abscess.
* Pterygomaxillary abscess: A term used to describe a retropharyngeal abscess.
* Pharyngeal space abscess: A general term for an abscess in the pharyngeal space.
* Posterior visceral space abscess: A term used to describe a retropharyngeal abscess.
* Retroesophageal space abscess: A term used to describe a retropharyngeal abscess.
* Retrovisceral space abscess: A term used to describe a retropharyngeal abscess.

**DEFINITION / DESCRIPTION**

A retropharyngeal (pronounced “reh-tro-fah-RIN-jee-ul”) abscess is a collection of pus deep in the back of your throat. It results from a bacterial infection affecting your throat’s lymph nodes.

As part of your immune system, your lymph nodes help fight infection. They can become swollen and irritated when you have an infection.

A retropharyngeal abscess is a serious condition that requires immediate medical attention. It’s most common in young children, but anyone can develop this abscess.

***Who do retropharyngeal abscesses affect?***

Retropharyngeal abscesses most commonly affect children between ages 2 and 4. Lymph nodes are larger at this age, making them more susceptible to infection.

Lymph nodes in your throat shrink as you age, decreasing your risk of an abscess. Retropharyngeal abscesses are slightly more common in males.

***How common is a retropharyngeal abscess?***

Retropharyngeal abscess is uncommon, but cases have increased in children and adults over the past 20 years. Some studies suggest that the rise is related to increased cases of tonsillitis.

Tonsillitis is an infection in your tonsils, the two lumps of tissue in the back of your throat. Another potential reason is the rise in infections involving antibiotic-resistant bacteria called MRSA.

**CAUSES**

A retropharyngeal abscess forms when your lymph nodes get infected. Your lymph nodes swell and eventually break down, forming pus that collects as an abscess in the back of your throat. Causes are different for children and adults.

* Children: Abscesses usually form following an upper respiratory infection (for example, a sinus infection or ear infection). The original infection spreads from the primary site to the lymph nodes in the back of your child's throat, forming an abscess.
* Adults: Abscesses usually form after an injury to the back of your throat. The injury may involve an object puncturing the back of your throat (like a chicken or fish bone). Or the injury may be a complication related to a procedure, like dental work, upper endoscopy or intubation.

***Is it viral or bacterial?***

Usually, the infection involves multiple types of bacteria. Bacteria that cause retropharyngeal abscesses include:

* Streptococcus pyogenes.
* Staphylococcus aureus (including MRSA).
* Bacteroides.
* Fusobacteria.
* Peptostreptococcus.
* Porphyromonas.
* Prevotella.
* Veillonella.
* Haemophilus influenzae.

**SIGNS / SYMPTOMS**

Having trouble swallowing or experiencing pain when you swallow are the most common symptoms of a retropharyngeal abscess. Other symptoms to look for include:

* Severe sore throat.
* Swollen lymph nodes.
* Difficulty breathing.
* Difficulty speaking.
* Noisy breathing.
* Severe headache.
* Stiff neck.
* Coughing.
* Fever.

Children with a retropharyngeal abscess may seem anxious or irritable. They may have so much trouble swallowing that they drool. A child who positions their head oddly (as if to clear their airway) may struggle to get enough air.

See your healthcare provider immediately if you notice any signs of a retropharyngeal abscess.

**DIAGNOSIS METHODS**

Your healthcare provider will assess your symptoms and perform a physical exam. An imaging test, like an X-ray or a CT scan, can show the abscess.

Your provider may also perform blood tests, including a complete blood count (CBC) and a bacteria culture test. High levels of white blood cells in your CBC indicate an infection. The culture test can show what types of bacteria are causing your infection.

You’ll likely work with a specialist in head and neck conditions called an ear, nose and throat (ENT) specialist during diagnosis and treatment.

**TREATMENT OPTIONS**

Retropharyngeal abscess is a serious infection that requires hospitalization. Treatment involves clearing your infection and, in some cases, draining the abscess. Treatments include:

* Antibiotics: You’ll receive broad-spectrum antibiotics intravenously (through a vein) to treat your infection. Broad-spectrum antibiotics kill multiple types of bacteria. You may receive antibiotics designed to treat a specific strain of bacteria if the culture test reveals particular bacteria that are causing your infection.
* Surgery: Your provider will decide whether to drain your abscess based on factors like whether it’s interfering with your breathing. Your provider will insert a breathing tube so you can breathe during the procedure. Then, they’ll cut the abscess open so the pus can escape.

Depending on the extent to which the abscess is blocking your airway, you may need to receive oxygen while in the hospital to help you breathe. You may need to receive fluids through an IV to prevent dehydration if you’re having trouble swallowing.

Once you’re discharged from the hospital, you’ll need to continue taking antibiotics by mouth to ensure the infection’s gone and doesn’t return.

***What medications are used?***

Antibiotics used to treat retropharyngeal abscesses include:

* Ampicillin sulbactam.
* Clindamycin.
* Vancomycin.
* Linezolid.
* Amoxicillin-clavulanate.

**PREVENTION TIPS**

Don’t delay seeking treatment if your child is sick, especially if they have an upper respiratory infection.

You can prevent complications from a retropharyngeal abscess by taking symptoms seriously. If you’re noticing signs of a retropharyngeal abscess — in either yourself or your child — see your healthcare provider immediately.

**OUTLOOK / PROGNOSIS**

The outlook is excellent with treatment. Treating the infection and draining the pus can cure the condition. Still, you should be aware of any symptoms that the abscess has returned. The abscess comes back in about 1% to 5% of cases. See your healthcare provider immediately if you notice signs of an abscess.

### **Is a retropharyngeal abscess life-threatening?**

It can be. A retropharyngeal abscess is potentially fatal without treatment. It’s essential to receive treatment as soon as you notice signs of this condition.

### **Is a retropharyngeal abscess an emergency?**

Yes. A retropharyngeal abscess can block your throat to the point that it cuts off your oxygen supply. See your provider well in advance of your condition worsening to this degree.

**POSSIBLE COMPLICATIONS**

A retropharyngeal abscess is serious largely because of its location. Without treatment, a retropharyngeal abscess can eventually block your airway, making breathing difficult. The infection can spread to nearby organs and your blood, causing life-threatening complications. Complications include:

* Blocked airway.
* Aspiration pneumonia.
* Swelling and inflammation in your chest.
* Blood clots in the major veins in your neck (jugular veins).
* Infection that spreads to nearby tissues and organs, like your lungs.
* Infection that spreads to your blood (septic shock).

**WHEN TO SEE A DOCTOR / RED FLAG**

* High fever and severe sore throat that does not improve or worsens
* Difficulty swallowing (dysphagia) or pain when swallowing (odynophagia)
* Drooling or refusal to eat/drink due to pain or inability to swallow
* Neck stiffness or difficulty turning the head (torticollis)
* Swelling in the neck or throat area, including a bulging at the back of the throat
* Hoarseness, muffled or "hot potato" voice
* Difficulty breathing, noisy breathing (stridor), or respiratory distress
* Trismus (difficulty opening the mouth)
* Agitation, lethargy, or signs of systemic illness
* Positioning of the head or neck to ease breathing, such as leaning forward or tilting the head

**DIFFERENTIAL DIAGNOSIS**

***Diagnostic Considerations***

Differentials include the following:

* Airway obstruction
* Calcific retropharyngeal tendinitis
* Apnea
* Sepsis
* Pneumonia
* Mediastinitis
* Epidural abscess
* Epiglottitis
* Croup
* Bacterial tracheitis
* Peritonsillar abscess
* Uvulitis
* Foreign body ingestion
* Angioedema
* Lymphoma
* Diphtheria

In a study in Germany, 20 of 234 patients with deep space infections of the neck had airway compromise requiring tracheostomy.In a study in Salt Lake City, 7 of 130 pediatric patients with RPA presented with airway obstruction requiring Pediatric Intensive Care Unit admission and/or intubation.

A case report of a 4-year-old boy with compression of his internal carotid artery and internal jugular vein by a retropharyngeal abscess was documented.

***Differential Diagnoses***

* Acute Torticollis
* Angioedema
* Bacterial Pneumonia
* Bacterial Sepsis
* Caustic Ingestions
* Dental, Infections
* Diphtheria
* Emergent Management of Acute Otitis Media
* Emergent Management of Croup (Laryngotracheobronchitis)
* Emergent Management of Pediatric Epiglottitis
* Emergent Management of Pediatric Patients with Fever
* Epidural Infections (Spinal Epidural Abscess) and Subdural Infections (Subdural Empyema)
* Epiglottitis
* Esophagitis
* Gastrointestinal Foreign Bodies
* Kawasaki Disease
* Lymphomas of the Head and Neck
* Mediastinitis in Emergency Medicine
* Meningitis
* Infectious Mononucleosis (IM) in Emergency Medicine
* Neonatal Sepsis
* Pediatric Foreign Body Ingestion
* Pediatric Pharyngitis
* Pediatric Pneumonia
* Pediatric Sepsis
* Pediatrics, Meningitis and Encephalitis
* Peritonsillar Abscess in Emergency Medicine
* Pharyngitis
* Sinusitis (Rhinosinusitis) Imaging
* Trachea Foreign Bodies

## **Procedures**

See the list below:

* Nasopharyngolaryngoscopy
  + A review of the literature did not reveal a role for nasopharyngolaryngoscopy use in the diagnosis of retropharyngeal abscess.
  + Safety of this procedure in the setting of retropharyngeal abscess is unclear.
  + Nasopharyngolaryngoscopy has been performed preoperatively in two adults, but no reports of its use in children have been found.
* Endotracheal intubation
  + Securing the airway may be required if the patient with retropharyngeal abscess is exhibiting signs of impending upper airway obstruction. Endotracheal intubation may be attempted, but it may be difficult because of distortion of the upper airway.
  + Prophylactic intubation for a patient with retropharyngeal abscess but without respiratory distress generally is not indicated unless an interhospital transfer is planned.
* A literature review did not reveal the role of surgical or needle cricothyrotomy in RPA, but If a patient with signs of upper airway obstruction cannot be intubated, a surgical or needle cricothyrotomy may be required; the procedure may be difficult to perform, however, due to tissue edema and distortion.
* A tracheostomy may be required as definitive airway management in patients with retropharyngeal abscess and respiratory distress, but the procedure may be difficult to perform due to tissue edema and distortion.
* Airway management in the operating room is preferred, with surgeon and anesthesiologist present, if clinical condition and time allow it

**EPIDEMIOLOGY**

***Frequency***

*United States*

The incidence of pediatric RPA in the United States more than doubled in the first decade of the 21st century, according to a study of pediatric deep neck space infections.

Deriving their statistics from the Kids’ Inpatient Database (KID), Novis et al found that between 2000 and 2009, the incidence of RPA increased from 0.1 cases per 10,000 to 0.22 cases per 10,000. They also found no significant change in the incidence of either peritonsillar or parapharyngeal abscess in those years.

A study by Woods et al, also using the KID, reported the incidence of RPA to have risen, among children under age 20 years, from 2.98 per 100,000 population in 2003 to 4.10 per 100,000 population in 2012.

A review of cases of RPA over an 11-year period at the Children's Hospital of Michigan revealed a 4.5-times increase in the incidence of RPA when compared with the previous 12 years.A later review at the same hospital revealed that the incidence increased 2.8-fold between 2004 and 2010, compared with the incidence from 1993-2003.

Similarly, an 11-year chart review of 162 pediatric patients with RPA at St. Louis Children's Hospital revealed that the number of RPA cases in children increased significantly from 1995 to 2006.

A study by Angajala et al determined that of 119 pediatric patients in the greater Los Angeles community with a neck abscess treated with incision and drainage, 10.1% had an RPA. Patients with neck abscesses requiring incision and drainage tended to reside in lower income neighborhoods.

*International*

A review of deep neck infections (DNI) in children over a 12-year period at a medical center in Taiwan revealed 50 children with DNI. Nine children had DNI in the retropharyngeal space, 17 in the parapharyngeal space, 21 in the peritonsillar region, and 3 were mixed.

Another study from Taiwan, by Huang et al, found that out of 52 children with DNI, the retropharyngeal space was the third most common site of infection (7 patients), after the parapharyngeal space (22 patients) and the submandibular space (12 patients).

A review of RPAs and parapharyngeal abscesses (PPAs) in children presenting to 2 pediatric tertiary care medical centers in Israel over an 11-year period revealed 39 children with RPA or PPA. The incidence increased during the course of the study.

A retrospective analysis of children diagnosed with RPA and PPA over a 9-year period in a tertiary care medical center in Spain revealed 17 children with RPA, 11 with PPA, and 3 with both.

Another Spanish study, a retrospective, single-center report by Sanz Sánchez and Morales Angulo, found the incidence of RPA over an approximately 25-year period to be 0.2 cases per 100,000 inhabitants per year.

A study by Yap et al found that in Wales, hospital admissions for RPA, as well as for tonsillitis, PPA, and peritonsillar abscess, rose between 1999 and 2014.

A retrospective review at a single center in Scotland revealed that the number of deep neck space abscesses grew between 2006 and 2015 from 1 to 15, respectively.

***Mortality/Morbidity***

Once mediastinitis occurs, mortality approaches 25%, even with antibiotic therapy. Retropharyngeal abscess can also cause internal jugular vein thrombosis, carotid artery erosion, pericarditis, and epidural abscess. In addition to invasion of contiguous structures, retropharyngeal abscess can cause sepsis and airway compromise.

Overall mortality rate was 1% in a review of deep cervical space infections in Taiwan.

In a study of 234 adults with deep space infections of the neck in Germany, the mortality rate was 2.6%. The cause of death was primarily sepsis with multiorgan failure.

In the United States, in 2003, a review of the Kids' Inpatient Database (KID) revealed 1321 pediatric admissions with RPA, with no fatalities.

A case series from Children's National Medical Center in Washington DC presents 4 children of ages ranging from 8 months to 18 months with RPA who developed mediastinitis.

All 4 were treated aggressively with antibiotics and surgical drainage of RPA, and 3 patients required thoracoscopic debridement. All 4 children survived without sequelae.

***Race***

In a 10-year review of retropharyngeal abscess cases treated at Kings County Hospital in Brooklyn, New York, 70% of patients were Black, 25% were White, and 5% were Hispanic.

A study of pediatric patients with retropharyngeal abscess at Wayne State University in Detroit revealed 43% of cases occurred in Blacks, 54% in Whites, 1% in Hispanics, and 1% in biracial.

In the United States, in 2003, a review of the Kids' Inpatient Database (KID) revealed 1321 pediatric admissions with retropharyngeal abscess, of which 37.4% were White, 11.7% were Black, 11.1% were Hispanic, 2% were Asian, and 3.8% were other races, with the race not being recorded in the rest of the patients.

***Sex***

Retropharyngeal abscess is more common in males than in females, with generally reported male preponderance of 53-55%.

* Children's Hospital of Michigan reports 54% of cases of RPA in males in a 2012 study.
* A study of children with retropharyngeal abscess in Toronto reported 67% of cases in males.
* A study of retropharyngeal abscess in children in Detroit found 56% of cases in males.
* A study of adults with deep space infections of the neck in Germany revealed that 56% of patients were male and 44% were female.
* A study of cases in Nigeria found a male-to-female ratio of 1:1.
* In the United States, in 2003, a review of the Kids' Inpatient Database (KID) revealed 1321 pediatric admissions with retropharyngeal abscess, of which 63% were male.

***Age***

Initially, retropharyngeal abscess was thought to be a disease limited to children, but now it is being encountered with increasing frequency in adults.

* In children, retropharyngeal abscesses develop most frequently between the ages of 2 and 4 years.
* This was supported by a review by Bochner, which found the incidence to be greatest in children younger than age 5 years and, by gender, in boys.
* A review of adults with deep space infections of the neck in Germany revealed a mean age (±standard deviation) of 44.5 (±21.8) years.
* A review of retropharyngeal abscess cases at the Hospital for Sick Children in Toronto revealed that 66% of pediatric cases occurred in children younger than 6 years.
* A review of 30 cases of retropharyngeal abscess over an 11-year period in Nigeria found the median age to be 21 months, and 77% of patients were younger than 5 years. Eighty-three percent of retropharyngeal abscesses occurred in children, and 17% occurred in adults.
* A 10-year review at Kings County Hospital in Brooklyn, New York, revealed that 30% of the cases were in pediatric patients aged 16 months to 8 years and 70% were in adults aged 21-64 years.
* A 35-year review of cases involving children who were treated for retropharyngeal abscess at the Children's Hospital of Los Angeles revealed that 50% of patients were younger than 3 years and 71% were younger than 6 years.
* A review or retropharyngeal abscess in children in Detroit found a mean age of 4.1 years, with a range from 2 months to 18 years.
* A review in Sydney, Australia, found that, in 55% of pediatric cases of retropharyngeal abscess, the children were younger than 1 year, with 10% diagnosed in the neonatal period.
* A review of RPA cases in children in Albuquerque revealed a median age of 36 months, with 75% of patients younger than 5 years and 16% of patients younger than 1 year.
* In the United States, in 2003, a review of the Kids' Inpatient Database (KID) revealed 1321 pediatric admissions with retropharyngeal abscess, with an average age of 5.1 years (SD, 4.4).
* An 11-year chart review of 162 pediatric patients with retropharyngeal abscess at St. Louis Children's Hospital revealed an average age of 4.9 years (range, 6 d to 17 y).
* A 5-year review of 11 children with parapharyngeal abscess in Portugal revealed an average age of 3.3 years (range, 0-12 y).
* A 12-year retrospective review of 50 pediatric patients with deep neck infections in Taipei revealed that all of the retropharyngeal abscesses occurred in children younger than 10 years.

**PREDEFINED Q & A SETS**

Q1: “How do I take care of myself?”

Make sure to take all prescribed antibiotics, even if you feel better. Retropharyngeal abscesses can return. Taking all your medicine as directed can reduce the risk that this occurs.

Q2: “What’s the difference between a retropharyngeal abscess and a peritonsillar abscess?”

A retropharyngeal abscess is a collection of pus in the back of your throat. In contrast, a peritonsillar abscess is a collection of pus that forms near your tonsils. Both result from an infection and require similar treatment approaches, including antibiotics and draining the abscess.

**DOCTOR-PATIENT CONVERSATIONS**

Doctor: Hello, what brings you in today?

Patient: Doctor, my child has been having a sore throat and fever for a few days, and now I noticed some difficulty swallowing and they seem to have a swollen neck. Could this be serious?

Doctor: Thank you for telling me. Your child's symptoms could be due to an infection behind the throat in an area called the retropharyngeal space. Sometimes, infections there can cause a collection of pus called a retropharyngeal abscess, which is a serious but treatable condition.

Patient: What exactly is a retropharyngeal abscess?

Doctor: It's a deep infection in the tissues behind the throat, which can cause swelling and sometimes block the airway or cause difficulty swallowing. This happens when bacteria from an upper respiratory infection or throat infection spread into that space.

Patient: What symptoms should I watch out for?

Doctor: Common symptoms include fever, painful swallowing, difficulty opening the mouth, neck stiffness or swelling, muffled voice, and sometimes noisy breathing or difficulty breathing. Children, especially those under 5 years old, are more at risk.

Patient: How do you diagnose it?

Doctor: We usually start with a physical exam. If we suspect an abscess, we confirm it using imaging such as a CT scan of the neck, which shows us the exact size and location of the abscess.

Patient: How is it treated?

Doctor: Treatment usually involves intravenous antibiotics to fight the infection. In many cases, surgery is needed to drain the abscess to relieve the pressure and remove the pus. This is typically done by an ENT specialist under general anesthesia.

Patient: Is it dangerous? What about recovery?

Doctor: If treated promptly, the prognosis is good. However, if left untreated, the infection can spread to nearby structures and become life-threatening. With proper treatment, most children recover fully, though hospital stay and close monitoring are necessary.

Patient: Are there any complications we should know about?

Doctor: Possible complications include airway obstruction, spread of infection to the chest, or sepsis, but these are rare with timely care. That's why early diagnosis and treatment are important.

Patient: What should we do at home after treatment?

Doctor: Follow all medication instructions carefully. Keep your child well-hydrated and rested. Attend all follow-up appointments to ensure the infection has fully resolved.

Patient: Thank you, doctor. I feel better understanding

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[Retropharyngeal Abscess: Causes, Symptoms & Treatment](https://my.clevelandclinic.org/health/diseases/24026-retropharyngeal-abscess#overview)

**SCARS**

**ALTERNATIVE NAMES:** Synonyms for scars include blemish, mark, disfigurement, deformity, blotch, flaw, defect, pockmark, blight, and imperfection. Other alternative names for scars are cicatrix, cicatrice, keloid, cat-face, discoloration, defect, scratch, and cicatrization. Additionally, scars can be referred to as ravages, aftereffects, consequences, effects, results, damaging effects, ill effects, blemishes, and disfigurement.

**DEFINITION / DESCRIPTION**

Scars form as part of the healing process after your skin has been cut or damaged. The skin repairs itself by growing new tissue to pull together the wound and fill in any gaps caused by the injury. Scar tissue is made primarily of a protein called collagen.

Scars develop in all shapes and sizes. Some scars are large and painful, while some are barely visible. People with dark skin (especially people with African, Asian or Hispanic heritage), as well as red-haired individuals, are more likely to develop keloid scars. Keloids are raised scars that grow and extend beyond the injured area. Depending on their size, type and location, your scars may look unsightly and may even make it difficult to move.

Not all scars require treatment, and many fade away over time. If a scar is bothering you or causing pain, treatments can help.

Nearly everyone develops some type of scar, whether from an accident, a surgical procedure, acne or an illness like chickenpox (varicella).

**CAUSES**

Scars are part of the body’s healing process. As part of your immune system, your skin is the barrier to protect you from germs and other harmful substances. When skin is injured, the body creates new tissue made of collagen to help reseal itself.

Collagen plays many important roles throughout your body, including plumping up your skin and helping your cartilage protect your joints. When a scar develops, collagen fibers repair damaged skin and close any open areas. The new tissue protects you from infection.

**SIGNS / SYMPTOMS**

When a scar first develops on lighter skin, it’s usually pink or red. Over time, the pinkish color fades, and the scar becomes slightly darker or lighter than the color of the skin. In people with dark skin, scars often appear as dark spots. Sometimes scars itch, and they may be painful or tender.

A scar’s appearance depends on several factors, including:

* Injury or event that caused the scar, such as surgery, a burn or severe acne.
* Size, severity and location of the wound.
* Treatment you received for the wound, such as stitches or bandages.
* Your age, genes, ethnicity and overall health.

***What are the types of scars?***

Scars can develop anywhere on the skin. There are several types of scars, including:

* Contracture: Often developing after a burn, a contracture scar causes the skin to tighten (contract). These scars can make it difficult to move, especially when the scarring gets into the muscles and nerves or occurs over a joint.
* Depressed (atrophic): These sunken scars often result from chickenpox or acne. They look like rounded pits or small indentations in the skin. Also called ice pick scars, they develop most often on the face. Acne scars may become more noticeable as you age because the skin loses collagen and elasticity over time.
* Flat: Although it may be slightly raised at first, this type of scar flattens out as it heals. Flat scars are often pink or red. Over time, they may become slightly lighter or darker than the surrounding skin.
* Keloids: These scars are raised above the skin’s surface and spread beyond the wounded area. The overgrown scar tissue can get large and may affect movement.
* Raised (hypertrophic): You can feel a hypertrophic scar when you run your finger over it. These raised scars may get smaller over time, but they never completely flatten out. Unlike keloids, they don’t grow or spread beyond the wounded area.
* Stretch marks: When skin expands or shrinks quickly, the connective tissues under the skin can be damaged. Stretch marks often develop during pregnancy, puberty or after gaining or losing a lot of weight. They usually appear on the breasts, stomach, thighs and upper arms.

Scar tissue can also build up inside the body. Internal scar tissue can result from surgery (like abdominal adhesions) and some health conditions, such as Asherman’s syndrome and Peyronie’s disease. An autoimmune disease such as scleroderma creates skin changes resembling scarring from the inflammation in the skin.

**DIAGNOSIS METHODS**

You can easily diagnose most scars yourself by keeping an eye on an area of skin that has healed from an injury. Scars often look darker, lighter or pinker than the surrounding skin.

Your healthcare provider will do a physical examination to evaluate a scar that’s causing problems. Your provider will look at the scar’s size, texture and color to determine its type. Treatments vary depending on the type of scar, its location, what caused it and how long you’ve had it.

**TREATMENT OPTIONS**

Several treatments can make scars smaller or less noticeable. Your healthcare provider may recommend one treatment or a combination. Scar treatment depends on several factors, including:

* Type, size and location of the scar.
* Whether the scar is causing you pain or affecting your ability to move.
* Your age and the age of the scar.

***What are the treatments for scars?***

Treatments can reduce a scar’s size or appearance, but the scar will never completely go away. Some treatments prevent a scar from forming as a wound heals. Scar treatments include:

* Dermabrasion: A common acne scar treatment, dermabrasion removes the top layer of skin by gently “sanding” the skin. The procedure softens and smooths the skin and can improve the appearance of scars.
* Injections: Your healthcare provider injects medication directly into the scar, making it smaller and flatter. Corticosteroid injections can reduce the size of keloid scars. Your healthcare provider may inject drugs that treat cancer, such as bleomycin (Bleo 15k™) and fluorouracil (Adrucil® or 5-FU), to flatten scars and reduce itching and pain.
* Laser treatments: Several types of laser and light treatments can make scars (including acne scars) less noticeable. Laser treatments use a particular wavelength of light to cause a particular action in the skin. The V beam is a pulsed dye laser at 595 nm (nanometers) that targets small blood vessels in the skin. Sometimes the scars remain pink or red because the new blood vessels that developed to heal the wound never receded once their job was done. This laser can cauterize the small vessels from the inside out to remove them from the scar and allow the pink or red color to fade. This action may also help the scar flatten if it’s too thick or thicken if it is too thin. Other lasers (such as the Fraxel laser) can vaporize small columns of tissue within the scar to break up the collagen fibers and allow the scar to remodel and become more flexible. The treatments can also help with pain, itching and sensitivity. Laser treatments may cause hyperpigmentation (skin darkening) or hypopigmentation (skin lightening) in people who have dark skin. Talk to your healthcare provider about side effects before starting treatments.
* Pressure therapy: An elastic bandage, dressing or stocking puts pressure on a wound during the healing process. The pressure prevents a scar from forming or decreases its size. Massage therapy can also help break up scar tissue and allow it to remodel.
* Scar-revision surgery: A range of surgical procedures can remove a scar, improve its appearance or transplant skin from another area (skin graft). This is an exchange of one type of scar for a different, more preferable scar.
* Topical creams and ointments: Applying silicone ointment to a scar may make it smaller or prevent it from forming. Or your healthcare provider may recommend applying corticosteroid cream or a silicone gel sheet to the area. If you have dark skin, ask your provider about using a skin-lightening cream with hydroquinone to lighten scars.

**PREVENTION TIPS**

Although you can’t always prevent injuries that cause scars, you can reduce the risk of a scar forming after an injury. If a scar does develop, careful care can make the scar less noticeable.

To reduce the risk of scarring, you should:

* See your healthcare provider: If you have a wound that may leave a scar, visit your provider for an examination. You may need stitches or special bandages to hold the skin together while it heals. Stitches can minimize scarring. Be sure to follow your provider’s instructions when caring for stitches. Depending on the type and location of the wound you may need oral or topical antibiotics to prevent infection.
* Clean the wound: Wash the area with soap and water. Clean out any dirt or dried blood, and apply a bandage over the wound to keep germs out. Be sure to change the bandage often as the wound heals.
* Keep the wound moist: Applying petroleum jelly or moist burn pads will keep the wound from becoming too dry and developing a scab. Scabs can make scarring worse.
* Protect it from the sun: Cover the scar or use sunscreen to protect it. Sun exposure can make a scar darker. Repeated exposure increases the risk of developing skin cancer.
* Keep up your nutrition: Having low levels of vitamin D or C in your system can make scarring worse and you need adequate high quality protein in your diet to help your skin make what is needed to heal.

**OUTLOOK / PROGNOSIS**

Most scars fade over time and don’t cause long-term health problems. How a scar changes depends on its location, size and type. A scar may fade so much that you can barely see it, but it never completely goes away.

Some scars cause problems months or years later. As nerve endings grow back, the scar may become painful or itchy. Skin cancer can develop in scars, especially in burn scars. To avoid skin cancer, wear sunscreen or keep your scar covered

**WHEN TO SEE A DOCTOR / RED FLAG**

If a scar’s appearance bothers you, talk to your provider about procedures that can make it less noticeable.

Also see your provider if the scar changes or is painful, tender, itchy or infected. And if you notice a mole, freckle or growth on or near the scar, call your provider right away. This may be a sign of skin cancer, which can grow in a scar.

If you’ve had a keloid scar, you’re more likely to develop another one. Talk to your provider before getting piercings, tattoos or elective surgery (such as cosmetic surgery). Your provider will recommend precautions (like wearing a pressure garment) if skin starts to thicken and turn into a keloid.

**DIFFERENTIAL DIAGNOSIS**

1. Hypertrophic Scar

* Raised, red, thickened scars but confined within the boundaries of the original wound.
* Usually develops within weeks after injury, often improves or flattens over time.
* May cause itching or discomfort.

2. Keloid

* Raised, firm, often itchy or painful scars that extend beyond the original wound margins.
* More common in darker skin types and certain body areas (e.g., chest, shoulders).
* Histologically, keloids may have thick hyalinized collagen bundles and a "tongue-like" advancing edge.
* Keloids do not regress spontaneously and are more resistant to treatment.

3. Atrophic Scar

* Depressed or sunken scars due to loss of underlying tissue, e.g., acne scars or scars after varicella.

4. Other Conditions Mimicking Scars

* Collagenoma: benign collagen nodules, usually multiple and skin-colored.
* Acne keloidalis: papules and keloid-like scarring especially on the scalp's nape.
* Keloidal dermatofibroma: benign fibrous tumor that may resemble keloids.
* Cutaneous neoplasms: such as dermatofibroma or dermatofibrosarcoma protuberans, which may mimic scar tissue clinically or histologically.
* Factitial (self-induced) scars: due to chronic picking or scratching, seen in neurodermatitis or prurigo nodularis.

5. Scar-related Pain Conditions

* Painful scars may be a feature of nerve entrapment or neuroma formation within the scar tissue.
* Complex regional pain syndrome (CRPS) can sometimes mimic scar pain syndromes

**EPIDEMIOLOGY**

* Prevalence of Scars: Approximately 48.5% of people report having at least one scar, with about 22% of scars appearing within the past year. The most common scar locations are the face and abdomen, varying slightly by sex (men often report facial scars, women more often abdominal scars).
* Hypertrophic Scars and Keloids:
  + Hypertrophic scars occur frequently after surgery and burn injuries, with incidence rates reported to be more than 40% following surgery or burns in the U.S..
  + The prevalence of keloids varies geographically and ethnically, with a higher prevalence in darker-skinned populations, especially among Africans.
  + Hypertrophic scars are reported in 32-72% of patients after burn injuries, with younger patients and larger, deeper burns being more at risk.
* Symptoms and Impact: Many scars cause symptoms such as itching, pain, and discomfort, reported by around a quarter to nearly half of patients depending on the study. These symptoms can affect physical comfort and quality of life.
* Risk Factors for Pathologic Scarring: Younger age, burn severity, burn location (thorax, upper limbs, feet), genetic predisposition, infection, delayed wound healing, and type of injury (flame burns higher risk) influence the likelihood of hypertrophic or keloid scarring.
* Global Data Gaps: Despite scars being very common worldwide, epidemiological data are still limited and mostly derived from specific populations or high-income countries. More comprehensive international studies are needed

**PREDEFINED Q & A SETS**

Q1: “What is a scar?”

A scar is a mark left on the skin after an injury, surgery, or illness has healed. It forms as part of the natural healing process, when the body produces new connective tissue (fibrous tissue) to repair the damaged skin.

Q2: “What causes scars?”

Scars develop following damage such as cuts, burns, infections, surgery, or conditions like acne. The depth, size, and location of the wound affect how noticeable a scar will be.

Q3: “What types of scars are there?”

Answer:

* Hypertrophic scars: Raised, red scars that stay within the boundaries of the original wound. They may improve over time.
* Keloid scars: Raised scars that grow beyond the original wound edges and can be itchy or painful. They are more common in darker-skinned individuals.
* Atrophic scars: Depressed or sunken scars, often from acne or chickenpox.
* Other scar-like conditions: Include scars with underlying nerve pain or scarring caused by repeated trauma.

Q4: “What symptoms might a scar cause?”

Scars can sometimes itch, feel tight, be painful, or cause discomfort. In some cases, they may restrict movement if they form over joints or large areas.

How are scars diagnosed?

A healthcare provider usually diagnoses scars by clinical examination. In some cases, a dermatologist may be consulted to distinguish scars from other skin conditions or to check for complications.

Q5: “Can scars be prevented?”

While it is not always possible to prevent scars, steps that promote good wound care—such as cleaning wounds properly, avoiding infection, and minimizing tension on healing skin—can reduce scarring risk.

Q6: “How can scars be treated or minimized?”

Answer:

* Massage: Gentle scar massage with moisturizers can soften and flatten scars.
* Topical treatments: Silicone gels or sheets help improve scar appearance.
* Medical treatments: Steroid injections, laser therapy, microneedling, or surgery may be options for problematic scars.
* Sun protection: Protecting scars from the sun with broad-spectrum sunscreen (SPF 30 or higher) prevents discoloration and reduces risk of skin cancer developing in scar tissue.

Q7: “When should I seek medical advice about a scar?”

Answer:

* If a scar becomes increasingly red, painful, swollen, or starts to itch persistently.
* If a scar seems to be growing beyond the original wound area.
* If you notice changes such as new lumps, sores, or unusual skin changes in or near the scar.
* If scars cause functional impairment or significant distress.

Q8: “What is the outlook for scars?”

Most scars improve and fade over time, usually within 12 to 18 months, though some may remain noticeable longer. Treatments can improve the appearance and symptoms of scars but cannot always remove them completely.

**DOCTOR-PATIENT CONVERSATIONS**

Doctor: Hello! I understand you have some concerns about a scar. Can you tell me a little about it?

Patient/Parent: Yes, the scar looks different from when it first healed. It’s red and raised, and sometimes it itches and hurts a bit. Should I be worried?

Doctor: Thank you for telling me. Scars often change over time as your skin heals and remodels. It’s common for scars to be red and raised for several months. Some scars can become thick or grow beyond the original wound — these are called keloid or hypertrophic scars.

Patient/Parent: What causes these kinds of scars to form?

Doctor: Keloid scars happen when the body produces too much collagen during healing. Some people are more prone to them because of genetic factors or skin type. Also, scars over joints or places that get stretched or irritated can be more noticeable.

Patient/Parent: What signs should make me see a doctor about the scar?

Doctor: You should come back if the scar:

* Keeps growing beyond the original injury
* Becomes painful, very itchy, or tender
* Shows signs of infection like redness, swelling, warmth, or oozing
* Limits movement if near a joint
* Shows new lumps or changes in color that concern you

Patient/Parent: How can we treat or manage the scar?

Doctor: Treatment depends on the scar type and symptoms. Options include:

* Silicone gel sheets or creams to reduce redness and thickness
* Steroid injections for raised or keloid scars
* Pressure therapy or massage
* Laser treatments in some cases
* Surgery only if necessary, but sometimes scars can come back worse after surgery
* Keeping the area moisturized and protected from the sun to improve healing

Patient/Parent: Will the scar ever go away completely?

Doctor: Scars usually improve over time and become less noticeable, but they rarely disappear completely. Our goal is to help the scar heal as well as possible and reduce any symptoms or cosmetic concerns.

Patient/Parent: Thank you, that’s helpful. When should I follow up?

Doctor: If you notice any changes I mentioned or if the scar causes discomfort or affects your daily activities, please come in for an evaluation. Otherwise, we can monitor it during regular visits.

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**SALIVARY GLAND TUMORS (DISORDERS)**

**ALTERNATIVE NAMES:** Alternative names for salivary gland tumors include "Tumor - salivary duct". Other references also mention similar terminology, but specific alternative names are not provided in the context.

**DEFINITION / DESCRIPTION**

Salivary gland tumors are growths of cells that start in the salivary glands. Salivary gland tumors are rare.

The salivary glands make saliva. Saliva aids in digestion, keeps the mouth moist and supports healthy teeth. There are three pairs of major salivary glands under and behind the jaw. These are the parotid, sublingual and submandibular glands. Many other tiny salivary glands are in the lips, inside the cheeks, and throughout the mouth and throat.

Salivary gland tumors can happen in any salivary gland. Most salivary gland tumors occur in the parotid gland. Of these, most aren't cancerous. For every five parotid gland tumors, on average, only one is found to be cancerous.

Treatment for salivary gland tumors is usually with surgery to remove the tumor. People with salivary gland cancers may need additional treatments.

**CAUSES**

The cause of many salivary gland tumors isn't known. Healthcare professionals have identified some things that raise the risk of salivary gland tumors. These include smoking and radiation therapy for cancer. Not everyone with a salivary gland tumor has these risk factors, though. More research is needed to find out exactly what causes these tumors.

Salivary gland tumors happen when cells in a salivary gland develop changes in their DNA. A cell's DNA holds the instructions that tell the cell what to do. In healthy cells, the DNA gives instructions to grow and multiply at a set rate. The instructions also tell the cells to die at a set time. In tumor cells, the changes give different instructions. The changes tell the tumor cells to make many more cells quickly. Tumor cells can keep living when healthy cells die. This causes too many cells.

Sometimes the changes in the DNA turn the cells into cancer cells. Cancer cells can invade and destroy healthy body tissue. In time, cancer cells can break away and spread to other parts of the body. When cancer spreads, it's called metastatic cancer.

### **Types of salivary gland tumors**

Many different types of salivary gland tumors exist. Salivary gland tumors are classified based on the type of cells involved in the tumors. Knowing the type of salivary gland tumor you have helps your healthcare team decide which treatment options are best for you.

Types of salivary gland tumors that aren't cancerous include:

* Pleomorphic adenoma.
* Basal cell adenoma.
* Canalicular adenoma.
* Oncocytoma.
* Warthin tumor.

Types of cancerous salivary gland tumors include:

* Acinic cell carcinoma.
* Adenocarcinoma.
* Adenoid cystic carcinoma.
* Clear cell carcinoma.
* Malignant mixed tumor.
* Mucoepidermoid carcinoma.
* Oncocytic carcinoma.
* Polymorphous low-grade adenocarcinoma.
* Salivary duct carcinoma.
* Squamous cell carcinoma.

**RISK FACTORS**

Factors that may increase the risk of salivary gland tumors include:

* **Older age.** Though salivary gland tumors can happen at any age, they most often happen in older adults.
* **Radiation exposure.** Radiation treatments for cancer, such as radiation used to treat head and neck cancers, may increase the risk of salivary gland tumors.
* **Smoking tobacco.** Smoking tobacco is shown to increase the risk of salivary gland tumors.
* **Viral infections.** People who have had viral infections such as Epstein-Barr virus, human immunodeficiency virus and human papillomavirus may have a higher risk of salivary gland tumors.
* **Workplace exposure to certain substances.** People who work with certain substances may have an increased risk of salivary gland tumors. Examples of industries associated with an increased risk include those that involve rubber manufacturing and nickel.

**SIGNS / SYMPTOMS**

Signs and symptoms of a salivary gland tumor may include:

* A lump or swelling on or near the jaw or in the neck or mouth.
* Muscle weakness on one side of the face.
* Numbness in part of the face.
* Ongoing pain near a salivary gland.
* Trouble opening the mouth widely.
* Trouble swallowing.

**DIAGNOSIS METHODS**

Salivary gland tumor diagnosis often begins with a physical exam of the area by a healthcare professional. Imaging tests and a biopsy may be used to find the location of the tumor and determine what type of cells are involved.

### **Physical exam**

A healthcare professional feels the jaw, neck and throat for lumps or swelling.

### **Imaging tests**

Imaging tests make pictures of the body. They can show the location and size of a salivary gland tumor. Tests might include MRI, CT and positron emission tomography, which is also called a PET scan.

### **Biopsy**

A biopsy is a procedure to remove a sample of tissue for testing in a lab. To collect a sample of tissue, a fine-needle aspiration or a core needle biopsy may be used. During the biopsy, a thin needle is inserted into the salivary gland to draw out a sample of suspicious cells. The sample is sent to a lab for testing. Tests can show what types of cells are involved and whether the cells are cancerous.

### **Determining the extent of salivary gland cancer**

If you're diagnosed with salivary gland cancer, you may have other tests to see if the cancer has spread. These tests help your healthcare team find out the extent of your cancer, also called the stage.

Cancer staging tests often involve imaging tests. The tests might look for signs of cancer in your lymph nodes or in other parts of your body. Your healthcare team uses the cancer staging test results to help create your treatment plan.

Imaging tests may include CT, MRI and PET scan. Not every test is right for every person. Talk with your healthcare professional about which procedures will work for you.

The stages of salivary gland cancer range from 0 to 4. A stage 0 salivary gland cancer is small and only in the gland. As the cancer gets larger and grows deeper into the gland and surrounding areas, such as the facial nerve, the stages get higher. A stage 4 salivary gland cancer has grown beyond the gland or has spread to the lymph nodes in the neck or to distant parts of the body.

**TREATMENT OPTIONS**

Treatment for salivary gland tumors usually involves surgery to remove the tumor. People with salivary gland cancers may need additional treatments. These additional treatments can include radiation therapy, chemotherapy, targeted therapy or immunotherapy.

### **Surgery**

Surgery for salivary gland tumors may include:

* **Removing part of the affected salivary gland.** If your tumor is small and located in an easy-to-access spot, your surgeon may remove the tumor and a small portion of healthy tissue around it.
* **Removing the entire salivary gland.** If you have a larger tumor, your surgeon may recommend removing the entire salivary gland. If your tumor extends into nearby structures, they also may be removed. Nearby structures may include the facial nerves, the ducts that connect the salivary glands, facial bones and skin.
* **Removing lymph nodes in your neck.** If your salivary gland tumor is cancerous, there may be a risk that the cancer has spread to the lymph nodes. Your surgeon may recommend removing some lymph nodes from your neck and testing them for cancer.
* **Reconstructive surgery.** After the tumor is removed, your surgeon may recommend reconstructive surgery to repair the area. If bone, skin or nerves are removed during your surgery, these may need to be repaired or replaced using reconstructive surgery.  
  During reconstructive surgery, the surgeon works to make repairs that improve your ability to chew, swallow, speak, breathe and move your face. You may need transfers of skin, tissue, bone or nerves from other parts of your body to rebuild areas in your mouth, face, throat or jaws.

Salivary gland surgery can be difficult because several important nerves are located in and around the glands. For example, a nerve in the face that controls facial movement runs through the parotid gland.

Removing tumors that involve important nerves may require working around and underneath the facial nerves. Sometimes the facial nerve gets stretched during surgery. This can cause loss of movement in the face muscles. Muscle movement often gets better over time. Rarely, the facial nerve must be cut in order to get all of the tumor. Surgeons can repair the facial nerve using nerves from other areas of the body or with other techniques.

### **Radiation therapy**

If you're diagnosed with salivary gland cancer, your healthcare team may recommend radiation therapy. Radiation therapy treats cancer with powerful energy beams. The energy can come from X-rays, protons or other sources. For salivary gland cancer, radiation therapy is most often done with a procedure called external beam radiation. During this treatment, you lie on a table while a machine moves around you. The machine directs radiation to precise points on your body.

Radiation therapy can be used after surgery to kill any cancer cells that might remain. If surgery isn't possible because a tumor is very large or is located in a place that makes removal too risky, your healthcare professional may recommend radiation alone or in combination with chemotherapy.

### **Chemotherapy**

Chemotherapy treats cancer with strong medicines. Chemotherapy isn't currently used as a standard treatment for salivary gland cancer, but researchers are studying its use.

Chemotherapy may be an option for people with advanced salivary gland cancer. It's sometimes combined with radiation therapy.

### **Targeted therapy**

Targeted therapy for cancer is a treatment that uses medicines that attack specific chemicals in the cancer cells. By blocking these chemicals, targeted treatments can cause cancer cells to die.

For salivary gland cancer, targeted therapy may be used when the cancer can't be removed with surgery. It also may be used for advanced cancers that spread to other parts of the body or cancer that comes back after treatment.

Some targeted therapies only work in people whose cancer cells have certain DNA changes. Your cancer cells may be tested in a lab to see if these medicines might help you.

### **Immunotherapy**

Immunotherapy for cancer is a treatment with medicine that helps the body's immune system kill cancer cells. The immune system fights off diseases by attacking germs and other cells that shouldn't be in the body. Cancer cells survive by hiding from the immune system. Immunotherapy helps the immune system cells find and kill the cancer cells.

For salivary gland cancer, immunotherapy may be used on cancer that can't be removed with surgery. It also may be used for advanced cancers that have spread to other parts of the body or cancer that has come back after treatment.

### **Palliative care**

Palliative care is a special type of healthcare that helps you feel better when you have a serious illness. If you have cancer, palliative care can help relieve pain and other symptoms. A healthcare team that may include doctors, nurses and other specially trained health professionals provides palliative care. The care team's goal is to improve the quality of life for you and your family.

Palliative care specialists work with you, your family and your care team. They provide an extra layer of support while you have cancer treatment. You can have palliative care at the same time you're getting strong cancer treatments, such as surgery, chemotherapy or radiation therapy.

The use of palliative care with other proper treatments can help people with cancer feel better and live longer

**Lifestyle and home remedies**

### **Coping with dry mouth**

People with salivary gland tumors that are cancerous may need to have radiation therapy. One side effect of radiation therapy to the head and neck area is having a very dry mouth, called xerostomia. Having a dry mouth can cause discomfort. It can lead to frequent infections in your mouth, cavities and problems with your teeth. Dry mouth also can make eating, swallowing and speaking difficult.

You may find some relief from dry mouth and its complications if you:

* **Avoid acidic or spicy foods and drinks.** Choose foods and drinks that won't irritate your mouth. Avoid caffeinated and alcoholic beverages.
* **Brush your teeth several times each day.** Use a soft-bristled toothbrush and gently brush your teeth several times each day. Tell your healthcare professional if your mouth becomes too sensitive to tolerate gentle brushing.
* **Choose moist foods.** Avoid dry foods. Moisten dry food with sauce, gravy, broth, butter or milk.
* **Keep your mouth moistened with water or sugarless candies.** Drink water throughout the day to keep your mouth moistened. Also try sugarless gum or sugarless candies to stimulate your mouth to produce saliva.
* **Rinse your mouth with warm salt water after meals.** Make a mild solution of warm water and salt. Rinse your mouth with this solution after each meal.

Tell your healthcare professional if you have dry mouth. Treatments may help you cope with more-severe symptoms of dry mouth. You also may be referred to a dietitian who can help you find foods that are easier to eat if you're experiencing dry mouth.

**Alternative medicine**

Complementary or alternative medicine treatments can't cure salivary gland tumors. But complementary and alternative treatments can be combined with your healthcare team's care to help relieve fatigue, pain and other symptoms.

Options may include:

* Acupuncture.
* Exercise.
* Guided imagery.
* Hypnosis.
* Massage.
* Relaxation techniques.

Ask your healthcare team whether these options are safe for you.

**PREVENTION TIPS**

There’s no way to prevent salivary gland cancer. You can reduce your overall cancer risk by avoiding certain risk factors, like smoking and drinking too much alcohol.

**OUTLOOK / PROGNOSIS**

Most people recover fully from salivary gland tumor treatment if the cancer is diagnosed and treated early. Your prognosis will depend on factors like:

* The tumor’s size.
* Whether the cancer’s spread.
* Whether the cancer has recurred.
* Which salivary gland contains the cancer cells.
* How abnormal the cancer cells appear when viewed with a microscope.
* Your overall health status.

### **What is the survival rate of salivary gland cancer?**

Cancer survival rates reflect research that tracks how many people with a particular cancer diagnosis are alive over a period of time, usually five years. With salivary gland cancer, survival rates depend on the type of cancer. For example, the survival rate for mucoepidermoid carcinoma ranges from 75% to 90% at five years. The location of the tumor matters, too. If it’s only in your salivary gland, the survival rate is 94%. The survival rate is lower if the cancer spreads.

It’s important to keep in mind that this data doesn’t consider factors unique to you — like your health, your response to treatment, etc.

Ask your healthcare provider about your likely outcomes based on your unique situation

**POSSIBLE COMPLICATIONS**

Benign salivary gland tumors may become malignant over time. The symptoms of salivary gland cancers include rapid enlargement of a pre-existing mass in or around your mouth, numbness, weakness and facial pain. These symptoms may interfere with your ability to speak and swallow properly.

**WHEN TO SEE A DOCTOR / RED FLAG**

If you have any symptoms of a salivary gland tumor, especially if your symptoms last for more than two weeks, make an appointment with a healthcare provider.

**DIFFERENTIAL DIAGNOSIS**

The differential diagnosis for malignant salivary gland tumors are as follows:

* Benign salivary lesions (pleomorphic adenoma, myoepithelioma, basal cell adenoma, Warthin's tumor, oncocytoma, canalicular adenoma, sebaceous adenoma, lymphadenoma, inverted ductal papilloma, intraductal papilloma, cystadenoma)
* Benign salivary cysts
* Sialadenitis
* Sialoliths
* Lymphadenopathy (from infectious or inflammatory causes)
* Tuberculosis
* Mononucleosis
* Chronic Sclerosing Sialadenitis (Küttner tumor, which is more common in the submandibular gland)
* First Branchial cleft cysts
* Lymphoepithelial cysts (especially in immunocompromised patients such as HIV patients)
* Metastases from tumors of other body sites

**EPIDEMIOLOGY**

Salivary gland malignancies comprise 0.5 to 1.2% of all cancers and 5% of head and neck cancers. They more commonly affect women with a male-to-female ratio of 1 to 1.5. Malignant lesions are found in about 21.7% of all salivary gland neoplasms.

Most malignant cases occur in the parotid, followed by the submandibular, sublingual, and minor salivary glands.The probability of malignancy in a parotid mass ranges from 15% to 32%, compared to 41% to 50% in a submandibular mass, 70% to 90% in minor salivary gland masses, and almost 100% in sublingual masses.

Salivary gland tumors in children are more likely to be malignant. Malignant tumors in children under 10 years old tend to be of a higher grade with poorer prognosis. Salivary tumors in children older than 10 were benign in 85% of cases, similar to that of the adult population. The most common pediatric malignant salivary gland tumors include MEC, adenocarcinoma, and ACC.

The parotid gland harbors 60 to 75% of all salivary gland tumors. The most common malignant tumors are MEC, AdCC, CExPA, adenocarcinoma, and SCC. The submandibular gland harbors 10-15% of all salivary gland tumors with an equal distribution of benign and malignant neoplasms.

AdCC is the most common malignant neoplasm in the submandibular gland, followed by MEC and CExPA. Less common tumors include ACC, salivary duct carcinoma, epi-myoepithelial carcinoma, carcinosarcoma, oncocytic carcinoma, and SCC.

Malignant submandibular tumors are more common in the 6th decade with a predilection for men. In the minor salivary glands, as many as 50% of tumors are malignant, most often located in the palate.

*Epidemiology of Specific Salivary Gland Malignancies*

* MEC is the most common salivary gland malignancy in adults and children. About 89% of cases are found in the parotid, followed by 8.4% in the submandibular gland and 0.4% in the sublingual gland.There is an equal distribution between the sexes with a predilection for the 4th to 5th decade.
* AdCC accounts for about 10% of all salivary gland neoplasms and 30% of all minor salivary gland tumors. It has a predilection for patients in the 5th to 6th decade with no difference in gender, although it tends to be more common in the submandibular gland in women.
* ACC is located in the parotid gland in more than 80% of all cases, submandibular glands in 4%, and intraoral minor salivary glands in 17%. Bilateral parotid involvement is seen in 3-5% of cases. It has a predilection for women and more commonly occurs in the 5th decade.
* CExPA accounts for 5% to 15% of all salivary gland malignancies and can arise in up to 25% of untreated pleomorphic adenomas.Malignant transformation is often seen in recurrent pleomorphic adenoma (PA), with the risk of transformation ranging from 5% to 10% for untreated pleomorphic adenomas over 15 years.About 82% of cases occur in the parotid and submandibular glands, followed by 18% in the intraoral minor salivary glands.
* Polymorphous Low-Grade Adenocarcinoma (PLGA) occurs almost exclusively in the minor salivary glands, with rare reports of it in the major salivary glands. It is the second most common intraoral minor salivary gland malignancy after MEC.
* Salivary Duct Carcinoma accounts for 7 to 10% of all salivary gland tumors and is often found in the parotid in older men in the 6th to 7th decades. It is a very aggressive malignancy of the salivary glands.
* SCC has a predilection for men and is usually found in the parotid. They are considered rare due to the infrequent occurrence of squamous metaplasia of ductal epithelium, which is thought to be responsible for the malignant transformation. The specific etiology of the malignant transformation is not known, although there is evidence implicating high-risk HPV viruses.

High-grade MEC and extension from an extra-parotid source are often misdiagnosed as primary SCC of the parotid. The true incidence of primary squamous cell carcinoma of the parotid is unknown due to its rarity and its frequency of being a misclassification of metastatic SCC. Evidence from the literature suggests the true incidence may be around 0.75-1%.Although a range of 0.3% to 4.3% has also been cited, these higher frequencies are thought to be due to misrepresentations or misclassification of these tumors.

* Primary melanoma of the salivary gland is extremely rare and accounts for 0.68% of malignant parotid neoplasms. There is a predilection for males in the 6th to 7th decade. Most melanoma in the parotid is due to cutaneous and mucosal metastasis from the head and neck.
* NHL of the salivary gland accounts for less than 10% of malignant salivary gland tumors. Although it encompasses less than 5% of all extranodal NHL, it is the most common extranodal lymphoma at the neck, comprising two-thirds of all cases. It preferentially occurs in women and patients over 50 years old. The most common variant of lymphoma associated with Sjogren Syndrome is mucosa-associated lymphoid tissue lymphoma (MALT), with 48% to 75% of all cases followed by diffuse large B-cell lymphoma (DLBCL) and follicular lymphoma.The majority of cases occur in the parotid gland

**PREDEFINED Q & A SETS**

Q1: “What are salivary gland tumors?”

Salivary gland tumors are abnormal growths that develop in the salivary glands, which produce saliva to aid digestion and keep the mouth moist. These tumors can be benign (non-cancerous) or malignant (cancerous).

Q2: “What causes salivary gland tumors?”

The exact cause of most salivary gland tumors is unknown. Genetic mutations and exposure to radiation may increase risk. Some benign tumors arise from normal gland tissue, while malignant tumors may develop after benign lesions or independently.

Q3: “What are the common types of salivary gland tumors?”

Answer:

* Benign: Pleomorphic adenoma (most common), Warthin tumor, and others.
* Malignant: Mucoepidermoid carcinoma, adenoid cystic carcinoma, acinic cell carcinoma, carcinoma ex pleomorphic adenoma, among others.

Q4: “What are the symptoms of salivary gland tumors?”

Answer:

* A painless lump or swelling in the area of a salivary gland (commonly the parotid gland near the ear)
* Facial numbness or weakness (if nerves are involved)
* Difficulty swallowing or opening the mouth
* Pain or discomfort in the affected area if advanced
* Rapid growth of a previously noticed lump

Q5: “How are salivary gland tumors diagnosed?”

Answer:

* Physical examination focusing on salivary glands and facial nerve function
* Imaging studies such as ultrasound, CT scan, or MRI to locate and assess the tumor
* Biopsy, usually fine needle aspiration (FNA), to determine if the tumor is benign or malignant

Q6: “What treatment options are available?”

Answer:

* Surgery: The main treatment, which may involve removing part or all of the affected salivary gland and possibly nearby lymph nodes
* Radiation therapy: May be recommended after surgery or if the tumor cannot be fully removed
* Chemotherapy or targeted therapy: Used less commonly, mainly for advanced or metastatic cancer
* Clinical trials: Participation may be an option for some patients exploring new therapies

Q7: “What are the risks or side effects of treatment?”

Answer:

* Surgery may affect facial nerve function causing weakness or numbness, which is often temporary but can sometimes be permanent
* Radiation can cause skin changes, dry mouth, or swallowing difficulties
* Chemotherapy side effects include fatigue, nausea, and decreased immunity
* Reconstructive surgery may be needed if large areas of tissue are removed

Q8: “What is the prognosis for salivary gland tumors?”

Prognosis depends on tumor type, size, whether it has spread, and how early it is treated. Many benign tumors have excellent outcomes after complete removal. Survival rates for malignant tumors vary by type and stage but can be favorable with early detection and appropriate treatment.

Q9: “How can I prepare for an appointment about a salivary gland tumor?”

Answer:

* Note any symptoms, such as lumps, pain, or facial changes
* Prepare a list of medications and medical history
* Write down questions about diagnosis, treatment options, risks, and prognosis
* Consider having a family member or friend accompany you for support and recall

**DOCTOR-PATIENT CONVERSATIONS**

Doctor: Hello, what brings you in today?

Patient: Doctor, I noticed a lump near my jaw and sometimes I feel discomfort. Could this be a tumor in my salivary gland?

Doctor: It’s good that you came in. Salivary gland tumors are growths that develop in the glands that produce saliva. They can be benign, meaning non-cancerous, or malignant, which means cancerous. Many salivary gland tumors are benign, but it’s important to evaluate this lump carefully.

Patient: What causes these tumors?

Doctor: The exact causes are often unknown. Sometimes genetic factors or exposure to radiation can increase the risk. Tumors can arise from normal gland tissues or rarely develop after prior benign growths.

Patient: How do you figure out what kind of tumor I have?

Doctor: We start with a thorough examination of your head and neck, including feeling around the lump and checking your facial nerve function. We will order imaging tests like an ultrasound, CT scan, or MRI to see the tumor's size and location. Then, a biopsy called fine needle aspiration (FNA) may be done to collect some cells from the lump to determine if it’s benign or malignant.

Patient: What treatments are available?

Doctor: Treatment depends on the tumor type and whether it’s cancerous. Surgery is the main treatment, where we usually remove the tumor or sometimes the whole salivary gland if needed. If the tumor is malignant, you may require additional therapies like radiation or chemotherapy to reduce the risk of recurrence.

Patient: Will surgery affect my facial muscles or appearance?

Doctor: Surgery carries some risks, especially because the facial nerve runs through the parotid gland. Most patients recover well, but temporary or occasionally permanent facial weakness can occur. We take great care to preserve your nerve function as much as possible.

Patient: What is the outlook after treatment?

Doctor: Many benign tumors have excellent outcomes after surgery. The prognosis for malignant tumors varies with the type, size, and stage at diagnosis. Early detection and treatment improve your chances for a good outcome.

Patient: Should I prepare anything before my appointment or treatment?

Doctor: Yes, it helps to note any symptoms you have experienced, your medical history, and any medications you take. You may also want to write down questions so you don’t forget to ask them during your visits. Bringing a family member or friend can help with support and remembering information.

Patient: Thank you, doctor. That was very helpful.

Doctor: You’re welcome. We will guide you through each step and answer any questions you have along the way.

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**SCHWANNOMATOSIS**

**ALTERNATIVE NAMES:** Schwannomatosis has several alternative names, including vestibular schwannomatosis , neurofibromatosis type II (NF2) , and central neurofibromatosis. It was previously referred to as neurofibromatosis type 2, or NF2. The condition is also sometimes called neurinomatosis.

**DEFINITION / DESCRIPTION**

Schwannomatosis is a condition that results in slow-growing tumors on nerve tissue. The tumors can grow on nerves in the ears, brain, spine and eyes. They also can grow on peripheral nerves, which are nerves located outside of the brain and spinal cord. Schwannomatosis is rare. It's usually diagnosed in early adulthood.

There are three types of schwannomatosis. Each type is caused by an altered gene.

In NF2-related schwannomatosis (NF2), tumors grow in both ears and can cause hearing loss. The altered gene that causes this type is sometimes passed down from a parent. NF2-related schwannomatosis was previously known as neurofibromatosis 2 (NF2).

The other two types of schwannomatosis are SMARCB1-related schwannomatosis and LZTR1-related schwannomatosis. The altered genes that cause these types usually are not passed down through families.

The tumors caused by schwannomatosis usually are not cancerous. Symptoms can include headaches, hearing loss, trouble with balance and pain. Treatment focuses on managing symptoms.

**CAUSES**

Schwannomatosis is caused by an altered gene. The specific genes involved depend on the type:

* **NF2-related schwannomatosis (NF2).** The NF2 gene produces a protein called merlin, also called schwannomin, that suppresses tumors. An altered gene causes a loss of merlin, leading to cell growth that's not controlled.
* **SMARCB1- and LZTR1-related schwannomatosis.** So far, two genes are known to cause these types of schwannomatosis. Changes of the genes SMARCB1 and LZTR1, which suppress tumors, are linked with these conditions.

**RISK FACTORS**

The gene that causes schwannomatosis is sometimes passed down from a parent. The risk of inheriting the gene differs based on the type of schwannomatosis.

For about half of people who have NF2-related schwannomatosis (NF2), they received an altered gene from a parent that caused the disease. NF2 has an autosomal dominant inheritance pattern. This means that any child of a parent who is affected by the disease has a 50% chance of having the gene change. People who have NF2 and whose relatives aren't affected are likely to have a new gene change.

In SMARCB1- and LZTR1-related schwannomatosis, the disease is less likely to be passed down from a parent. Researchers estimate that the risk of inheriting SMARCB1- and LZTR1-related schwannomatosis from an affected parent is about 15%.

**SIGNS / SYMPTOMS**

Schwannomatosis symptoms depend on the type.

### **NF2-related schwannomatosis**

Symptoms of NF2-related schwannomatosis (NF2) usually result from slow-growing tumors in both ears, known as acoustic neuromas or vestibular schwannomas. The tumors are benign, which means that they are not cancerous.

The tumors grow on the nerve that carries sound and balance information from the inner ear to the brain. These tumors can cause hearing loss.

Symptoms tend to appear during the late teen and early adult years, and can vary. Symptoms may include:

* Gradual hearing loss.
* Ringing in the ears.
* Poor balance.
* Headaches.

Sometimes NF2 can lead to the growth of tumors on other nerves, including in the brain, spine and eyes. They also can grow on peripheral nerves, which are located outside of the brain and spinal cord. People who have NF2 also may develop other benign tumors.

Symptoms of these tumors can include:

* Numbness and weakness in the arms or legs.
* Pain.
* Poor balance.
* Facial drop.
* Changes in vision or cataracts.
* Seizures.
* Headaches.

### **SMARCB1- and LZTR1-related schwannomatosis**

These two types of schwannomatosis typically affect people after age 20. Symptoms usually appear between ages 25 and 30.

SMARCB1- and LZTR1-related schwannomatosis can cause tumors to grow on nerves in the brain, spine and eyes. Tumors also may grow on peripheral nerves located outside of the brain and spinal cord.

Symptoms of SMARCB1- and LZTR1-related schwannomatosis include:

* Long-lasting pain, which can occur anywhere in the body and can be disabling.
* Numbness or weakness in various parts of the body.
* Loss of muscle, known as atrophy.

These types of schwannomatosis also can cause tumors to grow in the ear. But it happens rarely, and tumors usually grow in only one ear. This differs from NF2, which causes tumors to grow in both ears. For this reason, people with SMARCB1- and LZTR1-related schwannomatosis don't have the same hearing loss as people who have NF2.

**DIAGNOSIS METHODS**

To diagnose schwannomatosis, a healthcare professional begins with a review of your personal and family medical history and a physical exam. You also may need other tests to diagnose NF2-related schwannomatosis (NF2) or SMARCB1- and LZTR1-related schwannomatosis.

Other tests include:

* **Eye exam.** An eye exam can reveal cataracts and visual loss.
* **Hearing and balance exams.** These include a test that measures hearing called audiometry and a test that measures balance by recording eye movements, known as electronystagmography. Another test measures the electrical messages that carry sound from the inner ear to the brain, known as brain stem auditory evoked response.
* **Imaging tests.** X-rays, CT scans or MRIs can help identify bone changes, tumors in the brain or spinal cord, and very small tumors. Imaging tests also are used to monitor the condition after diagnosis.
* **Genetic tests.** Genetic tests won't always identify NF2 or SMARCB1- and LZTR1-related schwannomatosis because other genes that aren't known may be involved with the condition. However, some people choose genetic testing before having children.

**TREATMENT OPTIONS**

Treatment for schwannomatosis may include surgery or pain management. You may need regular exams and tests to monitor tumor growth. There is no cure for schwannomatosis.

### **Surgery and other procedures**

Surgery or other procedures may be needed to treat serious symptoms or complications.

* **Surgery to remove tumors.** People who have NF2-related schwannomatosis (NF2) and have hearing loss, brain stem compression or tumor growth, may need surgery to remove acoustic neuromas. Complete removal of the tumors can ease pain.
* **Stereotactic radiosurgery.** This procedure delivers radiation to the tumor without the need to cut into the body. Stereotactic radiosurgery might be an option to remove acoustic neuromas related to NF2 while preserving hearing.
* **Auditory brain stem implants and cochlear implants.** These devices might help improve your hearing if you have NF2 and hearing loss.

### **Cancer treatment**

If tumors become cancerous, they're treated with standard cancer therapies, such as surgery, chemotherapy and radiation therapy. Early diagnosis and treatment are the most important factors for a good outcome.

### **Pain medicines**

Managing pain is an important part of treatment for SMARCB1- and LZTR1-related schwannomatosis. Your healthcare professional might recommend:

* Medicines for nerve pain such as gabapentin (Neurontin, Gralise, Horizant) or pregabalin (Lyrica).
* Tricyclic antidepressants such as amitriptyline.
* Serotonin and norepinephrine reuptake inhibitors such as duloxetine (Cymbalta).
* Epilepsy medicines such as topiramate (Topamax, Qudexy XR, others) or carbamazepine (Carbatrol, Tegretol, others).

Researchers are studying medicines that can shrink noncancerous tumors that grow on the hearing and balance nerves in the ears.

**OUTLOOK / PROGNOSIS**

Your prognosis varies based on the size, quantity and location of schwannoma tumors in your body. You may have several tumors or very few. Tumors might exist in only one part of your body or many different parts. Symptoms vary from person to person.

The most identifiable symptom of schwannomatosis is chronic pain. It can be challenging to live with pain, especially if it’s severe. Pain can impact your mental and physical health. Talk to a healthcare provider if symptoms of schwannomatosis make you feel depressed or unable to meet personal or social commitments. You may find comfort in speaking to a mental health professional.

Treatment is available to help you manage symptoms. Many people see symptom improvement after taking medications. Others might need surgery to remove the tumors. There’s a risk that tumors could grow back after removal.

#### **What is the life expectancy of a person with schwannomatosis?**

Schwannomatosis doesn’t directly affect your life expectancy. If you’re concerned about your outlook, you should speak directly with your healthcare provider. Your situation is unique, so they can give you the most up-to-date information about your case.

**POSSIBLE COMPLICATIONS**

Complications can occur in schwannomatosis, and they depend on the type the person has.

### **NF2-related schwannomatosis complications**

NF2-related schwannomatosis (NF2) complications can include:

* Partial or total hearing loss.
* Facial nerve damage.
* Vision changes.
* Small benign skin tumors, known as skin schwannomas.
* Weakness or numbness in the legs or arms.
* Having many benign brain tumors or spinal tumors, known as meningiomas. These require frequent surgeries.

### **SMARCB1- and LZTR1-related schwannomatosis complications**

The pain caused by this type of schwannomatosis can be weakening. People with this type may need surgery or management by a pain specialist.

**WHEN TO SEE A DOCTOR / RED FLAG**

Visit a healthcare provider if you experience symptoms like pain and muscle weakness with an unknown cause. Always contact a provider if you notice any unusual lumps or bumps on your body.

If you have schwannomatosis and your symptoms change or get more intense, let your provider know.

**DIFFERENTIAL DIAGNOSIS**

* Neurofibromatosis type 2 (NF2)
  + Characterized by bilateral vestibular schwannomas (involving hearing and balance).
  + Genetic mutations in the NF2 gene.
  + Schwannomatosis patients do *not* have bilateral vestibular schwannomas or NF2 mutations.
* Neurofibromatosis type 1 (NF1)
  + Presents with café-au-lait spots, multiple neurofibromas, Lisch nodules.
  + Genetically distinct from schwannomatosis.
* Isolated (Solitary) Schwannomas
  + Single schwannoma without multiple tumor involvement or family history.
  + No associated genetic mutations typical of schwannomatosis.
* Other Peripheral Nerve Sheath Tumors
  + Neurofibromas, malignant peripheral nerve sheath tumors (MPNSTs), or other soft tissue tumors that may mimic schwannomas clinically or radiographically but differ histologically and genetically.
* Meningiomas without schwannomas
  + Appear without associated peripheral nerve tumors; distinct pathology.

## Diagnostic distinctions for schwannomatosis include:

* Multiple noncutaneous schwannomas without vestibular involvement on MRI.
* Absence of NF2 clinical or genetic features.
* Genetic mutations often in SMARCB1 or LZTR1 genes (distinct from NF2 mutations).
* Pain is a prominent symptom in schwannomatosis, differentiating it clinically.

**EPIDEMIOLOGY**

Schwannomatosis is a rare genetic disorder with an estimated birth incidence roughly between 1 in 40,000 and 1 in 70,000 live births, and a point prevalence around 1 in 126,000 in well-studied populations such as North-West England. This makes schwannomatosis less common than neurofibromatosis type 2 (NF2), whose prevalence is about twice as high (roughly 1 in 50,000), with a birth incidence of about 1 in 27,000 compared to 1 in 69,000 for schwannomatosis in that region.

Key epidemiological points include:

* Schwannomatosis is dominantly inherited but also has sporadic cases.
* It typically presents in adulthood, with peak incidence reported between ages 30 and 60 years.
* There is no clear predilection by sex or race.
* Schwannomatosis is characterized by multiple schwannomas mostly on peripheral and spinal nerves and does *not* feature bilateral vestibular schwannomas (unlike NF2).
* Life expectancy in schwannomatosis patients is generally better than in NF2, with mean age at death around 77 years compared with 66 years in NF2.
* Genetic mutations involving the SMARCB1 and LZTR1 genes are implicated in schwannomatosis, distinct from NF2 gene mutations.

**PREDEFINED Q & A SETS**

Q1: “What is Schwannomatosis?”

Schwannomatosis is a rare genetic disorder characterized by the development of multiple benign tumors called schwannomas on the coverings of peripheral nerves. These tumors can occur anywhere in the body and often cause chronic pain and neurological symptoms. It is distinct from other types of neurofibromatosis like NF1 and NF2.

Q2: “What causes Schwannomatosis?”

Schwannomatosis is caused by mutations in tumor suppressor genes, primarily SMARCB1 and LZTR1, located on chromosome 22. These genetic mutations lead to uncontrolled growth of Schwann cells, forming tumors. The condition may be inherited or occur spontaneously.

Q3: “Who is affected by Schwannomatosis?”

Schwannomatosis affects about 1 in 40,000 to 1 in 70,000 people. It typically presents in adults, with most diagnoses occurring between ages 20 and 40, though symptoms can appear at any age. There is no known gender or racial predilection.

Q4: “What are the symptoms of Schwannomatosis?”

Answer:

* Chronic pain is the hallmark symptom and can vary in location and severity.
* Numbness, tingling, or weakness if tumors press on nerves.
* Possible lumps under the skin due to superficial tumors.
* Less common symptoms depend on tumor location and may include difficulty with urination, bowel function, facial weakness, or headaches.

Q5: “How is Schwannomatosis diagnosed?”

Answer:

* Imaging (MRI): To identify schwannomas on peripheral nerves and spinal roots.
* Biopsy: Sample of tumor tissue examined under microscope to confirm diagnosis.
* Genetic testing: To identify mutations in SMARCB1 or LZTR1 genes and differentiate from NF2.

Q6: “How is Schwannomatosis treated?”

Answer:

* There is no cure currently.
* Treatment focuses on symptom management, especially pain control.
* Medications may be prescribed for pain relief.
* Surgical removal of schwannomas may be considered if tumors cause severe symptoms; however, surgery carries risk of nerve damage and tumors might recur.
* Clinical trials may be available for new therapies.

Q7: “What is the prognosis?”

Prognosis varies depending on the number, size, and location of tumors. Schwannomatosis can cause significant chronic pain but does not usually affect life expectancy as severely as NF2. With proper management, many live active lives.

Q8: “When should I see a doctor?”

Answer:

* If you experience unexplained chronic pain or numbness.
* If you develop lumps or swelling along nerves.
* If neurological symptoms worsen or new symptoms appear.

**DOCTOR-PATIENT CONVERSATIONS**

Doctor: Hello, I want to talk to you today about a condition called schwannomatosis. It’s a rare genetic disorder where multiple benign nerve tumors, called schwannomas, develop on different nerves in the body.

Patient/Parent: What causes schwannomatosis? Is it inherited?

Doctor: Schwannomatosis is typically caused by changes, or mutations, in specific genes that control nerve cell growth. It is different from other similar conditions like neurofibromatosis type 2, and usually doesn’t have the same pattern of inheritance, but a family history can sometimes play a role. We can discuss genetic testing if that’s appropriate.

Patient/Parent: What symptoms should we expect?

Doctor: The most common symptom is pain caused by the tumors pressing on nerves. People may also have numbness, tingling, or weakness. These tumors are benign, meaning they’re not cancerous, but because they affect nerves, they can cause discomfort or functional issues.

Patient/Parent: How is schwannomatosis diagnosed?

Doctor: Diagnosis usually involves imaging tests like MRI to see the tumors, as well as a thorough neurological exam. Genetic testing may be done to confirm the diagnosis and rule out related conditions. Sometimes it takes some time to diagnose because the symptoms can be similar to other nerve disorders.

Patient/Parent: What treatment options are there?

Doctor: Treatment mostly focuses on managing symptoms. Surgery can be done to remove tumors that are causing severe pain or neurological problems. Pain management includes medications or sometimes other therapies to help control discomfort. Because this is a chronic condition, we will monitor you regularly to watch for new tumors or changes.

Patient/Parent: What is the outlook for someone with this condition?

Doctor: Schwannomatosis is a lifelong condition that varies a lot from person to person. Many people live well with it by managing their symptoms. Surgery generally helps with troublesome tumors, but new tumors can develop, so ongoing care is important. Psychological support and counseling are also helpful for coping with the diagnosis.

Patient/Parent: Should we consider genetic counseling?

Doctor: Yes, genetic counseling can be very useful. It helps understand inheritance risks for family members and guides further testing and family planning decisions.

Patient/Parent: Is there anything specific we should watch for?

Doctor: Yes, if you notice worsening pain, numbness, weakness, or any new neurological symptoms, please let us know promptly. Early evaluation can help manage symptoms better.

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**SKULL BASE TUMORS**

**ALTERNATIVE NAMES:** Skull base tumors are also referred to by several alternative names, including cranial base tumors , skull base cancer , and sometimes specifically named based on their type, such as meningiomas, schwannomas, chordomas, and paragangliomas. Additionally, certain types of skull base tumors have specific names, such as acoustic neuromas (also known as vestibular schwannomas) , glomus tumors , and meningiomas.

**DEFINITION / DESCRIPTION**

Skull base tumors are growths that form at the area of the skull just behind the eyes and nose, beneath the brain. The base of the skull is where important blood vessels and nerves come together with the spinal cord. The base of the skull also separates the brain from the neck.

Tumors also may be called masses, growths or lesions. These tumors can grow inside or outside the skull. They may be benign (noncancerous) or malignant (cancerous). Some skull base tumors may not cause any symptoms, but others can cause symptoms that affect your senses, such as vision or hearing, and the way that your nervous system works.

Even noncancerous skull base tumors can be dangerous. These tumors may press on the blood vessels, nerves and brain tissue in the skull base. This may lead to hearing loss, vision problems, dizziness and other issues that affect safety and well-being.

### **Types of skull base tumors**

Types of skull base tumors are characterized by whether they are cancerous or noncancerous, and where the tumors grow in the base of the skull. Some tumors start growing somewhere else in the body and expand into the skull base.

There are many types of skull base tumors. Some of the more common types of noncancerous, also known as benign, skull base tumors include:

* **Acoustic neuroma.** Also called a vestibular schwannoma, this type of tumor grows on two of the nerves that help with hearing and balance. Acoustic neuromas usually grow slowly.
* **Meningioma.** This type of skull base tumor begins growing in the cells of the membrane lining the brain and spinal cord. This membrane is called the meninges. Rarely, meningiomas may be cancerous and grow aggressively.
* **Pituitary tumor.** This type of tumor forms in a pea-sized gland, called the pituitary gland, found in the skull base.
* **Paraganglioma.** This type of tumor usually grows in the head and neck area. While it's possible for a paraganglioma to be cancerous, it usually is not.
* **Osteoma.** This type of tumor is made of dense bone tissue. Osteomas grow slowly and form on the bones of the skull base. They also may be found in the sinuses.

Other rare types of benign skull base tumors include craniopharyngiomas, granular cell tumors and angiofibromas.

### **Malignant tumors**

Some of the more common types of cancerous, also known as malignant, skull tumors include:

* **Chordomas.** These types of tumors tend to grow slowly, beginning in the bones of the skull base or spine. Chordomas often cause problems with vision and hearing, balance, and headaches.
* **Chondrosarcomas.** These tumors often develop in bones but also may grow in the nearby soft tissue called cartilage. Chondrosarcomas can occur in the bones of the skull base. These tumors usually grow slowly.
* **Carcinomas.** There are several kinds of skull base carcinomas, including adenoid cystic carcinomas, nasopharyngeal carcinomas, squamous cell carcinomas and adenocarcinomas. These tumors may grow in the salivary glands, the nasal cavity, the sinuses, and the head and neck area. Adenoid cystic carcinomas often grow slowly, but they tend to spread to nearby nerves and tissues.
* **Olfactory neuroblastomas.** These are very rare tumors that grow in the nasal cavity. They also are called esthesioneuroblastomas. These tumors can cause nosebleeds and loss of smell and may spread into the areas around the eyes and brain.

**CAUSES**

Experts aren't sure what causes most skull base tumors. Most skull base tumors seem to develop without any clear cause. It's thought that some skull base tumors, particularly acoustic neuromas, likely are caused by genetic conditions passed down in families. But several other factors also may contribute. These factors include exposure to radiation and harmful chemicals.

It's likely that a combination of factors in genes and the environment cause skull base tumors.

**RISK FACTORS**

While experts don't know exactly what causes most skull base tumors, there are several factors that are thought to increase the risk of developing skull base tumors. The risk factors for skull base tumors include:

* **Genetic conditions.** Inheriting certain conditions through changes in genes can cause some types of skull base tumors. These changes in genes can cause the growth of tumor cells. Neurofibromatosis type 1 (NF1) and neurofibromatosis type 2 (NF2) are genetic conditions that can cause acoustic neuromas and other types of skull base tissues.
* **Exposure to certain chemicals.** Prolonged exposure to toxic substances such as vinyl chloride, herbicides and arsenic may increase the risk of developing skull base tumors called carcinomas, particularly in the nasal and sinus areas. Cancer-causing substances such as asbestos, formaldehyde and wood dust have been linked to the development of certain skull base cancers, particularly sinonasal cancers. Exposure to high particulate air pollution — where there are solid particles in the air that may be breathed into the lungs — and radon also are associated with some tumors, especially meningiomas. These chemicals can cause mutations in the cells lining the sinuses and nasal cavity, leading to cancer.
* **History of radiation exposure.** Having prior radiation therapy, especially targeted at the head and neck area, may contribute to the growth of skull base tumors.
* **Frequent sinus infections.** Having sinus infections can be a risk factor for certain types of tumors at the skull base, particularly in the nasal cavity and paranasal sinuses.
* **Hormonal changes.** These are thought to play a role in the development of certain skull base tumors. It's thought that the changes in hormones during puberty may contribute to the growth of some of these tumors. Pituitary gland tumors also are affected by changes in hormones.
* **Family history.** Having a family history of tumors or genetic conditions such as neurofibromatosis, multiple endocrine neoplasia or other hereditary cancer syndromes increases the risk of developing skull base tumors.
* **Previous history of tumors.** If you've previously been diagnosed with a tumor, particularly in the head or neck, you may be at increased risk of developing a secondary skull base tumor. This may be due to factors such as previous radiation treatment or shared risk factors for tumors.
* **Smoking and alcohol consumption.** These factors are linked to an increased risk of certain skull base tumors, such as squamous cell carcinoma of the sinuses or the nasopharynx. The nasaopharynx is the top part of the throat that connects it to the nasal cavity.

It's important to note that having any of these risk factors doesn't mean that you will have a skull base tumor. Not everyone with these risk factors has a skull base tumor. Some people develop tumors without having any known risk factors.

**SIGNS / SYMPTOMS**

Symptoms of skull base tumors are different based on the size of the tumor, the kind of tumor, and where the tumor is growing in the skull base.

Some of the most common symptoms of skull base tumors are:

* Headaches.
* Neck pain.
* Changes in vision — blurry or double vision, or loss of vision.
* Changes in hearing — hearing loss or ringing in the ears.
* Problems with walking and balance.
* Clumsiness or other problems with coordination.
* Hoarse voice.
* Problems with breathing or swallowing.
* Changes in the sense of smell.
* Nosebleeds.

Skull base tumors may not cause any symptoms. Or the symptoms may be mild and general. Some symptoms may be headaches or problems with balance. Skull base tumors may be found by accident during imaging scans or other tests to find the cause of other symptoms.

**DIAGNOSIS METHODS**

Your doctor or other healthcare professional will ask you about your medical history and any symptoms you have. You also will have a physical exam to check your vision, hearing, balance, coordination, sensation, strength and reflexes. This can help your healthcare professional figure out which part of your brain might be affected by the tumor.

Other tests to diagnose skull base tumors may include:

* **Imaging tests, such as a CT scan, MRI or PET scan.** These tests take pictures of your brain and surrounding structures to show the size and location of the tumor. They also can show if there is any pressure or blockage of the cerebrospinal fluid, which is the fluid that surrounds the brain and spinal cord.
* **Biopsy.** In this procedure, a healthcare professional takes a small sample of the tumor and sends it to a lab to be tested. A biopsy is done by removing a small part of your skull and using a needle to take a sample of the tumor.
* **Blood tests.** Your healthcare professional may draw a sample of blood and test your hormone levels and other relevant labs, which may indicate the presence of certain types of tumors.
* **Tests of hearing and vision.** Because skull base tumors can affect hearing and vision, these tests are needed to look for problems with function and to examine the nerves to see if the tumor is compressing the nerves.

Prognosis and life expectancy with skull base tumors is dependent on many factors, such as the type of tumor, tumor grade, location of the tumor, your age and your overall health.

**TREATMENT OPTIONS**

There are several options to treat a skull base tumor. Your healthcare team considers many factors when creating a treatment plan. These factors might include the tumor's location, how fast it's growing, whether it has spread to other parts of the brain. Other factors include the results of tests on the tumor cells, and whether the tumor is causing symptoms. Your care team also considers your age and your overall health.

The goal of treating skull base tumors is to remove the tumor or stop the growth of the tumor without causing damage to the nearby tissues.

Treatment options include:

* **Surgery.** Usually, surgery is the first treatment for skull base tumors. This is done to remove the skull base tumor, either partially or completely. Surgery to remove the tumor may not be the best option for skull base tumors that have more risk of complications with removal. Surgical techniques may include cutting a small section of the skull so that surgeons can access the brain to remove the tumor.  
  There are less invasive surgeries, such as endoscopic surgery. This technique allows surgeons to use a thin tube with a camera through the nostrils. The surgeon uses tiny surgical tools, placed into the nostril with the tube, to remove the tumor.  
  Surgeons may use advanced technologies, such as intraoperative MRI, to guide them during surgery.
* **Radiation therapy.** This type of therapy uses very focused, powerful beams of energy, usually X-rays, to destroy tumors. For skull base tumors, radiation therapy may be used after surgery to destroy any tumor cells not removed by surgery.  
  There are several types of radiation therapy available, including stereotactic radiosurgery. This type of therapy targets skull base tumors with very high doses of energy beams directed at the tumor, sparing nearby cells and tissues from damage. Stereotactic radiosurgery is usually used on small tumors.  
  Another type of radiation therapy is fractionated radiation therapy. Instead of one intense session, smaller doses of energy beams are delivered in several sessions over the course of a few weeks. Fractionated radiation therapy is usually used on larger or more complicated tumors.  
  Proton therapy also may be used to treat skull base tumors. This type of radiation uses particles called protons instead of X-ray (photon) beams. Proton therapy targets the tumor more precisely. It may be used to treat tumors located near the optic nerves, brainstem or other risky areas.
* **Chemotherapy.** This treatment involves very strong medicines designed to kill or stop the growth of tumor cells. There are many different types of chemotherapy medicines. These may be given through a vein or by a pill that you swallow. Chemotherapy may be used after surgery or in addition to other treatments.  
  Chemotherapy is less commonly used for skull base tumors unless they are cancerous and growing very quickly. It may be used in combination with other therapies for cancers such as sinonasal cancers.
* **Watchful waiting.** Some people may not need treatment for certain skull base tumors. These tumors may include small, noncancerous tumors that don't cause any symptoms. Instead, your healthcare professional may suggest monitoring the tumor with regular imaging tests to check for changes.

### **Potential future treatments**

Ongoing research in cancer treatment and neurosurgery may lead to potential future treatments for skull base tumors. While current treatment methods such as surgery, radiation and chemotherapy remain the standard methods, advances in technology, drug development and precision medicine offer new possibilities for more effective and less invasive treatments.

Gene therapies and immunotherapy show potential, as well as medicines that target and destroy specific cells in the skull base tumor. Researchers also are exploring new ways to use existing techniques, including proton beam therapy and endoscopic surgery.

**Alternative medicine**

There are no alternative medicines that can cure skull base tumors. But some complementary and alternative treatments may help with common tumor symptoms. These may include pain, fatigue and stress.

Some alternative and complementary therapies that you may find helpful include:

* Meditation, guided imagery and other mindfulness practices.
* Yoga.
* Relaxation exercises.
* Acupuncture.
* Massage therapy.
* Physical activity.

If you're interested in alternative medicines and complementary therapies for skull base tumors, talk about it with your healthcare team. Ask about the benefits and potential risks. And discuss with your care team what options might be helpful and safe for you.

**POSSIBLE COMPLICATIONS**

Complications of skull base tumors are usually caused by the tumors growing and pressing into blood vessels, nerves and other tissues, including the brain.

Complications of the nervous system may include:

* Problems with vision.
* Hearing loss.
* Weakness or numbness in the face.
* Difficulty swallowing or speaking.
* Difficulty breathing.
* Seizures.
* Headaches.
* Cerebrospinal fluid leaks.
* Water buildup on the brain, which can increase pressure inside the skull and cause headaches, nausea and vomiting.

Other complications may include:

* Changes in hormone levels.
* Infections, such as meningitis.
* Problems with memory, concentration, mood or behavior.
* Problems with balance, coordination and walking.
* Stroke or bleeding in the brain.
* Problems with the eustachian tube in the ear.

Sometimes skull base tumors can spread. Noncancerous skull base tumors generally do not spread. Cancerous skull base tumors can grow into nearby tissues and spread to the lymph nodes, lungs, liver or other parts of the body. Chordomas, chondrosarcomas, sinonasal carcinomas and olfactory neuroblastomas are more likely than other types of skull base tumors to spread throughout the body.

Whether or not skull base tumors spread, and how quickly they do so, depends on the type and grade of the tumor. High-grade cancerous tumors are more aggressive. They likely will spread more quickly than low-grade tumors. The grade of a tumor shows how different the tumor cells are from healthy cells. High-grade tumors have irregular cells and tend to grow and spread very quickly.

**WHEN TO SEE A DOCTOR / RED FLAG**

* Persistent or severe headaches
* Neck pain
* Changes in vision such as blurriness, double vision, or vision loss
* Changes in hearing like hearing loss or ringing in the ears (tinnitus)
* Problems with balance, dizziness, or clumsiness
* Facial pain, numbness, weakness, or paralysis
* Hoarseness or changes in voice
* Difficulty swallowing or breathing
* Nosebleeds or frequent sinus problems
* Loss or alteration of sense of smell
* Lumps or swelling in the neck or face
* Severe or worsening neurological symptoms such as weakness or sensory changes

**DIFFERENTIAL DIAGNOSIS**

## Skull Base Tumors Differential Diagnosis List

| **Tumor Type** | **Key Features / Notes** |
| --- | --- |
| Chordoma | Malignant tumor from notochord remnants; typically midline clivus region; locally aggressive. |
| Chondrosarcoma | Malignant cartilage-forming tumor; usually off-midline petroclival area; may be confused with chordoma. |
| Schwannoma | Benign nerve sheath tumor; most commonly vestibular schwannoma (acoustic neuroma) at cerebellopontine angle (CPA); also trigeminal and facial nerve schwannomas. |
| Meningioma | Usually benign, slow-growing tumor arising from meninges; presents with characteristic imaging features like dural tail. |
| Epidermoid cyst / Cholesteatoma | Benign cystic lesions predominantly in CPA or petrous apex areas; slow growing. |
| Plasmacytoma / Multiple Myeloma | Plasma cell tumors affecting bone, including skull base; typically lytic lesions. |
| Langerhans cell histiocytosis | Rare lesion affecting bone in children/young adults; can involve skull base. |
| Osteosarcoma | Primary malignant bone tumor, rare in skull base, aggressive. |
| Giant cell tumor | Occasionally affects temporal bone/skull base, usually benign but locally aggressive. |
| Fibrous dysplasia | Benign fibro-osseous lesion with ground-glass appearance on imaging, can mimic tumors. |
| Metastases | Secondary tumors commonly from breast, lung, prostate cancers involving skull base bone and soft tissue. |
| Lymphoma | Can involve skull base as soft tissue mass; mimics other malignancies. |
| Pituitary adenoma | Tumors of pituitary gland; may extend into skull base (sellar/suprasellar region). |
| Rhabdomyosarcoma | Malignant soft tissue tumor mostly in children; can involve skull base. |
| Paraganglioma (glomus tumors) | Vascular tumors often at jugular foramen or carotid body near skull base. |
| Sinonasal carcinomas / Esthesioneuroblastoma | Malignant tumors of sinonasal region encroaching skull base anteriorly. |

**EPIDEMIOLOGY**

### Frequency

See the list below:

* CPA tumors other than vestibular schwannoma (percentage of total CPA occurrence)
  + Meningiomas (3-13%)
  + Epidermoids (2-6%)
  + Facial and lower cranial nerve schwannomas (1-2%)
  + Arachnoid cysts (1%)
* Rare tumors
  + Lipomas
  + Dermoid tumors
  + Neuroepithelial cysts
  + Trigeminal schwannoma
  + Endolymphatic sac carcinomas
  + Ependymoma
  + Brainstem glioma
  + Astrocytoma
  + Medulloblastoma
  + Choroid plexus papilloma
  + Large or giant aneurysms of anterior inferior cerebellar artery (AICA), superior cerebellar artery (SCA), or the midbasilar artery
  + Brainstem arteriovenous malformation (AVM)
  + Brainstem cavernous malformation originating in the brain stem (may reach a pial surface along the CPA)
  + Petroclival chondrosarcoma
  + Clivus chordoma (usually arise in midline but may extend laterally into the CPA)
  + Osteosarcoma of the lateral skull base
  + Metastatic tumors (can reach CPA by hematogenous spread or direct extension)
  + Granulomatous inflammatory masses

**PREDEFINED Q & A SETS**

Q1: “What are skull base tumors?”

Skull base tumors are abnormal growths that develop at the bottom of the skull, an area housing many important nerves, blood vessels, and brain structures. They can be benign (noncancerous) or malignant (cancerous) and arise from bone, cartilage, nerve tissue, or surrounding soft tissues.

Q2: “What causes skull base tumors?”

The exact causes are often unknown. Some tumors, like acoustic neuromas, may be linked to genetic conditions inherited in families. Other contributing factors might include exposure to radiation or harmful chemicals. Most skull base tumors arise spontaneously without clear cause.

Q3: “What types of skull base tumors are there?”

The tumors vary by location within the skull base and tissue origin. Common types include:

* Vestibular schwannoma (acoustic neuroma): A benign tumor on the nerve from the inner ear to the brain, causing hearing loss and balance problems.
* Meningioma: Usually benign tumor from membranes covering the brain and spinal cord.
* Chordoma: Malignant tumor arising from notochord remnants, often midline in the clivus bone.
* Chondrosarcoma: Malignant cartilage tumor.
* Paraganglioma (glomus tumors): Usually benign tumors near jugular veins, can cause pulsatile tinnitus.
* Pituitary tumors, craniopharyngiomas, Rathke’s cleft cysts: Tumors in the middle skull base near the pituitary gland affecting hormonal function.
* Others: Osteosarcoma, lymphoma, epidermoid cysts, and metastatic tumors.

Q4: “What symptoms do skull base tumors cause?”

Symptoms depend on tumor size and location, and may develop gradually, including:

* Headaches and neck pain
* Changes in vision (blurred or double vision)
* Hearing loss or ringing in ears
* Balance or coordination problems
* Facial numbness or weakness
* Difficulty swallowing or speaking
* Persistent sinus congestion or nosebleeds
* Hoarseness or voice changes

Q5: “How are skull base tumors diagnosed?”

Answer:

* Medical history and neurological exam to assess symptoms and nerve function
* Imaging tests: MRI and CT scans are critical to locate tumors, assess extent, and plan treatment. MRI is especially useful for soft tissue detail; CT shows bone involvement.
* Biopsy: May be needed in some cases to confirm tumor type.

Q6: “What treatments are available?”

Answer:

* Surgery: The mainstay treatment aiming to remove or reduce tumor size while preserving neurological function. Surgical approaches depend on tumor location.
* Radiation therapy: Often used after surgery or for tumors that cannot be fully removed surgically. Techniques include conventional radiation and focused methods like stereotactic radiosurgery.
* Chemotherapy: Less commonly used but may be employed for certain malignant tumors.
* Symptom management: Pain control, physical therapy, and supportive care.

Q7: “What is the prognosis?”

Outcomes vary widely based on tumor type, size, location, and whether the tumor is benign or malignant. Early diagnosis and treatment improve outcomes. Many benign tumors have excellent prognosis, while malignant or aggressive tumors require comprehensive care and may have more guarded outlooks.

**DOCTOR-PATIENT CONVERSATIONS**

Doctor: Hello. I’d like to talk with you about the results of your imaging and explain what a skull base tumor is.

Patient/Parent: Yes, please. I don’t really understand what that means.

Doctor: The skull base is the area at the bottom of your skull near the brain and includes many important nerves and blood vessels. A skull base tumor is an abnormal growth that develops in this area. These tumors can be benign, meaning non-cancerous, or malignant, which means cancerous, and they can affect nearby structures causing various symptoms.

Patient/Parent: What kind of symptoms should we expect?

Doctor: Symptoms vary depending on the tumor’s location and size. Common signs include headaches, changes in your vision or hearing, facial numbness or weakness, difficulty swallowing or speaking, dizziness, or sometimes swelling in the neck or face. Sometimes, tumors are found before symptoms appear during scans for other reasons.

Patient/Parent: How do you find out exactly what it is?

Doctor: Usually, we begin with imaging tests like CT scans or MRIs to see the tumor’s size and exact location. Sometimes, we need a biopsy, which is a small tissue sample, to determine the exact type of tumor. This helps us plan the best treatment.

Patient/Parent: What treatment options are there? Is it dangerous?

Doctor: Treatment depends on the type of tumor, its size, symptoms, and overall health. Options include surgery to remove the tumor, radiation therapy, and sometimes chemotherapy. Some benign tumors may be watched over time if they aren’t causing problems. Early diagnosis and treatment usually give the best outcomes, but because this area is so delicate, management is always carefully planned by a team of specialists.

Patient/Parent: Will the tumor come back?

Doctor: Some tumors can recur, so regular follow-up with imaging is important. If the tumor comes back or grows, we reassess and may need additional treatment.

Patient/Parent: What should we watch out for at home?

Doctor: Watch for any new or worsening symptoms like increased headaches, changes in vision or hearing, facial weakness, or difficulty swallowing. If these happen, please seek medical care promptly.

Patient/Parent: How can we handle it emotionally?

Doctor: It’s normal to feel overwhelmed. We have support services including counseling and patient support groups to help you and your family cope. We will work closely with you every step of the way.

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**SINUS INFECTION**

**ALTERNATIVE NAMES:** Alternative names for a sinus infection include sinusitis, chronic rhinosinusitis without nasal polyps, chronic rhinosinusitis with nasal polyps, and rhinosinusitis. Other terms used to refer to a sinus infection are sinus infection, nasal inflammation, stuffy nose, hay fever, allergic rhinitis, head cold, and nasal catarrh.

**DEFINITION / DESCRIPTION**

Sinusitis is an inflammation, or swelling, of the tissue lining your sinuses. Sinuses are structures inside your face that are normally filled with air. Bacterial infections, viral infections and allergies can irritate them, causing them to get blocked and filled with fluid. This can cause pressure and pain in your face, nasal congestion (a stuffy nose) and other symptoms.

Sinusitis is also sometimes called rhinosinusitis.

#### **Sinuses**

Your sinuses are four paired cavities (spaces) in your head. Narrow passages connect them. Sinuses make mucus that drains out of the passages in your nose. This drainage helps keep your nose clean and free of bacteria, allergens and other germs (pathogens).

#### **Types of sinusitis**

We describe types of sinusitis based on how long it’s been going on (acute, subacute, chronic or recurrent) and what’s causing it (bacteria, virus or fungus).

##### **Acute, subacute, chronic and recurrent sinusitis**

* Acute sinusitis symptoms (nasal congestion, drainage, facial pain/pressure and decreased sense of smell) last less than four weeks. It’s usually caused by viruses like the common cold.
* Subacute sinusitis symptoms last four to 12 weeks.
* Chronic sinusitis symptoms last at least 12 weeks. Bacteria are usually the cause.
* Recurrent acute sinusitis symptoms come back four or more times in one year and last less than two weeks each time.

##### **Bacterial and viral sinusitis**

Viruses, like the ones that cause the common cold, cause most cases of sinusitis. Bacteria can cause sinusitis, or they can infect you after a case of viral sinusitis. If you have a runny nose, stuffy nose and facial pain that don’t go away after ten days, you might have bacterial sinusitis. Your symptoms may seem to improve but then return and are worse than the initial symptoms. Antibiotics and decongestants usually work well on bacterial sinusitis.

##### **Fungal sinusitis**

Sinus infections caused by fungus are usually more serious than other forms of sinusitis. They’re more likely to happen if you have a weakened immune system.

### **How do I know if I have sinusitis, COVID, a cold or an allergy?**

Colds, COVID-19, allergies and sinus infections all have similar symptoms. It can be difficult to tell them apart. The common cold typically builds, peaks and slowly disappears. It lasts a few days to a week. Nasal allergies cause sneezing, itchy nose and eyes, congestion, runny nose and postnasal drip (mucus in your throat). They usually don’t cause the facial pain that sinus infections do. COVID-19 can cause additional symptoms, like fever and shortness of breath.

A cold, COVID or allergies can all cause sinus infections. You can test yourself or have a provider test you for some viral infections, like COVID-19 and the flu.

**CAUSES**

Viruses, bacteria, fungi and allergens can cause sinusitis. Specific triggers for sinusitis include:

* The common cold.
* The flu (influenza).
* *Streptococcus pneumoniae* bacteria.
* *Haemophilus influenza* bacteria.
* *Moraxella catarrhalis* bacteria.
* Nasal and seasonal allergies.

#### **Risk factors for sinusitis**

Some people are more likely to get sinusitis than others. Risk factors include:

* Nasal allergies.
* Asthma.
* Nasal polyps (growths).
* Deviated septum. Your septum is a line of tissue that divides your nose. A deviated septum isn’t straight, narrowing the passage on one side of your nose. This can cause a blockage.
* A weakened immune system. This can be from illnesses like HIV or cancer, or from certain medications.
* Smoking.

#### **Is sinusitis contagious?**

Sinusitis itself isn’t contagious. But the viruses and bacteria that can cause it are. Remember to follow good handwashing practices, avoid other people if you’re sick and sneeze or cough into your elbow.

### **What happens if sinusitis is left untreated?**

You don’t necessarily need to treat sinusitis — it often goes away on its own. Very rarely, untreated sinus infections can lead to life-threatening infections. This happens if bacteria or fungi spread to your brain, eyes or nearby bone.

**SIGNS / SYMPTOMS**

Common symptoms of a sinus infection include:

* Postnasal drip (mucus dripping down your throat).
* Runny nose with thick yellow or green mucus.
* Stuffy nose.
* Facial pressure (particularly around your nose, eyes and forehead). This might get worse when you move your head around or bend over.
* Pressure or pain in your teeth.
* Ear pressure or pain.
* Fever.
* Bad breath (halitosis) or a bad taste in your mouth.
* Cough.
* Headache.
* Tiredness.

**DIAGNOSIS METHODS**

Healthcare providers diagnose sinusitis based on your symptoms and health history. A provider will check your ears, nose and throat for swelling, draining or blockage. They might use an endoscope (a small, lighted instrument) to look inside your nose.

A primary care provider may also refer you to a specialist, like an otolaryngologist (also called an ENT — an ear, nose and throat specialist).

#### **Specific tests to diagnose sinusitis**

Specific tests your provider might order to diagnose sinus infection include:

* Nasal endoscopy.
* Nasal swabs. Your provider may use a soft-tipped stick to get a fluid sample from your nose. They’ll test it for viruses or other germs that might be causing your symptoms.
* Imaging. In some cases, your provider might order a computed tomography (CT) scan to better understand what’s happening inside your sinuses.
* Allergy testing. If you have chronic sinusitis, your provider may test you for allergies that could be triggering it.
* Biopsy. Rarely, a provider may take a tissue sample from your nose for testing.

**TREATMENT OPTIONS**

There are many treatment options for sinusitis, depending on your symptoms and how long you’ve had them. You can treat a sinus infection at home with:

* Decongestants.
* Over-the-counter (OTC) cold and allergy medications.
* Nasal saline rinses.
* Drinking plenty of fluids.

If symptoms of sinusitis don’t improve after 10 days, a provider may prescribe:

* Antibiotics.
* Oral or topical decongestants.
* Prescription intranasal steroid sprays. (Don’t use nonprescription sprays or drops for longer than three to five days — they may actually increase congestion.)

Providers treat chronic sinusitis by focusing on the underlying condition. Treatments can include:

* Intranasal steroid sprays.
* Topical antihistamine sprays or oral pills.
* Leukotriene antagonists, like montelukast.
* Surgery to treat structural issues, polyps or fungal infections.

#### **Medications for a sinus infection**

If you need an antibiotic, which one your provider prescribes depends on your specific situation. Some options include:

* Augmentin (amoxicillin/clavulanate).
* Amoxicillin.
* Doxycycline.
* Levofloxacin.
* Cefixime.
* Cefpodoxime.
* Clindamycin.

#### **Are complementary and alternative therapies useful for treating sinusitis?**

You might find acupressure, acupuncture or facial massage helpful in reducing symptoms of sinusitis, including draining, pressure and pain. Ask a provider if these therapies might help in your specific case.

#### **Do I need antibiotics for every sinus infection?**

No. Providers often wait to see how long your symptoms last before prescribing antibiotics. Many sinus infections are caused by viruses. You can’t cure viral infections with antibiotics. Overusing antibiotics or using them to treat viral infections can lead to unnecessary side effects or antibiotic resistance. This may make future infections harder to treat.

**PREVENTION TIPS**

Depending on the cause, there are a few ways to reduce your risk of getting sinus infections, including:

* Rinsing your nose with saline (salt water) as directed by your provider.
* Taking steps to prevent allergies. This includes medications, allergy shots and avoiding your known allergy triggers (like dust, pollen or smoke).
* Using steroid nasal sprays if your provider recommends them.
* Establishing good handwashing and other habits that reduce your risk of getting sick with infectious diseases.
* Avoiding smoke. There are ways to help you quit smoking, if you do.

**OUTLOOK / PROGNOSIS**

Sinusitis usually only lasts a week to 10 days. You can usually treat it with over-the-counter medicines and at-home treatments. If you have chronic sinusitis or if it keeps coming back, there may be underlying causes that you’ll need to treat.

**WHEN TO SEE A DOCTOR / RED FLAG**

You can usually care for sinus conditions on your own. But if you continue to have symptoms that concern you or if you get infections repeatedly, talk to a healthcare provider. They can help you understand your next steps.

Go to the nearest emergency room or seek medical attention right away if you experience symptoms of a serious infection, including:

* High fever (over 103 degrees Fahrenheit/40 degrees Celsius).
* Confusion or other mental changes.
* Vision changes, especially if you have pain or swelling around your eyes.
* Seizures.
* Stiff neck.

**DIFFERENTIAL DIAGNOSIS**

* Acute Viral Rhinosinusitis  
  Most common causes; symptoms usually resolve within 7-10 days without antibiotics. It often follows a cold.
* Acute Bacterial Sinusitis  
  Symptoms persist beyond 10 days or worsen after initial improvement, with localized pain, purulent nasal discharge, and sometimes fever.
* Allergic Rhinitis  
  Nasal congestion, sneezing, itchy nose/eyes, clear nasal discharge; usually related to allergies without purulent discharge.
* Chronic Sinusitis  
  Symptoms last more than 12 weeks; may involve persistent inflammation or infection, sometimes with nasal polyps.
* Dental Abscess / Periapical Abscess  
  Can mimic maxillary sinusitis due to pain and swelling near the upper teeth; dental examination and imaging are needed.
* Nasal Polyps / Sinonasal Polyposis  
  Chronic nasal obstruction and congestion, often associated with chronic sinusitis or allergic rhinitis; may cause loss of smell (anosmia).
* Migraine or Other Headache Syndromes  
  Facial pain or headache without nasal signs; features of migraine or tension headaches.
* Facial Pain Syndromes (e.g., Trigeminal Neuralgia)  
  Sharp, shooting facial pain unrelated to sinus inflammation or discharge.
* Nasal Foreign Body  
  Especially in children; unilateral foul-smelling nasal discharge and obstruction.
* Structural Abnormalities  
  Deviated nasal septum, bone spurs, or anatomic variations causing nasal obstruction and secondary sinus issues.
* Fungal Sinusitis  
  Less common; chronic symptoms especially in immunocompromised patients; diagnosed with imaging and fungal cultures.
* Upper Respiratory Tract Infection (common cold)  
  Overlapping symptoms but typically includes cough, sore throat, and systemic viral illness features.

**EPIDEMIOLOGY**

* In the United States, approximately 28.9 million adults (around 11.6%) are diagnosed with sinusitis, with over 30 million diagnosed annually overall.
* Sinusitis affects about 1 in 7 adults annually, leading to around 16 million physician office visits per year.
* Acute bacterial rhinosinusitis (ABRS) accounts for roughly 16% of adults diagnosed annually, while viral sinus infections are more common but less likely to require antibiotics.
* The prevalence of chronic sinusitis is also significant, with millions of visits to healthcare providers each year (about 2.7 million visits for chronic sinusitis in 2018).
* Sinus infections show a seasonal pattern, being more common from early fall to early spring, which correlates with the typical cold and flu seasons.
* Women tend to have more episodes of sinus infection than men, possibly due to greater exposure to children who commonly carry respiratory infections.
* Children typically have multiple colds per year (~6-8), and about 6-13% of viral upper respiratory infections in children can develop into bacterial sinusitis.
* Globally, the prevalence varies, with acute sinusitis affecting 3 per 1,000 people and chronic sinusitis about 1 per 1,000 in some populations like the UK.
* Sinusitis leads to substantial healthcare burden and costs, estimated in the US at billions of dollars annually

**PREDEFINED Q & A SETS**

## Q1: “What is a sinus infection?”

A sinus infection, or sinusitis, is an inflammation or infection of the sinus cavities—air-filled spaces in the bones around your nose and eyes. It can be caused by viruses, bacteria, fungi, allergies, or other irritants.

## Q2: “What causes a sinus infection?”

Answer:

* Most commonly caused by viruses (like the common cold).
* Bacterial infections can develop if mucus buildup blocks the sinuses.
* Allergies or nasal structural problems (like a deviated septum or nasal polyps) can also trigger inflammation.
* Fungal infections occur rarely, mainly in immunocompromised people.

Q3: “What are the symptoms of a sinus infection?”

Answer:

* Facial pain or pressure, especially around cheeks, forehead, or between the eyes.
* Nasal congestion or stuffiness.
* Thick, discolored nasal discharge (green or yellow).
* Decreased sense of smell or taste.
* Headache.
* Cough, which may worsen at night.
* Fatigue, bad breath, or dental pain.
* Fever may occur in bacterial sinusitis.

Q4: “How is a sinus infection diagnosed?”

Answer:

* Based on symptom history and physical examination.
* Nasal endoscopy or imaging (CT scan) may be used in chronic or complicated cases.
* Distinguishing viral from bacterial sinusitis relies mostly on symptom duration and severity.

Q5: “How long does a sinus infection last?”

Answer:

* Viral sinus infections usually improve within 7 to 10 days.
* Bacterial infections may last longer and can worsen after 5, then persist beyond 10 days.
* Chronic sinusitis lasts more than 12 weeks and may require further evaluation.

Q6: “How is a sinus infection treated?”

Answer:

* Viral sinusitis often resolves without antibiotics.
* Supportive care includes nasal saline irrigation, decongestants, pain relievers, and rest.
* Antibiotics are prescribed if bacterial infection is suspected or confirmed (symptoms worsen after 5–10 days or are severe).
* Chronic or recurrent cases may require specialist evaluation, nasal steroids, or surgery.

Q7: “Can sinus infections lead to complications?”

Answer:

* Yes, untreated bacterial sinus infections can sometimes spread to nearby structures causing ear infections, orbital cellulitis, or rarely brain abscess.
* Prompt medical attention is needed for severe symptoms like vision changes, severe headache, swelling around eyes, or neurological signs.

Q8: “When should I see a doctor?”

Answer:

* If symptoms last longer than 10 days without improvement.
* If symptoms worsen after initial improvement.
* If you have high fever, severe facial pain, swelling around the eyes, vision changes, or confusion.
* If sinus infections recur frequently or become chronic.

Q9: “How can I prevent sinus infections?”

Answer:

* Manage allergies and avoid triggers.
* Practice good hand hygiene to reduce viral infections.
* Use humidifiers or nasal saline sprays as needed.
* Avoid smoking and irritants that dry the nasal passages.

**DOCTOR-PATIENT CONVERSATIONS**

Doctor: Hello! It looks like you have symptoms of a sinus infection, which is an inflammation or infection of the sinus cavities around your nose and eyes.

Patient: What causes sinus infections?

Doctor: Sinus infections often start as a viral cold or allergy that causes swelling and blockage in the sinus openings. This can trap mucus and sometimes lead to a bacterial infection, though most sinus infections are viral and improve on their own.

Patient: What symptoms should I expect?

Doctor: Common symptoms include nasal congestion, thick nasal discharge, facial pain or pressure—especially around the cheeks, forehead, or between the eyes—headache, and sometimes fever. You might also notice a decreased sense of smell.

Patient: How should I treat it?

Doctor: For most sinus infections, treatment is about relieving symptoms. That includes rest, staying well hydrated, using saline nasal sprays or rinses to clear mucus, warm compresses to reduce facial pain, and over-the-counter pain relievers like acetaminophen or ibuprofen. If your symptoms last more than 10 days or worsen after initial improvement, antibiotics might be necessary if a bacterial infection is suspected.

Patient: When should I come back or see a doctor urgently?

Doctor: You should come back if you have:

* High fever lasting more than a few days
* Severe facial pain or swelling
* Difficulty breathing or vision changes
* Symptoms that last longer than 10 days without improvement
* Or if the sinus infection keeps coming back

These could signal complications or the need for specialized care.

Patient: Can sinus infections go away without antibiotics?

Doctor: Yes, most sinus infections are viral and get better without antibiotics. Using antibiotics unnecessarily can cause side effects and antibiotic resistance. We reserve antibiotics for cases where bacterial infection is clear or symptoms worsen.

Patient: Is there anything I should do at home to feel better?

Doctor: Yes, nasal saline rinses can help clear out mucus. Avoid smoke and allergens, rest well, use warm compresses, and take pain relievers if needed. Don’t use nasal decongestant sprays for more than three days to avoid rebound swelling.

*REFERENCES:*

<https://www.cdc.gov/nchs/fastats/sinuses.htm>

<https://emedicine.medscape.com/article/232670-overview>

[Sinus Infection (Sinusitis): Causes, Symptoms & Treatment](https://my.clevelandclinic.org/health/diseases/17701-sinusitis#overview)

**STREP THROAT**

**ALTERNATIVE NAMES:** Strep throat is also known by several alternative names, including streptococcal pharyngitis , group A strep infection , and streptococcus throat infection. Additionally, it is sometimes referred to as strep throat.

**DEFINITION / DESCRIPTION**

Strep throat is a bacterial infection that can make your throat feel sore and scratchy. Strep throat accounts for only a small portion of sore throats.

If untreated, strep throat can cause complications, such as kidney inflammation or rheumatic fever. Rheumatic fever can lead to painful and inflamed joints, a specific type of rash, or heart valve damage.

Strep throat is most common in children, but it affects people of all ages. If you or your child has signs or symptoms of strep throat, see your doctor for prompt testing and treatment.

**CAUSES**

Strep throat is caused by infection with a bacterium known as Streptococcus pyogenes, also called group A streptococcus.

Streptococcal bacteria are contagious. They can spread through droplets when someone with the infection coughs or sneezes, or through shared food or drinks. You can also pick up the bacteria from a doorknob or other surface and transfer them to your nose, mouth or eyes.

**RISK FACTORS**

Several factors can increase your risk of strep throat infection:

* Young age. Strep throat occurs most commonly in children.
* Time of year. Although strep throat can occur anytime, it tends to circulate in winter and early spring. Strep bacteria flourish wherever groups of people are in close contact.

**SIGNS / SYMPTOMS**

Signs and symptoms of strep throat can include:

* Throat pain that usually comes on quickly
* Painful swallowing
* Red and swollen tonsils, sometimes with white patches or streaks of pus
* Tiny red spots on the area at the back of the roof of the mouth (soft or hard palate)
* Swollen, tender lymph nodes in your neck
* Fever
* Headache
* Rash
* Nausea or vomiting, especially in younger children
* Body aches

It's possible for you or your child to have many of these signs and symptoms but not have strep throat. The cause of these signs and symptoms could be a viral infection or some other illness. That's why your doctor generally tests specifically for strep throat.

It's also possible for you to be exposed to a person who carries strep but shows no symptoms.

**DIAGNOSIS METHODS**

If you think you may have strep throat, you should see a healthcare provider. The provider will ask about your symptoms and perform a physical exam. They may also give you a strep test. Viral illnesses can have the same symptoms as strep throat. That’s why it’s important that you take a strep test to confirm the presence of group A *Streptococcus* bacteria in your throat.

### **What is a strep test?**

A strep throat test will check for group A *Streptococcus* bacteria in your throat. The test is painless — although it can be uncomfortable for some — and takes very little time. A healthcare provider will use the tip of a specialized long cotton swab to wipe the back of your throat. Then, the provider will either use the swab to perform a rapid strep test or send the swab to a laboratory for testing (throat culture).

A rapid strep test takes about 20 minutes. If the test is positive (the swab contained *Streptococcus* bacteria), you have strep throat. If the test is negative (no signs of *Streptococcus* on the swab), you may not have strep throat. However, the provider may send the throat swab to a laboratory to double-check the results with a throat culture.

With a throat culture, a lab technician smears your swab onto a culture dish. It takes one to two days to see if *Streptococcus* bacteria grow from it. Throat cultures can sometimes find bacteria that rapid strep tests miss.

**TREATMENT OPTIONS**

Strep throat treatment includes antibiotics. An antibiotic is a type of medicine that kills the bacteria that cause an infection.

Penicillin and amoxicillin are common antibiotics healthcare providers use to treat strep throat. If you’re allergic to penicillin, the provider can prescribe another antibiotic.

A healthcare provider may give you an antibiotic shot, or they may prescribe an antibiotic in either pill or liquid form. You’ll usually take the pills or liquid for 10 days. Follow your provider’s instructions. You should take all of the medication, even if you feel better. The bacteria can still be alive even if you’re feeling well.

#### **How can I get rid of strep throat overnight?**

Unfortunately, you can’t cure strep throat overnight. A type of bacteria causes strep throat. Therefore, you need an antibiotic to clear it up. After starting on an antibiotic, you should start to feel better within a day or two. Until then, there are things you can do to manage your symptoms, such as drinking warm liquids and taking pain relievers.

**PREVENTION TIPS**

To prevent strep infection:

* **Wash your hands.** Proper hand-washing is the best way to prevent all kinds of infections. That's why it's important to wash your own hands regularly with soap and water for at least 20 seconds. Teach your children how to wash their hands properly using soap and water or to use an alcohol-based hand sanitizer if there is no soap and water available.
* **Cover your mouth.** Teach your children to cover their mouths with an elbow or tissue when they cough or sneeze.
* **Don't share personal items.** Don't share drinking glasses or eating utensils. Wash dishes in hot, soapy water or in a dishwasher.

**OUTLOOK / PROGNOSIS**

You should start to feel better within a day or two of beginning an antibiotic. Most symptoms of strep throat should be gone within seven to 10 days.

***When can I go back to work/school?***

If you don’t have a fever, you can return to regular activities 24 hours after taking your first dose of an antibiotic.

***Does strep throat go away on its own?***

No, strep throat won’t go away on its own. A type of bacteria causes strep throat, and antibiotics treat infections caused by bacteria. Strep throat can lead to more serious illnesses if not treated, so it’s important to start on antibiotics immediately.

**POSSIBLE COMPLICATIONS**

Strep throat can lead to serious complications. Antibiotic treatment reduces the risk.

### **Spread of infection**

Strep bacteria may spread, causing infection in:

* Tonsils
* Sinuses
* Skin
* Blood
* Middle ear

### **Inflammatory reactions**

Strep infection may lead to inflammatory illnesses, including:

* Scarlet fever, a streptococcal infection characterized by a prominent rash
* Inflammation of the kidney (poststreptococcal glomerulonephritis)
* Rheumatic fever, a serious inflammatory condition that can affect the heart, joints, nervous system and skin
* Poststreptococcal reactive arthritis, a condition that causes inflammation of the joints

A possible relationship has been suggested between strep infection and a rare condition called pediatric autoimmune neuropsychiatric disorder associated with group A streptococci (PANDAS). Children with this condition experience worsened symptoms of neuropsychiatric conditions, such as obsessive-compulsive disorder or tic disorders, with strep. This relationship currently remains unproved and controversial.

**WHEN TO SEE A DOCTOR / RED FLAG**

Call your doctor if you or your child has any of these signs and symptoms:

* A sore throat accompanied by tender, swollen lymph glands
* A sore throat that lasts longer than 48 hours
* A fever
* A sore throat accompanied by a rash
* Problems breathing or swallowing
* If strep has been diagnosed, a lack of improvement after taking antibiotics for 48 hours

**DIFFERENTIAL DIAGNOSIS**

* Other causes of pharyngitis:
  + *Viral pharyngitis* (most common cause overall): caused by viruses such as adenovirus, rhinovirus, coronavirus, influenza, Epstein-Barr virus (EBV, infectious mononucleosis), cytomegalovirus, herpes simplex virus.
  + *Group C streptococcal pharyngitis* (less common bacterial cause).
  + *Neisseria gonorrhoeae* pharyngitis (gonococcal pharyngitis).
  + *Corynebacterium diphtheriae* (diphtheria).
* Abscesses and deep infections:
  + *Peritonsillar abscess* (quinsy): unilateral throat pain, muffled voice, "hot potato" voice, trismus.
  + *Retropharyngeal abscess*: more common in children; may present with neck stiffness and respiratory distress.
  + *Epiglottitis*: severe sore throat, difficulty swallowing, drooling, stridor; medical emergency.
* Infectious mononucleosis (EBV infection):
  + Pharyngitis with marked lymphadenopathy, fatigue, splenomegaly, and sometimes rash.
  + May have overlapping symptoms with strep throat but often has cough and conjunctivitis absent in strep.
* Other viral infections:
  + Coxsackie virus (herpangina), influenza, and others.
* Non-infectious causes:
  + Allergic rhinitis with postnasal drip.
  + Gastroesophageal reflux disease (GERD).
  + Foreign bodies or trauma to the throat.
  + Neoplasms of head and neck presenting with sore throat.
  + Referred pain from dental abscess or otitis media.

**EPIDEMIOLOGY**

* In the United States, acute pharyngitis results in approximately 12 million outpatient visits annually, making it one of the most common primary diagnoses.
* GAS pharyngitis is more common in children aged 3 to 15 years, with the highest incidence typically in school-age children, especially those 5-15 years old.
* The infection exhibits a seasonal pattern, occurring most frequently in winter and early spring in temperate climates.
* Globally, there are an estimated 616 million cases of GAS pharyngitis annually.
* Children with GAS pharyngitis typically experience symptoms for an average of around 4.5 days, with about 1.9 days of school absence per episode and parents missing about 1.8 days of work caring for the child.
* The prevalence of asymptomatic GAS carriage in children is approximately 12%, while the prevalence of streptococcal infection among symptomatic children presenting with sore throat is around 37%.
* Complications such as rheumatic heart disease (RHD) remain a significant consequence primarily in developing countries, causing substantial morbidity and mortality globally

**PREDEFINED Q & A SETS**

#### **What does strep throat look like?**

If you have strep throat, your throat and tonsils may appear red, sore and swollen. You may also have white patches, spots or streaks of pus on your throat and tonsils. In addition, you may develop tiny, red spots on the roof of your mouth called petechiae.

Depending on the strain of bacteria, you may also develop a strep throat rash known as scarlet fever. It shows up on your neck and chest first, but it may spread to other parts of your body.

Group A *Streptococcus* bacteria can also infect your skin and cause sores. When this occurs, the infection is called impetigo.

#### **What does strep throat feel like?**

Strep throat is typically a mild condition, but the infection can be very painful. Your sore throat may be severe and very uncomfortable. The lymph nodes in your neck may be very tender and swollen. You may have pain when swallowing. If you have a sore throat rash, it may feel rough like sandpaper.

### **How do you get strep throat?**

A type of bacteria known as group A *Streptococcus* (group A strep) causes strep throat. This makes strep throat different from most other cases of sore throat, which occur due to viruses.

### **Is strep throat contagious?**

Yes. Strep throat is very contagious. Some people with the infection don’t have symptoms or look sick. But even if you don’t have symptoms, you can still easily spread the infection to others. However, people who exhibit symptoms or appear sick are more contagious than people who don’t have symptoms.

### **How does strep throat spread?**

Strep throat spreads from person to person very easily, especially among members of the same household. You can spread the bacteria that causes strep throat to other people through respiratory droplets and direct contact.

#### **Respiratory droplets**

The bacteria that cause strep throat often live in your nose and throat. When you sneeze, cough or talk, you can spread the infection through respiratory droplets. Other people can get the infection if they:

* Breathe in the droplets.
* Touch something that contains the droplets and then touch their nose or mouth.
* Share personal items (such as drinking from the same glass).

#### **Direct contact**

The bacteria that cause strep throat may live in infected sores on your skin. Other people can get the infection if they touch your sores or come into contact with the fluid from them.

### **How long is strep throat contagious?**

The incubation period for strep throat is two to five days. An incubation period is the time between when you get infected and when symptoms develop. You can spread the infection to others during this time. If you’re taking antibiotics, you won’t be contagious after the first 24 to 48 hours of treatment.

#### **What kind of complications can strep throat cause?**

Left untreated, the bacteria that cause strep throat can spread to other parts of your body. This can cause serious complications, including:

* Ear or sinus infections: Infections can occur when bacteria cause swelling and inflammation in your ears or sinuses.
* Abscesses: Pockets of pus (abscesses) can form around your tonsils.
* Guttate psoriasis: A skin condition that causes small, red and scaly teardrop-shaped spots to develop on your arms, legs and torso.
* Scarlet fever: An infection that causes a red, bumpy rash all over your body.
* Rheumatic fever: A disease that mainly affects your heart valves.
* Post-streptococcal glomerulonephritis: A kidney disease that can develop after strep infections.
* Invasive streptococcal infection: An infection that can occur when bacteria enter your tissue. It can be life-threatening, with conditions occurring such as streptococcal toxic shock syndrome, necrotizing fasciitis and other severe infections. This can ultimately lead to rapid decline and death.

### **What are some home remedies for strep throat?**

There are many sore throat home remedies that can help relieve your symptoms. Strep throat self-care tips that you can try include:

* Eat soft foods, such as applesauce.
* Drink soothing liquids, such as warm tea.
* Gargle with warm salt water.
* Suck on age-appropriate throat lozenges or ice pops.
* Get plenty of sleep.
* Stay hydrated by drinking plenty of water.
* Use a cool-mist vaporizer or humidifier.
* Take a pain reliever, such as acetaminophen (Tylenol®) or ibuprofen (Advil®).

Don’t give your child aspirin. Aspirin can cause a life-threatening illness called Reye’s syndrome in children and adolescents who have fevers.

### **Can you get strep throat without tonsils?**

Strep throat usually affects your throat and tonsils. But you can still get the infection if you don’t have tonsils — it doesn’t make you immune. However, if you’ve had your tonsils removed, your risk of getting strep throat decreases. You may get the infection less often. In addition, your symptoms may not be as severe.

### **Other sore throats don't need special medicine, so why does strep throat?**

Viruses cause most sore throats, and medicine (like antibiotics) can’t cure viruses. You can only relieve your aches and pains. Viruses heal without antibiotics on their own with time. Because strep throat is a bacterial infection, you need antibiotics to clear it up.

### **Can I take antibiotics I already have in the house for strep throat?**

No. You should never take medicines left over from an earlier illness or give leftover medicine to anyone else. Leftover antibiotics can also make strep throat more difficult to treat and can cause serious side effects.

### **What’s the difference between strep throat and tonsillitis?**

Tonsillitis occurs when your tonsils are inflamed or infected. The condition can be viral or bacterial. Viruses cause most cases of tonsillitis. Bacteria such as group A *Streptococcus* cause the remaining cases.

Only group A *Streptococcus* bacteria can cause strep throat. It’s never viral.

**DOCTOR-PATIENT CONVERSATIONS**

Doctor: Hello. What brings you in today?

Patient/Parent: My child has a sore throat and fever.

Doctor: I see. How long has the sore throat been going on?

Patient/Parent: About two days now.

Doctor: Has your child had any difficulty swallowing, swollen glands, or white spots on their tonsils?

Patient/Parent: Yes, swallowing is painful, and I noticed some white patches in the throat.

Doctor: Do they have a cough or runny nose?

Patient/Parent: No, no cough or cold symptoms.

Doctor: That’s helpful. These signs—sore throat with white spots, fever, but no cough—suggest it could be strep throat, which is a bacterial infection caused by group A streptococcus.

Patient/Parent: How serious is it?

Doctor: It’s usually treatable with antibiotics, but it’s important to treat it to prevent complications like rheumatic fever or kidney problems.

Patient/Parent: What will the treatment involve?

Doctor: I will prescribe a course of antibiotics. It’s very important to give all the medicine for the full duration, even if your child feels better after a couple of days. Also, make sure they rest, drink plenty of fluids, and avoid irritants like smoke.

Patient/Parent: How soon will they feel better?

Doctor: Most children start to improve within 2 to 3 days of starting antibiotics. Fever and throat pain usually improve first.

Patient/Parent: Should they stay home from school?

Doctor: Yes, they should stay home at least 24 hours after starting antibiotics and until they feel well enough to participate.

Patient/Parent: When should we come back or seek urgent care?

Doctor: If your child develops difficulty breathing, severe pain, swelling of the neck, rash, or doesn’t improve after 48 to 72 hours of treatment, you should come back immediately.

Patient/Parent: Thank you, doctor. That helps.

Doctor: You’re welcome. Let me know if you have any questions. I hope your child feels better soon.

***REFERENCES:***

[Strep Throat Symptoms, Causes & Treatment](https://my.clevelandclinic.org/health/diseases/4602-strep-throat#signs-and-symptoms)

[Strep throat - Symptoms & causes - Mayo Clinic](https://www.mayoclinic.org/diseases-conditions/strep-throat/symptoms-causes/syc-20350338)

**SNORING**

ALTERNATIVE NAMES

**DEFINITION / DESCRIPTION**

Snoring refers to a rattling, snorting or grumbling sound some people make during sleep. It happens when there’s an obstruction in your airway.

#### **Is snoring normal?**

Snoring is common (and normal) for many people. In fact, nearly everyone snores at some point, including babies and young children.

But loud, jarring snoring may indicate sleep apnea — a condition that causes you to pause breathing during sleep. If snoring occurs in combination with apneic episodes (gasping for air in your sleep) and other symptoms like fatigue or irritability, then you should talk to a healthcare provider.

**CAUSES**

When you breathe, you push air through your nose, mouth and throat. A blockage in your airway can cause these tissues to vibrate against each other as air moves through your:

* Soft palate (the back of the roof of your mouth).
* Tonsils.
* Adenoids.
* Tongue.

The vibrations make a rumbling, rattling noise (what we know as snoring).

Several different factors can cause this airway blockage, including:

* Age. Snoring is more common as we age because muscle tone decreases, causing our airways to constrict (shrink).
* Alcohol and sedatives. Beverages containing alcohol and certain medications relax your muscles, restricting airflow through your nose, mouth and throat.
* Anatomy. Enlarged adenoids, big tonsils or a large tongue can make it hard for air to flow through your nose and mouth. A deviated septum (when the cartilage that separates your nostrils is off-center) can also block the flow of air.
* Sex. Snoring is more common in men.
* Family history. Snoring runs in families. If you have a biological parent who snores, you’re more likely to snore, too.
* Overall health. Nasal congestion due to allergies and the common cold blocks airflow through your mouth and nose. Pregnancy can also cause snoring due to hormonal changes.
* Weight. Snoring and sleep-related breathing disorders are more common in people who have overweight (a body mass index, or BMI, greater than 25) or obesity (a BMI greater than 30).

#### **Is snoring bad?**

Snoring isn’t necessarily bad. Most of us snore at some point during our lives. But it’s time to see a healthcare provider if you snore loudly, or if snoring disrupts your sleep quality.

**SIGNS / SYMPTOMS**

Snoring sounds vary from person to person. Snores might sound like:

* Quiet vibrations.
* Whistling.
* Grumbling.
* Snorting.
* Rumbling.

People who snore may also:

* Toss and turn during sleep.
* Wake up with a dry or sore throat.
* Feel tired during the day (fatigue).
* Have headaches.
* Feel moody or irritable.
* Have difficulty focusing.

**DIAGNOSIS METHODS**

A healthcare provider will perform a physical examination of your nose, mouth and throat. They’ll also ask you (and maybe even your partner or partners) several questions, including:

* How often do you snore?
* What does your snoring sound like?
* Do you feel rested during the day?

#### **Sleep study**

If your provider thinks you might have a sleep disorder (like sleep apnea), they may recommend a sleep study (polysomnography). You might be able to do a sleep study at home, or you may need to spend the night in a sleep center.

A sleep study evaluates:

* Brain wave activity.
* Breathing patterns, including any periods when you stop breathing or gasp for air.
* Heart rate and oxygen levels.
* Movements during sleep, such as arm or leg movements or tossing and turning.
* Sleep cycles and snoring.

**TREATMENT OPTIONS**

Healthcare providers use a wide range of treatments to reduce snoring. The option that’s right for you depends on several factors, including the severity of your snoring, your health history and your personal preferences.

#### **Nonsurgical snoring treatments**

Nonsurgical snoring remedies focus on improving your sleep posture or opening your airways. These treatments may include:

* Lifestyle changes. Changing your sleep position, avoiding beverages containing alcohol and maintaining a weight that’s healthy for you can reduce snoring.
* Medications. Cold and allergy medications relieve nasal congestion and help you breathe freely.
* Nasal strips. Wearing nasal strips (flexible bands that stick to the outside of your nose) can help keep your nasal passages open.
* Oral appliances. Wearing an oral appliance when you sleep keeps your jaw in the proper position so air can flow. Your healthcare provider might call it a mouth device or mouth guard. A mouth guard used for other purposes, like sports, won’t resolve snoring.

#### **Surgical snoring treatments**

Healthcare providers may use surgery to treat severe snoring. The goal of surgery is to shrink or remove excess tissue or correct a structural issue (like a deviated septum). Surgical treatments may include:

* Laser-assisted uvulopalatoplasty (LAUP). LAUP reduces tissue in your soft palate and improves airflow.
* Ablation therapy. Also called Somnoplasty®, this technique uses radiofrequency energy to shrink excess tissue in your soft palate and tongue.
* Septoplasty. If you have a deviated septum, your provider may recommend septoplasty. A septoplasty improves airflow through your nose by reshaping the cartilage and bone.
* Tonsillectomy or adenoidectomy.A surgeon removes excess tissue from the back of your throat (tonsillectomy) or the back of your nose (adenoidectomy).

**PREVENTION TIPS**

Certain lifestyle changes may help you stop or reduce snoring. Here are some things to try:

* Avoid sedatives (like zolpidem, clonazepam and eszopiclone) or beverages containing alcohol before bedtime.
* Ask your provider about medications to relieve nasal congestion.
* Stay active, get plenty of exercise and maintain a weight that’s healthy for you.
* Elevate your head during sleep to improve airflow.
* Try sleeping on your side instead of your back.
* Purchase a snore-reducing pillow that keeps your head in the proper position when you sleep.

Talk to your provider for more tips on how to stop snoring. They can offer personalized recommendations based on your needs.

OUTLOOK / PROGNOSIS

Occasional snoring is usually harmless. But loud, disruptive or frequent snoring can be a symptom of sleep apnea, a serious disorder. Long-term snoring increases your risk of health issues, including:

* Decreased blood oxygen levels (hypoxia).
* Difficulty concentrating.
* Fatigue (feeling very tired during the day).
* Heart attack.
* High blood pressure.
* Stroke.
* Type 2 diabetes.

**WHEN TO SEE A DOCTOR / RED FLAG**

* Loud, frequent snoring that disrupts your or your partner’s sleep
* Witnessed pauses in breathing, gasping, choking, or snorting sounds during sleep
* Excessive daytime sleepiness or fatigue despite seemingly adequate sleep
* Morning headaches, sore throat upon waking, or restless sleep
* Difficulty concentrating during the day
* High blood pressure or chest pain at night
* Your snoring is very loud and disturbing others
* If a child snores with poor attention span, behavioral problems, or poor school performance
* Any symptoms of airway obstruction such as difficulty breathing or changes in voice

**DIFFERENTIAL DIAGNOSIS**

* Primary (Simple) Snoring
  + Snoring without significant airway obstruction or sleep disruption; no daytime sleepiness.
* Obstructive Sleep Apnea (OSA)
  + Repeated airway collapse during sleep causing apnea, hypopnea, oxygen desaturation, and fragmented sleep; often with loud snoring, witnessed apnea, daytime sleepiness.
* Upper Airway Resistance Syndrome (UARS)
  + Increased upper airway resistance causing arousals without frank apnea/hypopnea; snoring and daytime sleepiness present.
* Nasal Obstruction Causes
  + Deviated nasal septum
  + Nasal polyps
  + Allergic or nonallergic rhinitis
  + Chronic sinusitis
* Oropharyngeal and Soft Tissue Abnormalities
  + Enlarged tonsils and adenoids (common cause in children)
  + Enlarged tongue (macroglossia)
  + Long or thick soft palate and uvula
  + Obesity with excess neck/throat tissue
* Craniofacial Abnormalities
  + Retrognathia (receded jaw)
  + Micrognathia (small jaw)
  + High-arched palate
  + Large neck circumference (>40 cm)
* Lifestyle and Other Contributing Factors
  + Alcohol or sedative use (muscle relaxation)
  + Supine sleep position
  + Smoking (airway inflammation)
  + Sleep deprivation
* Other Medical Conditions Associated with Snoring
  + Hypothyroidism
  + Gastroesophageal reflux disease (GERD)
  + Pregnancy (due to hormonal airway changes)
  + Down syndrome (due to anatomical predisposition)
* Rare or Miscellaneous Causes
  + Hypopharyngeal cysts or tumors
  + Abnormal epiglottis

**EPIDEMIOLOGY**

* Around 30-45% of adults snore at least occasionally, with approximately 25-33% snoring regularly. Habitual snoring prevalence is typically higher in men than women (e.g., 33-38% in men vs. 17-20% in women).
* Snoring prevalence increases with age, affecting about 50% of people over 60 years old, including approximately 60% of older men and 40% of older women.
* Risk factors associated with snoring include male sex, older age, increased body mass index (BMI)/obesity, smoking, alcohol use, hypothyroidism, allergic rhinitis, hypertension, and certain medications.
* Habitual snoring may be present without obstructive sleep apnea (OSA), but many people with OSA are habitual snorers. Studies estimate OSA prevalence around 3.8% in adults, often overlapping with snoring cases.
* Snoring prevalence rates vary worldwide and have been reported to range widely due to different study populations and definitions—for example, from about 15% to over 30% in adults in various countries.
* In children, snoring affects around 6% with some having coexisting apnea, impacting memory, concentration, and behavior.
* Snoring may be linked to increased risks of hypertension, cardiovascular disease, stroke, and reduced quality of life due to daytime sleepiness and social consequences

**PREDEFINED Q & A SETS**

### **When should I be worried about snoring?**

See a healthcare provider if snoring disrupts your sleep or leads to issues like fatigue, headaches or gasping for air. They can run tests to see if you have sleep apnea or another type of sleep-disordered breathing.

### **What questions should I ask my doctor?**

If you plan to see a healthcare provider about your snoring, here are some questions you might want to ask:

* What’s causing me to snore?
* Is my snoring causing any serious health issues?
* Do I need treatment? If so, what kind?
* Are there lifestyle changes I can make that might reduce snoring?

### **How do you know if you snore?**

If you live with a partner or roommate (or several), they may have already told you that you snore. If you live by yourself, it can be more difficult to know for sure. To find out, you can get a noise-activated voice recorder and turn it on before you go to sleep.

### **Does snoring mean sleep apnea?**

Snoring doesn’t necessarily mean you have sleep apnea. Snoring is a common sleep apnea symptom, but there are many other reasons why people snore.

### **My baby is snoring. Should I worry?**

Snoring in babies is common and rarely indicates a serious condition. Most of the time, a stuffy nose causes it. But if you’re worried or concerned, you should schedule a visit with your baby’s pediatrician. They can find out why your baby is snoring and whether they need treatment.

**DOCTOR-PATIENT CONVERSATIONS**

Doctor: Hello! You mentioned that you’ve been having trouble with snoring. Can you tell me a bit more about it?

Patient: Yes, I’ve been snoring quite loudly at night, and my partner says sometimes I seem to stop breathing.

Doctor: Thank you for sharing that. Loud snoring is common, but when it’s accompanied by pauses in breathing, it could suggest a condition called obstructive sleep apnea. This means the airway gets partially or fully blocked during sleep, causing interruptions in breathing.

Patient: Oh, that sounds serious. What symptoms should I watch for?

Doctor: Besides loud snoring and witnessed breathing pauses, other symptoms include feeling very tired or sleepy during the day, morning headaches, difficulty concentrating, and sometimes waking up gasping or choking. Snoring that disrupts your sleep or your partner’s sleep is also important to pay attention to.

Patient: What causes snoring and sleep apnea?

Doctor: Snoring happens when the tissues in your throat vibrate as you breathe during sleep. Factors like being overweight, nasal congestion, alcohol use before bed, or sleeping on your back can make snoring worse. Sleep apnea occurs when those tissues actually block your airway temporarily.

Patient: How do you find out if I have sleep apnea?

Doctor: We often start with a detailed history and physical exam. If sleep apnea is suspected, we might recommend a sleep study, which monitors your breathing, oxygen levels, and other body functions overnight.

Patient: What treatments are available?

Doctor: Treatment depends on how severe the snoring or sleep apnea is. Lifestyle changes like losing weight, avoiding alcohol before bedtime, and sleeping on your side can help. For sleep apnea, devices like a CPAP machine can keep your airway open at night. In some cases, surgery or dental devices may be options.

Patient: When should I come back or see someone urgently?

Doctor: If you or your partner notice loud snoring with pauses in breathing, excessive daytime sleepiness affecting your daily activities, or symptoms like chest pain and severe morning headaches, it’s important to see a doctor promptly.

Patient: Thanks, doctor. It helps to understand what to look for.

Doctor: You’re welcome! Let me know if you have any more questions or want help arranging a sleep study.

REFERENCES:

<https://www.mayoclinic.org/diseases-conditions/snoring/symptoms-causes/syc-20377694>

[Snoring: Causes, Remedies & Prevention](https://my.clevelandclinic.org/health/diseases/15580-snoring#overview)

<https://www.hopkinsmedicine.org/health/wellness-and-prevention/why-do-people-snore-answers-for-better-health>

**SOFT PALATE CANCER**

ALTERNATIVE NAMES: The alternative names for soft palate cancer include oropharyngeal cancer and squamous cell carcinoma of the soft palate.

**DEFINITION / DESCRIPTION**

Your soft palate is the back part of the roof of your mouth. Soft palate cancer is one of several kinds of oropharyngeal cancer. It happens when squamous cell carcinoma cells multiply and create cancerous tumors. Soft palate cancer is often found before it can spread, making it easier to treat with surgery to remove tumors.

#### **Is soft palate cancer common?**

No, it’s not. Soft palate cancer is a type of oropharyngeal cancer. About 53,000 people in the U.S. are diagnosed with oropharyngeal cancer each year. Soft palate cancer accounts for about 5% to 12% of all oropharyngeal cancer. Given that data, soft palate cancer affects 2,000 to 6,000 people. For comparison, lung cancer affects more than 236,000 people annually.

**CAUSES**

Like most types of oropharyngeal cancer, the most common type of soft palate cancer is squamous cell carcinoma (SCC). This cancer starts in flat cells of tissue that line the inside of your mouth and throat. In soft palate cancer, something causes your healthy squamous cells to mutate, or change, into cancerous cells that multiply and become tumors.

Medical researchers have identified some risk factors that may increase the chance you’ll develop soft palate cancer. Risk factors include:

* Heavy tobacco use, specifically smoking.
* Regular consumption of beverages that contain alcohol.

**RISK FACTORS**

The risk factors for soft palate cancer include heavy tobacco use, specifically smoking, and regular consumption of beverages that contain alcohol. Medical research shows that HPV infections increase the risk of developing most types of oropharyngeal cancer, including soft palate cancer.

Additionally, using alcohol and tobacco together increases the risk even more. Reverse smoking, where the lit end of the cigarette or cigar is placed inside the mouth, has been linked to the development of malignant lesions of the hard palate.

Other risk factors include poor oral hygiene, chronic oral mucosal trauma, ill-fitting dentures, Human Papilloma Virus (HPV) infection, irritative mouthwashes, and vitamin A deficiency. People with a weakened immune system, such as those taking medicines to control the immune system, also have a higher risk of soft palate cancer.

**SIGNS / SYMPTOMS**

Symptoms may include:

* A sore on your soft palate. The sore may hurt or bleed.
* A white patch that doesn’t go away.
* Bad breath (halitosis).
* Trouble swallowing (dysphagia).
* A lump in your neck.
* Trouble opening your mouth (trismus).
* A sore throat (pharyngitis) that doesn’t go away.

**DIAGNOSIS METHODS**

Healthcare providers will ask you about your symptoms. For example, they may ask some of the following questions:

* When did you first notice a sore or any other change in the area?
* Has the sore gotten bigger?
* Does it bleed?
* Does it hurt?
* Do you have trouble swallowing?
* Have you noticed any neck swelling or swollen lymph nodes?
* Do you have ear pain? (That is a common sign of oropharyngeal cancer.)

They may ask you about personal habits, such as whether you use tobacco, including smokeless tobacco. They may also ask how often you drink beverages containing alcohol.

They’ll use a small mirror or light to examine your soft palate and check lymph nodes in your neck for signs of swelling.

#### **What tests do healthcare providers use?**

Providers use biopsies to obtain tissue samples. Medical pathologists examine tissue samples under microscopes to determine if the tissues contain cancerous cells.

Providers may also do computed tomography (CT) scans to determine tumor size and if it’s spread.

#### **Soft palate cancer stages**

Cancer staging is the process of determining if cancer is present and, if so, how far it has spread. It helps your healthcare team develop your treatment plan.

Soft palate cancer is a type of oropharyngeal cancer. The stages of oropharyngeal cancer span from Stage I (best prognosis) to Stage IV (worst prognosis). In this instance, providers use the cancer staging system developed by the American Joint Committee on Cancer. This system uses specific information about the tumor’s size, whether it’s spread to nearby lymph nodes and/or to distant organs, and whether or not the cancer is associated with the HPV virus.

Cancer staging is a complicated process to complete, much less explain. It’s understandable if you feel intimidated, confused or unnerved by a process that seems to reduce your illness to a formula of letters and numbers. Your providers understand why you may feel this way. If you’re confused or concerned by what you’re hearing, ask your healthcare provider to explain how the cancer staging system works in your situation.

**TREATMENT OPTIONS**

Providers use several types of treatment depending on the cancer stage:

* Surgery: Providers remove the tumor with surgery, including a minimally invasive surgery called transoral robotic surgery (TORS). They may also remove lymph nodes in your neck affected by the tumor. This is neck dissection.
* Radiation therapy: Radiation therapy may be first-line or initial treatment for larger tumors that have spread into nearby tissues and/or lymph nodes.
* Chemotherapy: Providers may use chemotherapy as an initial treatment for soft palate cancer that has spread to other areas of your body.
* Chemoradiation: As it sounds, chemoradiation combines chemotherapy and radiation therapy. This treatment is for soft palate cancer that’s spread to your lymph nodes but not to other areas of your body.
* Reconstructive surgery: You may need additional surgery. You may need a prosthetic device (soft palate obturator) that replaces your soft palate. A soft palate obturator is like the dental retainer you may have used if you had braces on your teeth.
* Psychological therapy (psychotherapy): Some people have trouble adjusting to changes in their bodies after soft palate treatment. They may feel self-conscious about needing a soft palate obturator or other prosthetic devices. They may feel depressed about their situation. If that’s your situation, psychological therapy may help.

#### **What are treatment complications?**

Your soft palate helps you swallow and speak. Surgery and radiation therapy to treat soft palate cancer may affect your ability to swallow and speak as you did before treatment.

**PREVENTION TIPS**

You may not be able to prevent soft palate cancer. However, you may be able to reduce your risk by avoiding certain activities that increase your risk of developing the condition.

* If you smoke cigarettes or use smokeless tobacco, try to quit. Talk to a healthcare provider about programs to help you stop using tobacco.
* If you regularly drink beverages that contain alcohol, try to limit how much and how often you drink. If you think you may have an issue with alcohol, ask a provider for help.
* Eat a healthy diet that includes fruits and vegetables.
* Protect yourself against HPV. An HPV infection may increase your risk. Reduce your risk of HPV infection by being vaccinated against the virus and by avoiding high-risk sexual practices such as unprotected oral sex and intercourse

**OUTLOOK / PROGNOSIS**

That depends on your situation. Healthcare providers may be able to cure many early-stage soft palate cancers with surgery to remove tumors or radiation therapy. (Early-stage cancer refers to small tumors that haven’t spread.) Some late-stage cancers are also curable.

### **What is the soft palate cancer survival rate?**

Survival rates are estimates based on the experiences of other people who have the same condition. Soft palate survival rates vary. Studies show that 75% to 100% of people with early-stage soft palate cancer were alive five years after diagnosis. About 33% to 47% of people with large tumors that spread were alive five years after diagnosis.

When you think about survival rates, it’s important to remember these rates reflect what happened to other people. Prognosis or expected outcome and survival rates depend on many factors. Your healthcare provider is your best resource for prognosis and survival rate information.

**POSSIBLE COMPLICATIONS**

Possible complications for soft palate cancer include velopharyngeal insufficiency, hypernasal speech, dysphagia, and middle ear effusion from scarring at the eustachian tube opening or loss of function of tensor and/or levator palatini muscles. Surgery for soft palate cancer can cause difficulties with talking, chewing, and swallowing, and may require additional reconstructive surgery to treat these complications. Complications from hard palate resection depend on the extent of resection, and may include difficulty speaking, swallowing, and eating. Additionally, surgical resection of oral structures can cause difficulties in speech and swallowing, potentially requiring long-term enteral feeding, recurrent aspiration, and communication deficits.

**WHEN TO SEE A DOCTOR / RED FLAG**

* A lump, ulcer, patch, or thickening on the soft palate (the muscular part at the roof of the mouth toward the throat) that does not heal or goes away
* Persistent or unexplained bleeding in the mouth
* Difficulty swallowing (dysphagia) or painful swallowing
* Difficulty speaking, hoarseness, or changes in voice
* A sensation of something caught in your throat
* Mouth sores that do not heal
* Bad breath (halitosis) without obvious cause
* Mouth or throat pain that persists
* Loose teeth or dentures that no longer fit properly
* Ear pain, especially on the same side as the lesion
* Swollen lymph nodes in the neck, which may be painful or growing
* Unexplained weight loss
* White or red patches in the mouth that persist

**DIFFERENTIAL DIAGNOSIS**

* Squamous Cell Carcinoma (SCC): This is the most prevalent malignant tumor found on the soft palate. It often appears as a non-healing ulcer, a lump, or an irregular mucosal patch. Human papillomavirus (HPV) infection is a significant risk factor for SCC in this area .
* Other Malignant Tumors:
  + Adenoid cystic carcinoma: A malignant tumor originating from minor salivary glands. It is known for its slow growth and tendency for perineural invasion .
  + Mucoepidermoid carcinoma: Another malignant salivary gland tumor that can present as a mass, with varying degrees of aggressiveness .
  + Adenocarcinoma: A less common malignant tumor of glandular origin .
  + Lymphoma: Can involve the soft palate as part of lymphoid tissue, typically appearing as a diffuse swelling rather than a discrete mass.
  + Melanoma: A rare but aggressive malignant tumor, which may present with noticeable pigmentation.
* Benign Tumors and Lesions:
  + Papilloma: A benign epithelial tumor that may manifest as an exophytic or wart-like growth.
  + Pleomorphic adenoma: The most common benign salivary gland tumor, usually presenting as a slow-growing, firm swelling.
  + Schwannoma (Neurilemmoma): A benign nerve sheath tumor that typically forms a painless mass .
* Infectious and Inflammatory Conditions:
  + Odontogenic infections or abscesses: Swelling or ulcerations near the palate can be caused by dental issues, often accompanied by signs of infection and dental symptoms .
  + Cystic Lesions: Such as mucoceles or retention cysts, which are typically benign fluid-filled sacs .
  + Other Soft Tissue Masses or Inflammatory Lesions: Including granulomas, traumatic ulcers, or reactive lesions that can mimic cancer clinically

**RECENT GUIDELINES OR UPDATES**

The most recent guidelines and updates for soft palate cancer, as of the latest available information, include the following:

* The 5-year relative survival rate for oropharyngeal cancers, including soft palate cancer, is around 52% at all stages. This means that just over half of people with soft palate cancer will be alive 5 years after diagnosis when compared to people who don’t have soft palate cancer. The survival rate depends on how far it’s spread from where it started: 59% for localized cancer, 62% for regional cancer, and 29% for distant cancer.
* According to a 2018 study of 4,366 people with soft palate cancer, overall survival was 68.7 months (5 years, 8.7 months). However, in people who died specifically from soft palate cancer, survival was 161.3 months (13 years, 5.3 months).
* The Cleveland Clinic states that soft palate cancer is often found before it can spread, making it easier to treat with surgery to remove the tumor. The survival rates vary, with studies showing that 75% to 100% of people with early-stage soft palate cancer were alive five years after diagnosis, while about 33% to 47% of people with large tumors that spread were alive five years after diagnosis.
* The American Cancer Society reports that the 5-year relative survival rate for oropharyngeal cancer, like soft palate cancer, is 59% if the cancer is localized (hasn’t spread) and 29% for cancers that have spread to other parts of the body.
* The Cleveland Clinic also mentions that healthcare providers may use several types of treatment depending on the cancer stage, including surgery, radiation therapy, chemotherapy, chemoradiation, and reconstructive surgery.

**EPIDEMIOLOGY**

* Oral cavity and pharynx cancers combined have an incidence rate of about 11.6 per 100,000 men and women per year (age-adjusted).
* The lifetime risk of developing oral cavity and oropharyngeal cancer overall is about 1.2% for men and women, with higher incidence in men.
* Incidence of oropharyngeal cancers (including soft palate) has been increasing in recent decades, particularly those associated with human papillomavirus (HPV) infection. HPV is found more frequently in oropharyngeal cancers (including soft palate) than in other oral cancers. The rise in HPV-related cases contributes to the increasing incidence in sites like the soft palate.
* The average age at diagnosis for oropharyngeal cancers is around 64 years, but cases can occur in younger adults as well.
* Prognosis and survival rates depend on the stage at diagnosis: the overall 5-year relative survival rate for oropharyngeal cancers like soft palate cancer is approximately 52%, with higher survival for localized disease (around 59%) and lower survival for distant metastases (around 29%

**PREDEFINED Q & A SETS**

Q1: “Does human papillomavirus (HPV) cause soft palate cancer?”

It may. Medical research shows HPV infections increase the risk of developing most types of oropharyngeal cancer, including soft palate cancer.

Q2: “What are the complications of soft palate cancer?”

Soft palate cancer that’s not treated can spread (metastasize) to other areas of your body. Cancer that spreads is more difficult to treat.

Q3: “What does soft palate cancer look like?”

It may look like a white patch or bump on your soft palate. Your soft palate is the squishy section between your hard palate (the bony section on the roof of your mouth) and your uvula (a tiny bit of tissue that hangs down at the back of your throat.)

Q4: “How do I take care of myself?”

People with soft palate cancer often need help managing eating and speaking after surgery. Your healthcare provider will help you manage any side effects or concerns you may have about regaining your speech or ability to eat.

Some people continue using tobacco products even after going through surgery and other soft palate cancer treatment. If that’s your situation, ask your provider for ways to stop using tobacco products

**DOCTOR-PATIENT CONVERSATIONS**

Doctor: Hello. I’d like to talk with you about the findings and explain what soft palate cancer is.

Patient: Yes, please. I don’t really understand what it means.

Doctor: The soft palate is the soft, muscular part at the back of the roof of your mouth, just before the throat. Soft palate cancer occurs when abnormal cells in this area grow uncontrollably and form a tumor. The most common type is called squamous cell carcinoma, which arises from the thin, flat cells lining the soft palate.

Patient: What symptoms usually happen with this type of cancer?

Doctor: Symptoms can include a sore or lump on the soft palate that doesn’t heal, pain or difficulty when swallowing, persistent bad breath, bleeding in the mouth, white or red patches on the soft palate, ear pain on the same side, swelling in your neck due to lymph nodes, and sometimes weight loss. Because of the location, it can also affect your voice or cause a feeling of something caught in your throat.

Patient: How do you confirm it’s cancer, and what testing is needed?

Doctor: We would perform a thorough exam of your oral cavity and throat. A biopsy, where a small tissue sample is taken from the lesion, is necessary to confirm cancer under a microscope. We may also do imaging tests like CT scans or MRIs to determine the size of the tumor and whether it has spread to nearby areas or lymph nodes. These help us stage the cancer and plan treatment.

Patient: What treatment options are there?

Doctor: Treatment depends on the stage and exact location of the cancer. It usually involves surgery to remove the tumor, radiation therapy to destroy any remaining cancer cells, and sometimes chemotherapy. We tailor the treatment plan to balance removing the cancer and preserving function like speech and swallowing as much as possible.

Patient: Is soft palate cancer serious? What’s the outlook?

Doctor: Early detection and treatment improve outcomes significantly. The prognosis depends on factors like tumor size, spread to lymph nodes, and overall health. HPV infection is sometimes involved and can affect prognosis. Regular follow-up is important to detect any recurrence early.

Patient: What can I do to prepare for treatment and recovery?

Doctor: We will guide you through the process, including nutrition support and rehabilitation if needed. You may have some side effects like soreness, swallowing difficulty, or voice changes, but many improve with time and therapy. It’s helpful to have family support and communicate openly with your care team.

Patient: When should I ask for help or come back for issues?

Doctor: Please reach out if you experience new or worsening pain, difficulty swallowing or breathing, signs of infection, or if you notice any lumps or swelling. We are here to support you throughout treatment and recovery.

*REFERENCES:*

<https://www.mayoclinic.org/diseases-conditions/soft-palate-cancer/symptoms-causes/syc-20354183>

<https://my.clevelandclinic.org/health/diseases/24791-soft-palate-cancer>

<https://www.cancer.org/cancer/types/oral-cavity-and-oropharyngeal-cancer/about/key-statistics.html>

**SPEECH DISTURBANCES**

ALTERNATIVE NAMES: Alternative names for speech disturbances include speech disorder, speech impediment, stutter, stammer, impairment, impediment, faltering, hesitancy, speech defect, lisp, and hesitant speech. These terms are often used interchangeably, although they may have slightly different meanings. For instance, "impediment" is considered obsolete in medicine and refers to an illness or defect that hinders functioning. Other terms like "speech disorder" and "speech impairment" are more commonly used today. Additionally, specific types of speech disturbances may have their own names, such as dysarthria, which is characterized by slowed, slurred speech, abnormal rhythm when speaking, limited jaw and tongue movement, labored speech, and difficulty articulating.

**DEFINITION / DESCRIPTION**

A speech impairment (sometimes called a speech impediment or speech disorder) happens when you have trouble saying sounds so that people don’t understand what you’re saying. Some people are born with conditions that affect their speech. But you can develop a speech impairment at any time in your life if you’re injured or have a medical condition that affects your voice or your ability to speak.

Without treatment, children with speech impairments may have difficulty learning to read and write. Research suggests children with speech impairments may develop mental health issues like anxiety and depression. Adults may feel anxious about trying to communicate and develop social isolation that can lead to depression.

#### **Types of speech impairments**

There are different classifications for speech impairments:

* Fluency disorders.
* Orofacial myofunctional disorders (OMD).
* Speech sound disorders.
* Voice disorders.

##### **Fluency disorders**

Fluency is the flow of a person’s speech. A person is fluent when they speak continuously and smoothly. A fluency disorder involves chronic and repeated interruptions to speech flow. Examples of fluency disorders are:

* Stuttering: If you stutter, you may sound like you’re trying to say a sound, a word or a syllable, but it’s not coming out.
* Cluttering: If you clutter, you may speak quickly, merging words or cutting off parts of words.

##### **Orofacial myofunctional disorders (OMD)**

An orofacial myofunctional disorder is when something affects how you use or move your face, mouth and tongue muscles. OMDs may affect how you speak, making it hard for you to make sounds like “s” as in sun or “sh” as in ship.

##### **Speech sound disorders**

A speech sound disorder affects your ability to say sounds clearly. Examples of speech sound disorders are:

* Apraxia in adults.
* Articulation disorders.
* Childhood apraxia of speech.
* Dysarthria.

##### **Voice disorders**

Some voice disorders may affect your ability to speak. Examples include:

* Hoarseness.
* Laryngitis.
* Spasmodic dysphonia.

#### **Are speech impairments common?**

Experts estimate that 5% of children in the U.S. ages 3-17 have a speech impairment that lasts for a week or longer. By the time children are in first grade, 5% have noticeable issues. About 2% of children in the U.S. have voice disorders that affect their ability to speak.

The overall picture is less clear when we’re talking about adults who have speech impairments. In adults, speech impairments are often classified by the specific condition.

For example, more than 3 million people in the U.S. stutter, or about 10% of the total U.S. population. While most people outgrow stuttering during their childhood, 1 in 4 people experience stuttering as adults. Voice disorders affect 4% of adults in the U.S.

**CAUSES**

Healthcare providers don’t know all the reasons why children and adults have speech impairments (sometimes called speech impediments). In general, anything that may cause brain damage or nerve damage may affect your ability to speak, such as:

* Neurodevelopmental conditions like autism spectrum disorder, attention-deficit/hyperactivity disorder (ADHD) or epilepsy.
* Traumatic brain injury (TBI).
* Medical conditions like stroke, brain tumor or dementia.

**RISK FACTORS**

The risk factors for speech disturbances in children include a variety of biological, environmental, and developmental factors. Key risk factors identified in multiple studies include:

* Male gender: Several studies have found that males are more likely to experience speech and language delays compared to females.
* Family history of speech and language disorders: A positive family history, particularly with parents or siblings having speech and language difficulties, is a significant risk factor.
* Hearing problems: Otitis media (middle ear infections) and other hearing issues can lead to speech and language delays due to reduced auditory input.
* Pre- and perinatal problems: Factors such as maternal stress or infections during pregnancy, complications during delivery, preterm birth, and low birthweight are associated with speech and language delays.
* Poor home environment: A negative or unsupportive home environment, including low parental education and limited interaction, can contribute to speech and language delays.
* Sucking habits: Prolonged use of pacifiers, dummies, thumbs, or bottles can lead to oral sensory issues and oro-motor dysfunction, which may affect speech development.
* Low socioeconomic status: Children from lower socioeconomic backgrounds are at higher risk for speech and language delays.
* Delayed gross motor milestones: Children who experience delays in gross motor development are more likely to have speech and language delays.
* Excessive screen time: More than 2 hours of screen time per day has been identified as a risk factor for delayed speech.
* Impulsive behavior: Impulsive behavior in children is associated with an increased risk of speech and language delays.
* Reactive temperament: A more reactive temperament in children is a consistent risk factor for speech and language impairment.
* Low maternal well-being: Lower levels of maternal well-being are associated with an increased risk of speech and language impairment.

These factors highlight the complex interplay of genetic, environmental, and developmental influences on speech and language development in children.

**SIGNS / SYMPTOMS**

Speech impairment symptoms vary depending on the cause. For example, if you have stuttering, you can’t control the muscles that you use to speak, so you repeat sounds or syllables, or hold or draw out certain sounds or syllables. If you have a voice disorder, your voice may sound uneven or shaky, strangled or breathy.

**DIAGNOSIS METHODS**

Healthcare providers usually begin diagnosis with a comprehensive physical examination. A pediatrician may do a developmental evaluation. A provider or pediatrician may refer you or your child to a speech-language pathologist (SLP), a specialist who diagnoses and treats conditions that affect your ability to communicate.

**TREATMENT OPTIONS**

Treatment varies depending on your situation. For example, speech therapy is a common treatment for many speech impairments (speech disorders). If you have a voice disorder that affects your speech, your provider may refer you to specialists for voice therapy.

#### **Can you fix a speech impairment?**

In some cases, yes, speech therapy can help people overcome speech impairments. But everyone is different. You can develop a speech impairment if you have an underlying condition that affects your ability to speak. In that case, speech therapy may help, but it may not fix the issue.

**PREVENTION TIPS**

There’s no way to prevent most speech impairments. You may be able to prevent some voice disorders by protecting your voice from overuse.

**OUTLOOK / PROGNOSIS**

There are many types of speech impairments, which makes it hard to say exactly what you can expect. For example, some speech impairments improve as children grow older. If a medical condition causes speech issues, your speech is likely to improve as you get better. But there are situations when people need long-term speech therapy so they can communicate. If you have a speech impairment, your healthcare provider is your best resource for information.

**POSSIBLE COMPLICATIONS**

Speech disturbances can lead to various complications, including difficulties in social interactions, academic performance, and overall communication effectiveness.

Without treatment, children with speech impairments may have difficulty learning to read and write, and may develop mental health issues like anxiety and depression. Adults may feel anxious about trying to communicate and develop social isolation that can lead to depression.

Additionally, speech problems can cause difficulties in social situations, leading to strain at work, school, and in relationships, which can result in mental health issues like depression. The stress of struggling to communicate and be understood can have significant impacts on an individual's quality of life.

**WHEN TO SEE A DOCTOR / RED FLAG**

Visit a healthcare provider as soon as you feel like something is affecting your ability to speak, hear or communicate with others. Visit a provider if you think your child might have difficulties with language or comprehension.

A sudden change in your ability to speak may be a symptom of a serious medical condition that needs immediate care. A stroke or head injury can affect speech. If you’re with someone who has the following symptoms, call 911 or your local emergency service number right away. To recognize the warning signs of a stroke, remember to think BE FAST:

* B. Be watchful for a sudden loss of balance.
* E. Look out for sudden loss of vision in one or both eyes. Are they experiencing double vision?
* F. Ask the person to smile. Look for a droop on one or both sides of their face, which is a sign of muscle weakness or paralysis.
* A. A person having a stroke often has muscle weakness on one side. Ask them to raise their arms. If they have one-sided weakness (and didn’t have it before), one arm will stay higher while the other will sag and drop downward.
* S. Strokes often cause a person to lose their ability to speak. They might slur their speech or have trouble choosing the right words.
* T. Time is critical, so don’t wait to get help! If possible, look at your watch or a clock and remember when symptoms start. Telling a healthcare provider when symptoms started can help the provider know what treatment options are best.

**DIFFERENTIAL DIAGNOSIS**

* Dysarthria  
  Motor speech disorder caused by weakness, incoordination, or paralysis of speech muscles due to neurological injury or disease (e.g., stroke, Parkinson’s disease, ALS). Speech is typically slurred, slow, or soft.
* Apraxia of Speech (Childhood or Acquired)  
  Impaired motor planning and programming of speech movements despite normal muscle strength. Characterized by inconsistent speech errors, groping for sounds, and abnormal prosody.
* Phonological Disorders  
  Developmental speech sound disorders with patterned errors or simplifications affecting phoneme use, common in children.
* Articulation Disorders  
  Difficulty producing specific speech sounds correctly due to functional or structural issues (not neurological).
* Stuttering (Fluency Disorder)  
  Disruptions in speech flow such as repetitions, prolongations, or blocks, often developmental but sometimes acquired after brain injury or stress.
* Voice Disorders (Dysphonia)  
  Abnormalities in voice quality caused by vocal cord pathology, neurological disorders, or muscle dysfunction. Symptoms include hoarseness, breathiness, or weak voice.
* Neurodevelopmental Disorders  
  Speech impairments associated with autism spectrum disorder, intellectual disability, cerebral palsy, and other developmental conditions.
* Structural Abnormalities  
  Cleft lip or palate, craniofacial anomalies causing hypernasal speech or articulation issues.
* Hearing Impairment  
  Reduced auditory feedback leading to delayed or unclear speech development.
* Psychogenic (Functional) Speech Disorders  
  Speech disturbances without organic cause, often related to psychological or psychiatric conditions.
* Other Neurological Disorders  
  Progressive diseases such as multiple sclerosis, Huntington’s disease, dementia can cause speech disturbance.

**RECENT GUIDELINES OR UPDATES**

The Centers for Disease Control and Prevention (CDC) updated their developmental milestones in 2024, which include changes to speech and language development guidelines.

These updates have sparked debate among professionals and parents, with some concerns about the potential impact on early identification and intervention for speech delays.

The CDC revised the milestones to reflect the age at which most children reach certain skills, rather than the average age, which has led to some lowering of expectations for vocabulary development, such as the expectation of 30 words at 24 months instead of 50 words.

* CDC Developmental Milestones Update: The CDC updated their developmental milestones for children aged 2 months to 5 years, with changes aimed at helping parents and healthcare providers better identify potential delays. The update includes new milestones for speech and language development, as well as social-emotional skills.
* Speech Sound Disorders: The American Speech-Language-Hearing Association (ASHA) provides guidelines for speech sound disorders, including articulation and phonology. These guidelines address the assessment and treatment of speech sound disorders in children, emphasizing the importance of early intervention and individualized therapy.
* Unspecified Speech Disturbances: Cortica Care outlines the symptoms, causes, and management of unspecified speech disturbances, which can include articulation errors, voice abnormalities, and disruptions in fluency. The organization emphasizes the importance of comprehensive evaluations by speech-language pathologists and the use of speech therapy as a primary treatment.

**EPIDEMIOLOGY**

* In the United States, approximately 7.2% of children aged 3 to 17 years have a voice, speech, or language disorder within a 12-month period. The prevalence is highest among younger children ages 3 to 6 (about 10.8%), decreases with age (8.8% in 7-10 years, 4.3% in 11-17 years), and is more common in boys (9.1%) than girls (5.2%).
* Among children with communication disorders, about one-third have multiple communication problems (speech and language combined). Intervention services are utilized by roughly 60% of affected children, with some variation by race and sex.
* Globally and in various studies, the prevalence of communication disorders ranges broadly, with estimates from around 5% to 10% of children having significant speech or language impairment.
* In specific regional studies, the prevalence rates for speech/language disorders range from about 7% to 11.5% among children, with speech disorders often making up a smaller portion compared to language or auditory processing disorders.
* Boys show higher rates of speech/language/fluency disorders compared to girls in many studies, sometimes by a ratio of about 2:1.
* Speech disturbances in adults are less frequently studied epidemiologically but are often related to neurological causes (stroke, degenerative diseases), developmental conditions persisting from childhood, or voice disorders. Precise prevalence data for adults varies depending on the specific speech disturbance type.

**PREDEFINED Q & A SETS**

### **What’s the difference between a speech impairment and a language disorder?**

A speech impairment happens when your mouth, jaw, tongue and vocal tract can’t work together to produce recognizable words. A language disorder happens when you have trouble understanding what others are saying. You may have trouble expressing your thoughts in ways that people understand.

### **Why do we call it a speech impairment instead of a speech impediment?**

Language changes. Over the years, people have called speech conditions by many names, including speech disorders, speech impediment and speech impairment. But these terms all have slightly different meanings.

The word “impediment” is obsolete in medicine. It’s from the 1600s and comes from the verb “to impede.” It refers to an illness or defect that hinders or gets in the way of (obstructs) functioning. Essentially, a speech impediment is something that blocks or hinders. Like a loose tooth or a new tongue piercing, it feels like something temporary or external that physically affects your ability to speak.

Now, people use terms like “speech disorder” and “speech impairment” instead. They both mean that something is going on that changes (disorder) or weakens (impairs) your ability to function or perform a task on a physical, psychological or structural (anatomical) level.

### **How do I take care of myself?**

A speech impairment can affect your quality of life. Some suggestions for living with a speech impairment are:

* Consider support groups: Connecting with a national or local support group can help you find people who understand what it’s like to experience speech issues.
* Consider mental health support: People who have speech impairments may experience depression or anxiety. Working with a counselor or psychologist may help.

**DOCTOR-PATIENT CONVERSATIONS**

Doctor: Hello, I understand you’ve been experiencing some difficulties with your speech. Can you tell me more about what you’ve been noticing?

Patient: Yes, I’ve been slurring my words lately, and sometimes I have trouble pronouncing certain sounds. It’s been happening for a few weeks now.

Doctor: Thank you for sharing that. Speech difficulties can arise from various causes like muscle weakness, nerve problems, or issues with how your brain plans the speech movements. Based on what you describe, it may be what we call dysarthria or possibly apraxia of speech. Have you noticed if your speech problems come and go, or have they been persistent?

Patient: It’s been pretty constant, and I sometimes get frustrated because people ask me to repeat myself.

Doctor: That’s understandable, and you’re not alone. There are therapies that can help improve your speech, including working with a speech-language pathologist who can provide exercises tailored to your needs. We might also consider some tests like imaging or neurological evaluations to better understand the underlying cause.

Patient: Will this get better?

Doctor: It depends on the cause, but many people improve significantly with therapy. Early intervention can make a big difference. We will work together to find the best plan for you.

Patient: Okay, I’m willing to try.

Doctor: Great. I’ll refer you to a speech therapist, and we’ll schedule some further tests. Also, please let me know if you notice any new symptoms like weakness in other parts of your body or difficulty swallowing.

Patient: I will. Thank you, doctor.

REFERENCES:

[Speech Impairment: Types, Signs & Causes](https://my.clevelandclinic.org/health/diseases/speech-impairment#overview)

<https://www.medicalnewstoday.com/articles/324764>

**STRIDOR**

ALTERNATIVE NAMES: Alternative names for stridor include jangle, noise, cacophony, din, racket, discordance, stridency, discord, caterwauling, and dissonance. Other synonyms listed include stridency, jangle, cacophony, and dissonance. Additionally, stridor is sometimes referred to as noisy breathing.

**DEFINITION / DESCRIPTION**

Stridor is an abnormal high-pitched sound you make when you inhale or exhale. This sound happens when you have a blockage in your throat (pharynx), voice box (larynx) or windpipe (trachea).

#### **Types of stridor**

There are three types of stridor characterized by where there is a blockage in your airway:

* Inspiratory: A blockage in your throat or voice box (extrathoracic region) that causes symptoms when you breathe in.
* Expiratory: A blockage above your lungs in your windpipe (intrathoracic region) that causes symptoms when you breathe out.
* Biphasic (laryngomalacia or laryngeal stridor): Narrowing of the cartilage below the vocal cords that causes symptoms when you breathe in and out.

**CAUSES**

There are several causes of stridor. The most common causes of stridor include:

* An upper airway infection (viral croup). This is the most common cause in children and accounts for approximately 90% of causes of stridor.
* Airway blockage by a foreign object (foreign body aspiration).
* Injury to your airway.
* Swelling (tonsillitis epiglottitis).

Other causes of stridor include:

* Allergic reaction.
* A tumor (laryngeal cancer).
* An abscess.
* Vocal cord dysfunction or paralysis.
* Smoke or chemical inhalation.
* Neck surgery.

**RISK FACTORS**

Stridor is an abnormal, high-pitched breathing sound caused by a blockage in the throat or voice box. Several risk factors can increase the likelihood of developing stridor.

* Age is a significant factor, as stridor is more common in children, particularly those under the age of 5.
* Males are more frequently affected than females.
* Geographic location can also play a role, as certain regions may have higher incidences of respiratory infections that can lead to stridor. Underlying conditions such as asthma or chronic obstructive pulmonary disease (COPD) can also increase the risk.
* Environmental factors such as exposure to smoke, allergens, or pollutants can contribute to airway irritation and stridor.
* Lifestyle factors, including smoking and obesity, can exacerbate airway issues.

For children, the duration of mechanical ventilation (MV) is a critical risk factor for post-extubation stridor. The risk increases if the duration of MV is greater than 72 hours.

Other risk factors for post-extubation stridor include the presence of a cuffed orotracheal cannula, although this association is not statistically significant. In a neurocritical care population, female sex and weight on admission are risk factors for post-extubation stridor.

Children are at greater risk for stridor because they have narrower, shorter, and softer airways than adults. Infections such as croup, epiglottitis, and retropharyngeal abscesses are common causes of stridor in children.

Foreign body aspiration, which is more common in toddlers and young children, can also lead to stridor. Additionally, congenital anomalies such as laryngomalacia are frequent causes of stridor in infants.

**SIGNS / SYMPTOMS**

The main sign of stridor is a high-pitched whistle that can sound like a squeak when you breathe in or out.

Depending on the cause, you might experience additional symptoms that could include:

* Coughing.
* Drooling.
* Hoarseness.
* Pain in your throat and/or neck.
* Fever.
* Swelling in your throat and/or neck.
* Discomfort when you swallow.

Severe symptoms that need immediate medical attention include:

* Difficulty breathing.
* Your lips, fingertips or skin turn blue to purple color.
* You can’t eat or drink.
* You cough up blood.

If you have severe symptoms, visit the emergency room immediately or call 911.

**DIAGNOSIS METHODS**

Your provider will diagnose the cause of your stridor after gathering more information about your medical history and your symptoms. They will perform a physical exam and will recommend tests to verify your diagnosis, which could include:

* An imaging test like a neck or chest X-ray.
* A blood gas test to check the oxygen and carbon dioxide levels in your blood.
* A pulse oximeter to check your blood oxygen levels.
* Laryngoscopy or bronchoscopy to look inside your airways and lungs with a small tube (scope) with a light on it.

**TREATMENT OPTIONS**

Treatment for stridor focuses on opening your airways and/or removing the blockage in your airways. Treatment varies based on what caused your stridor but could include:

* Surgery to remove the blockage or foreign object or expand your airways.
* Medicine to reduce swelling (inflammation), treat an infection or reduce pain.
* Performing the Heimlich maneuver to dislodge airway blockage if you choked on an object.
* Receiving supplemental oxygen through a mask or a nasal tube.
* Inserting a tube into your mouth, nose or surgically inserting a tube into your trachea to allow air to flow past the blockage until your provider can remove it.

After treatment, your provider will monitor your symptoms to make sure it was successful and that you’re safely recovering.

**PREVENTION TIPS**

You can’t prevent all cases of stridor because there are several different causes, but you can take steps to reduce your risk of getting stridor by:

* Supervise your child when they’re playing or eating.
* Cut your food into small, bite-sized pieces to prevent choking.
* Chew your food completely before swallowing it.
* Keep small objects away from children or away from your mouth.
* Avoid areas where there is smoke.
* Clean and sanitize frequently used surfaces and objects like toys.
* Wash your hands with soap and water regularly.

**OUTLOOK / PROGNOSIS**

Quick treatment to remove the airway blockage leads to a positive prognosis. Your provider will diagnose the cause of your stridor and treat you immediately to prevent any complications.

Stridor can be life-threatening if you don’t receive treatment right away, which can cause respiratory failure. Respiratory failure happens when your blood doesn’t have enough oxygen to keep your body functioning.

**POSSIBLE COMPLICATIONS**

Stridor can lead to several complications, depending on its underlying cause and the promptness of treatment. If left untreated, it can result in respiratory failure due to severe airway obstruction, which can be life-threatening.

Additionally, aspiration pneumonia may occur, especially in cases of foreign body aspiration, where food or objects enter the lungs, leading to infection. Chronic respiratory issues may also develop, particularly in cases of recurrent stridor, which may indicate underlying chronic conditions requiring ongoing management.

Children with laryngomalacia may be at risk for failure to thrive, while patients with tracheomalacia may face the possibility of aspiration pneumonia. In some cases, stridor can lead to chronic airway issues, such as recurrent infections or the development of asthma.

In severe cases, complications such as hypoxia, feeding problems, and aspiration may require surgical intervention, including supraglottoplasty. Quick treatment to remove the airway blockage is essential to prevent complications, as stridor can be life-threatening if not treated promptly.

**WHEN TO SEE A DOCTOR / RED FLAG**

Visit your healthcare provider if you have stridor. Stridor is a sign that something life-threatening is occurring to you. Stridor will lead to life-threatening complications if left untreated, so don’t wait to see your provider.

Visit the emergency room or call 911 immediately if you have a new onset of stridor or any trouble breathing.

**DIFFERENTIAL DIAGNOSIS**

* Bilateral Vocal Fold Paralysis
* Croup
* Glottic Stenosis
* Infantile Hemangioma
* Laryngomalacia
* Pediatric Airway Foreign Body
* Pediatric Epiglottitis
* Pediatric Gastroesophageal Reflux
* Subglottic Stenosis in Children
* Multidisciplinary Management of Vascular Anomalies
* Tracheal Stenosis Imaging
* Tracheomalacia

## **Common Drug Treatments for Stridor**

1. Corticosteroids (Steroids)
   * Examples: Dexamethasone, Prednisolone
   * Use: Reduce airway swelling in conditions like croup, laryngitis, or other inflammatory causes of stridor. Given orally, intravenously, or by injection.
   * Side Effects: Irritation at injection site (if injected), increased blood sugar, mood changes, gastrointestinal upset, rare immunosuppression. Short courses typically have minimal side effects.
2. Nebulized Epinephrine (Adrenaline)
   * Use: Rapid reduction of upper airway swelling in severe croup or acute airway obstruction; causes vasoconstriction and decreased mucosal edema.
   * Side Effects: Increased heart rate (tachycardia), anxiety, tremor, pallor, hypertension, headache. Usually safe under medical supervision in an acute setting.
3. Antibiotics
   * Use: Reserved for bacterial infections causing stridor (e.g., epiglottitis, bacterial tracheitis, abscess).
   * Side Effects: Allergic reactions, gastrointestinal symptoms (nausea, diarrhea), antibiotic resistance if overused.
4. Oxygen Therapy
   * Not a drug but often administered to improve oxygenation while more definitive treatment is initiated.

## Additional Medical and Surgical Management

* In severe or unresponsive cases, airway management may require intubation, tracheostomy, or surgical removal of obstructions (e.g., foreign body, tumors).
* Sedation and close monitoring are critical in dangerous airway compromise.

**RECENT GUIDELINE OR UPDATES**

Stridor is a high-pitched, wheezing sound caused by disrupted airflow, often indicating significant upper airway obstruction. Recent updates and guidelines highlight the importance of timely diagnosis and management, particularly in pediatric patients. The Scottish Palliative Care Guidelines and other resources provide detailed information on assessing and treating stridor, emphasizing the need for urgent intervention when necessary.

* Scottish Palliative Care Guidelines: The latest update from the Right Decisions Service (RDS) includes information on stridor, emphasizing the importance of recognizing it as a sign of significant airway obstruction and the need for prompt assessment and management.
* GGC Medicines - Management of Stridor: This guideline outlines the management of stridor, including initial assessment, monitoring, and treatment options such as oxygen therapy, dexamethasone, nebulized salbutamol, and considerations for more severe cases, including tracheostomy or nebulized adrenaline.
* StatPearls - Stridor in Children: This resource provides an in-depth overview of stridor in children, including its causes, diagnosis, and treatment. It emphasizes the importance of considering the patient's age and symptom acuity in determining the underlying cause.

**EPIDEMIOLOGY**

The epidemiology of stridor varies based on the underlying cause. Stridor is a more frequent presentation among pediatric patients than in adults. Croup reaches its highest incidence between 6 and 36 months, leading to 350,000 to 400,000 croup-related emergency department visits annually. Croup affects 2% to 6% of infants and children annually, with a slightly higher prevalence among males than females, at a ratio of 1.4:1.

Foreign body aspiration accounts for more than 17,000 emergency department visits per year in the United States, with most cases occurring in children younger than 3 years old.

**PREDEFINED Q & A SETS**

### **How soon after treatment will I feel better?**

Your recovery time depends on the cause of your stridor. It depends on what type of treatment your provider used to remove or treat the blockage in your airway. Medicine could make you feel better within two to three days. Surgery has a longer healing time and could take several weeks before you feel back to normal. Regardless of what type of treatment you need, you will feel immediate relief after your provider removes the blockage.

### **Is stridor the same as wheezing?**

While you might hear stridor referred to as wheezing, there are differences between the two symptoms. Stridor and wheezing, both sound like a high-pitched whistle.

Stridor occurs when you have a blockage in your central (larger) airway (extrathoracic region). Wheezing occurs when your airways tighten (intrathoracic airways)

Wheezing is common among people diagnosed with allergies or asthma. Stridor happens among children or adults who have a blockage in their airway, most often by a foreign object or infection.

### **What is the difference between stridor and stertor?**

Stertor is noisy breathing that happens above your voice box (above your larynx). It is typically low-pitched.

Stridor is noisy breathing in your airway that includes your voice box (larynx) and below, which includes your windpipe (trachea).

### **How common is stridor?**

Stridor can affect anyone at any age, but it’s more common among children since their airways are narrower than adults, which makes them more at risk of getting airway blockages. Sometimes stridor can be present at birth (congenital).

The exact rate of occurrence varies by the cause. In children, stridor caused by an upper airway infection (viral croup) is the most common, and affects an estimated 5 to 6 toddlers out of 100, between 6 months to 3 years of age. Stridor caused by objects stuck in the larynx or trachea (foreign body aspiration) accounts for an estimated 17,000 emergency room visits per year in the United States.

### **How does stridor affect my body?**

Stridor is the result of a blockage in your airway. This can make breathing difficult since air isn’t able to smoothly pass in and out of your airways like it normally would. Stridor can cause other life-threatening symptoms if left untreated because your body relies on oxygen intake and carbon dioxide outtake to survive and keep your organs functioning.

### **What happens to my body if I have stridor?**

Your respiratory system airways consist of your throat (pharynx), voice box (larynx) or windpipe (trachea and bronchi). Your mouth and nose help your airways bring oxygen into your lungs when you inhale and release carbon dioxide when you exhale. Your body needs oxygen to survive.

Like a trumpet, your airways produce sound when air travels through them by creating vibrations. When you press the keys on a trumpet, the sound that the instrument makes changes. This happens because the pressed key blocks the movement of air and changes the vibration. If you have a blockage in your airways, the sound that you produce when you inhale and exhale will change in the same way that a pressed key changes the sound of a trumpet.

**DOCTOR-PATIENT CONVERSATIONS**

Doctor: Hello, I understand you’ve noticed a noisy, high-pitched sound when you breathe, called stridor. Can you tell me more about when you hear this sound and if you have any difficulty breathing?

Patient: Yes, I hear this wheezing or squeaking noise mostly when I breathe in, and sometimes it feels hard to get enough air. It’s been getting worse over the past few days.

Doctor: Thank you for sharing that. Stridor is a sign that the airway—usually around the throat or windpipe—is narrowed or partially blocked. This can happen from swelling, an infection, a foreign body, or other causes.

Patient: That sounds serious. What should I watch out for?

Doctor: If you develop trouble breathing, become very sleepy, or start turning blue around your lips or face, that is an emergency and you need immediate care. For now, I’ll do a physical exam and may order some imaging or refer you to a specialist to find out the exact cause and how to treat it.

Patient: What kind of treatments can help?

Doctor: Treatment depends on what’s causing the stridor. For example, if it’s an infection causing swelling, steroids or medications may reduce the swelling. If there’s a foreign object, it might need to be removed. In severe cases, we may need to secure the airway to help you breathe safely.

Patient: Is this something that can get better quickly?

Doctor: Sometimes yes, especially if the cause is inflammation or minor swelling. But we need to be cautious and identify the cause because stridor always means the airway is at risk.

Patient: Thank you. What can I do in the meantime?

Doctor: Try to stay calm and sit upright to help your breathing. Avoid any irritants like smoke. If you feel your breathing worsens, go straight to emergency services.

*REFERENCES:*

<https://emedicine.medscape.com/article/995267-medication>

<https://www.ncbi.nlm.nih.gov/books/NBK525995/#article-29547.s4>

[Stridor (Noisy Breathing) Symptoms & Causes](https://my.clevelandclinic.org/health/diseases/23303-stridor#overview)

**SUBGLOTTIC STENOSIS**

Alternative names and related terms for Subglottic Stenosis include:

· Subglottic narrowing

· Subglottic airway stenosis

· Subglottic obstruction

· Subglottic stricture

· Laryngotracheal stenosis (when involving both the larynx and subglottic area)

· Subglottic laryngeal stenosis

· Subglottic constriction

· Congenital subglottic stenosis (if present from birth)

· Acquired subglottic stenosis (due to trauma, intubation, infection, etc.)

**DEFINITION AND DESCRIPTION**

Subglottic stenosis is when the upper section of your trachea (windpipe) is narrower than usual so that there’s a whistling noise when you breathe, or you feel short of breath. “Subglottic” means the part of your trachea just below your vocal cords and “stenosis” means there’s a narrowing.

Anyone can have subglottic stenosis. Your newborn baby may be born with an unusually narrow airway (congenital subglottic stenosis). Or you could develop it after you’re born (acquired subglottic stenosis). Several things may cause subglottic stenosis in adults, but it can happen for no known reason (idiopathic subglottic stenosis).

Without treatment, severe subglottic stenosis can be life-threatening. But treatment to widen the narrow area in your windpipe can effectively treat the condition.

Subglottic stenosis is a rare condition, affecting 1 in every 400,000 people.

**SIGNS / SYMPTOMS**

Subglottic stenosis symptoms can be mild, moderate or severe, depending on the narrowness of your airway. Think of sipping water through a flexible straw. You only get a sip of water when the straw is wide open. Squeeze it a little, and there’s less to drink. The more you squeeze, the less water flows through the straw.

In mild or moderate subglottic stenosis, part of your airway is narrower than usual. In severe subglottic stenosis, that part of your airway is nearly closed shut so that very little air that comes in through your nose makes it through your windpipe and into your lungs.

If your baby has this condition, they may have:

· Stridor, which is a high-pitched whistling noise when your baby breathes in or out.

· Dyspnea (difficulty breathing).

· Recurring croup.

If you or your older child has subglottic stenosis, symptoms may include:

· A cough that doesn’t go away or you bring up more mucus than usual when you cough.

· Shortness of breath (dyspnea) that doesn’t improve with inhalers or other treatments

· Stridor.

**CAUSES**

**What causes subglottic stenosis?**

The condition can happen if your baby’s airway is narrower than usual (congenital subglottic stenosis). A child or an adult may develop it if they:

· Need intubation on a ventilator. People who need intubation for more than two weeks can sometimes develop subglottic stenosis.

· Have an injury that damages their airway.

· Have a rare autoimmune disorder or vasculitis that causes scar tissue to build up in their airway.

Some of the time, however, there’s no known reason why people develop subglottic stenosis. These cases tend to affect females between the ages of 30 and 60.

**DIAGNOSIS METHODS**

Your healthcare provider will perform a physical examination. They’ll ask about your medical history and your symptoms. If they suspect subglottic stenosis, they may refer you to an ear, nose and throat specialist (ENT or otolaryngologist). The specialist may perform tests, including:

· **Flexible laryngoscopy:** Your provider inserts a flexible tube down your throat. There’s a tiny camera on the tube that allows your provider to look at your larynx (voice box) and windpipe**.**

· **Pulmonary function test:** This test measures the amount of air that you can breathe in or out. Reduced airflow is a sign of subglottic stenosis.

· **Computed tomography (CT) scan:** Your provider may order a CT scan so they can look at the structure of your airway.

**TREATMENT OPTIONS**

Subglottic stenosis is rare, so there’s no standard or common treatment for it. Treatments may include checking on your condition, steroid injections and surgery.

#### **Monitoring**

If tests show subglottic stenosis is causing mild narrowing of your airway, your provider may schedule regular appointments so they can check your airway. They may do more tests to see if your airway is narrowing.

#### **Steroid injections**

Your provider may do steroid injections as initial or first-line treatment. The treatment involves injecting a steroid through your neck into your airway. (Your provider will use local anesthesia to numb your neck, airway and nose so you won’t have pain during treatment.) Typically, people who have this treatment have a series of injections every four to six weeks.

#### **Surgery for subglottic stenosis**

There are a few different surgical procedures to treat subglottic stenosis:

· **Endoscopic dilation:** In this treatment, your provider uses an endoscope to place a tiny balloon in the narrow area of your windpipe. The balloon dilates (stretches) the section. An endoscopic dilation isn’t a permanent solution: at some point, you’ll need to have the procedure again to keep your airway open.

· **Cricotracheal resection:** This surgery involves removing the narrowed part of your airway just below your voice box and then reconnecting your airway and voice box.

· **Endoscopic laryngotracheoplasty**: In this surgery, providers widen your airway by placing pieces of cartilage in your airway. Surgeons may call this surgery laryngotracheal reconstruction.

## **OUTLOOK / PROGNOSIS**

Often, prompt treatment can effectively treat the condition. But subglottic stenosis can come back, which means you or your child will need more treatment.

### **WHEN TO SEE A DOCTOR / RED FLAG**

If you or your child has treatment for subglottic stenosis, contact your provider as soon as you notice changes or symptoms, like stridor or breathing issues, that may mean the condition is coming back. You or your child may need more treatment to prevent symptoms from getting worse.

## **EPIDEMIOLOGY**

Frequency

The frequency of congenital subglottic stenosis (SGS) is unknown.

The incidence of acquired subglottic stenosis (SGS) has greatly decreased since the late 20th century. In the late 1960s, when endotracheal intubation and long-term ventilation for premature infants began, the incidence of acquired subglottic stenosis (SGS) was as high as 24% in patients requiring such care. In the 1970s and 1980s, estimates of the incidence of subglottic stenosis (SGS) were 1-8%.

In 1998, Choi and Zalzal reported that the incidence of subglottic stenosis (SGS) had remained constant at the Children's National Medical Center in Washington, DC; it was approximately 1-2% in children who had been treated in the neonatal intensive care unit (ICU).Walner reported that, among 504 neonates who were admitted to the level III ICU at the University of Chicago in 1997, 281 were intubated for an average of 11 days, with no patients developing subglottic stenosis (SGS) over a 3-year period. Moreover, in a systematic review published in 2001, Walner et al reported a decreasing incidence of neonatal subglottic stenosis in the literature, with studies published after 1983 finding an incidence of less than 4% and studies after 1990 having an incidence of under 0.63%. In 1996, a report from France described no incidence of subglottic stenosis (SGS) in the neonatal population who underwent intubation with very small endotracheal tubes (ie, 2.5-mm internal diameter) in attempts to prevent trauma to the airway.

Morbidity

Using the Kids’ Inpatient Database (KID), Arianpour et al found that in addition to gastroesophageal reflux (GER), comorbidities more likely to be diagnosed in inpatients aged 20 years or younger with acquired subglottic stenosis (SGS) include trisomy 21, asthma, and additional upper airway anomalies. However, the chance of prematurity and dehydration was indicated to be lower in pediatric acquired SGS.

Diagnostic Procedures

In a child with mild or moderate airway obstruction, perform flexible fiberoptic nasopharyngoscopy and laryngoscopy in the clinic or the emergency department (ED). If extreme airway obstruction exists or if an active supraglottic infectious process is suspected in a young child, flexible endoscopy may be deferred in favor of formal rigid bronchoscopy in the operating room (OR). However, flexible fiberoptic nasopharyngoscopy may be performed in a controlled setting in the OR, because determination of the nature of the supraglottis and glottis in awake, unsedated patients is crucial. The procedures are described as follows.

Flexible fiberoptic nasopharyngoscopy and laryngoscopy

During flexible fiberoptic nasopharyngoscopy and laryngoscopy, topical anesthesia and decongestion can be accomplished in older infants and children with topical Afrin and lidocaine. A 3-mm endoscope can be used, even in an infant. Pass the endoscope into both nasal cavities to access pyriform aperture stenosis, midnasal stenosis, choanal atresia or stenosis, lesions of the nose and nasopharynx, and the adenoid pad.

Pass the endoscope into the superior oropharynx and hypopharynx. The hypopharynx and larynx can be assessed. Identify the structure and position of the supraglottis. Evaluate the epiglottis and arytenoids for malacia or stenosis. Evaluate the position and movement of the true vocal cords. Evaluate edema or erythema of the true vocal cords, epiglottis, and arytenoids.

Flexible endoscopy

This can be performed with the patient in the supine or sitting position. The supine position often results in the obstruction of certain supraglottic processes. If the goal is to obtain the best visualization of the true vocal cords and supraglottis, place a child (even an infant) in the sitting position with his or her neck extended.

If the child is older, the voice can be evaluated, and videostroboscopy can be performed to assess the vocal cord waveform and vocal cord mobility.

Occasionally, the subglottis can be visualized with flexible endoscopy; however, rigid laryngoscopy and bronchoscopy are the safest procedures and offer the best visualization for the subglottis and tracheobronchial tree.

Rigid laryngoscopy and bronchoscopy

Rigid laryngoscopy and bronchoscopy is the best single test for evaluating airway obstruction in children. The otolaryngologist must have knowledge of the pediatric airway, and the OR must have adequate bronchoscopes and telescopes of various sizes. Prepare all equipment for bronchoscopy, including laryngoscopes, light sources, video documentation equipment, telescopes, and bronchoscopes prior to the child's arrival in the OR. Throughout the procedure, maintain good communication between anesthesiologists, surgical nursing staff, and physicians, so that any potential airway obstruction can be quickly assessed and addressed.

Do not further injure the pediatric airway—this point is of paramount importance. Use the smallest bronchoscope or telescope alone for evaluation of the subglottis in a child who does not require ventilation throughout the procedure. This practice allows good visualization without iatrogenic injury to the area. If ventilation is required throughout the evaluation, use a bronchoscope-telescope combination.

If a child has a tracheotomy or is not in extreme distress, the child can breathe spontaneously and inhale oxygen and anesthetics through an endotracheal tube in the pharynx while the airways are visualized with a laryngoscope and large telescope. Frequently, the true vocal cords are anesthetized with lidocaine prior to evaluation to help prevent laryngospasm.

Determine the size of the child's airway by using endotracheal tubes. As previously mentioned, Myer and Cotton established a scale for subglottic stenosis (SGS) severity that is based on the child's age and the size of the endotracheal tube that can be placed in the airway with an air leak pressure of less than 20 cm of water.

Evaluate the subglottis and glottis for fixation, scarring, granulation, edema, paralysis or paresis, and other abnormalities. Evaluate the distance and caliber of the stenosis. Apply the Myer and Cotton staging system only to circumferential subglottic stenosis (SGS).Glottic stenosis and SGS often coexist and must be considered when reconstruction is planned.

Evaluate the maturity of the stenosis. If a firm white scar is present, the stenosis is mature. If the stenosis has a granular or erythematous appearance, GERD, viral infection, allergic esophagitis, or another inflammatory process may be present.

Examine the area below the subglottis into the trachea and bronchi for secondary lesions. The suprastomal area is important because pathologic stenosis or malacia can influence the choice of surgical procedure. In severe subglottic stenosis (SGS), viewing the suprastomal area requires the passage of a tiny telescope through a narrow subglottis or a telescope or bronchoscope through a tracheotomy site, if available.

**pediatric differential diagnosis (DDx) of subglottic stenosis**

· Congenital subglottic stenosis: Narrowing present at birth due to thickened soft tissue or cartilage in the subglottic airway.

· Acquired subglottic stenosis: Often due to prolonged or repeated endotracheal intubation causing scarring and fibrosis.

· Laryngomalacia: Most common cause of stridor in infants, caused by floppy supraglottic structures collapsing during inspiration.

· Subglottic hemangioma: Vascular tumor in the subglottic region causing airway obstruction and biphasic stridor.

· Vocal cord paralysis: Unilateral or bilateral immobility leading to airway compromise and voice changes.

· Laryngotracheobronchitis (Croup): Viral infection causing inflammation and narrowing of the subglottic airway.

· Epiglottitis: Acute bacterial infection causing supraglottic swelling and airway obstruction.

· Bacterial tracheitis: Severe bacterial infection causing airway inflammation and narrowing.

· Foreign body aspiration: Sudden onset airway obstruction with history of choking.

· Laryngeal webs or cysts: Congenital or acquired membranes causing partial airway obstruction.

· Granulomatosis with polyangiitis: Autoimmune vasculitis causing subglottic inflammation and stenosis.

· Extrinsic airway compression: From vascular rings or masses compressing the airway externally.

· Gastroesophageal reflux disease (GERD): Can exacerbate airway inflammation and contribute to stenosis or edema.

**PREDEFINED Q AND A**

1. How severe is my child’s condition?

The severity of subglottic stenosis (SGS) is usually graded based on how much the airway is narrowed:

· Mild (Grade 1-2): Less than 50% narrowing; many children may have few or no symptoms and sometimes improve as they grow.

· Moderate (Grade 3): 50–70% narrowing; symptoms like stridor and breathing difficulty may be more noticeable.

· Severe (Grade 4): More than 70% narrowing; significant breathing problems often require urgent intervention.

Your doctor will determine severity through endoscopic airway evaluation and imaging.

2. Will my child need treatment right away?

· Mild cases: Often monitored closely without immediate intervention. Many improve with growth and airway maturation.

· Moderate to severe cases: Usually require treatment to prevent breathing difficulties. Timing depends on symptoms and overall health.

· If your child has significant breathing problems or recurrent infections, more urgent treatment may be needed.

3. What treatment do you recommend for my child?

Treatment depends on severity and symptoms:

· Observation: For mild stenosis without symptoms, regular monitoring is key.

· Endoscopic surgery: Minimally invasive procedures like balloon dilation or laser removal of scar tissue are often used for mild to moderate stenosis.

· Open surgery: For moderate to severe cases, procedures like laryngotracheal reconstruction (LTR) or partial cricotracheal resection (PCTR) are performed to widen the airway.

· Tracheostomy: In some cases, a temporary tracheostomy may be needed to secure the airway before or during treatment.

Your child’s care team will tailor treatment based on the grade of stenosis, airway anatomy, and your child’s overall condition.

**GENOMIC DATA**

· Congenital SGS and Genetic Syndromes:  
 Congenital subglottic stenosis is rare and can be associated with genetic syndromes such as Trisomy 21 (Down syndrome) and CHARGE syndrome, indicating a developmental genetic component affecting airway cartilage formation.

· Idiopathic Subglottic Stenosis (iSGS) and Genetics:  
 Idiopathic SGS primarily affects middle-aged Caucasian women, suggesting a possible hormonal influence rather than a classic inherited genetic disorder. Studies investigating HLA genotyping found no significant genetic association within the major histocompatibility complex, indicating that if genetic susceptibility exists, it likely lies outside the HLA locus. Familial cases are rare but reported, supporting a possible genetic or familial predisposition in some patients.

· Molecular Mechanisms:  
 Emerging research points to a complex interplay of genetic susceptibility, growth factors, cytokines, fibroblast activity, hypoxia, and biomechanical stress contributing to the fibrotic scarring and airway narrowing seen in SGS

**DOCTOR PATIENT CONVERSATION**

Doctor: Hello! I want to discuss the results of your child’s airway evaluation. The findings show that your child has subglottic stenosis, which means there is narrowing just below the vocal cords.

Parent/Patient: What does that mean exactly? How serious is it?

Doctor: The subglottic area is the narrowest part of the airway in children. When it’s narrowed, it can make breathing more difficult and cause symptoms like noisy breathing or stridor. The severity can vary—from mild narrowing that may improve over time, to more significant narrowing that needs treatment.

Parent/Patient: What caused this? Was it something we did?

Doctor: Subglottic stenosis can be congenital, meaning present from birth, or acquired, often from prolonged intubation or injury to the airway. Sometimes, we don’t find a clear cause. It’s not your fault, and we have effective treatments available.

Parent/Patient: What kind of treatments are there? Will my child need surgery?

Doctor: Treatment depends on how severe the narrowing is and how much it affects breathing. Mild cases might just need monitoring. Moderate cases can sometimes be treated with minimally invasive procedures like balloon dilation. More severe cases may require surgery to widen the airway. We’ll tailor the plan to your child’s needs.

Parent/Patient: How soon do we need to start treatment?

Doctor: If your child is having significant breathing difficulties, we’ll want to start treatment soon to prevent complications. If symptoms are mild, we can monitor closely and plan treatment accordingly.

Parent/Patient: Are there risks with the treatments?

Doctor: Like any procedure, there are risks such as bleeding, infection, or need for repeat treatments. But these are relatively uncommon, and we take every precaution to keep your child safe.

Parent/Patient: What should I watch for at home?

Doctor: Watch for worsening noisy breathing, difficulty feeding, fatigue, or bluish color around the lips. If you notice these, seek medical attention promptly.

Parent/Patient: Will this affect my child long-term?

Doctor: Many children do very well after treatment and can breathe normally. Some may need ongoing follow-up, but with proper care, the outlook is good.

Parent/Patient: Thank you for explaining everything. What’s the next step?

Doctor: We’ll schedule the appropriate treatment or monitoring and provide you with information and support. Please feel free to call anytime with questions or concerns.

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**SUPERIOR CANAL DEHISCENCE SYNDROME**

ALTERNATIVE NAMES: Superior Canal Dehiscence Syndrome: A rare inner ear disorder characterized by an abnormal opening in the superior semicircular canal, leading to symptoms such as dizziness, vertigo, and hearing issues.

* SCDS: An abbreviation for Superior Canal Dehiscence Syndrome.
* Superior Semicircular Canal Dehiscence Syndrome: Another name for Superior Canal Dehiscence Syndrome, emphasizing the specific location of the dehiscence in the superior semicircular canal.
* SSCDS: An abbreviation for Superior Semicircular Canal Dehiscence Syndrome.
* Third Mobile Window Syndrome: A collective term for the symptoms caused by various types of dehiscences around the labyrinth, including Superior Canal Dehiscence Syndrome.
* Labyrinthine Dehiscence: A term used to describe dehiscences around the labyrinth, which can include Superior Canal Dehiscence Syndrome.
* Labyrinthine Fistula: Another term for dehiscences around the labyrinth, which can be associated with Superior Canal Dehiscence Syndrome.
* Otic Capsule Dehiscence: A term referring to dehiscences involving the otic capsule, which can be related to Superior Canal Dehiscence Syndrome.
* Minor's syndrome: An alternative name for Superior Canal Dehiscence Syndrome, named after its discoverer, Lloyd B. Minor. However, this term is also used for an unrelated condition.

**DEFINITION / DESCRIPTION**

Superior canal dehiscence syndrome is a rare condition where there’s a hole in the bone that covers a key inner ear structure. This structure — the superior semicircular canal — is one of three coiled tubes inside your inner ear. An important part of your vestibular system, this canal is filled with fluid that shifts when you move. Your brain uses this information to regulate your balance.

But with SCDS, an opening (dehiscence) or thinning bone interferes with balance signals. It can allow sounds to travel to your brain. This can change your hearing.

The good news is that SCDS is treatable. If you’re having symptoms, a healthcare provider can likely help.

**CAUSES**

Superior canal dehiscence syndrome happens when there’s a hole in the bone covering your superior semicircular canal. The opening causes a communication breakdown between your brain and your inner ear.

Doctors don’t know for sure what causes problems with the bone. But it’s likely that many factors play a role. It may be that the bone near your superior canal doesn’t thicken enough during fetal development. Infections or head trauma (injury) may also cause SCDS. Or a bone that’s already thin may get even thinner with age.

**SIGNS / SYMPTOMS**

### **Symptoms of superior canal dehiscence syndrome**

SCDS symptoms can impact both your hearing and balance. They include:

* Hearing loss (especially at low frequencies)
* Heightened sensitivity to sound (hyperacusis)
* Hearing your voice or internal sounds, like breathing or blinking, as unusually loud
* Hearing noises in your head that happen in time with your pulse or heartbeat (pulsatile tinnitus)
* The feeling that objects are moving when they shouldn’t be (oscillopsia)
* Feeling like you’re spinning when you’re not (vertigo)

Activities that change the pressure in your brain or ear can trigger vertigo and oscillopsia. Examples include coughing, exercising, heavy lifting, hearing loud noises and sneezing.

**DIAGNOSIS METHODS**

You’ll likely see a neurotologist to diagnose and treat your condition. A neurotologist is a doctor who specializes in brain- and nerve-related concerns that also involve your ears. They may order several tests, including:

* Computed tomography (CT scan): This imaging procedure can show the opening in your bone covering the superior semicircular canal.
* Hearing test: An audiologist will test your hearing.
* VEMP test: This test provides information about how the parts of your inner ear are working to control your balance. You’ll move your head to the left or right, or look up, while staring at a target and listening to a series of tones.

**TREATMENT OPTIONS**

Treatment for mild symptoms usually involves management techniques. Options include lifestyle changes or vestibular therapy to help with balance issues. If your symptoms are severe, you may need surgery to repair the bone.

#### **Management techniques**

You can manage symptoms by avoiding activities that trigger balance issues. You can also protect your ears from excess noise. Things you can do include:

* Wearing earplugs
* Avoiding loud environments when possible
* Whispering (if your voice sounds too loud)
* Avoiding activities that trigger balance problems, like heavy lifting
* Avoiding activities that can cause pressure changes in your ear, like flying

Seeking treatment from a certified vestibular physical therapist can help manage symptoms. They can teach you exercises to help improve your balance and reduce your risk of falls.

#### **Surgery**

You may need surgery if management isn’t helping enough. The most common surgical repairs for SCDS include:

* Plugging the opening: This technique closes the opening using bone or connective tissue. This procedure involves accessing your inner ear through an opening in your skull or behind your ear.
* Resurfacing: This repair technique covers the opening without closing it completely. It’s less common than plugging.

Your healthcare provider may use both plugging and resurfacing techniques during surgery.

**OUTLOOK / PROGNOSIS**

Most people are able to manage symptoms once they learn techniques to manage them. But the actual opening won’t go away on its own. Unlike other bones in your body, the bone that surrounds your superior semicircular canal doesn’t repair itself.

SCDS surgery decreases or relieves symptoms for most people. Some studies show that certain symptoms resolve more quickly following surgery than others. For example, hearing loss and vertigo may improve faster than headaches and brain fog.

Talk to your healthcare provider about what you should expect from surgery.

**POSSIBLE COMPLICATIONS**

With severe SCDS, you may develop brain-related symptoms, like brain fog and headaches. You may also have mental health issues. Isolating to avoid exposing yourself to unpleasant sounds can lead to anxiety and depression.

This is why it’s so important to see a healthcare provider if you’re experiencing issues. You don’t have to accept a reduced quality of life just because you have SCDS.

**WHEN TO SEE A DOCTOR / RED FLAG**

* Sudden or severe hearing loss
* Intense or persistent vertigo (spinning sensation or dizziness) that does not subside
* Severe headaches or neurological symptoms such as visual changes or confusion
* Signs of infection like fever or discharge/drainage from the ear
* Persistent or worsening symptoms that affect your daily life, such as hearing difficulties, balance problems, or tinnitus (ringing in the ears)
* Symptoms triggered by pressure changes or loud noises that interfere significantly with activities

**DIFFERENTIAL DIAGNOSIS**

***Diagnostic Considerations***

These include the following:

* Patulous eustachian tube
* Ménière disease
* Vestibular neuritis
* Vestibular schwannoma of the skull base

***Differential Diagnoses***

* Benign Paroxysmal Positional Vertigo
* Inner Ear, Evaluation of Dizziness
* Inner Ear, Labyrinthitis
* Inner Ear, Meniere Disease, Medical Treatment
* Migraine-Associated Vertigo
* Multiple Sclerosis
* Otosclerosis
* Ototoxicity
* Perilymphatic Fistula
* Syphilis

**EPIDEMIOLOGY**

### Frequency

*United States*

The true incidence of persons with symptomatic SCDS is currently unknown. One study of 1000 cadaveric temporal bones revealed that a dehiscence of bone that overlies the superior canal was present in approximately 0.5% of temporal bone specimens. In an additional 1.4% of the specimens, the bone was markedly thin (≤ 0.1 mm) compared with the normal bone.

### Race

SCDS has no racial bias.

### Sex

SCDS appears to affect males and females equally.

### Age

In 2000, Minor reported that, in his original series of 17 patients, the median age at diagnosis was 40 years (range, 27-70 y)

**PREDEFINED Q & A SETS**

Q1: What is Superior Canal Dehiscence Syndrome (SCDS)?  
A1: SCDS is a condition caused by a thinning or absence of bone overlying the superior semicircular canal in the inner ear. This “third window” in the ear creates abnormal sound and pressure sensitivity, leading to symptoms like dizziness, vertigo, hearing loss, and tinnitus.

Q2: What causes SCDS?  
A2: The exact cause is not always clear. It may be congenital (present from birth due to thin bone), or develop after head trauma, chronic pressure changes, or increased intracranial pressure. It involves a defect in the bony covering of the superior semicircular canal.

Q3: What are the common symptoms of SCDS?  
A3: Typical symptoms include:

* Vertigo or dizziness triggered by loud sounds, pressure changes, or straining
* Autophony (hearing your own voice or body sounds unusually loud)
* Conductive hearing loss, especially in low frequencies
* Pulsatile tinnitus (hearing heartbeat-like sounds)
* Ear fullness or discomfort

Q4: How is SCDS diagnosed?  
A4: Diagnosis is based on clinical symptoms, audiometric hearing tests showing conductive hearing loss, vestibular tests (like VEMP), and confirmed by high-resolution CT imaging demonstrating the bony defect over the superior semicircular canal.

Q5: What other conditions can mimic SCDS?  
A5: Conditions like Ménière’s disease, perilymphatic fistula, vestibular migraine, benign positional vertigo, otosclerosis, and acoustic neuroma can have similar symptoms. Imaging and specialized tests help differentiate them.

Q6: When should I see a doctor about SCDS?  
A6: See a doctor if you have persistent or severe dizziness, hearing loss, pulsatile tinnitus, or symptoms triggered by sound or pressure that interfere with daily life. Sudden worsening or neurological symptoms require urgent evaluation.

Q7: What treatments are available for SCDS?  
A7: Treatment may include avoiding triggers (like loud noises and straining), using hearing aids, or in more severe cases, surgery to repair or resurface the dehiscent canal to eliminate symptoms.

Q8: Is surgery for SCDS safe?  
A8: Surgery can be effective, but as with any operation near the inner ear and brain, there are risks including hearing loss, dizziness, or infection. Careful evaluation by a specialist is essential before considering surgery.

Q9: Can SCDS get worse over time?  
A9: Symptoms can fluctuate, and some patients experience progressive symptoms, especially if exposed to head trauma or increased intracranial pressure. Regular monitoring is important.

Q10: Can I live a normal life with SCDS?  
A10: Many people manage their symptoms with lifestyle adjustments and medical care. With proper diagnosis and treatment, most patients have a good quality of life.

### **Is superior canal dehiscence syndrome a disability?**

It may be, depending on your symptoms and the type of work you do. If you have a hearing issue, you can request reasonable accommodations to do your job, according to the Americans with Disabilities Act. Ask your healthcare provider for guidance on how to apply for accommodations for work.

**DOCTOR-PATIENT CONVERSATIONS**

Doctor: Hello! I want to talk with you today about a condition called Superior Canal Dehiscence Syndrome, or SCDS for short. Have you heard of it before?

Patient: No, I haven’t. What is it?

Doctor: SCDS is a rare condition that affects the inner ear. Normally, the superior semicircular canal—a part of your balance system—is covered by a thin bone. In SCDS, there is a small hole or thinning in this bone. This opening lets sound and pressure move where they shouldn’t, causing symptoms affecting your hearing and balance.

Patient: What kind of symptoms should I expect?

Doctor: Common symptoms include dizziness or vertigo triggered by loud noises, sudden pressure changes like coughing or sneezing, and sometimes hearing your own heartbeat or your voice unusually loudly — this is called autophony. You may also notice hearing loss, especially for low tones, or ringing in your ears known as tinnitus.

Patient: That sounds uncomfortable. How do you find out if I have SCDS?

Doctor: To diagnose SCDS, we start with a detailed history and hearing and balance tests. A special hearing test called audiometry may show a particular pattern of hearing loss. We also use high-resolution CT scans of your inner ear to look for that bone opening. Sometimes balance tests help us understand how the inner ear is affected.

Patient: Is it something that can get worse over time?

Doctor: Symptoms can fluctuate and sometimes worsen, especially if you have head injuries or pressure changes in your skull. But many people live well with careful management. If symptoms are mild, we often just monitor and avoid triggers. For more severe cases, surgery can help close the opening and relieve symptoms.

Patient: What treatment options do I have?

Doctor: Treatment depends on how much your symptoms affect you. Avoiding loud noises and sudden pressure changes can help. If hearing loss is significant, hearing aids may be useful. Surgery is considered if symptoms are severe and disabling; it involves repairing the bone defect to restore normal ear function.

Patient: When should I definitely see a doctor urgently?

Doctor: If you experience sudden severe hearing loss, persistent vertigo that doesn’t improve, worsening balance problems, or new neurological symptoms like headaches, confusion, or vision changes, you should seek care promptly. These could be signs of complications.

Patient: Is there anything I can do at home to feel better?

Doctor: Yes, protecting your ears from loud noises, avoiding heavy lifting or straining that changes pressure in your head, and managing stress can help reduce symptoms. Regular follow-up is important so we can adjust your care as needed.

Patient: Thank you, doctor. It’s helpful to understand what’s going on.

Doctor: You’re welcome! We will work together to manage your symptoms and maintain your quality of life. Please feel free to ask any questions as we go along.

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<https://emedicine.medscape.com/article/857914-clinical>

**SENSORINEURAL HEARING LOSS (SNHL)**

ALTERNATIVE NAMES: Alternative names for sensorineural hearing loss include SNHL , which stands for sensorineural hearing loss. It is also referred to as inner ear hearing loss , and it is sometimes called sensory hearing loss. Additionally, it is known as presbycusis when it is related to age-related hearing loss.

**DEFINITION / DESCRIPTION**

Sensorineural (*sen-suh-ree-NUR-uhl)* hearing loss, also known as SNHL, occurs because of damage to your inner ear. It can happen suddenly due to an illness, injury or exposure to loud noise, like an explosion. Alternatively, SNHL can develop gradually over time as part of the aging process. And in some cases, people are born with it.

SNHL has a significant impact on your hearing ability. You may not be able to hear soft sounds, and even loud noises may seem muffled. That’s because tiny receptors in your inner ear called hair cells are damaged. These hair cells are responsible for converting sound waves into signals that your brain interprets as sound.

Unfortunately, these cells can’t repair themselves, making SNHL a permanent condition in most cases. But wearing hearing aids or other hearing devices can greatly improve your hearing and quality of life.

#### **Types of sensorineural hearing loss**

There are a few different types of sensorineural hearing loss:

* Unilateral sensorineural hearing loss affects one ear.
* Bilateral sensorineural hearing loss affects both ears.
* Asymmetrical sensorineural hearing loss affects both ears, but hearing loss is worse in one ear.
* Sudden sensorineural hearing loss is a medical emergency that happens within 72 hours of an injury, illness or exposure to loud noise (like fireworks or gunshots).

#### **How common is sensorineural hearing loss?**

Sensorineural hearing loss, particularly the age-related variant, is one of the most common types of hearing loss in adults. It often develops between the ages of 50 and 70.

Sudden sensorineural hearing loss — which happens immediately or over the course of a few days — affects between 1 and 6 out of 5,000 people every year.

**CAUSES**

Sensorineural hearing loss happens because of damage to your inner ear. Specifically, it occurs when the tiny hair cells in your cochlea or vestibulocochlear nerve get damaged or destroyed.

Some people are born with SNHL because of a lack of oxygen or other complications during fetal development. Others develop it later in life due to illnesses, trauma, exposure to loud noises or typical aging.

#### **RISK FACTORS**

A risk factor is something that increases your chance of developing a certain condition. Diseases and conditions that raise your risk for SNHL include:

* Acoustic neuroma. This is a noncancerous tumor that affects your inner ear.
* Aging. Simply growing older increases your risk for hearing loss, as the tiny hair cells in your inner ear deteriorate over time.
* Ménière’s disease. This chronic ear disorder causes symptoms like vertigo and tinnitus (ringing in your ears).
* Ototoxicity. This is inner ear damage that results from taking certain medications.
* Systemic conditions. Some conditions — like diabetes and meningitis — can disrupt blood flow to your inner ear or inflame your vestibulocochlear nerve.
* Traumatic brain injury. Brain trauma can cause fractures, blood flow disruption and increased intracranial pressure — all of which can lead to SNHL.

In some cases, healthcare providers can’t identify an exact cause. When this happens, they call it idiopathic SNHL.

**SIGNS / SYMPTOMS**

If you have sensorineural hearing loss, you might notice:

* It’s easier to hear deep voices as opposed to high-pitched voices.
* It’s harder to hear in noisy environments.
* Other people sound like they’re mumbling.
* You have ringing in your ears.
* You have trouble following conversations with multiple people.
* Your hearing is muffled.

**DIAGNOSIS METHODS**

A healthcare provider (usually an otolaryngologist) will start with a physical exam. They’ll gently feel around the outside of your ear. Then, they’ll look inside your ear with a lighted tool called an otoscope. This helps them find any abnormalities that might interfere with your hearing.

Your healthcare provider may also ask questions like:

* Did you lose your hearing suddenly or gradually?
* If it was gradual, when did it start?
* Do you hear better out of one ear?
* Have you had any recent illnesses or traumas?
* Do you have a family history of hearing loss?

#### **What tests will be done to diagnose this condition?**

An audiologist will run some hearing tests to determine whether you have SNHL. These tests might include:

* Acoustic reflex test, which measures how tightly your stapedial muscle (a small muscle in your middle ear) contracts in response to loud sounds.
* Auditory brainstem response, which uses electrodes to track your brain waves in response to sounds.
* Bone conduction test, which sends sound directly to your inner ear and helps determine which type of hearing loss you have.
* Otoacoustic emissions (OAEs), which measures sound-related vibrations from your inner ear and can show whether you have a blockage or hearing loss.
* Pure-tone audiometry, which determines the quietest sounds you can hear at different frequencies (pitches).
* Speech audiometry, which tests how well you understand words and records the softest speech you can repeat.
* Tuning fork exams, exams like Weber’s test and the Rinne test, which determine whether you have sensorineural or conductive hearing loss (can be done by your otolaryngologist).
* Tympanometry, which tells your healthcare provider how well your eardrum moves in response to sounds.

Your healthcare provider may also need to examine your cranial nerves or take imaging tests, like an MRI or CT scan.

**TREATMENT OPTIONS**

Sensorineural hearing loss treatments include:

* Hearing aids. These devices amplify sounds like tiny speakers and make it easier for you to hear.
* Cochlear implants. These surgically placed devices bypass your inner ear and create a new pathway for sounds to reach your brain.
* Medications. If inflammation or disease causes SNHL, your healthcare provider might prescribe medications like corticosteroids.
* Bone-anchored hearing aids (BAHA). These surgically implanted devices use vibrations to send sounds to your inner ear. BAHA might be helpful if you have single-sided (unilateral) sensorineural hearing loss.
* Active surveillance. Your healthcare provider may recommend “watchful waiting.” This is especially true if you have mild sensorineural hearing loss.

**PREVENTION TIPS**

You can’t always prevent SNHL because many factors are out of your control. But there are things you can do to reduce your risk:

* Discuss any medication side effects with your healthcare provider.
* Get your hearing tested regularly.
* Wear ear protection (like earplugs or earmuffs) in noisy environments. Hearing loss from noise exposure is 100% preventable.

**OUTLOOK / PROGNOSIS**

Overall outlook depends on the underlying cause and severity of hearing loss. In most cases, SNHL is permanent. But hearing aids or cochlear implants can improve your hearing abilities and keep you from missing out on what’s happening around you.

If you have sudden sensorineural hearing loss, it’s important to seek medical treatment right away. Early intervention may result in better outcomes.

**WHEN TO SEE A DOCTOR / RED FLAG**

You should tell your healthcare provider if you:

* Develop vertigo or other balance issues.
* Have trouble hearing in loud environments or when multiple people are speaking.
* Hear ringing in your ears.
* Need to turn up the volume on a regular basis.
* Notice a sudden change in your hearing.

**DIFFERENTIAL DIAGNOSIS**

**Congenital**

Hearing loss is the most common congenital sensory disorder, often requiring pediatricians' expertise in conjunction with medical geneticists and pediatric otolaryngologists.Congenital causes of hearing loss can be divided into genetic and environmental. Genetic causes can be further divided into non-syndromic (70%) and syndromic (30%).

**Genetic Non-syndromic**

Of the nonsyndromic causes, the most common inheritance pattern is autosomal recessive, followed by autosomal dominant. More than 60 autosomal recessive genes have been identified, the most common being the gap junction beta 2 (GJB2) gene, which accounts for half of nonsyndromic hearing loss cases. GJB2 encodes connexin 26, a protein important for the proper functioning of the cochlea's potassium ion channels. In these patients, due to an absence of other physical findings or relevant history, diagnostic workup remains a challenge but primarily consists of genetic testing, electrocardiogram (ECG), and testing for cytomegalovirus infection.

**Genetic Syndromic**

In children who have hearing loss due to syndrome, determining the underlying cause is often more important as the other clinical features can be severe. More than 400 syndromes have been identified with hearing loss as a feature; however, only a small number of these account for most cases of SNHL. Outlined below are the key features of the most common syndromes seen in children.

1. Waardenburg syndrome is the most common, with SNHL being a significant feature found in over two-thirds of patients. The other key feature is pigmentation abnormalities of the eyes, skin, and cochlea.
2. Usher syndrome is one of the most common autosomal recessive causes of syndromic hearing loss. This condition is characterized by hearing loss and visual loss due to a progressive SNHL and retinitis pigmentosa.
3. Pendred syndrome classically presents with a varying degree of SNHL, vestibular dysfunction, and a thyroid goiter. Along with Usher syndrome, it is another one of the most common autosomal recessive causes of syndromic hearing loss. A specific mutation in SLC26A4 occurs in around half of the affected patients.
4. Jervell and Lange-Nielsen syndrome is also inherited with an autosomal recessive pattern. The key feature, along with SNHL, is a prolonged QT interval seen on the ECG. These patients can suffer from or have a family history of syncope, sudden death, or long QT syndrome.
5. Alport syndrome is inherited in an X-linked manner and occurs due to a defect in type IV collagen. It classically presents with glomerulonephritis, end-stage kidney disease, eye abnormalities, and a bilateral SNHL. The hearing loss is initially in high frequency, but the lower frequencies begin to get affected as it worsens. Hematuria and proteinuria are key signs as the condition progresses.

***Environmental***

1. Intrauterine infection (toxoplasmosis, cytomegalovirus, herpes, rubella)
2. Alcohol, smoking
3. Ototoxic drugs
4. Premature births, hypoxia, neonatal jaundice

**Sensorineural Hearing Loss**

Acute sensorineural hearing loss is defined as a hearing loss greater than 30 dB in at least three consecutive audiometric frequencies over 72 hours.This is usually classified as an otolaryngologic emergency condition, which requires prompt management. There are several possible causes of SNHL, including trauma, infection, malignancy, and Meniere's disease; however, there is no identifiable cause of their hearing loss in most patients and will be classed as idiopathic. As part of the work-up, patients should have a pure tone audiogram as soon as is possible. Often routine blood tests and an autoimmune screen are sent off, although practice varies between departments.

**Presbycusis**

Presbycusis or age-related hearing loss can be defined as a progressive bilateral SNHL of mid to late adult-onset. The diagnosis of presbycusis is one of exclusion, and primary causes such as otosclerosis, Meniere's disease, and cytotoxicity, amongst many others, must be excluded first.

It is commonly associated with degeneration of cochlear hair cells, mainly OHCs in the cochlea's basal portion, and the changes begin in the basal end of the cochlea and spread towards the apex as the condition worsens. Patients typically present with a slowly deteriorating hearing loss, especially in the presence of background noise. It is often a lack of clarity rather than a loss of volume that the patient describes. Tinnitus is frequently an accompanying symptom and can be the most challenging aspect for the patient.

A typical PTA will show a gradual downsloping hearing loss towards higher frequencies. It has been shown that once a certain amount of hearing loss has occurred (roughly 70-80 dB), further progression is slow, especially in the higher frequencies.In terms of management options, hearing aids often benefit patients and prevent social isolation and depression.

**Noise-induced Hearing Loss**

This condition occurs when a patient experiences hearing loss due to excessive noise exposure, either recreational or occupational. Occupational noise exposure is one of the most prevalent, potentially preventable health conditions. It has a slight male predominance and usually affects the middle-aged population.

The symptoms present similar to most SNHL conditions, with the insidious progression of worsening hearing loss over many years, often accompanied by tinnitus. Hyperacusis is found in 40% of tinnitus sufferers, and its severity can be determined using a hyperacusis questionnaire.

Bedside otological examination is usually normal, and the diagnosis is generally based on the history combined with the classical finding of a notched appearance at 4kHz, which appears to start recovering at 8kHz on a pure tone audiogram.

This is known as the Carharts notch; however, it is not always present. Without a previous noise exposure history, it is not indicative of NIHL. Once a diagnosis is reached, it is essential to reduce further noise exposure as much as possible using ear protection.

The Control of Noise at Work Regulations of 2005 sets out a framework for employers to ensure their employees' safety based on their average occupational sound exposure.

**Others**

Meniere's disease is characterized by a triad of spontaneous episodic vertigo, hearing loss, and tinnitus. Patients may also experience aural fullness. Their PTA usually shows an up sloping curve indicating a low-frequency moderate SNHL. PTA remains the most useful investigation, but a brain MRI is done to rule out lesions such as a vestibular schwannoma.

Treatment ranges from conservative such as alcohol, coffee, and salt restriction, to treatments targeted at the symptoms most affecting the patients. Vestibular suppressants such as prochlorperazine can be helpful. Hearing aids and tinnitus retraining therapy can also be used. Psychological support can be key in those patients who suffer from the psychological complications of the condition.

Autoimmune SNHL was first described in 1979 by McCabe as a rapidly progressive bilateral SNHL that responded to steroid therapy. Several antigens have been implicated in the etiology. Many systemic autoimmune disorders have been reported, such as Wegener's granulomatosis, rheumatoid arthritis, and systemic lupus erythematosus.

Patients usually present in their early twenties. The condition has a female predominance. Symptoms often start in one ear before becoming bilateral in the majority of patients. Aside from audiometric evaluation, blood tests (erythrocyte sedimentation rate, antinuclear antibody) looking specifically at autoimmune conditions are sent off.

Treatment is directed by rheumatologists and includes steroid therapy, long-term intratympanic steroid injections, cyclophosphamide, and IL-1 receptor antagonists.

A head injury that results in a temporal bone fracture can lead to conductive hearing loss or mixed SNHL. Otic capsule fractures cause a severe SNHL through various mechanisms, including disruption of the membranous labyrinth, hemorrhage into the cochlea, perilymph fistula, and avulsion or trauma of the cochlear nerve. In those cases that result in bilateral deafness, a cochlear implant is a treatment option.

Ototoxic agents can cause SNHL in many ways. Aminoglycosides such as gentamicin cause hair cell death resulting in permanent hearing loss and balance dysfunction. This can occur after repeated administration of systemic therapy. The hearing loss initially affects the higher frequencies but continues progressively to the lower frequencies as more hair cells are damaged. Loop diuretics are thought to affect the stria vascularis and cause acute but completely reversible effects.

**EPIDEMIOLOGY**

The incidence of sensorineural hearing loss varies in different countries. In the United States, sudden SNHL affects between 5-27 per 100,000 people each year, with approximately 66,000 new annual cases. Due to different studies using varying thresholds when classifying hearing loss, there is little consensus in the literature regarding the epidemiology of age-related hearing known as presbycusis. In presbycusis, hearing loss prevalence doubles every decade of life from the second through to the seventh decade, and is nearly universal past the eighth decade of life. Another important cause of hearing loss in the adult population is noise-induced hearing loss (NIHL). It has been estimated that 16% of adults worldwide disabling hearing loss is occupational noise related. This remains a common occupational disease despite legislation in place in most developed countries to prevent NIHL.

Congenital hearing loss is nearly always sensorineural in nature, and can have various etiologies. In patients with robust prenatal care, congenital infectious causes such as cytomegalovirus are rare and the most common causes are genetic. There are many genetic syndromes with hearing loss as a component, and SNHL developing in childhood warrants a thorough workup

**PREDEFINED Q & A SETS**

Q1: What is sensorineural hearing loss?  
A1: Sensorineural hearing loss occurs when there is damage to the inner ear hair cells or the auditory nerve pathways. This damage prevents sound signals from being properly transmitted to the brain, causing permanent hearing impairment.

Q2: What causes sensorineural hearing loss?  
A2: Causes include aging (presbycusis), exposure to loud noise, infections, trauma, certain medications (ototoxic drugs), genetic factors, and sudden sensorineural hearing loss (which can be idiopathic).

Q3: What are the common symptoms?  
A3: Symptoms include difficulty hearing soft sounds, muffled hearing, trouble understanding speech especially in noisy environments, tinnitus (ringing in the ears), and sometimes dizziness or balance issues.

Q4: Is sensorineural hearing loss permanent?  
A4: Yes, it is generally permanent because the inner ear hair cells cannot regenerate. However, symptoms can be managed effectively with hearing aids and other assistive devices.

Q5: How is sensorineural hearing loss diagnosed?  
A5: Diagnosis is made through hearing tests (audiometry) to differentiate sensorineural from conductive hearing loss, combined with medical history and sometimes imaging like MRI to exclude other causes.

Q6: What treatment options are available?  
A6: While there is no cure, treatments include hearing aids to amplify sound, cochlear implants for severe cases, and strategies to manage tinnitus. Avoiding further noise exposure is important.

Q7: Can sensorineural hearing loss get worse over time?  
A7: It can be progressive, especially age-related loss or due to ongoing noise exposure. Early intervention with hearing aids improves long-term outcomes and quality of life.

Q8: When should I see a doctor for sudden hearing loss?  
A8: Sudden sensorineural hearing loss occurring over hours to days is an emergency. Seek medical care promptly, as early treatment with steroids improves chances of hearing recovery.

Q9: Can sensorineural hearing loss affect daily life?  
A9: Yes, it can cause communication difficulties, social isolation, fatigue from straining to hear, and impact relationships. Supportive therapies and hearing rehabilitation help.

Q10: Are there any preventive measures?  
A10: Protect your ears from loud noise by using earplugs or reducing exposure, avoid ototoxic medications when possible, and have regular hearing check-ups if at risk

**DOCTOR-PATIENT CONVERSATIONS**

Doctor: Hello! I want to talk with you about your recent hearing test results and explain what sensorineural hearing loss means. Have you heard the term before?

Patient: Not really, doctor. What is sensorineural hearing loss?

Doctor: Sensorineural hearing loss happens when there is damage to the inner ear—specifically to the tiny hair cells that help convert sound into nerve signals—or to the auditory nerve pathways leading to the brain. This type of hearing loss is usually permanent because these hair cells do not regenerate.

Patient: What symptoms might I have?

Doctor: Common symptoms include difficulty hearing soft sounds, trouble understanding speech—especially in noisy environments—muffled hearing, and sometimes ringing in your ears, called tinnitus. You may also notice hearing is better for some pitches than others.

Patient: What causes sensorineural hearing loss?

Doctor: Several things can cause it. Age-related changes, known as presbycusis, are a common cause. Exposure to loud noises over time, certain medications that can affect hearing (called ototoxic drugs), infections, head trauma, or genetic factors can also lead to this. Sometimes, sudden hearing loss can occur without a clear cause, which requires urgent evaluation.

Patient: Is there a cure for it?

Doctor: Unfortunately, sensorineural hearing loss is generally not reversible. However, we have many ways to help you manage it. Hearing aids are the most common treatment, amplifying sounds so you can hear better. In severe cases, cochlear implants may be an option. Also, protecting your hearing from further damage is very important.

Patient: How do you confirm this diagnosis?

Doctor: We do hearing tests—called audiometry—that help us understand the degree and pattern of your hearing loss. Sometimes we use imaging like MRI to rule out other causes like tumors. Your test results showed sensorineural loss, meaning the problem lies in the inner ear or neural pathways.

Patient: What happens next? Do I need more tests?

Doctor: If no other concerning symptoms or risk factors are present, hearing aids may be the next step. If your hearing loss appeared suddenly or worsens, or if you have balance problems or vertigo, we would investigate further. We’ll also provide advice on communication strategies to make daily life easier.

Patient: How can I protect my remaining hearing?

Doctor: Avoiding loud noises, using ear protection, and not using medications that can harm hearing unnecessarily are key. Regular check-ups help us monitor your hearing. Also, avoiding smoking and controlling chronic conditions like diabetes can help protect your ears.

Patient: Thank you, doctor. This helps me understand what’s going on.

Doctor: You’re welcome. Please ask any questions anytime. We’ll work together to manage your hearing and maintain your quality of life.

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Sialadenitis

ALTERNATIVE NAMES

**DEFINITION / DESCRIPTION**

Sialadenitis” is the medical term for inflammation of one or more of your salivary glands. Salivary glands are the glands that make your saliva (spit). Saliva helps you swallow, digest food and protects your teeth from harmful bacteria. There are three major salivary glands:

* Parotid glands, located in front of each ear.
* Submandibular glands, located in the very back of your mouth, below your jaw.
* Sublingual glands, located under your tongue on the floor of your mouth.

In addition to your major salivary glands, you have hundreds of minor salivary glands. These smaller glands are in your lips, inner cheeks and all throughout the linings of your mouth and throat.

Sialadenitis mostly affects your parotid and submandibular glands.

It can be an acute (sudden), chronic (long-term), or recurrent (returning) condition. Sialadenitis is rare.

**CAUSES**

Sialadenitis causes include:

* Bacterial or viral infection.
* Dehydration.
* Recent illness.
* Certain medications, such as diuretics, antihistamines and beta-blockers.
* Sjögren’s syndrome, an autoimmune disease.
* Poor oral hygiene

**SIGNS / SYMPTOMS**

Symptoms of sialadenitis include:

* Enlargement, tenderness and discoloration of one or more salivary glands.
* Fever (when the inflammation leads to an infection).
* Decreased saliva (a symptom of both acute and chronic sialadenitis).
* Pain while eating.
* Dry mouth (xerostomia).
* Swelling in your cheek and neck region.

If you notice any of these symptoms, see a healthcare provider right away.

**DIAGNOSIS METHODS**

A healthcare provider will diagnose sialadenitis through a physical examination and a history of your symptoms. They may also examine your salivary glands with an endoscope (a tiny camera with a light attached).

Your healthcare provider might use a salivary gland scan to find blockages. They’ll use ultrasound or computed tomography (CT) scans for this test.

In some cases, your provider might refer you to an otolaryngologist (ENT) for further evaluation. An ENT is a provider who specializes in diagnosing and treating ear, nose and throat conditions.

**TREATMENT OPTIONS**

There are several ways to manage sialadenitis, including:

* Antibiotics.
* Home remedies.
* Nonsurgical treatments.
* Surgery.

Treatment depends on the severity of your condition.

#### **Antibiotics**

Antibiotics are usually the first line of sialadenitis treatment. The most common antibiotics for sialadenitis include dicloxacillin, cephalosporin or clindamycin.

#### **Home remedies**

Your healthcare provide may also recommend home remedies to help decrease pain and increase saliva flow, including:

* Staying hydrated.
* Drinking lemon juice or sucking on sour candy.
* Using warm compresses.
* Massaging your salivary glands.
* Improving your oral hygiene.

#### **Nonsurgical treatments**

In some cases, your healthcare provider may deliver intravenous (IV) medications through a vein in your arm. They’ll give you saline or dextrose solutions to keep you hydrated and antibiotics to fight infection.

#### **Surgical treatments**

If sialadenitis doesn’t respond to other treatments, your healthcare provider may recommend surgery. If you’ve developed an abscess (a pocket of pus), they’ll drain the infection and remove any stones or other blockages. They may also use an endoscope to examine your salivary gland. Healthcare providers call this procedure a sialendoscopy.

**PREVENTION TIPS**

You may not be able to prevent sialadenitis completely. But there are a few things you can do to reduce your risk:

* Drink plenty of water.
* Practice good oral hygiene.
* Eat a diet that’s healthy for you.
* Avoid smoking and using other tobacco products.

**OUTLOOK / PROGNOSIS**

Generally, people with sialadenitis feel better in about one week with conservative treatment. If you need surgery, it could take two weeks for swelling and bruising to subside.

Sialadenitis can recur (return) in some people. While you can’t always prevent recurrence, you can reduce your risk by practicing good oral hygiene, staying hydrated and avoiding smoking.

#### **Is sialadenitis fatal?**

Left untreated, a salivary gland infection can spread to the tissues of your head and neck. This is a life-threatening event. If you have difficulty breathing or swallowing, call 911 (or your local emergency number) or go to the emergency room immediately.

**WHEN TO SEE A DOCTOR / RED FLAG**

If you have lingering sialadenitis symptoms, such as facial swelling and fever that lasts for more than three days, schedule an appointment with your healthcare provider. They can find out what caused your swollen salivary gland and recommend appropriate treatment.

If you develop severe sialadenitis symptoms — such as difficulty breathing or swallowing — call 911 (or your local emergency number) or go to the nearest emergency room. You may need emergency treatment to avoid life-threatening complications.

**DIFFERENTIAL DIAGNOSIS**

| **Cause Type** | **Conditions Included & Notes** |
| --- | --- |
| Infectious Causes | - Bacterial sialadenitis (most common: *Staphylococcus aureus*, *Streptococcus viridans*, anaerobes) often secondary to obstruction (e.g., stones) or decreased salivary flow.  - Viral infections: Mumps virus (classic cause), coxsackievirus, parainfluenza, HIV-associated parotitis.  - Fungal infections (rare, usually immunocompromised patients). |
| Obstructive Causes | - Sialolithiasis (salivary stones): Leading cause of obstructive sialadenitis, most often in submandibular gland due to anatomy.  - Ductal strictures or stenosis.  - External duct compression. |
| Autoimmune/Inflammatory Causes | - Sjögren’s syndrome: Chronic bilateral, painless or mildly painful swelling.  - Sarcoidosis (Heerfordt’s syndrome includes parotid swelling).  - Systemic lupus erythematosus, granulomatosis with polyangiitis.  - Chronic sclerosing sialadenitis (Kuttner tumor) mimicking tumors. |
| Neoplastic Causes | - Salivary gland tumors can present similarly and cause secondary sialadenitis-like symptoms.  - Obstructive sialadenitis from tumor blocking ducts. |
| Other Causes | - Radiation-induced sialadenitis following radiotherapy (often chronic).  - Juvenile recurrent parotitis (idiopathic chronic inflammation in children).  - Idiopathic or drug-induced xerostomia leading to secondary infection.  - Post-operative or traumatic inflammation.  - Sialectasis and diverticula causing stasis and inflammation |

**EPIDEMIOLOGY**

* Overall incidence rates for salivary gland pathologies (including sialadenitis) have been reported around 59.94 per 100,000 population per year, with non-malignant inflammatory lesions like sialadenitis comprising a large majority.
* Acute suppurative parotitis, a common form of bacterial sialadenitis, accounts for about 0.01–0.02% of all hospital admissions in some populations.
* The submandibular gland accounts for approximately 10% of all sialadenitis cases of the major salivary glands.
* There are no strong race or sex predilections, although sialadenitis tends to affect older, debilitated, or dehydrated patients more frequently.
* Chronic sialadenitis is relatively common, whereas bacterial sialadenitis is less frequent today due to antibiotics and supportive care.
* Viral causes, such as mumps virus, have been historically significant causes of sialadenitis, especially in countries or populations without widespread vaccination.
* Hospital admission data from the UK showed an incidence of 27.5 per million population for sialadenitis admissions.
* The prevalence of salivary stones (sialolithiasis), a major cause of obstructive sialadenitis, is around 1.2%, with microcalculi forming throughout life but often passed without symptoms.
* Sialadenitis generally presents more frequently in adults but can occur at a wide age range

**PREDEFINED Q & A SETS**

#### **Who gets sialadenitis?**

Anyone can get sialadenitis. But it’s most common among adults older than 50, especially those who have salivary gland stones.

Sialadenitis can also occur in other age groups, including:

* Infants during the first few weeks of their life.
* People who are sick or recovering from surgery.
* People with dehydration, malnutrition or immunosuppression (when your body can’t fight disease).

#### **Is sialadenitis serious?**

Without proper treatment, sialadenitis can spread into the deep tissues of your head and neck, causing a severe infection. It’s important to see a healthcare provider right away if you have any sialadenitis symptoms, such as pain, swelling and fever.

#### **Is sialadenitis contagious?**

It depends on the cause. If sialadenitis is the result of an infection, then you can spread the infection to others through sneezing, coughing or coming into contact with infected saliva.

If a salivary gland stone caused sialadenitis, then there’s no risk of passing the condition on to someone else.

### **How long does it take to recover from sialadenitis?**

In most cases, salivary gland infections resolve after one week with conservative treatment.

If you had surgery, it could take about two weeks to fully recover.

### **Can sialadenitis go away on its own?**

Infected or swollen salivary glands often go away on their own or with conservative treatment. But if you have lingering symptoms, such as swelling, pain, fever or difficulty swallowing, call your healthcare provider right away. You may need emergency treatment.

**DOCTOR-PATIENT CONVERSATIONS**

Doctor: Hello! What brings you in today?

Patient: I've had pain and swelling under my jaw for a few days. It's quite uncomfortable, especially when I eat.

Doctor: That sounds like it could be sialadenitis, an inflammation of one of your salivary glands. The glands produce saliva, which helps with digestion and oral health. When they get blocked or infected, they can swell and become painful.

Patient: What causes this inflammation?

Doctor: It can be caused by a bacterial or viral infection, salivary gland stones blocking the ducts, or sometimes other conditions like autoimmune diseases. The blockage or infection causes saliva to build up, which leads to swelling and pain.

Patient: How do you find out if I have sialadenitis?

Doctor: We'll start with a physical exam, checking the swollen area. Sometimes we use imaging tests like ultrasound or CT scans to see if there are stones or other blockages. In some cases, we might take a sample if there's pus to identify the bacteria.

Patient: What treatment will I need?

Doctor: Depending on the severity, mild cases can improve with home care: staying well hydrated, warm compresses to the area, gently massaging the gland to help saliva flow, and good oral hygiene. If bacteria are involved, antibiotics are often necessary. In some severe cases, fluids might be given through an IV. If there is an abscess or persistent blockage, a minor procedure might be needed to drain it or remove stones.

Patient: Is this serious?

Doctor: Most cases respond well to treatment and get better in a few days to weeks. But if untreated, it can cause complications like abscesses or spread of infection, so it's important to follow treatment and come back if symptoms worsen.

Patient: What should I watch for that means I need to come back or see someone urgently?

Doctor: If the swelling becomes very large or hard, you have a high fever, difficulty opening your mouth or swallowing, severe pain, or if you feel short of breath, please seek medical help right away.

Patient: How can I prevent this from happening again?

Doctor: Drinking plenty of fluids, practicing good oral hygiene, and stimulating saliva flow with sour candies or lemon can help prevent blockages. If you have stones, sometimes they may need to be removed. If you have frequent infections, we might investigate further.

Patient: Thank you, doctor. That makes me feel better.

Doctor: You're welcome! We'll monitor your progress and I'm here if you have any questions.

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**Sinus headaches**

ALTERNATIVE NAMES

**DEFINITION / DESCRIPTION**

A sinus headache is a symptom of sinus infections (sinusitis). Sinus headaches make your face hurt. You may feel a constant, dull ache behind the eyes or in your cheekbones, forehead and the bridge of your nose. The pain gets worse when you move your head suddenly or you bend over. Typically, sinus headaches go away once a sinus infection runs its course

**CAUSES**

If you have a sinus headache, a viral or bacterial infection in your sinuses may be to blame. Sinuses are a series of connected hollow spaces behind your cheekbones, forehead and nose. Air that comes in through your nose travels through your sinuses on its way to your lungs.

Your sinuses are lined with tissue. They also make mucus that keeps your nose moist and traps intruders like bacteria, viruses, fungi and dust-carrying allergens.

Normally, free-flowing mucus carries off intruders before they can make trouble in your sinuses. But sometimes your sinuses’ reaction to intruders starts a chain reaction that leads to sinus headaches.

First, your sinuses start making more mucus. Mucus building up in your sinuses creates a place where intruders like bacteria and viruses can settle and grow. Growing intruders make sinus tissue swell, trapping mucus so it can’t flow from your sinuses. The result is swollen, irritated, fluid-filled sinuses that make your face feel achy and tender.

#### **What are sinus headache risk factors?**

Sinus headaches stem from sinus infections. Understanding sinus infection risk factors may reduce your risk of sinus headaches. According to the U.S. Center for Disease Control and Prevention, those risk factors include:

* Having the common cold.
* Seasonal allergies.
* Smoking and exposure to secondhand smoke.
* Structural issues within your sinuses. For example, nasal polyps or a deviated septum may trap mucus in your sinuses.
* Having a weak immune system or taking drugs that weaken your immune system increases your risk of infections, including sinus infections.

**SYMPTOMS**

Signs and symptoms of sinus headaches may include:

* Pain, pressure and fullness in the cheeks, brow or forehead
* Worsening pain if you bend forward or lie down
* Stuffy nose
* Fatigue
* Achy feeling in the upper teeth

### 

### **Sinusitis or migraine?**

Migraines and headaches from sinusitis are easy to confuse because the signs and symptoms of the two types of headaches may overlap.

Both migraine and sinusitis headache pain often get worse when you bend forward. Migraine can also be accompanied by various nasal signs and symptoms — including congestion, facial pressure and a clear, watery nasal discharge. These are due to involvement of the autonomic nervous system in a migraine attack. In fact, studies have shown that most people who see a health care provider for sinus headaches are found to have migraines instead.

Sinusitis, however, usually isn't associated with nausea or vomiting or aggravated by noise or bright light — all common features of migraines.

Sinusitis usually:

* Occurs after a viral upper respiratory infection or cold
* Includes thick, discolored nasal mucus
* Is associated with a decreased sense of smell
* Causes pain in one cheek or upper teeth

Headaches due to sinus disease often last days or longer, and migraines most commonly last hours to a day or two.

**DIAGNOSIS METHODS**

Your healthcare provider will perform a physical exam and ask about your symptoms. If your symptoms are severe or ongoing, you may also need imaging tests like X-rays or computed tomography (CT) scans.

Imaging tests show if your sinuses are blocked. If they aren’t, it may mean you have a different issue like a migraine or a tension headache. Migraine headaches and sinus headaches have common symptoms. Studies suggest 80% of people who thought they had sinus headaches had migraines.

**TREATMENT OPTIONS**

Sinus headaches happen because you have a sinus infection. Healthcare providers may treat bacterial infections with antibiotics. Viral infections typically go away without treatment.

Your healthcare provider may also recommend other medications to ease discomfort, like:

* Antihistamines to prevent allergy symptoms.
* Decongestants to reduce swelling in your nose and sinuses.
* Pain relievers to ease headache pain.
* Steroids to reduce inflammation.

**PREVENTION TIPS**

Whether or not you take preventive medications, you may benefit from lifestyle changes that can help reduce the number and severity of headaches. One or more of these suggestions may be helpful for you:

* **Avoid triggers.** If certain foods or odors seem to have triggered your headaches in the past, avoid them. Your provider may recommend you reduce your caffeine and alcohol intake and avoid tobacco.  
  In general, establish a daily routine with regular sleep patterns and regular meals. In addition, try to control stress.
* **Exercise regularly.** Regular aerobic exercise reduces tension and can help prevent headaches. If your provider agrees, choose any aerobic exercise you enjoy, including walking, swimming and cycling.  
  Warm up slowly, however, because sudden, intense exercise can cause headaches.  
  Obesity is also thought to be a factor in headaches, and regular exercise can help you maintain a healthy weight or lose weight.
* **Reduce the effects of estrogen.** If estrogen seems to trigger or make your headaches worse, you may want to avoid or reduce the medications you take that contain estrogen.  
  These medications include birth control pills and hormone replacement therapy. Talk with your provider about the appropriate alternatives or dosages for you.

**OUTLOOK / PROGNOSIS**

That depends on what caused you to have sinusitis. For example, viruses cause most sinus infections. When the viral infection clears up, the sinus headache goes away. That may take a week or so. Sinus issues that don’t go away may mean you have a bacterial or fungal sinus infection that requires treatment like an antibiotic or antifungal.

**WHEN TO SEE A DOCTOR / RED FLA**G

Most sinus headaches go away when sinus infections clear. Talk to a healthcare provider if your sinus issues don’t go away within a week or so.

**DIFFERENTIAL DIAGNOSIS**

* sinusitis (acute or chronic): Inflammation or infection (viral, bacterial, or fungal) of the paranasal sinuses causing mucosal swelling, obstruction, and pressure buildup. Symptoms include facial pain/pressure (especially with bending forward), nasal congestion, purulent nasal discharge, and sometimes fever.
* Allergic rhinitis: Chronic allergies causing nasal inflammation, congestion, and headache or facial pain mimicking sinus headache.
* Nasal polyps or anatomical variations: Nasal polyps, deviated septum, or narrow sinus ostia can cause sinus drainage issues and headaches.
* Migraine headache: Migraines often cause facial pain and symptoms resembling sinus headache, including congestion or watery nasal discharge. Migraines typically include nausea, photophobia, and phonophobia—features usually absent in sinus headache.
* Tension-type headache: Can cause bilateral or facial pain commonly mistaken for sinus headache.
* Cluster headache: Severe unilateral headache with autonomic symptoms including lacrimation and nasal congestion that may mimic sinus disease.
* Dental causes: Dental abscess or temporomandibular joint disorders (TMJ) causing facial pain.

**EPIDEMIOLOGY**

* Sinusitis affects roughly 11–15% of adults annually in the United States, with over 30 million people diagnosed each year. This includes acute and chronic sinusitis cases, which can cause sinus headaches as part of their symptoms.
* Globally, sinusitis is a common condition that leads to millions of healthcare visits yearly, especially in cooler seasons; for example, acute sinusitis predominates in fall and winter due to viral upper respiratory infections.
* Studies show that among patients who self-report "sinus headaches," a large majority (up to 80-88%) actually meet migraine diagnostic criteria, indicating that true sinus headache prevalence is likely lower than perceived.
* Age-wise, sinus-related headaches commonly affect young adults between 20-40 years old, with some studies indicating a slight male predominance for sinusitis-related headache, though women predominated in migraine-related headache populations.
* Sinus headache prevalence data can be complex due to overlapping symptoms with migraine and tension-type headaches and varying diagnostic criteria, but sinusitis remains a significant contributor to headache burden when infection symptoms are present.

**PREDEFINED Q & A SETS**

### **Is there a way to get rid of my sinus headache instantly?**

Unfortunately, there’s no quick fix for sinus headaches. You need treatment for the underlying cause to get rid of a sinus headache. But there are things you can do to ease sinus pressure and pain:

* Apply a warm compress to painful areas of your face.
* Use a decongestant to reduce sinus swelling and allow mucus to drain.
* Try a saline nasal spray or drops to thin the mucus.
* Use a vaporizer or inhale steam from a pan of boiled water. Warm, moist air may help relieve sinus congestion.

### **How do I know if my headache is sinus-related?**

There are several kinds of headaches that may make your head hurt in different ways. For example, people often confuse migraine headaches and sinus headaches because they both cause pain that pinpoints certain parts of your aching head. The difference is where pain happens:

* Sinus headaches make your face hurt. The pain affects both sides of your head.
* Migraine headaches typically cause pain high in your forehead, around your temples or in the back of your head. The pain typically affects one side of your head.
* A sinus headache may feel like it’s lasting an eternity.

### **What’s the difference between sinusitis and a sinus headache?**

The difference is that a sinus headache is just one symptom of sinusitis. If you have sinusitis, you may also have the following symptoms:

* Fever.
* Fatigue.
* Postnasal drip.
* Cough.

**GENOMIC DATA**

* Chronic rhinosinusitis (CRS), especially with nasal polyps, has been associated with genetic variants in over 70 genes related to innate and adaptive immunity. These genes influence inflammation and mucosal barrier function, which can predispose to persistent sinus inflammation and symptoms like sinus headache.
* Genetic conditions such as cystic fibrosis (CF) and primary ciliary dyskinesia (PCD), which cause impaired mucociliary clearance and thick mucus, have well-characterized mutations linked to chronic sinus disease and sinus infections.
* Variations in the CDHR3 gene have been identified to increase susceptibility to CRS, highlighting the role of viral interactions (like common cold viruses) along with genetics in sinus disease development.
* Studies show a genetic correlation between CRS and autoimmune diseases such as allergic rhinitis, asthma, rheumatoid arthritis, and hypothyroidism. Shared genes have been found predominantly in tissues of the endocrine/exocrine, digestive, skin, and nervous systems, suggesting systemic immune influences on sinus inflammation

**DOCTOR-PATIENT CONVERSATIONS**

Doctor: Hello! I understand you are having headaches around your forehead and face. Can you tell me more about what you’re experiencing?

Patient: Yes, I have this constant pressure and pain around my cheeks, forehead, and sometimes behind my eyes. It gets worse when I bend over or lie down. I thought it might be a sinus headache.

Doctor: That’s a common concern. Sinus headaches happen when the sinus passages behind your forehead, cheeks, and nose get congested or inflamed, leading to pressure and pain. Do you also have any nasal stuffiness, thick nasal discharge, or fever?

Patient: I do have a blocked nose and feel some congestion, but no fever.

Doctor: Those symptoms can fit with sinus inflammation or sinusitis, which may be caused by infections, allergies, or nasal polyps. If your sinuses are blocked, mucus can build up and cause the pressure you feel. Sometimes this also affects your upper teeth or causes facial tenderness.

Patient: How do you know if it’s really a sinus headache or something else?

Doctor: That’s a great question. Many people who think they have sinus headaches actually have migraines or tension headaches, which can cause similar facial pain and nasal symptoms like a runny nose or congestion. Migraines often come with throbbing pain, sensitivity to light or sound, and nausea, which usually aren’t part of sinus headaches. We look at your overall symptoms, medical history, and sometimes do imaging or nasal exams to figure out the cause.

Patient: What can I do to feel better?

Doctor: Treatment depends on the cause. For sinus headaches caused by sinus congestion or infection, we can try decongestants, nasal sprays, and pain relievers. Drinking plenty of fluids, using a humidifier or steam inhalation, and applying warm compresses can help relieve pressure. If there's a bacterial infection, antibiotics might be needed. For allergy-related symptoms, antihistamines or allergy treatments help.

Patient: Are there any risks with these medicines?

Doctor: Yes, some decongestant nasal sprays shouldn’t be used for more than a few days because they can worsen congestion later. Also, some decongestants may raise blood pressure, so we need to be careful if you have hypertension. Always follow the dosing instructions and let me know if you have other health conditions.

Patient: What if my headaches don’t get better?

Doctor: If your symptoms persist beyond 10 days, worsen, or if you get a high fever, severe facial pain, or vision changes, we should reassess. Chronic sinus issues sometimes need further evaluation or treatment like imaging or referral to a specialist. Also, if headaches recur often or have migraine features, we may explore other causes and treatments.

Patient: Thank you, doctor. It’s helpful to understand what’s going on and what I can do.

Doctor: You’re welcome! Remember, we’ll work together to manage your symptoms and get you feeling better. Please reach out if your symptoms change or don’t improve.

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**Skin cancer**

ALTERNATIVE NAMES

**DEFINITION / DESCRIPTION**

Skin cancer is a disease that involves the growth of abnormal cells in your skin tissues. Normally, as skin cells grow old and die, new cells form to replace them. When this process doesn’t work as it should — like after exposure to ultraviolet (UV) light from the sun — cells grow more quickly. These cells may be noncancerous (benign), which don’t spread or cause harm. Or they may be cancerous.

Skin cancer can spread to nearby tissue or other areas in your body if it’s not caught early. Fortunately, if skin cancer is identified and treated in early stages, most are cured. So, it’s important to talk with your healthcare provider if you think you have any signs of skin cancer.

#### **Types of skin cancer**

There are three main types of skin cancer:

* Basal cell carcinoma, which forms in your basal cells in the lower part of your epidermis (the outside layer of your skin).
* Squamous cell carcinoma, which forms in your squamous cells in the outside layer of your skin.
* Melanoma, which forms in cells called melanocytes. Melanocytes produce melanin, a brown pigment that gives your skin its color and protects against some of the sun’s damaging UV rays. This is the most serious type of skin cancer because it can spread to other areas of your body.

Other types of skin cancer include:

* Kaposi sarcoma.
* Merkel cell carcinoma.
* Sebaceous gland carcinoma.
* Dermatofibrosarcoma protuberans.

#### **How common is skin cancer?**

Skin cancer is the most common cancer diagnosed in the U.S. In fact, about 1 in 5 people develop skin cancer at some point in their life.

**CAUSES**

The main cause of skin cancer is overexposure to sunlight, especially when you have sunburn and blistering. UV rays from the sun damage DNA in your skin, causing abnormal cells to form. These abnormal cells rapidly divide in a disorganized way, forming a mass of cancer cells.

#### **What are the risk factors for skin cancer?**

Anyone can get skin cancer, regardless of race or sex. But some groups get it more than others. Before the age of 50, skin cancer is more common in women. After 50, though, it’s more common in men. And it’s about 30 times more common in non-Hispanic white people than non-Hispanic Black people or people of Asian/Pacific Islander descent. Unfortunately, skin cancer is often diagnosed in later stages for people with darker skin tones. This makes it more difficult to treat.

Although anyone can develop skin cancer, you’re at increased risk if you:

* Spend a considerable amount of time working or playing in the sun.
* Get easily sunburned or have a history of sunburns.
* Live in a sunny or high-altitude climate.
* Tan or use tanning beds.
* Have light-colored eyes, blond or red hair and fair or freckled skin.
* Have many moles or irregularly shaped moles.
* Have actinic keratosis (precancerous skin growths that are rough, scaly, dark pink-to-brown patches).
* Have a family history of skin cancer.
* Have had an organ transplant.
* Take medications that suppress or weaken your immune system.
* Have been exposed to UV light therapy for treating skin conditions such as eczema or psoriasis.

**SIGNS / SYMPTOMS**

The most common warning sign of skin cancer is a change on your skin — typically a new growth or a change in an existing growth or mole. Skin cancer symptoms include:

* A new mole. Or a mole that changes in size, shape or color, or that bleeds.
* A pearly or waxy bump on your face, ears or neck.
* A flat, pink/red- or brown-colored patch or bump.
* Areas on your skin that look like scars.
* Sores that look crusty, have a depression in the middle or bleed often.
* A wound or sore that won’t heal, or that heals but comes back again.
* A rough, scaly lesion that might itch, bleed and become crusty.

#### **What does skin cancer look like?**

Skin cancer looks different depending on what type of skin cancer you have. Thinking of the ABCDE rule tells you what signs to watch for:

* Asymmetry: Irregular shape.
* Border: Blurry or irregularly shaped edges.
* Color: Mole with more than one color.
* Diameter: Larger than a pencil eraser (6 millimeters).
* Evolution: Enlarging, changing in shape, color or size. (This is the most important sign.)

If you’re worried about a mole or another skin lesion, make an appointment and show it to your healthcare provider. They’ll check your skin and may ask you to see a dermatologist and have the lesion further evaluated.

**DIAGNOSIS METHODS**

First, a dermatologist may ask you if you’ve noticed changes in any existing moles, freckles or other skin spots, or if you’ve noticed any new skin growths. Next, they’ll examine all of your skin, including your scalp, ears, palms of your hands, soles of your feet, between your toes, around your genitals and between your buttocks.

#### **What tests will be done to diagnose skin cancer?**

If your provider suspects skin cancer, they may perform a biopsy. In a biopsy, a sample of tissue is removed and sent to a laboratory where a pathologist examines it under a microscope. Your dermatologist will tell you if your skin lesion is skin cancer, what type you have and discuss treatment options.

#### **Skin cancer stages**

Cancer stages tell you how much cancer is in your body. The stages of skin cancer range from stage 0 to stage IV. In general, the higher the number, the more cancer has spread and the harder it is to treat. But the staging for melanoma is different from non-melanoma skin cancers that start in your basal or squamous cells.

Melanoma staging

* Stage 0 (melanoma in situ): The melanoma is only in the top layer of your skin.
* Stage I: The melanoma is low risk and there’s no evidence that it has spread. It’s generally curable with surgery.
* Stage II: It has some features that indicate that it’s likely to come back (recur), but there’s no evidence of spread.
* Stage III: The melanoma has spread to nearby lymph nodes or nearby skin.
* Stage IV: The melanoma has spread to more distant lymph nodes or skin, or has spread to internal organs.

Non-melanoma staging

* Stage 0: Cancer is only in the top layer of your skin.
* Stage I (1): Cancer is in the top and middle layers of your skin.
* Stage II (2): Cancer is in the top and middle layers of your skin and moves to target your nerves or deeper layers of skin.
* Stage III (3): Cancer has spread beyond your skin to your lymph nodes.
* Stage IV (4): Cancer has spread to other parts of your body and your organs like your liver, lungs or brain.

**TREATMENT OPTIONS**

Treatment depends on the stage of cancer. Sometimes, a biopsy alone can remove all the cancer tissue if it’s small and limited to the surface of your skin. Other common skin cancer treatments, used alone or in combination, include:

* Cryotherapy: Your dermatologist uses liquid nitrogen to freeze skin cancer. The dead cells slough off after treatment.
* Excisional surgery: Your dermatologist removes the tumor and some surrounding healthy skin to be sure all the cancer is gone.
* Mohs surgery: Your dermatologist removes only diseased tissue, saving as much surrounding normal tissue as possible. Providers use this to treat basal cell and squamous cell cancers and, sometimes, other skin cancers that develop near sensitive or cosmetically important areas, like your eyelids, ears, lips, forehead, scalp, fingers or genital area.
* Curettage and electrodesiccation: Your dermatologist uses an instrument with a sharp, looped edge to remove cancer cells as it scrapes across the tumor. Then, they use an electric needle to destroy any remaining cancer cells. Providers often use this to treat basal cell and squamous cell cancers and precancerous skin tumors.
* Chemotherapy: Your dermatologist or oncologist uses medications to kill cancer cells. Anticancer medications can be applied directly on the skin (topical chemotherapy) if limited to your skin’s top layer or provided through pills or an IV if the cancer has spread to other parts of your body.
* Immunotherapy: Your oncologist gives you medications to train your immune system to kill cancer cells.
* Radiation therapy: Your radiation oncologist uses radiation (strong beams of energy) to kill cancer cells or keep them from growing and dividing.
* Photodynamic therapy: Your dermatologist coats your skin with medication, which they activate with a blue or red fluorescent light. This therapy destroys precancerous cells while leaving normal cells alone.

#### **Complications/side effects of the treatment**

The side effects of skin cancer treatment depend on what treatments your healthcare provider thinks will work best for you. Chemotherapy for skin cancer can lead to nausea, vomiting, diarrhea and hair loss. Other side effects or complications of skin cancer treatment include:

* Bleeding.
* Pain and swelling.
* Scars.
* Nerve damage that results in loss of feeling.
* Skin infection.
* Regrowth of the tumor after it’s been removed.

**PREVENTION TIPS**

In most cases, skin cancer can be prevented. The best way to protect yourself is to avoid too much sunlight and sunburns. UV rays from the sun damage your skin, and over time, this may lead to skin cancer.

#### **How can I lower my risk?**

Ways to protect yourself from skin cancer include:

* Use a broad-spectrum sunscreen with a skin protection factor (SPF) of 30 or higher. Broad-spectrum sunscreens protect against both UV-B and UV-A rays. Apply the sunscreen 30 minutes before you go outside. Wear sunscreen every day, even on cloudy days and during the winter months.
* Wear hats with wide brims to protect your face and ears.
* Wear long-sleeved shirts and pants to protect your arms and legs. Look for clothing with an ultraviolet protection factor label for extra protection.
* Wear sunglasses to protect your eyes. Look for glasses that block both UV-B and UV-A rays.
* Use a lip balm with sunscreen.
* Avoid the sun between 10 a.m. and 4 p.m.
* Avoid tanning beds. If you want a tanned look, use a spray-on tanning product.
* Ask your healthcare provider or pharmacist if any of the medications you take make your skin more sensitive to sunlight. Some medications known to make your skin more sensitive to the sun include tetracycline and fluoroquinolone antibiotics, tricyclic antibiotics, the antifungal agent griseofulvin and statin cholesterol-lowering drugs.
* Regularly check all your skin for any changes in size, shape or color of skin growths or the development of new skin spots. Don’t forget to check your scalp, ears, the palms of your hands, soles of your feet, between your toes, your genital area and between your buttocks. Use mirrors and even take pictures to help identify changes in your skin over time. Make an appointment for a full-body skin exam with your dermatologist if you notice any changes in a mole or other skin spot.

**OUTLOOK / PROGNOSIS**

Nearly all skin cancers can be cured if they’re treated before they have a chance to spread. The earlier skin cancer is found and removed, the better your chance for a full recovery. It’s important to continue following up with your dermatologist to make sure cancer doesn’t come back. If something seems wrong, call your doctor right away.

Most skin cancer deaths are from melanoma. If you’re diagnosed with melanoma:

* The five-year survival rate is 99% if it’s detected before it spreads to your lymph nodes.
* The five-year survival rate is 66% if it has spread to nearby lymph nodes.
* The five-year survival rate is 27% if it has spread to distant lymph nodes and other organs

**POSSIBLE COMPLICATIONS**

Skin cancer can lead to several complications, many of which can be prevented with early detection and management. However, the complications vary based on the subtype of skin cancer. General complications include:

* Further invasion of the cancer locally, either peripherally or deeply, causing tissue damage and symptoms such as pain.
* Metastasis of skin cancer to other areas of the body, including the brain, lungs, lymph nodes, or skin.
* Infection of skin cancer, particularly in ulcerated lesions.
* Scarring and tissue destruction from the cancer or its surrounding skin, which could lead to temporary or permanent physical debility or disfigurement.
* Unpleasant patient-reported symptoms, such as pain, itching, bleeding, or discomfort in the area of the skin cancer.
* Recurrence of skin cancer, even after treatment.
* Psychiatric conditions associated with skin cancer, such as depression or anxiety.
* Psychological complications, such as distress or fear.
* Effects of treatments, such as adverse drug events.
* Impaired immune function.

Early evaluation and management of skin cancer are crucial to prevent potential complications. For example, recurrence rates of skin cancer vary widely depending on the subtype. Basal cell carcinoma has a rare recurrence rate after treatment with micrographic dermatologic surgery. In contrast, mucinous carcinoma of the skin, which can be either primary or metastatic from other areas (eg, colon and breast), has a local recurrence rate estimated to be between 20% and 40%. Recurrence is most commonly associated with the tumor's size. The interprofessional team's coordination throughout a patient's care continuum is essential in communicating skin cancer prevention, early detection of skin cancer, tailored treatment, and early detection of complications.

**WHEN TO SEE A DOCTOR / RED FLAG**

Make an appointment to see a healthcare provider or dermatologist as soon as you notice:

* Any changes to your skin or changes in the size, shape or color of existing moles or other skin lesions.
* The appearance of a new growth on your skin.
* A sore that doesn’t heal.
* Spots on your skin that are different from others.
* Any spots that change, itch or bleed.

Your provider will check your skin, take a biopsy (if needed), make a diagnosis and discuss treatment. Also, see a dermatologist annually for a full skin review.

**DIFFERENTIAL DIAGNOSIS**

Skin neoplasms can be either benign or malignant, with each type of skin cancer presenting in a variety of ways. Due to this diversity, the differential diagnosis includes all lesions that may appear as macules, patches, papules, plaques, or nodules. In rare cases, they may also present as vesicles, bullae, or pustules. The differential diagnoses should consider relevant vascular, infectious, neoplastic (eg, metastases to the skin from other organs), inflammatory, traumatic, metabolic, mechanical, allergic, autoimmune, and iatrogenic lesions.

Common lesions to distinguish from true skin cancer include:

* Psoriasis
* Atopic dermatitis
* Tinea corporis (or other body area)
* Acne vulgaris
* Warts
* Lupus erythematosus
* Actinic keratosis
* Metastatic skin tumors
* Sebaceous hyperplasia
* Nevus
* Benign melanocytic lesions
* Dysplastic nevi

Cysts should also be considered in the differential diagnosis of skin cancer.

**EPIDEMIOLOGY**

Skin cancers are more frequent than any other type of cancer worldwide and in the United States. The economic burden of risk factors, such as indoor tanning, contributes to the rising incidence of skin cancer, resulting in an annual economic burden of at least 8.1 billion dollars in the United States. The increasing number of people diagnosed with skin cancer highlights a significant health challenge, both in terms of patient well-being and healthcare expenditures.

Skin cancer occurs in all races worldwide but is more common in individuals with lighter skin, likely due to the reduced photoprotective effects of epidermal melanin. In individuals with lighter skin, approximately 75% to 80% of nonmelanoma skin cancers are basal cell carcinomas, whereas up to 25% are squamous cell carcinomas. Heritable defects in DNA repair mechanisms, such as those seen in xeroderma pigmentosum and Muir-Torre syndrome, also increase the risk of cutaneous carcinomas in some patients.

Among skin cancers, melanoma has the highest disability-adjusted life year, basal cell carcinoma has the highest incidence rate, and squamous cell carcinoma has the highest prevalence. The incidence of melanoma in the United States, from 1990 to 2019, increased to 17 per 100,000 individuals, and prevalence increased to 138 per 100,000 individuals. Meanwhile, nonmelanoma skin cancer had an incidence of 787 per 100,000 individuals and a prevalence of 359 per 100,000 individuals. Males had a higher incidence, prevalence, and mortality rate than females from 1990 to 2019. Although skin cancer occurs in all age groups, it is more common in older adults, likely due to increased cumulative UV light exposure. Geographically, melanoma rates are higher in the northern half of the United States compared to the southern half, while nonmelanoma skin cancer incidence is highest in the southeastern, western, and northeastern regions of the country.

Other malignancies, such as Merkel cell carcinoma or Kaposi sarcoma, have unique epidemiological information outside the scope of this resource. Merkel cell carcinoma is far more common in men and among individuals with lighter skin. Kaposi sarcoma is more common in individuals with HIV. Worldwide, skin cancer rates are highest in populations with significant UV light exposure and lighter skin tones, such as those in Europe and Australia

**PREDEFINED Q & A SETS**

### **How does skin cancer become a life-threatening cancer?**

You may wonder how cancer on the surface of your skin becomes a life-threatening cancer. It seems logical to think you could just scrape off the skin with the cancer cells or even remove the cancerous skin lesion with a minor skin surgery and that’s all that would be needed. These techniques are successfully used if cancer is caught early.

But if skin cancer isn’t caught early, something that’s “just on my skin” can grow and spread beyond the immediate area. Cancer cells can break away and travel through your bloodstream or lymph system. They can settle in other areas of your body and begin to grow and develop into new tumors. This travel and spread is called metastasis.

The type of cancer cell where cancer first started — called primary cancer — determines the type of cancer. For example, if malignant melanoma metastasized to your lungs, the cancer would still be called malignant melanoma. This is how that superficial skin cancer can turn into life-threatening cancer.

### **Why does skin cancer occur in more non-sun-exposed body areas in people of color?**

Scientists don’t fully know why people with darker skin tones develop cancer in non-sun-exposed areas like the palms of your hands and feet. They think that the sun is less of a factor, though. That said, dermatologists still see plenty of UV sunlight-induced melanomas and squamous cell skin cancer in people with skin tones ranging from fair to very dark.

### **Are all moles cancerous?**

Most moles aren’t cancerous. Some moles are present at birth. Others develop up to about age 40. Most adults have between 10 and 40 moles.

In rare cases, a mole can turn into melanoma. If you have more than 50 moles, you have an increased chance of developing melanoma.

1. What type of skin cancer do I have?  
   Your dermatologist will tell you if it is basal cell carcinoma, squamous cell carcinoma, melanoma, or another less common type based on biopsy and examination.
2. What stage is my skin cancer?  
   The stage depends on the size, depth of invasion, and whether it has spread to lymph nodes or other parts of the body. Early stages are usually localized and easier to treat.
3. What tests will I need?  
   Typical tests include a skin biopsy to confirm diagnosis. Depending on the type and stage, imaging such as ultrasound, CT, or MRI may be done to check for spread.
4. What’s the best treatment for my skin cancer?  
   Treatment depends on type and stage but may include surgical removal (excision, Mohs surgery), topical treatments, radiation, or systemic therapies for advanced cases.
5. What are the side effects of that treatment?  
   Side effects vary by treatment but can include scarring, infection, changes in skin pigmentation, fatigue (with systemic treatments), and others depending on therapy used.
6. What are the potential complications of this cancer and the treatment for it?  
   Complications include recurrence, spread (metastasis), and side effects of treatment such as wound healing issues or secondary cancers in rare cases.
7. What outcome can I expect?  
   Many early-stage skin cancers have excellent outcomes with treatment. Melanoma prognosis depends on stage at diagnosis, with early detection improving survival rates.
8. Do I have an increased risk of additional skin cancers?  
   Yes. Having one skin cancer raises your risk of developing others in the future, so ongoing skin monitoring and sun protection are critical.
9. How often should I be seen for follow-up checkups?  
   Follow-up schedules depend on risk factors, cancer type, and stage. For low-risk basal cell carcinoma, often just one follow-up appointment is needed after treatment. For higher-risk squamous cell carcinoma or melanoma, follow-ups commonly occur every 3 to 6 months for the first few years, then yearly if no recurrence is seen. Follow-ups allow your doctor to check for recurrence and new cancers.

## **Genomic Subtypes of Cutaneous Melanoma**

According to large-scale genomic studies such as The Cancer Genome Atlas (TCGA), cutaneous melanomas are mainly divided into four genomic subtypes defined by mutations in the following genes:

| **Genomic Subtype** | **Common Mutations and Features** |
| --- | --- |
| BRAF-mutant | Mutations primarily in *BRAF* gene (e.g., V600E mutation) causing activation of the MAPK pathway. More common in younger patients and superficial spreading melanoma subtype. These tumors often respond to BRAF inhibitors. |
| RAS-mutant | Mutations in *NRAS*, *HRAS*, or *KRAS* genes, also activating MAPK pathway. More frequent in older patients, associated with nodular melanoma and chronic sun damage. |
| NF1-mutant | Loss-of-function mutations in the tumor suppressor *NF1* gene, leading to MAPK pathway activation. This subtype shows distinct molecular features and may respond differently to therapy. |
| Triple-WT | Wild-type for *BRAF*, *RAS*, and *NF1*. Enriched for mutations or amplifications in *KIT* gene and complex structural genomic rearrangements. This group includes melanomas without common driver mutations and often comprises acral, mucosal, and some rare subtypes. |

Genomic profiling identifies additional alterations such as *KIT* amplifications in acral and mucosal melanomas, and highlights the role of immune-related gene expression linked to better prognosis in some cases.

## Other Skin Cancers

* Basal Cell Carcinoma (BCC): Some inherited syndromes like basal cell nevus syndrome (mutations in *PTCH1*/*PTCH2*) increase risk. Sporadic BCCs usually arise from UV-induced mutations but less characterized by broad genomic subtyping compared to melanoma.
* Squamous Cell Carcinoma (SCC): Linked to inherited conditions (e.g., albinism, epidermolysis bullosa) and UV damage but genomic classifications are less well established than for melanoma.

**DOCTOR-PATIENT CONVERSATIONS**

Doctor: Hello! I understand you have a suspicious spot on your skin that we’ve just biopsied. I want to talk with you about the results and what to expect next. Is that okay?

Patient: Yes, please. I’m a little nervous and want to understand what this means.

Doctor: Of course. The biopsy shows that you have skin cancer. Specifically, it’s a basal cell carcinoma, which is the most common type of skin cancer. It usually grows slowly and rarely spreads to other parts of the body.

Patient: What exactly is basal cell carcinoma?

Doctor: It’s a type of cancer that starts in the basal cells, which are found in the lowest layer of the skin’s epidermis. These cells normally produce new skin cells, but in basal cell carcinoma, they grow uncontrollably forming a tumor.

Patient: Is it serious? Will it spread?

Doctor: Basal cell carcinoma is usually not life-threatening because it rarely spreads beyond the skin. However, if left untreated, it can grow and damage nearby tissues, and can be more difficult to treat.

Patient: What treatment options do I have?

Doctor: Treatment depends on the size and location, but the most common method is surgical removal. We can use techniques like simple excision or Mohs surgery, which removes the cancer layer by layer and preserves as much healthy tissue as possible. Other treatments include topical medications or radiation in some cases.

Patient: What are the side effects of the treatment?

Doctor: Surgery may leave a scar, and there can be some redness or swelling afterward. Most patients heal well with minimal cosmetic impact, especially if the cancer is caught early.

Patient: Will I need more tests or follow-ups?

Doctor: For basal cell carcinoma, additional imaging is usually not needed unless the cancer is large or aggressive. Follow-up visits are important to check for any new skin cancers or recurrence. Typically, we recommend exams every 6 to 12 months initially.

Patient: Am I at risk for more skin cancers?

Doctor: Yes, having one skin cancer increases your risk of developing more in the future. That’s why ongoing skin checks and protecting your skin from the sun with sunscreen and protective clothing is very important.

Patient: What if I notice new or changing spots?

Doctor: Please contact us promptly if you see any new bumps, sores that won’t heal, or any skin changes. Early detection is key to successful treatment.

Patient: Thank you, doctor. This helps me understand what to expect.

Doctor: You’re welcome. We’ll work closely together to manage your care, and I’m here to answer any questions you have along the way.

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**Spasmodic dysphonia**

**ALTERNATIVE NAMES**

LARYNGEAL DYSTONIA

**DEFINITION / DESCRIPTION**

Spasmodic dysphonia — also called laryngeal dystonia — is a voice disorder that affects your larynx (voice box) and vocal cords. Your voice may break, sound tight and strained or very breathy when you speak. A rare condition, it affects around 500,000 people in the U.S.

Typically, your vocal cords vibrate when you talk, producing the sound of your voice. But with spasmodic dysphonia, the muscles controlling your vocal cords go into spasms without your control. The spasms can make your vocal cords come together too tightly, making your voice strained. Or, the spasms may make them too loose, making your voice breathy and weak. This condition can make it harder to speak and be understood.

Spasmodic dysphonia usually only affects conversational speech. Your voice will likely feel and sound normal when you’re singing, laughing or whispering.

#### **Types of spasmodic dysphonia**

The three types of spasmodic dysphonia are:

* Adductor spasmodic dysphonia (most common). This type makes your voice sound strained, tight and hoarse. It happens when spasms force your vocal cords too close together.
* Abductor spasmodic dysphonia. This type makes your voice sound weak or breathy. It happens when spasms keep your vocal cords too far apart.
* Mixed spasmodic dysphonia. Rarely, people have symptoms of both types of spasmodic dysphonia.

Any of these may also present with vocal tremor, which makes your voice sound shaky.

**CAUSES**

Spasmodic dysphonia starts in an area of your brain called the basal ganglia. This part of your brain helps coordinate involuntary muscle movements. Conditions that cause uncontrollable muscle movements because of faulty brain signals are called dystonias. This is why this condition is also called “laryngeal dystonia.”

The faulty signaling makes the muscles in your larynx and your vocal cords go into spasms.

#### **Risk factors**

Researchers don’t know what triggers spasmodic dysphonia. In some cases, it may be related to gene variants you inherit from your biological parents. About 1 in 4 people diagnosed have a family history of dystonia. It’s also possible that it occurs after an injury or sickness.

It’s three times more common in females.

**SIGNS / SYMPTOMS**

Symptoms usually start in midlife (between ages 30 to 60). Voice changes may come and go, so you can’t predict when your voice will suddenly sound different. Your voice may sound:

* Strained and tight
* Hoarse and raspy
* Breathy, soft or like you’re whispering
* Broken because certain sounds cut off while you’re speaking
* Shaky or trembling

Stress, including the stress of talking on the telephone or with groups of people, or in noisy places, can make symptoms worse.

**DIAGNOSIS METHODS**

Diagnosis usually involves a team of healthcare providers. An otolaryngologist and a speech-language pathologist (SLP) will listen to you speak. They’ll check for breaks in your speech and other signs of the condition. They’ll also do a videostroboscopy. This imaging test shows how your vocal cords move when you talk.

A neurologist may check for other types of movement disorders

**TREATMENT OPTIONS**

Healthcare providers can’t cure spasmodic dysphonia. But there are treatments to ease vocal cord spasms. Treatments include:

* Botox® injections (most common). Botox® blocks the nerve signals that cause spasms, making it easier to talk. The effects usually last up to four months. You’ll need regular injections to keep seeing benefits.
* Voice therapy. An SLP can teach you exercises to improve how you use your voice. Voice therapy works best when you’re also getting Botox.
* Communication aids. You can use technology that makes your voice sound louder or that translates typed text into speech.

#### **Surgery**

There are two types of surgeries providers use to treat adductor spasmodic dysphonia (the type where your vocal cords are too close):

* Type II thyroplasty sets your vocal cords farther apart. A surgeon readjusts the supportive tissue in your voice box that holds your vocal cords in place.
* Selective laryngeal adductor denervation-reinnervation (SLAD-R) changes the pathway that nerve signals use to travel from your brain to your vocal cords. A surgeon disconnects some of the nerves you use to speak and reconnects them to a different nerve.

Surgery for spasmodic dysphonia is controversial because the benefits may not last very long for everyone. More research is needed to show that the benefits of surgery outweigh the risks.

**OUTLOOK / PROGNOSIS**

Spasmodic dysphonia is a lifelong condition. Typically, symptoms develop gradually, level off and then remain the same for the rest of your life. Sometimes, the spasms disappear for a period. But they usually return at some point.

The best way to deal with spasmodic dysphonia is to work with your care team to manage it. For most people, regular Botox injections and voice therapy help.

Your healthcare provider can explain your options.

**WHEN TO SEE A DOCTOR / RED FLAG**

Contact your healthcare provider if you have unexplained voice changes. Voice strain or breaks can be a sign of something temporary, like laryngitis. But if things don’t improve within a couple of weeks, it’s time to see a provider.

**DIFFERENTIAL DIAGNOSIS**

| **Condition** | **Key Differentiating Features** |
| --- | --- |
| Muscle Tension Dysphonia (MTD) | Sustained hyperfunctional voice use without spasmodic breaks; often improves with voice therapy; lacks task-specific spasms seen in SD. |
| Psychogenic Dysphonia | Voice symptoms linked to psychological factors; inconsistent voice changes; voice may normalize with distraction or non-voice tasks. |
| Essential Voice Tremor | Rhythmic, regular tremor of voice during sustained phonation; differs from irregular spasms in SD; may coexist with SD. |
| Vocal Fold Paralysis or Paresis | Weak, breathy voice with immobile vocal folds visible on laryngoscopy; no spasms. |
| Laryngeal Pathology | Structural abnormalities like nodules, polyps, granulomas causing hoarseness without spasmodic breaks. |
| Laryngitis or Inflammatory Conditions | Acute inflammation with medical treatment response; absence of spasmodic voice breaks. |
| Functional or Habitual Dysphonia | Voice altered due to behavioral causes, no neurological spasms. |
| Other Neurological Disorders | Broader dystonias, Parkinson’s disease, multiple sclerosis, which have other neurological signs beyond voice symptoms. |

## Diagnostic Considerations:

* SD is characterized by involuntary, task-dependent spasms during speech that are not present during other vocal activities (e.g., whispering, coughing).
* Laryngoscopy during speech can show spasms, but spasms may be too rapid to see easily.
* Diagnosis typically involves a multidisciplinary approach including otolaryngology, neurology, and speech pathology.
* Differentiating SD from MTD or essential tremor can be challenging; trial treatments and detailed voice assessments help.
* Absence of other neurological signs points more towards focal laryngeal dystonia (SD).

**EPIDEMIOLOGY**

* Prevalence: SD affects approximately 1 to 7 per 100,000 people, with some studies citing about 2 per 100,000 in the general population. It is the third most common form of focal dystonia after cervical dystonia and blepharospasm.
* Gender: Women are more frequently affected than men, comprising about 60-85% of cases, with female-to-male ratios reported ranging from roughly 2:1 up to 7:1 in certain populations.
* Age of Onset: SD typically develops in mid-adulthood, with onset commonly between 30 and 50 years of age, averaging around 39-45 years but with a wide range from teenage years up to 70s reported.
* Types: The adductor type is far more common (~90%) than the abductor type. Mixed types and whispering dysphonia are much rarer.
* Risk Factors: Although the exact cause is unknown, studies indicate possible associations with:
  + Prior upper respiratory tract infections (about 30% report this before onset)
  + Major life stress (about 20% report this)
  + Essential tremor and writer’s cramp commonly co-occur.
  + No clear genetic cause identified yet, though about 10% report family history of dystonia.
* Demographics: SD seems more common among persons of European descent. Epidemiologic data from Japan shows lower prevalence, possibly due to underdiagnosis.
* Other Notes: SD is a chronic condition with gradual onset and typically lifelong course

**PREDEFINED Q & A SETS**

### **Is there anything I can do to feel better?**

Here are some things you can do if you’re living with spasmodic dysphonia:

* Learn your voice. Some people notice symptoms ease when they’re chewing or supporting their head while talking or singing. It’s possible that there are similar “hacks” that work for you.
* Practice self-care. Being too tired or stressed can worsen the symptoms of spasmodic dysphonia. Getting enough rest and caring for your mental health can help.
* See a counselor. A counselor can help you cope with the impact your condition has on your ability to work and socialize.
* Be open with others. You may feel self-conscious about your voice. If your voice changes mid-conversation, consider explaining why it happens. Most people will be understanding.
* Join support groups. Support groups can connect you with others who understand the challenges of spasmodic dysphonia or other voice disorders.

### **Is spasmodic dysphonia a symptom of Parkinson’s?**

No. Spasmodic dysphonia is different from Parkinson’s disease. Parkinson’s causes you to gradually lose muscle control throughout your body. But spasmodic dysphonia only affects your voice. Instead of getting worse over time, symptoms usually level off.

### **Is spasmodic dysphonia a disability?**

It can be. Spasmodic dysphonia may be considered a disability if it interferes with your ability to do your job. Working with a counselor who specializes in disability-related issues can help you understand what may be involved.

## **Botulinum Toxin Treatment for Spasmodic Dysphonia**

* Mechanism: Botox temporarily weakens or paralyzes the overactive muscles controlling the vocal cords, thereby reducing spasms that cause voice breaks or strain.
* Injection sites: Depends on SD type—usually the thyroarytenoid muscles for adductor type or posterior cricoarytenoid for abductor type.
* Duration of effect: Typically lasts 3–4 months; injections are repeated regularly as needed.

## Common Side Effects of Botox Treatment in SD

* Voice-related:
  + Excessive breathiness or weak voice (due to muscle weakening)
  + Hoarseness
  + Voice fatigue or mild dysphonia
  + Temporary worsened voice right after injection
* Swallowing and airway:
  + Mild dysphagia (difficulty swallowing), which can occasionally lead to aspiration of fluids
  + Weak cough reflex
  + Rarely, delayed upper airway obstruction due to vocal cord adduction requiring emergency care—very uncommon but serious
* Local:
  + Mild pain or discomfort at injection site
  + Flu-like symptoms such as headache, nausea
  + Bruising or swelling
* Other reported side effects:
  + Drop in blood pressure or fainting episode during injection (rare)
  + Anxiety or emotional distress related to voice changes **post-injection**

**DOCTOR-PATIENT CONVERSATIONS**

Doctor: Hello! I understand you’ve been having some trouble with your voice. Can you tell me what you’ve been experiencing?

Patient: Yes, doctor. My voice sounds shaky, tight, and sometimes it just breaks or falters when I try to speak. It’s making it hard for people to understand me.

Doctor: That sounds like it could be spasmodic dysphonia. It’s a neurological voice disorder where the muscles that control your vocal cords have involuntary spasms. These spasms cause the voice interruptions and changes you’re describing.

Patient: What causes these spasms? Is it permanent?

Doctor: The exact cause isn’t fully understood, but it involves abnormal nerve signals from the brain to the vocal muscles. It’s not something you caused or caught, and it’s not related to psychological issues. It usually is a chronic condition, but there are effective treatments to help control the symptoms.

Patient: How do you diagnose spasmodic dysphonia?

Doctor: We evaluate your voice and how it behaves during different speaking tasks, often using a special camera called a laryngoscope to look at your vocal cords in action. The pattern of spasms and breaks helps us distinguish it from other voice disorders.

Patient: What treatments are available?

Doctor: The most common and effective treatment is injecting botulinum toxin, or Botox, directly into the affected vocal muscles. This weakens the muscles temporarily, reducing the spasms and improving your voice. The injections usually last a few months and need repeating. Speech therapy is also important to teach you techniques to manage your voice better.

Patient: Are there any other options if Botox doesn’t work?

Doctor: In some cases, surgery can be considered, but it’s less common and depends on your individual situation. Your care team may also include neurologists and speech therapists who work together to find the best approach for you.

Patient: Will my voice ever go back to normal?

Doctor: While there’s no permanent cure yet, many people have marked improvement and can communicate more easily with treatment. The goal is to help you regain control and confidence in your speech.

Patient: This sounds overwhelming. How can I cope day-to-day?

Doctor: It can be challenging, and emotional support is important. Counseling or joining support groups can help you adjust. Also, practicing the speech techniques your therapist teaches, and pacing your conversations can reduce stress on your voice.

Patient: Thank you, doctor. It helps to know there are treatments and ways to manage this.

Doctor: You’re welcome. We’ll work together as a team to support you. Please ask any questions anytime, and we’ll adjust your care plan as needed.

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Swimmer's ear (Otitis externa)

ALTERNATIVE NAMES

**DEFINITION / DESCRIPTION**

Swimmer’s ear (otitis externa) is an ear canal infection. Your ear canal is the pathway between your outer ear and your middle ear. Swimmer’s ear can be a bacterial infection or fungal infection.

We call it “swimmer’s ear” because avid swimmers commonly experience it. But anyone can get it — especially during the warmer months when many people spend more time in the water.

Left untreated, a swimmer's ear may muffle your hearing. In some cases, it can even cause temporary hearing loss. Most of the time, treatment solves any infection-related hearing issues. Healthcare providers treat swimmer’s ear with ear drops.

#### **How common is swimmer’s ear?**

A swimmer's ear is common. About 10% of people in the U.S. will have it at some point. A swimmer's ear can affect anyone. But kids between the ages of 7 and 14 are most likely to get it.

**SIGNS / SYMPTOMS**

Swimmer’s ear symptoms can be mild or severe and may include:

* A feeling of fullness in your ear.
* Ear pain that may increase when gently tugging on your earlobe.
* Fever.
* Fluid draining from your ear.
* Itchiness inside of your ear.
* Muffled hearing.
* Redness and swelling in your outer ear.
* Swollen lymph nodes around your ear or upper neck.

### **Causes swimmer’s ear**

Many things can cause swimmer’s ear, but activity that traps water in your ear canal is the most common cause. Bacteria and fungi thrive in warm, moist places. So, water pooling in your ear canal is the perfect environment for bacteria and fungi to settle in, start multiplying and eventually cause infection.

It’s much more common to get swimmer’s ear from a bacterium than a fungus. In rare cases, a combination of pathogens (germs that make you sick) can cause swimmer’s ear. Bacteria that can cause swimmer’s ear include *Pseudomonas aeruginosa* and *Staphylococcus aureus*. Fungi that can cause the condition include *Candida* and *Aspergillus*.

#### **Risk factors for otitis externa**

You’re more likely to develop swimmer’s ear if you:

* Have certain skin conditions: Eczema or psoriasis in your ear canal increases your risk of developing swimmer’s ear.
* Injure your ear: You can damage your ear canal by sticking objects in your ear to try to remove earwax. (Think cotton swabs, pens, bobby pins or paper clips.) These objects can scratch your ear canal and make infections more likely to develop.
* Live in the tropics: Humid environments can increase your risk of swimmer’s ear.
* Lose earwax: Earwax protects your ear canal from bacteria and fungi. You may lose earwax because there’s too much water in your ear or because you accidentally remove too much earwax when you clean your ears.
* Swim in fresh water: You can get swimmer’s ear from swimming in pools. But freshwater swimming — swimming in lakes, ponds, rivers, streams and oceans — increases your risk because fresh water may have pollution and contain more bacteria than you’d find in a pool.
* Use earbuds or hearing aids: Dirt and other substances can contaminate hearing aids or earbuds, making ear infections more likely.

**DIAGNOSIS METHODS**

A provider will examine your ears for redness, swelling or other signs of damage. If you have drainage coming from your ears, providers may take a sample of the fluid to determine what caused the infection. This is valuable information because bacterial infections and fungal infections require different treatments.

#### **What can be mistaken for swimmer’s ear?**

It’s easy to confuse swimmer’s ear with a middle ear infection — another common childhood condition. Typically, kids with a middle ear infection have pain that gets worse when laying down. They might also develop vomiting, diarrhea or a decreased appetite.

These two conditions require different treatments. What works for swimmer’s ear won’t work for a middle ear infection, and vice versa. That’s why it’s so important to see your healthcare provider for a proper diagnosis.

**TREATMENT OPTIONS**

In most cases, healthcare providers prescribe ear drops to eliminate the infection. Depending on your situation, your provider might recommend these medications for otitis externa:

* Antibiotic ear drops to fight bacteria.
* Antifungal ear drops to fight fungi.
* Steroid ear drops to reduce inflammation.

You can also take over-the-counter (OTC) pain relievers like acetaminophen (Tylenol®) or ibuprofen (Advil®) to ease any discomfort.

**PREVENTION TIPS**

Yes, keeping your ears dry is the most effective way to prevent swimmer’s ear.

To further reduce your risk, follow these otitis externa self-care tips:

* Avoid swimming in polluted water.
* Don’t stick anything into your ear canal, including cotton swabs.
* Drain water from your ears by tipping your head from side to side and gently pulling your earlobe in different directions.
* Dry your ears after bathing, swimming or being in the water. Use a clean towel or a hairdryer set on low.
* Use cotton balls to absorb excess water in your ears when you shower.
* Wear earplugs when you swim or spend time in the water.

**OUTLOOK / PROGNOSIS**

A swimmer's ear usually isn’t serious when you get treatment. In most cases, you can expect the infection to go away in about a week. Without treatment, complications — like the spread of infection — can occur. These complications usually require stronger antibiotics or antifungals.

#### **Will swimmer’s ear (otitis externa) go away by itself?**

No, it won’t. Swimmer’s ear is an infection in your ear canal that won’t go away unless you treat it. Left untreated, a swimmer’s ear infection may spread to the base of your skull, your brain or your cranial nerves.

Because infection can spread beyond your outer ear, it’s important to schedule an appointment with your healthcare provider whenever you notice symptoms. The sooner you treat it, the better.

**POSSIBLE COMPLICATIONS**

Swimmer’s ear complications are rare, especially if you treat your symptoms quickly. But they can still happen. Possible issues include:

* Chronic (long-term) ear infections. If you have swimmer’s ear for more than three months, providers call it a chronic ear infection. This can happen if you have a combination of a bacterial and fungal infection. It’s also more common in people with skin conditions (like eczema or psoriasis) and people who have an allergic reaction to their ear drops.
* Cellulitis (deep tissue infection). In rare cases, swimmer’s ear can spread into deeper layers of your skin.
* Bone or cartilage damage (early osteomyelitis). This rare complication can happen if your swimmer’s ear infection turns into an outer ear infection. The infection can spread into nearby cartilage and bone. It’s most common in people with weakened immune systems.
* Advanced skull base osteomyelitis. If infection from your ear continues to spread into the lower bones of your skull, it can affect nearby nerves, your brain and other areas of your body. It’s rare for swimmer’s ear to worsen into osteomyelitis. But once it does, it can be life-threatening.

**WHEN TO SEE A DOCTOR / RED FLAG**

Schedule an appointment with your healthcare provider if you develop any swimmer’s ear symptoms like ear pain, drainage, itchiness or muffled hearing.

If a healthcare provider has already prescribed ear drops for a swimmer’s ear infection, call them if you still have symptoms 10 days later. You might need a stronger medication.

## **Diagnostic Considerations**

Failure to recognize necrotizing (ie, malignant) otitis externa (OE) is a significant pitfall. A patient who is diabetic or immunocompromised with severe pain in the ear should have necrotizing OE excluded by an otolaryngologist.

Problems to be considered include the following:

* Ear canal trauma
* Ear canal carcinoma
* Otitis media with a perforation or ventilation tube present
* Chondritis
* Cranial nerve palsy
* Hearing loss
* Wisdom tooth eruption
* Intracranial abscess
* Cavernous sinus thrombosis
* Ramsay Hunt syndrome
* Furuncle
* Skull base osteomyelitis
* Preauricular cyst and fistula
* Lacerations
* Atopic dermatitis
* Cerumen impaction
* Exostosis and osteoma
* Foreign body
* Acute (bullous) and chronic (granular) myringitis

Although malignant tumors of the ear canal are rare, they do occur and sometimes are misdiagnosed as OE.If the condition does not respond to treatment as expected, an otolaryngologist should evaluate the patient.

Ramsay Hunt syndrome, more accurately known as herpes zoster oticus, is caused by varicella-zoster virus (VZV) infection. It is characterized by facial nerve paralysis and sensorineural hearing loss, with bullous myringitis and a vesicular eruption of the concha of the pinna and the external auditory canal (EAC). Painful OE may be present as well. Treatment includes use of an antiviral agent (eg, valacyclovir) and systemic steroids. The role of facial nerve decompression remains controversial.

A furuncle is usually caused by staphylococcal infection of a hair follicle. This infection occurs in the lateral cartilaginous hair-bearing portion of the EAC. On otoscopic examination, a furuncle appears as a localized process, which may develop into an abscess, rather than as a diffuse inflammatory process, as is characteristic of OE.

Skull base osteomyelitis occurs most often in patients who are diabetic or immunocompromised. The usual bacterial pathogen is *Pseudomonas aeruginosa*. Other predisposing conditions include arteriosclerosis, immunosuppression, chemotherapy, steroid use, and other immunodeficient states. The diagnosis is strongly suggested by a history of diabetes mellitus, severe otalgia, cranial neuropathies, and characteristic EAC findings.

The EAC may be filled with friable granulation tissue, which is primarily found inferiorly. Because this presentation may be identical to that of a soft tissue malignancy, prudence dictates a tissue biopsy even if a history of diabetes mellitus is present. Bare bone of the EAC floor may be exposed; small bony sequestra may be observed as well.

Computed tomography (CT) demonstrates bone erosion, and gallium scanning can be performed at points throughout treatment to monitor resolution. Treatment consists of administration of an antipseudomonal intravenous (IV) antibiotic such as ceftazidime (in some cases) or oral ciprofloxacin (in less dramatic cases). Extended treatment for at least 6 weeks is most appropriate. Hyperbaric oxygen therapy may also be effective. Surgical debridement is reserved for granulation tissue and bony sequestra.

A preauricular cyst or fistula may form as the result of abnormal development of the first and second branchial arch and may manifest as persistent discharge or recurrent infection. A draining sinus may be present anterior to the tragus; when infected, the cyst distends with pus, and the overlying skin is erythematous. Complete excision is indicated if these lesions become repeatedly infected. The facial nerve is at risk for injury during excision because of the close relation of the cyst or fistula to the superior branches of the nerve within the parotid gland.

First branchial cleft anomalies have a more complex embryologic origin than preauricular cysts and fistulas do. These lesions may not have an obvious sinus tract on the skin and may manifest as an abscess extending deeply into the EAC, the parotid, or the neck.

Full-thickness auricular lacerations may be observed after blunt or sharp trauma. These injuries are managed surgically by closing both the perichondrium and the skin. In contrast, external canal lacerations may occur after attempts to clean the ear canal with cotton-tipped applicators. These lacerations are usually managed by microscopically placing any skin flaps in their normal position, packing the ear canal, and administering topical antibiotic drops.

Atopic dermatitis resulting from sensitivity to topical antibiotic solutions is well known. Neomycin allergy occurs in as many as 5% of patients treated with the medication. Suspect drug sensitivity if worsening of symptoms associated with skin excoriation and weeping occurs in the distribution of the topical medication exposure after the application of drops.

Metal sensitivity also manifests as excoriation, erythema, and edema around the exposure site (eg, a piercing hole). A common allergen is nickel, an impurity that may be present in precious metals. Atopic dermatitis is managed by removal of the allergen (eg, an earring) and beginning topical steroids and antibiotics if the wound is secondarily infected. The diagnosis of metal sensitivity is confirmed by performing a skin patch test.

Cerumen impaction is the most common abnormality found on otoscopic examination, yet only a small proportion of the general population requires regular disimpaction because the EAC has the innate ability both to produce and to clear itself of cerumen. Cerumen may vary in color and consistency, and cerumen impaction may coexist with other pathologic conditions.

Debris in the EAC from cholesteatoma or tumors may be confused with cerumen; accordingly, considerable care is required when debridement of the EAC is attempted. Debridement may be accomplished by using microinstruments or by aspirating ear canal contents with a No. 5 or No. 7 Barton suction device under direct vision through the otoscope or microscope. Irrigation of the ear canal is another option, but use of a pressurized irrigation system entails the risk of trauma.

Exostoses and osteomas, the two most common bony lesions of the EAC, differ both histologically and clinically. Exostoses tend to arise from the anterior or posterior floor of the medial EAC (or from both simultaneously), have a sessile base, and are covered with normal-appearing skin. Osteomas may arise from any region of the bony EAC, are often pedunculated, may be single or multiple, and are covered by normal skin. Exostosis and osteomas require surgical treatment only if they are so large that they lead to a conductive hearing loss or intractable OE.

Foreign bodies in the EAC are not infrequently encountered. In children, the appearance of these foreign bodies is variable; parts of toys or even food may be found in the EAC. In adults, fragments of cotton swabs are the most common finding. Erythema and edema surrounding the foreign body are commonly present. Depending on the patient’s ability to cooperate, the foreign body may be removed under a microscope with the aid of microinstruments.

Hearing aid or watch batteries can sometimes end up in the ear canal accidentally, both in the pediatric and in the adult population. Time to onset of symptoms can range from a few hours to a day. Like batteries accidentally placed anywhere in the body, batteries in the ear canal represent a medical emergency. Therefore, expeditious identification and removal are absolutely necessary.

Acute myringitis is usually caused by a mycoplasmal or viral infection and has been observed in both adults and children. It is characterized by hemorrhagic bullae involving the tympanic membrane and a flulike syndrome. It is self-limiting; treatment involves pain control and fever management. Chronic myringitis is defined as deepithelization of the tympanic membrane, granulation tissue formation, and discharge. Treatment includes topical application of eardrops, a caustic solution in unresponsive cases, and mechanical removal of polypoidal granulations.

## **Differential Diagnoses**

* Otitis Media

**RECENT GUIDELINES OR UPDATES**

The recommendations included the following:

* Differentiate diffuse acute OE from other possible causes of otalgia, otorrhea, and inflammation of the external auditory canal (EAC)
* Assess patients with diffuse acute OE for factors that may modify therapeutic management (eg, nonintact tympanic membrane, immunocompromised state)
* Assess for pain in patients with acute OE; base the analgesia recommendation on the patient’s pain severity
* Administer topical medications as initial therapy for diffuse, uncomplicated acute OE
* Do not administer systemic antimicrobial agents as initial therapy for diffuse, uncomplicated acute OE; reserve such treatment for cases in which there is extension outside of the EAC or there are specific host factors that require systemic agents
* Instruct patients on how to administer topical drops; perform an aural toilet, place a wick, or both, when the ear canal is obstructed
* Use a nonototoxic topical agent in patients with a known or suspected perforation of the tympanic membrane (eg, tympanostomy tube)
* Confirm the diagnosis of acute OE and reassess the differential diagnosis within 48-72 hours in cases refractory to the initial therapy

**EPIDEMIOLOGY**

### United States and international statistics

OE is found in all regions of the United States, occurring in 4 of every 1000 people annually.The infection is believed to be more prevalent in hot and humid conditions such as during the summer months, presumably because participation in aquatic activities is higher.Acute, chronic, and eczematous OE are also common. Necrotizing OE is rare.

The international frequencies of OE have not been fully determined; however, the incidence is increased in tropical countries.

### Age-, sex-, and race-related demographics

Although the infection can affect all age groups, OE appears to be most prevalent in the older pediatric and young adult population, with a peak incidence in children aged 7-12 years.A single epidemiologic study from the United Kingdom found a similar 12-month prevalence for individuals aged 5-64 years and a slight increase in prevalence for those older than 65 years.This was postulated to occur secondary to an increase in comorbidities, as well as an increase in the use of hearing aids, which may cause trauma to the EAC.

OE affects both sexes equally. No racial predilection has been established, though people in some racial groups have small ear canals, which may predispose them to obstruction and infection.

**PREDEFINED Q & A SETS**

Q1: What is swimmer’s ear?  
A1: Swimmer’s ear is an infection of the outer ear canal, often caused by bacteria. It can also be caused by fungi or viruses, but bacteria are the most common culprit. It typically happens when water remains in the ear, creating a moist environment that helps bacteria grow.

Q2: What causes swimmer’s ear?  
A2: Causes include water trapped in the ear after swimming or bathing, trauma or irritation from inserting objects (like cotton swabs), and damage to the skin lining the ear canal. Swimming in polluted water or having skin conditions can increase risk.

Q3: What are the symptoms of swimmer’s ear?  
A3: Symptoms usually start mild and worsen, and may include itching inside the ear, redness, pain (especially when pulling on the earlobe or pushing on the tragus), swelling, fluid or pus draining from the ear, muffled hearing, and sometimes fever in severe cases.

Q4: How is swimmer’s ear diagnosed?  
A4: Diagnosis is made clinically by a healthcare provider examining the ear canal with an otoscope to look for redness, swelling, debris, and to check the eardrum status. Sometimes samples of ear discharge are sent for testing if infection is persistent or severe.

Q5: How is swimmer’s ear treated?  
A5: Treatment typically involves ear drops containing antibiotics and sometimes steroids to reduce inflammation. Keeping the ear dry and avoiding inserting objects is important. Severe infections may require systemic antibiotics or specialist care.

Q6: Can swimmer’s ear be prevented?  
A6: Yes. Preventive measures include keeping ears dry, using ear plugs when swimming, drying ears thoroughly after water exposure, avoiding inserting objects into the ear, and not swimming in polluted water.

Q7: Is swimmer’s ear contagious?  
A7: No, swimmer’s ear is not contagious from person to person, but people swimming together can get infections simultaneously due to shared environmental exposure.

Q8: When should I see a doctor urgently?  
A8: Seek prompt care if you have severe pain that spreads to the face or neck, high fever, swelling around the ear, persistent or worsening symptoms despite treatment, or hearing loss.

Q9: What are possible complications of untreated swimmer’s ear?  
A9: Complications include spread of infection to surrounding tissues and bone (malignant otitis externa), abscess formation, chronic infection, and rare but serious airway or systemic infections, especially in people with diabetes or immune problems

**DOCTOR-PATIENT CONVERSATIONS**

Doctor: Hello! What brings you in today?

Patient: I’ve been having some pain and itching in my ear. It feels swollen, and it hurts especially when I pull on my earlobe.

Doctor: That sounds like it could be swimmer’s ear, which is an infection or inflammation of the outer ear canal. It’s quite common, especially if your ear has been exposed to water recently.

Patient: Yes, I’ve been swimming a lot lately. How does swimming cause this?

Doctor: When water stays trapped in your ear canal, it creates a moist environment that allows bacteria to grow and cause infection. Sometimes even small scrapes or irritation from cleaning your ear can make it easier for germs to infect the skin in the canal.

Patient: What other symptoms should I look out for?

Doctor: You might notice redness, swelling inside the ear, pain that worsens when moving the ear or chewing, itchiness, discharge or fluid drainage, muffled hearing, and in severe cases, fever or swollen lymph nodes in the neck.

Patient: How do you confirm it’s swimmer’s ear?

Doctor: I’ll examine your ear canal with an otoscope, a special lighted instrument. If I see redness, swelling, and possibly discharge, it supports the diagnosis. Sometimes we take a sample if the infection is severe or not responding to treatment.

Patient: What treatment will I need?

Doctor: Usually, we treat it with antibiotic ear drops that also reduce inflammation. It’s important to keep the ear dry while it heals. If your pain is severe, pain relievers can help. In rare or severe cases, oral antibiotics or more specialized care might be necessary.

Patient: How long does it usually take to get better?

Doctor: With proper treatment, most people start feeling better within a few days, and the infection clears up in a week to 10 days.

Patient: Can I keep swimming?

Doctor: It’s best to avoid getting water in your ears until your ear has completely healed to prevent worsening or recurrence. You can use ear plugs or a swim cap when you return to swimming.

Patient: Is swimmer’s ear contagious?

Doctor: No, it’s not contagious from person to person, but swimming in polluted water or environments with a lot of bacteria increases risk.

Patient: How can I prevent this from happening again?

Doctor: Dry your ears thoroughly after water exposure by tilting your head and gently wiping the outer ear. Avoid sticking objects in your ear canal, and consider using ear drops to help dry moisture after swimming. Protect your ears with plugs if you swim frequently.

Patient: When should I come back or see a doctor urgently?

Doctor: If your pain worsens, you develop a fever, swelling extends beyond the ear, or symptoms don’t improve after treatment, please come back right away. Also, if you notice discharge with a foul smell or hearing loss, seek prompt care.

Patient: Thank you, doctor. I feel better knowing what to do.

Doctor: You’re welcome! Let me know if you have any questions or concerns while you’re healing.

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**Saccular cysts**

ALTERNATIVE NAMES

**DEFINITION / DESCRIPTION**

Saccular cysts are benign fluid-filled cysts that arise from the laryngeal saccule, a mucous gland-containing pouch located in the larynx between the false vocal cords and the thyroid cartilage. They may be congenital (present at birth) or acquired later in life.

* A saccular cyst is a dilated mucus-filled sac of the laryngeal saccule, which normally produces mucus to lubricate the vocal cords.
* It is located between the false vocal cords (ventricular folds) and the thyroid cartilage.
* The cyst does not communicate with the laryngeal lumen (unlike laryngoceles which contain air and connect with the laryngeal lumen).

**CAUSES**

1. Congenital Causes:
   * Caused by atresia (blockage) or obstruction of the laryngeal saccule’s orifice during development. This leads to mucus retention and cyst formation.
   * The obstruction may result from abnormal migration of embryonic tissues arising from the fourth branchial arch or mesenchymal cell anomalies during fetal development.
   * Congenital saccular cysts are rare and can cause airway obstruction in neonates or present later with voice changes.
2. Acquired Causes and Risk Factors:
   * Obstruction of the saccule opening due to:
     + Prolonged orotracheal intubation: Particularly in infants, intubation can cause inflammation, scarring, or blockage of the saccule opening leading to cyst formation.
     + Laryngeal trauma or surgery: Injury or surgery around the larynx may cause scarring that blocks the saccule.
     + Infection or inflammation: Chronic laryngeal inflammation can lead to blockage of the saccule orifice and mucus retention.
     + Neoplasms: Tumors near the laryngeal ventricle can obstruct drainage of the saccule, causing cyst formation.
     + Recurrent increased intralaryngeal pressure: Some suggest that repeated high pressures (e.g., chronic coughing or vocal strain) could contribute, though evidence is limited.

**SIGNS / SYMPTOMS**

* Symptoms depend largely on the size, location, and patient age.
* In infants, symptoms often include:
  + Airway obstruction causing stridor (noisy breathing), respiratory distress, noisy breathing, or difficulty breathing.
  + Feeding difficulties and weak or hoarse cry.
* In adults, symptoms tend to be related to voice changes such as hoarseness, voice fatigue, or breathing difficulty depending on size.
* Large cysts may cause airway obstruction requiring urgent attention.

## Types

* Two types described:
  + Anterior cysts: Submucosal masses dependent on the false vocal cord protruding into the ventricle.
  + Lateral cysts: More common, occupy the ventricular band and may extend to the pharynx via aryepiglottic fold.

**DIAGNOSIS METHODS**

1. Flexible or Rigid Laryngoscopy (Laryngoscopy or Microlaryngoscopy)
   * The gold standard diagnostic tool.
   * Allows direct visualization of the cystic lesion in the larynx, typically between the false vocal cords and thyroid cartilage.
   * Rigid laryngoscopy is often performed under general anesthesia and allows detailed assessment and planned surgical intervention if needed.
   * Video laryngeal stroboscopy can help characterize mucosal vibration and cyst impact.
   * Typical findings include a smooth, cystic swelling in the supraglottic region.
2. Imaging Studies
   * Computed Tomography (CT) Scan:
     + Useful to show a well-defined, fluid-filled cystic lesion without air content (distinguishing saccular cyst from air-filled laryngocele).
     + Helps determine the size, extent, and presence of any extralaryngeal extension (especially for lateral cysts).
     + Commonly used when endoscopic visualization is insufficient or in larger cysts.
   * Magnetic Resonance Imaging (MRI):
     + Provides excellent soft tissue resolution to define cyst content and relations.
     + Preferred in complex cases or when malignancy needs to be ruled out.
   * Imaging confirms the nature (fluid-filled), size, and adjacent tissue involvement.
3. Clinical Examination and History
   * Symptoms such as stridor, airway obstruction, dysphonia, or foreign body sensation prompt further laryngeal examination.
   * History of prior intubation may predispose to acquired cysts.
4. Differential Diagnosis
   * Important to distinguish saccular cysts from laryngoceles (air-filled cysts communicating with laryngeal lumen), ductal cysts (smaller mucosal cysts), and neoplasms.
   * Definitive diagnosis may require biopsy or excision to rule out rare malignancies.

**TREATMENT OPTIONS**

* Endoscopic CO2 Laser Excision:
  + The cyst is accessed under general anesthesia via direct laryngoscopy.
  + A CO2 laser creates a small mucosal incision over the cyst, and the cyst wall is carefully dissected and removed with microsurgical instruments.
  + This approach is effective for most anterior and some lateral cysts up to about 3 cm in size.
  + Allows good visualization, minimal intraoperative bleeding, and avoids external incisions.
* Endoscopic Marsupialization or Unroofing:
  + Sometimes used but carries a higher risk of cyst recurrence because the cyst wall is left partially intact.
* External Surgical Approaches:
  + Reserved for very large, laterally extending cysts, recurrent cysts after endoscopic treatment, or cases with extralaryngeal extension.
  + Approaches include lateral cervical incision via the thyrohyoid membrane or laryngofissure.
  + More invasive, longer recovery, higher risk to nerves (superior laryngeal nerve) and tissues.

## Side Effects and Complications of Treatment:

* Common post-operative effects:
  + Temporary hoarseness or voice changes due to surgical manipulation of laryngeal tissues.
  + Mild throat discomfort or soreness after surgery.

**OUTLOOK / PROGNOSIS**

* Effective treatment: Complete excision of saccular cysts via minimally invasive endoscopic approaches, such as CO2 laser excision, is the preferred treatment. This method achieves good symptom resolution with minimal damage to surrounding laryngeal structures.
* Symptom relief: Patients typically experience improvement or resolution of symptoms such as airway obstruction, stridor, dysphonia (voice changes), and foreign body sensation after surgery.
* Low recurrence rate: Studies show no recurrence of cysts during follow-up periods averaging around 20 months after complete endoscopic excision. Recurrence is more common if incomplete removal or marsupialization (unroofing) is performed instead of full excision.
* Good voice outcomes: Voice function generally improves after surgery, and major voice complications are uncommon with endoscopic techniques that preserve vocal cord integrity.
* Complications: Serious complications are rare but can include airway obstruction from very large cysts if untreated. Early detection and intervention prevent progression to respiratory distress or need for tracheotomy.
* Infants and children: Congenital saccular cysts presenting with respiratory distress in neonates can be treated successfully with timely intervention, resulting in good recovery and low mortality.

**POSSIBLE COMPLICATIONS**

* Risk of cyst recurrence if cyst wall is not fully removed, especially with marsupialization techniques.
* Scarring or stenosis of the supraglottic airway if excessive tissue damage occurs, potentially leading to voice changes or airway problems.
* Injury to nearby structures like the vocal cords or superior laryngeal nerve, causing voice weakness or sensory changes.
* General risks of anesthesia and surgery such as bleeding, infection, or airway compromise (rare).
* Endoscopic CO2 laser excision has been shown to achieve complete cyst removal with minimal recurrence and good voice preservation.
* Patients typically recover well with improvement in symptoms like airway obstruction or voice changes.

**WHEN TO SEE A DOCTOR / RED FLAG**

* In infants or children:
  + Noisy breathing (stridor) or difficulty breathing, especially if it worsens or seems severe
  + Respiratory distress or difficulty feeding
  + Weak, muffled, or hoarse cry
* In adults:
  + Persistent or worsening voice changes such as hoarseness or voice fatigue
  + Feeling of a lump or foreign body sensation in the throat
  + Symptoms of airway obstruction or breathing difficulty, especially during exertion
* Any signs of airway compromise like labored breathing or cyanosis (bluish discoloration) require immediate emergency care.

Saccular cysts present with symptoms depending on their size and location; airway obstruction is the common symptom in infants, while voice changes are more typical in adults. Early consultation with a doctor—usually an ear, nose, and throat specialist—is important for diagnosis by laryngoscopy and imaging, and to plan appropriate treatment to relieve symptoms and prevent airway obstruction.

**DIFFERENTIAL DIAGNOSIS**

| **Condition** | **Distinguishing Features** |
| --- | --- |
| Ductal cysts | Smaller mucous retention cysts arising from superficial mucosal glands. Usually <1 cm and lined by mucinous epithelium, located superficially in the laryngeal mucosa. |
| Laryngoceles | Air-filled dilatations of the laryngeal saccule that *communicate* with the laryngeal lumen. They can cause intermittent swelling and are visible on imaging as air-containing. |
| Laryngeal web | A membranous tissue across the glottis causing airway obstruction, distinct from cystic lesions. |
| Vocal fold paralysis | Presents with breathy voice and glottic insufficiency rather than a cystic mass. |
| Laryngomalacia | Congenital collapse of supraglottic structures causing stridor, often in infants; no cystic mass. |
| Subglottic stenosis | Narrowing of the airway below the vocal cords, recognized by direct visualization but not cystic. |
| Branchial cleft cyst | Usually located in the neck rather than inside the larynx; congenital cystic lesion. |
| Thyroglossal duct cyst | Typically midline neck cyst, not laryngeal. |
| Laryngeal neoplasms (e.g., oncocytic cystadenoma) | Rare tumors that may appear cystic; require biopsy to exclude malignancy. |
| Other cystic masses or pseudocysts | Rare benign lesions that may mimic cysts but differ histologically |

**EPIDEMIOLOGY**

* The incidence of congenital saccular cysts is estimated at approximately 1.8 cases per 100,000 live births. Some sources report even lower rates, such as less than 1 per 300,000 live births, highlighting the rarity of this condition in neonates and infants.
* Saccular cysts represent about 25% of all laryngeal cysts and account for 2-5% of all benign laryngeal lesions overall.
* Cases can occur at any age from infancy to older adulthood. In a retrospective series of seven patients, ages ranged from 1 to 81 years, with a mean age of about 34 years. Both pediatric and adult cases occur, with two infants presenting with respiratory distress and five adults presenting with voice symptoms in that series.
* There appears to be no strong gender predilection, though small case series have had variable male-to-female ratios.
* Congenital cysts are often detected in neonates with respiratory distress or stridor; acquired cysts can appear later and may be related to factors such as prior intubation or laryngeal trauma.
* In adults, saccular cysts are rare and are part of the broader category of benign laryngeal lesions, which are uncommon in the general population.

**PREDEFINED Q & A SETS**

Q1: What are saccular cysts?  
A1: Saccular cysts are benign, mucus-filled cysts that arise from the laryngeal saccule, a small pouch in the larynx located between the false vocal cords and the thyroid cartilage. They can be present from birth (congenital) or develop later (acquired).

Q2: What symptoms do saccular cysts cause?  
A2: Symptoms depend on cyst size and location. In infants, they may cause noisy breathing (stridor) and difficulty breathing. In adults, they often cause voice changes such as hoarseness or voice fatigue. Large cysts can obstruct the airway.

Q3: How are saccular cysts diagnosed?  
A3: Diagnosis is usually made by direct visualization of the cyst through flexible or rigid laryngoscopy. Imaging tests like CT or MRI scans are used to assess size and extent and to differentiate saccular cysts from similar lesions like laryngoceles.

Q4: What causes saccular cysts?  
A4: Congenital cysts result from developmental blockage of the laryngeal saccule’s opening during fetal growth. Acquired cysts may form due to prolonged intubation, laryngeal trauma, infection, inflammation, or tumors obstructing the saccule.

Q5: How are saccular cysts treated?  
A5: The primary treatment is surgical removal. Most cysts can be removed endoscopically using a CO2 laser or microsurgical instruments. Larger or laterally located cysts might require open neck surgery. Complete excision reduces recurrence risk.

Q6: What are the risks or side effects of treatment?  
A6: Treatment risks include temporary hoarseness, throat discomfort, potential scarring, and rare injury to nearby nerves affecting voice. Incomplete removal can lead to cyst recurrence. Surgical risks and anesthesia complications are also possible.

Q7: What is the prognosis for saccular cysts?  
A7: Prognosis is excellent with timely diagnosis and complete surgical removal. Symptoms usually improve, and recurrence is rare when fully excised. Untreated large cysts can cause airway obstruction.

Q8: Who is at risk of developing saccular cysts?  
A8: Congenital cysts are rare and present at birth. Acquired cysts may occur in people with a history of prolonged intubation, laryngeal injury, or chronic inflammation.

Q9: Can saccular cysts recur after treatment?  
A9: Recurrence is uncommon if the cyst is completely removed. Partial removal procedures, like marsupialization, carry a higher recurrence risk.

Q10: When should I see a doctor about a possible saccular cyst?  
A10: See a doctor if you or your child has persistent voice changes, breathing difficulties (especially noisy or labored breathing), swallowing problems, or if airway obstruction symptoms appear.

**DOCTOR-PATIENT CONVERSATIONS**

Doctor: Hello! I understand you've been experiencing some voice changes and possibly some breathing difficulties. Can you tell me more about what you’ve noticed?

Patient: Yes, doctor. I've been feeling like there’s something in my throat, and my voice sounds hoarse and tired sometimes. I also sometimes have trouble breathing, especially when I exert myself.

Doctor: Thank you for sharing that. Based on your symptoms and examination, it looks like you have a saccular cyst. This is a benign — or non-cancerous — fluid-filled cyst that forms in an area of your larynx called the laryngeal saccule, which is a small pouch that normally makes mucus to keep your vocal cords moist.

Patient: How did this cyst form? Is it serious?

Doctor: Saccular cysts can be present from birth, or they might develop later due to something blocking the mucus drainage, like irritation or injury. While the cyst itself isn't cancerous, if it grows large it can cause symptoms like hoarseness, feeling of a lump, or even difficulty breathing. So, it's important to treat it.

Patient: How do you know for sure I have this cyst?

Doctor: We did a flexible laryngoscopy, where a small camera looks inside your throat and vocal cords. It showed a smooth, cyst-like swelling in the larynx. Sometimes, if needed, we also use imaging like a CT or MRI scan to understand its size and exact location.

Patient: What can be done to fix it?

Doctor: The best treatment is surgical removal. Usually, we perform a minimally invasive procedure using a laser through your mouth while you are under general anesthesia. This allows us to remove the cyst precisely with minimal damage to surrounding tissues. For larger or more complicated cysts, an open surgery approach might be needed, but that’s less common.

Patient: Are there any risks or side effects from the surgery?

Doctor: You might experience some temporary hoarseness or throat soreness after the procedure, which typically improves with time. There's a small risk of scarring or voice changes if the surgery affects nearby tissues, but modern techniques keep these risks very low. Recurrence is uncommon if the cyst is fully removed.

Patient: How long is recovery, and what should I expect after surgery?

Doctor: Most people recover well within a few weeks. You'll likely see improvement in your voice and breathing as the cyst is no longer causing obstruction. Follow-up appointments help us monitor healing and make sure there’s no recurrence.

Patient: Is this cyst life-threatening?

Doctor: No, it’s not cancerous or life-threatening. But if left untreated and it grows, it could block your airway, which would be a medical emergency. That’s why we recommend surgery to remove it safely and improve your symptoms.

Patient: Thank you, doctor. That helps me understand what’s going on and what to expect.

Doctor: You’re welcome! If you have any questions before or after the surgery, feel free to reach out. We’ll work together to help you get better

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**Salivary duct stones**

**DEFINITION / DESCRIPTION**

Sialolithiasis is a benign condition where stones form in the salivary ducts. These stones are called calculi and are mostly composed of calcium. They can develop in any of the salivary glands, including the parotid, submandibular, or sublingual, but are most likely to form in the submandibular gland.

Children rarely develop sialolithiasis. They are most commonly found in men between the ages of 30 and 60.

## **Saliva's Function**

Saliva is mostly made up of water but also contains small amounts of electrolytes, calcium, phosphate, important antibacterial compounds, and digestive enzymes. The antibacterial properties of saliva protect against:

* Oral infections
* Chronic dry mouth
* Gum disease
* Tooth decay

The digestive enzymes in saliva begin breaking down your food before you have even swallowed it and saliva is most commonly released in response to the smell and taste of food. Additional functions of saliva include helping us to swallow and talk.

Saliva is produced by several glands located in the mouth and throat. The major salivary glands then transport the saliva through tiny tubes called salivary ducts which eventually release the saliva into various places in your mouth, especially under your tongue and on the floor of your mouth. The three pairs of major salivary glands are called the parotid, submandibular, and sublingual glands.

In addition to the major saliva glands, there are multiple smaller glands, called minor salivary glands, located in your lips, cheeks, and throughout the tissue lining your mouth.

**CAUSES**

Conditions such as dehydration which cause thickening, or decreased water content of the saliva can cause the calcium and phosphate in saliva to form a stone. The stones often form in the salivary ducts and can either totally obstruct the salivary duct, or partially occlude it. You may develop sialolithiasis even if you are healthy, and a cause may not always be able to be pinpointed. However, conditions that may cause thick saliva and subsequent sialolithiasis include:

* Dehydration
* Use of medications or conditions which cause dry mouth (diuretics and anticholinergics)
* Sjorgen's syndrome, lupus, and autoimmune diseases in which the immune system may attack the salivary glands
* Radiation therapy of the mouth
* Gout
* Smoking
* Trauma

Submandibular stones account for 80 to 92 percent of all sialolithiasis, while parotid stones account for most of the remaining cases at 6 to 20 percent. The sublingual and minor glands have relatively low risk for development of a stone. Most stones only occur in one gland, but it is possible to have multiple stones form at once.

Small stones that do not block the flow of saliva can occur and cause no symptoms. However, when the flow of saliva becomes completely blocked it may cause the associated salivary gland to become infected

**SIGNS / SYMPTOMS**

Symptoms usually occur when you try to eat (since that's when the flow of saliva is stimulated) and may subside within a few hours after eating or attempting to eat. This is important to tell your healthcare provider since it may help differentiate sialolithiasis from other conditions. Symptoms of sialolithiasis may include:

* Swelling of the affected saliva glands which normally occurs with meals
* Difficulty opening the mouth
* Difficulty swallowing
* A painful lump under the tongue
* Gritty or strange tasting saliva
* Dry mouth
* Pain and swelling usually around the ear or under the jaw

Severe infections of a salivary gland may cause profound symptoms including fever, fatigue, and sometimes noticeable swelling, pain, and redness around the affected gland.

**DIAGNOSIS METHODS**

An otolaryngologist, or ENT, is a physician qualified to diagnose and treat sialolithiasis. Although healthcare providers in other specialties may also diagnose or treat this condition.

Your healthcare provider will consider your medical history and examine your head and neck, including the inside of your mouth. Sometimes the stone can be felt as a lump. Historically a sialograph, where dye is injected into the salivary duct followed by an X-ray, was used, however, this is more invasive than modern MRI or CT scans which are now more likely to be used.

**TREATMENT OPTIONS**

The treatment of sialolithiasis depends on where the stone is and how large it is. Small stones may be pushed out of the duct and you may be able to facilitate this by drinking plenty of water, or massaging and applying heat to the area. Sometimes a healthcare provider can push the stone out of the duct and into the mouth by using a blunt object and gently probing the area.

Large salivary duct stones may be more difficult to remove and sometimes require surgery. Sometimes a thin tube called an endoscope can be inserted into the duct. If the stone can be seen with the endoscope the healthcare provider may be able to insert another tool that is then used to pull the stone out.

Sometimes removal of the stone can be achieved with a small incision, in severe cases the entire gland and the stone may have to be surgically removed.

In the case of an infected gland, your healthcare provider may prescribe an oral antibiotic. Never take antibiotics without seeing a healthcare provider.

**PREVENTION TIPS**

You can’t prevent sialolithiasis altogether. But you can reduce your risk by staying hydrated, avoiding smoking and practicing good oral hygiene. If you start to feel pain or tenderness around your salivary ducts, try sucking on sour candies and starting gland massage to stimulate the flow of saliva. This could encourage any stones to pass naturally

**OUTLOOK / PROGNOSIS**

Sialolithiasis has an excellent prognosis, and the majority of patients can be managed conservatively with sialogogues and nonsteroidal anti-inflammatory drugs. The minimally invasive procedures discussed above have excellent success rates with minimal morbidity compared to traditional surgical techniques. Sialadenectomy for the treatment of sialolithiasis is rarely necessary with modern treatment techniques

**POSSIBLE COMPLICATIONS**

The primary complications of sialolithiasis are the development of sialadenitis, acute or chronic, and atrophy of the affected salivary gland. Obstruction of the salivary glands by a sialolith blocks the flow of saliva resulting in swelling and pain. Additionally, this blockage of flow prevents the removal of bacteria and debris from the salivary duct, resulting in bacterial infection. If the obstruction is chronic, the blocked flow of saliva will damage the salivary glands' acinar cells, resulting in local inflammation. Without proper treatment, it can result in permanent fibrosis of the gland and atrophy

**WHEN TO SEE A DOCTOR / RED FLAG**

If your symptoms continue for more than two weeks with no improvement or if you develop severe pain and redness of the skin overlying the gland, you should call a healthcare provider for further instructions.

**DIFFERENTIAL DIAGNOSIS**

The differential diagnosis for salivary gland swelling includes:

* Sialolithiasis
* Sialadenitis (inflammatory or infectious)
* Neoplasm

However, the clinical presentation can vary, and the differential diagnosis for the pathology of the oral cavity and face can be extensive and relies heavily on physical exam findings and clinical history. Conditions that can have a similar presentation include:

* Cellulitis
* Poor dentition and dental abscess formation
* Infection of the buccal or masticator space
* Herpes zoster
* Neoplasm

**EPIDEMIOLOGY**

The incidence of sialolithiasis is estimated at 1 in 10,000 to 1 in 30,000 individuals. The primary age of diagnosis is between 30 and 60, with a higher incidence in men. Approximately 85% of sialoliths occur within the submandibular gland, making it the most common location for sialolithiasis. One of the reasons is that the submandibular duct ascends towards its opening in the oral cavity, resulting in a stagnant flow of saliva. Additionally, the submandibular gland produces predominately mucinous saliva, which is more viscous than the secretions created by the parotid gland, resulting in a more stagnant flow of secretions. The submandibular gland also produces more alkaline saliva, which predisposes to the precipitation of inorganic salts (eg, calcium and phosphate), further leading to salivary stone formation. Approximately 15% of salivary stones occur within the parotid gland, and less than 5% occur within the sublingual and minor salivary glands.

**PREDEFINED Q & A SETS**

#### **Who does sialolithiasis affect?**

Anyone can get sialolithiasis. But the condition is most common in males aged 30 to 60. Sialolithiasis rarely affects children.

##### **How common are salivary stones?**

Even though sialolithiasis is the most common reason why people develop sialadenitis (salivary gland swelling), the condition is still rare overall. In fact, salivary gland stones only occur in about 1 in every 30,000 people.

#### **Is sialolithiasis contagious?**

No, salivary stones themselves aren’t contagious. However, if you have a viral or bacterial salivary gland infection, you could potentially pass it on to other people.

### **What tests can help diagnose sialolithiasis?**

In some cases, your healthcare provider may request imaging tests to confirm your diagnosis or to see the location of any salivary gland stones. These tests may include:

* X-rays.
* CT (computed tomography) scans.
* MRI (magnetic resonance imaging).

Your provider may also request a salivary gland scan. During this test, they’ll inject a radioactive tracer into a vein in your arm. Next, they’ll position you in front of a special device — a gamma scintillation camera — which detects radiation and captures images. About 45 minutes into the test, your provider will give you a lemon drop or other sour substance to stimulate saliva production. As your saliva glands release saliva, your provider will take more pictures to see how much saliva remains in your ducts.

### **What can I expect if I have sialolithiasis?**

If you have a blocked salivary gland, you may experience pain or swelling around your salivary ducts during mealtimes. Usually, these symptoms last about one to two hours, then diminish until your next meal.

### **Does sialolithiasis go away?**

In many cases, sialolithiasis goes away on its own or with conservative, at-home treatments, such as heat application, gentle massage and sucking on sour candy. If your symptoms continue after trying these home remedies, you should call a healthcare provider.

### **What will happen if sialolithiasis is not treated?**

Left untreated, sialolithiasis can result in infections or abscesses that can spread to deeper spaces in your neck. It’s important to call a healthcare provider if you have persistent facial pain and swelling.

### **Sialolithiasis vs. sialadenitis: What’s the difference?**

“Sialolithiasis” refers to calculi, or salivary gland stones. “Sialadenitis” refers to inflammation or infection of your salivary gland. Salivary gland stones can result in sialadenitis, though this isn’t always the case.

Q5: How are salivary duct stones diagnosed?  
A5: Diagnosis is usually clinical, supported by imaging tests such as ultrasound, X-rays, CT scan, or sialography to locate the stone and assess duct blockage.

Q6: Can salivary duct stones go away on their own?  
A6: Small stones may sometimes be dislodged by home measures such as massaging the gland, staying well hydrated, and stimulating saliva flow (e.g., sucking on sour candies). However, larger stones usually require medical treatment.

Q7: How are salivary duct stones treated?  
A7: Treatment options range from conservative management (hydration, massage, heat application) to medical procedures like minimally invasive removal using sialendoscopy (small camera and tools) or surgery if necessary. Antibiotics may be prescribed if infection occurs.

Q8: Where do these stones commonly occur?  
A8: Most stones develop in the submandibular glands (under the jaw), but they can also occur in the parotid glands (in front of the ears).

Q9: What are potential complications?  
A9: Complications include chronic gland swelling, infections, abscess formation, and rarely, spread of infection to nearby tissues if not promptly treated.

Q10: How can salivary duct stones be prevented?  
A10: Maintain good hydration, practice good oral hygiene, avoid medications that reduce saliva flow if possible, and promptly treat any gland infections.

**DOCTOR-PATIENT CONVERSATIONS**

Doctor: Hello! What brings you in today? Are you having any pain or swelling in your face or mouth?

Patient: Yes, doctor. I've noticed some swelling and pain under my jaw, especially when I'm eating. It feels like something is blocking my saliva flow.

Doctor: That sounds like it could be a salivary duct stone, which is a hard deposit that forms in one of the salivary gland ducts and blocks saliva from flowing into your mouth. This blockage causes swelling and pain, particularly during meals when your glands try to release saliva.

Patient: How do these stones form? Is it serious?

Doctor: Salivary stones usually develop when minerals like calcium build up and form a lump inside the duct. Factors like dehydration, reduced saliva production, or duct narrowing can contribute. While they are usually not dangerous, they can cause discomfort, swelling, and sometimes infections if untreated.

Patient: How do you confirm if I have a stone?

Doctor: We usually diagnose salivary stones through a physical exam and imaging tests, like an ultrasound or sometimes a CT scan, to locate the stone and assess its size and position.

Patient: What are the treatment options?

Doctor: If the stone is small, you might be able to help it pass on its own by massaging the area, staying well hydrated, applying warm compresses, or sucking on sour candies to stimulate saliva flow. If it doesn’t pass or if it's larger, we can remove it using minimally invasive techniques such as sialendoscopy—an endoscopic procedure where a tiny camera and instruments remove the stone through the duct.

Patient: Is surgery necessary? Are there risks?

Doctor: Surgery is usually reserved for stones that can't be removed with less invasive methods or stones located deep in the gland. Though surgery involves some risks like swelling or injury to nearby nerves, most patients do well. The goal is always to preserve your salivary gland and relieve symptoms with minimal discomfort.

Patient: What should I expect after treatment?

Doctor: After removal, pain and swelling usually improve quickly. We'll monitor you for any infections or complications, and you might need antibiotics if an infection occurred. Drinking plenty of water and good oral hygiene help prevent future stones.

Patient: Can stones come back?

Doctor: They can, but maintaining good hydration and oral care reduces the risk. If symptoms recur, you should see us promptly.

Patient: Thank you, doctor. That makes me feel more comfortable about the treatment.

Doctor: You’re welcome. Let me know if you have any questions or if your symptoms change. We’re here to help you.

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**Temporal bone cancer**

**ALTERNATIVE NAMES**

1. Malignant tumors of the temporal bone
2. Temporal bone carcinoma
3. Squamous cell carcinoma of the temporal bone (most common histologic type)
4. Temporal bone neoplasm
5. External auditory canal carcinoma (when arising from the ear canal)
6. Middle ear carcinoma (if arising primarily in the middle ear portion of the temporal bone)
7. Temporal bone malignancy
8. Temporal bone cancerous tumor

**DEFINITION / DESCRIPTION**

Carcinoma of the temporal bone is rare, accounting for fewer than 0.2% of all tumors of the head and neck. Only 200 new cases of temporal bone cancer may be diagnosed each year across the United States. This number includes cancers arising from skin of the pinna that spread to the temporal bone; primary tumors of the external auditory canal (EAC), middle ear, mastoid, or petrous apex; and metastatic lesions to the temporal bone.

## **Staging**

Staging systems are intended to help classify patients preoperatively into groups whereby decisions regarding treatment may be made on the basis of comparison to previously treated patients with similar tumor characteristics (ie, the stage). To date, no staging system for temporal bone malignancies is universally accepted. Several factors impede the process of developing a staging system, including the rarity of the tumors, the impossibility of evaluating disease extent by physical examination alone, and the unreliability of radiographic studies to determine the extent of disease in certain situations.

Many authors have proposed staging systems concurrent with a review of patient series from major institutions; however, the small number of patients per group, the disparity of staging criteria, the diversity in management protocols, and the use of nonstandardized surgical nomenclature prohibits meaningful comparison of outcomes. In addition, some patients are reportedly classified into groups with variability in histology types and sites of tumor origin, which further confounds analysis of outcome by stage.

Numerous staging systems have been proposed; however, to date, no universally accepted staging system for temporal bone cancers exists. A staging system for squamous cell cancers of the EAC proposed by the University of Pittsburgh has been shown useful and has gained support in the literature.This staging system is based on clinical, radiologic, and pathologic findings. In general, tumors that are limited to the EAC are defined as early disease, and those that extend beyond the external canal to invade the surrounding soft tissues, the middle ear, the mastoid, or CNs are recognized as advanced disease.

In the original staging system proposed by Pittsburgh, lesions were defined as follows:

* T1 - Tumor limited to the EAC without bony erosion or evidence of soft tissue involvement
* T2 - Tumor with limited EAC bone erosion (not full thickness) with limited (< 0.5 cm) soft tissue involvement
* T3 - Tumor eroding the osseous EAC (full thickness) with limited (< 0.5 cm) soft tissue involvement or tumor involving the middle ear, mastoid, or both
* T4 - Tumor eroding the cochlea, petrous apex, medial wall of the middle ear, carotid canal, or jugular foramen of dura; or with extensive soft tissue involvement (>0.5 cm), such as involvement of the temporomandibular joint or stylomastoid foramen; or with evidence of facial paresis

The Pittsburgh staging system has become widely applied in case reports of temporal bone cancer.

The Pittsburgh staging system was modified by the authors after further review of patients from an extended series.In the modified staging system, facial nerve weakness is considered a criterion for a T4 lesion. The authors observed that facial nerve paresis did not occur in lesions otherwise classified as limited T1, T2, or T3 lesions. Involvement of the facial nerve would be otherwise classified as T4 based on the anatomical area of involvement, including the medial wall of the middle ear (horizontal segment), extensive bony erosion within mastoid (vertical segment), or involvement of stylomastoid foramen. In the T4 group, survival was similar between patients with and without facial paralysis (unpublished). A few reports have used the modified staging system.

Nodal involvement and stage can be classified as it is for other cancers of the head and neck.

* N1 - Single ipsilateral lymph node, size less than 3 cm
* N2 - Single ipsilateral node, size 3-6 cm
* N2b - Multiple ipsilateral nodes, all less than 6 cm
* N2c - Bilateral or contralateral nodes, all less than 6 cm
* N3 - Nodes involved greater than 6 cm

Cancer is staged as follows:

* Stage 0 - Tis N0 M0
* Stage I - T1 N0 M0
* Stage II - T2 N0 M0
* Stage III - T3 N0 M0, T1 N1 M0, T2 N1 M0, T3 N1 M0
* Stage IV - T4 N0 M0, T4 N1 M0, any T N2 M0, any T N3 M0, any T any N M1

**CAUSES**

* Chronic inflammation and infections: Chronic otitis media (middle ear infection) and cholesteatoma (abnormal skin growth in the middle ear) have been implicated as important etiologic factors due to persistent inflammation that may lead to malignant transformation.
* Sun exposure and skin cancer: Prolonged and excessive exposure to ultraviolet (UV) rays from the sun is a major risk factor, particularly for skin cancers such as basal cell carcinoma and squamous cell carcinoma of the ear skin (pinna) that can involve or spread to the temporal bone. Fair-skinned individuals are at greater risk.
* Previous radiation therapy: Prior radiation treatment to the head and neck area increases the risk, possibly due to radiation-induced DNA damage leading to cancer development in the temporal bone.
* Human papillomavirus (HPV) infection: Some studies suggest an association between high-risk HPV strains (e.g., HPV16/18) and squamous cell carcinoma of the middle ear and temporal bone, possibly through oncogenic mechanisms.
* Parotid gland tumors: Tumors originating from the parotid gland, which lies adjacent to the temporal bone, can invade or involve the bone secondarily.
* Other factors: Chronic skin infections, fair complexion, and male sex have been noted as risk factors.

**SIGNS / SYMPTOMS**

Tumors often start as scaly areas or white bumps on the outside of the ear. The area might ooze or drain. A tumor also might start inside the ear canal. The patient might notice drainage from the canal or pain inside the ear.

**An ear, nose and throat (ENT) specialist must examine any ear infection that does not go away.**

**Basal skin carcinoma** is the most common type of ear and temporal bone cancer. A scaly area of skin on the ear, which does not improve with the application of moisturizer, is usually the first sign. Then, a pearly white bump appears which grows slowly. The lump can be painless or an ulcer might develop in the center of the lump. The ulcer later bleeds and becomes painful. These tumors can spread to the inside of the ear but rarely other parts of the body.

**Squamous cell cancer** grows deeper into the body and is more likely to spread. If the tumor grows into the temporal bone it can cause hearing loss, dizziness, and facial paralysis.

**DIAGNOSIS METHODS**

1. Clinical Examination
   * Evaluation by an otolaryngologist (ENT specialist) who examines the ear canal, external ear, and surrounding structures for suspicious lesions.
   * Symptoms such as ear pain, hearing loss, discharge, bleeding, or facial weakness raise suspicion.
   * Biopsy of visible lesions in the ear canal or skin can be performed in-office to obtain tissue for histopathologic confirmation of cancer.
2. Imaging Studies
   * Computed Tomography (CT) Scan of the temporal bone and neck is essential to assess:
     + Bone erosion or destruction caused by the tumor,
     + Extent of soft tissue involvement,
     + Involvement of nearby structures including middle ear, mastoid, skull base,
     + Presence of cervical lymphadenopathy (lymph node involvement).
     + Often performed with contrast for better delineation.
   * Magnetic Resonance Imaging (MRI)
     + Provides superior soft tissue detail,
     + Helps differentiate tumor from inflammation,
     + Evaluates possible involvement of the brain, dura, facial nerve, and vascular structures.
   * Positron Emission Tomography (PET) Scan (sometimes used)
     + Helps detect distant metastases or assess metabolic activity of the tumor.
3. Biopsy
   * Tissue sample from the lesion confirms the diagnosis and histological type (e.g., squamous cell carcinoma, basal cell carcinoma).
   * Biopsy is often done under local anesthesia in clinic or under general anesthesia depending on lesion accessibility.
4. Laboratory Tests
   * Routine preoperative labs include complete blood count, coagulation studies, liver and renal function tests to evaluate overall patient health before surgery.
   * No specific blood markers for temporal bone cancer exist.
5. Staging Systems
   * The Pittsburgh Staging System is commonly used for temporal bone squamous cell carcinoma, based on tumor size, bone invasion, and lymph node involvement, guiding prognosis and treatment.

**TREATMENT OPTIONS**

## Medical Therapy

Primary radiation is ineffective for curative treatment. In the most extreme cases in which contraindications to surgery are serious deterrents to an operation, palliative radiation and chemotherapy may be offered. The literature supports a beneficial effect of adjunctive radiation on survival, but no well-controlled studies have been performed. Postoperative radiation treatment may be indicated in advanced disease. Most authors advocate full course postoperative radiation to stage T3 or T4 tumors as defined by the University of Pittsburgh staging system. Some authors also recommend radiation for T2 disease.

The literature supports a beneficial effect of postoperative radiation on survival.The temporal bone and neck should be treated with 50-60 Gy for tumors staged T3 and T4. Radiation may also be indicated for smaller lesions.

Preoperative chemotherapy was reviewed by Nakagawa et al in a retrospective series of patients treated with preoperative chemoradiation or with chemoradiation alone).Four of 8 patients treated with chemoradiation (5-flourouracil or a fluoropyrimidine complex during external beam radiation with a dose of 40Gy), followed by chemotherapy in one case, were free of disease at 24-47 months. Pemberton et al reported 53% cancer-specific survival of 123 patients treated with radiotherapy alone (55 Gy).

A study by Noda et al indicated that nivolumab, an immune checkpoint inhibitor used against platinum-refractory metastatic or recurrent head and neck squamous cell carcinoma, has some therapeutic effect in temporal bone squamous cell carcinoma. The drug was administered to nine patients in whom recurrent or residual temporal bone squamous cell carcinoma was found following platinum-including chemotherapy and/or chemoradiotherapy. The investigators determined that one patient showed a partial response, two had stable disease, and four demonstrated progressive disease, with the size having not been assessed in two cases. Moreover, the 1-year overall survival rate was greater in the patients on nivolumab than in five patients who were not treated with the agent (33.3% vs 20.0%, respectively).

Based on a retrospective study and literature review of patients with temporal bone paragangliomas, Prasad et al recommended that elderly patients with advanced forms of these tumors be managed with a wait-and-scan approach, with radiotherapy reserved for the treatment of fast-growing tumors. In the study, among patients with class C or D tumors who underwent wait-and-scan management, the investigators found that the tumor remained stable in 22 of 24 patients who were followed up for less than 3 years and that the paraganglioma remained stable or regressed in 10 of 12 patients who were followed up for 3-5 years. In addition, the tumor remained stable or regressed in five of 11 patients who were followed up for more than 5 years.

A literature review, Prasad and colleagues stated, revealed no conclusive evidence that radiotherapy is an effective primary modality for the treatment of either class C or D temporal bone paragangliomas

## Surgical Therapy

In general, all patients who are medically able should undergo surgical treatment. The optimal surgery removes all of the cancer en bloc because positive margins are associated with poor survival rates.The resection procedures that can be performed for the temporal bone include a modified lateral temporal bone resection, lateral temporal bone resection, subtotal temporal bone resection, and total temporal bone resection. The specific procedures and nomenclature vary among surgeons. Adjunctive surgical procedures, including neck dissection, parotidectomy, and craniotomy, should be performed when indicated. Advanced tumors with intracranial invasion have a grave prognosis, and treatment should probably be limited to palliation with less extensive (and less morbid) surgical procedures

**OUTLOOK / PROGNOSIS**

Prognosis is related to the primary site, the histologic type, and treatment.The outcome of treatment for early stage tumors is favorable, with most series reporting an 80-100% survival rate. Later stage cancers after adequate surgery and radiation treatment have a survival between 50% and 80%. Advanced lesions have a poor prognosis despite aggressive treatment, with 2-year survival of 0-40%.Salvage surgery after an incomplete resection is associated with a poorer prognosis compared to a definitive procedure in an undisrupted field.A mastoidectomy or limited excision performed by an outside institution prior to referral complicates preoperative assessment and staging, disrupts margins, and forces the surgeon to operate in an inflamed field with interrupted landmarks. Positive margins, nodal disease, dural involvement, and advanced disease are associated with poorer survival.

A literature review by Sioufi et al found that most patients with temporal bone squamous cell carcinoma present in an advanced stage of the disease, with almost 22% demonstrating nodal metastasis. The 5-year overall survival (OS) rate in the study differed according to T stage, being 81.9% for patients with T1/T2-stage cancer, and 47.5% for those with T3/T4 disease. Five-year OS for patients with T1/T2 cancer who underwent surgery alone did not significantly differ from OS for those who were treated with radiation alone (100% vs 81.3%, respectively); the same was true for patients with T3/T4 disease treated with surgery with postoperative chemoradiotherapy, surgery with postoperative radiotherapy, or chemoradiotherapy alone (50% vs 53.3% vs 58.1%, respectively). However, the investigators stated that low cohort size limited their ability to draw strong conclusions of the effect of various therapies on OS in early and late-stage disease.

A retrospective study by Komune et al of outcomes and prognostic factors in temporal bone squamous cell carcinoma found the 5-year disease-specific survival rate to be higher in stage T1 to T3 cases than in patients with stage T4 tumors, the rates being 100% (T1), 92% (T2), 86% (T3), and 51% (T4). No correlation was found between poor survival and extension of the tumor to the middle ear cavity, as occurred in 13 of 17 stage T3 cases.

Another study by Komune et al found, through univariate Cox regression analysis, that in patients with temporal bone squamous cell carcinoma, a poor prognosis is associated with high ratios of neutrophils to lymphocytes and platelets to lymphocytes, a low ratio of lymphocytes to monocytes, a Glasgow Prognostic Score of 2, and a Systemic Inflammation Score of 2. Thus, the report indicated a link between inflammation-based prognostic markers and survival in temporal bone squamous cell carcinoma.

A study by Hongo et al indicated that in temporal bone squamous cell carcinoma, the tumor’s immune microenvironment can help to predict prognosis. For example, the investigators reported an association between a high density of CD8+ tumor-infiltrating lymphocytes (TILs) and a complete response to chemoradiotherapy. On the other hand, a poorer prognosis was associated with expression of programmed death ligand 1 (PD-L1) (1% or more) and a high density of Foxp3+ TILs.

A study by Marioni et al indicated that in patients with advanced, aggressive temporal bone squamous cell carcinoma, the risk for tumor recurrence after postoperative radiotherapy is increased in those with a primary tumor classification of 4, a pathologic grade of 2 or 3, and dura mater involvement.

**POSSIBLE COMPLICATIONS**

Complications of treatment depend on the extent of resection and the use of adjunctive radiation. Postoperative hearing loss, facial nerve paralysis, vertigo, and other CN deficits (eg, CN V, VII, VIII, IX, X, XI) may occur. Dural resection may predispose to cerebral spinal fluid leaks, meningitis, or intracranial complications. Significant complications can result from trauma to or resection of the carotid artery. Radiation has known complications of fibrosis of soft tissues, destruction of salivary gland tissue, osteoradionecrosis of the temporal bone, and possibly central nervous system effects if the field of radiation extends to intracranial tissues.

**DIFFERENTIAL DIAGNOSIS**

| **Condition** | **Key Features and How to Differentiate** |
| --- | --- |
| Chronic and Granulomatous Otitis Media/Cholesteatoma | Chronic inflammation, bone erosion, and middle ear involvement; usually history of chronic ear infections; imaging shows non-neoplastic mass with bone erosion but no tumor cells. |
| Benign Tumors: |  |
| - Vestibular Schwannoma (Acoustic Neuroma) | Arises from vestibulocochlear nerve; usually in cerebellopontine angle; MRI shows characteristic nerve sheath tumor. |
| - Paragangliomas (Glomus Tumors) | Highly vascular, pulsatile masses; seen in jugular foramen region; characteristic "salt and pepper" MRI appearance. |
| - Osteomas and Exostoses | Slow growing, benign bony growths; usually asymptomatic unless large. |
| - Schwannomas of Facial or Trigeminal Nerves | Nerve sheath tumors with distinct MRI features. |
| Malignant Tumors: |  |
| - Squamous Cell Carcinoma (SCC) | Most common temporal bone cancer; often originates in external auditory canal or skin; aggressive bone destruction seen on imaging. |
| - Basal Cell Carcinoma | Less aggressive than SCC; can involve ear skin and temporal bone. |
| - Adenoid Cystic Carcinoma | Rare; arises from ceruminous glands; perineural invasion common. |
| - Sarcomas (e.g. Rhabdomyosarcoma) | More common in children; aggressive lesions with bone destruction. |
| - Metastases | Secondary deposits from distant cancers (breast, lung, kidney); consider patient history. |
| Infectious/Inflammatory Lesions: |  |
| - Skull Base Osteomyelitis | Infection of bone with bone destruction, systemic signs of infection. |
| - Tuberculosis or Other Granulomatous Disease | Chronic infection with lytic lesions; systemic symptoms. |
| Other Cystic or Mass Lesions: |  |
| - Endolymphatic Sac Tumors | Rare, benign but locally aggressive; arise in petrous bone; imaging shows characteristic cystic mass. |
| - Langerhans Cell Histiocytosis | May cause lytic bone lesions including temporal bone in children. |

## Diagnostic Tools to Differentiate:

* Imaging: CT scan is key to assess bone destruction, extent of lesion, and involvement of adjacent structures. MRI is essential for soft tissue characterization and to detect perineural spread or intracranial extension.
* Biopsy: Histopathologic confirmation is essential to differentiate between infection, benign tumors, primary malignancies, and metastases.
* Clinical History: Prior infections, skin lesions, systemic symptoms, and patient age guide suspicion.

**EPIDEMIOLOGY**

* The annual incidence is estimated at about 1 to 6 cases per 1 million people worldwide, making it a very uncommon cancer.
* It accounts for approximately 0.2% of all head and neck cancers.
* It can affect all ages but is more commonly diagnosed in middle-aged to older adults, typically people in their 50s to 60s on average.
* The gender distribution is roughly balanced, with some studies showing slightly more male patients, but many report close to equal male-to-female ratios. For example, one pooled dataset showed about 31% male and 29% female, with a large portion unknown.
* The majority of temporal bone cancers are squamous cell carcinomas (around 60-75%), followed by basal cell carcinomas and rare other histologic types such as sarcomas.
* Most patients present with advanced-stage disease: approximately 40-50% present at stage IV, with relatively few detected at early stage.
* Metastatic involvement of lymph nodes or distant sites is relatively uncommon at diagnosis in many cases (~10-12% nodal metastasis, ~7% distant metastasis).
* The prognosis and survival vary widely depending on stage and treatment, with 5-year overall survival rates reported around 46% overall, higher (up to 60-80%) for early-stage disease and much lower for advanced tumors.

**GENOMIC DATA**

* The most frequently mutated gene in temporal bone squamous cell carcinoma is TP53, a tumor suppressor gene commonly altered in many squamous cell carcinomas (SCCs) across head and neck cancers .
* Other important mutated genes and molecular pathways involved include:
  + EGFR (epidermal growth factor receptor) and related tyrosine kinase receptor genes affecting pathways like PI3K, Notch, and FAT1 .
  + Mutations in CTNNB1 (β-catenin 1) and VEGFR-2 (vascular endothelial growth factor receptor 2) have been identified and linked to tumor progression and angiogenesis. These mutations have also led to successful targeted therapies in some cases .
  + Additional genes reported include DNMT1 (DNA methyltransferase 1), which is related to epigenetic regulation, and oncogenes like ZDHHC11B and TARP, possibly associated with tumor proliferation and invasion .
  + Somatic mutations in other genes such as NOTCH1, FREM3, GLIS3, COL15A1, FSCB, and INF2 have also been reported in cell line studies derived from temporal bone SCC, although their specific roles are not yet fully understood .
* Chromosomal amplifications have been observed in regions 3q, 5p, and 8q, which are common in squamous cell carcinomas of the head and neck .
* Molecular markers like overexpression of p53 and EGFR proteins have been correlated with disease progression and prognosis, including risk of lymph node metastasis .
* Novel transcriptome analyses suggest potential prognostic biomarkers such as the long non-coding RNA MMP3-1, although further validation is needed

**PREDEFINED Q & A SETS**

Q1: What is temporal bone cancer?  
A1: Temporal bone cancer is a rare malignant tumor that arises from the temporal bone, which is the part of the skull surrounding the ear canal. It can originate from the skin of the ear, ear canal, middle ear, or nearby glands like the parotid salivary gland.

Q2: What are the common symptoms of temporal bone cancer?  
A2: Early symptoms often include ear pain, hearing loss, and ear discharge. More advanced signs can be bleeding from the ear, facial weakness or paralysis, jaw pain, inability to open the mouth, tinnitus (ringing in the ear), and neurological symptoms if the cancer spreads.

Q3: How is temporal bone cancer diagnosed?  
A3: Diagnosis involves physical examination by an ENT specialist, biopsy of suspicious lesions for cancer confirmation, and imaging tests like CT and MRI scans to evaluate the tumor’s location, size, bone involvement, and spread to adjacent structures and lymph nodes.

Q4: What causes temporal bone cancer?  
A4: Major risk factors include chronic ear infections (like cholesteatoma), prolonged sun exposure leading to skin cancers on the ear, previous radiation therapy to the head and neck, viral infections (such as HPV), and tumors spreading from nearby glands like the parotid.

Q5: How common is temporal bone cancer?  
A5: It is very rare, with an incidence of about 1 to 6 cases per million people annually, representing less than 1% of head and neck cancers. It affects mainly middle-aged to older adults with no strong gender bias.

Q6: What are the treatment options for temporal bone cancer?  
A6: Treatment usually involves surgical removal of the tumor and affected structures, possibly including the ear canal, eardrum, parotid gland, and lymph nodes. Surgery is often followed by radiation therapy. Chemotherapy may be added in some cases.

Q7: What is the prognosis for someone with temporal bone cancer?  
A7: Prognosis depends on the stage at diagnosis. Early-stage tumors confined to the ear canal have better survival rates, while advanced tumors involving bone and surrounding tissues have a more guarded prognosis. Five-year survival rates vary widely but are improved with complete surgical resection.

Q8: Are there any side effects or risks from treatment?  
A8: Surgery and radiation can affect hearing, facial nerve function, and appearance. Risks include hearing loss, facial paralysis, wound complications, and the general risks associated with surgery and radiation.

Q9: Can temporal bone cancer spread to other parts of the body?  
A9: Yes, it can spread locally to nearby tissues and lymph nodes and, less commonly, metastasize to distant organs.

Q10: When should I see a doctor about symptoms related to temporal bone cancer?  
A10: See a doctor promptly if you have persistent ear pain, unexplained hearing loss, ear discharge or bleeding, facial weakness, or trouble opening your mouth, especially if these symptoms do not improve with standard ear treatments.

**DOCTOR-PATIENT CONVERSATIONS**

Doctor: Hello, thank you for coming in today. I wanted to discuss the results of your recent tests. The biopsy and imaging show that you have a type of cancer in the temporal bone, which is the bone around your ear.

Patient: Oh, I see. What exactly is temporal bone cancer?

Doctor: Temporal bone cancer is a rare malignancy that starts in the tissues of the temporal bone, which includes the ear canal and surrounding areas. It can affect hearing and nearby nerves, so early diagnosis and treatment are important.

Patient: What symptoms should I expect or watch out for?

Doctor: Common symptoms include ear pain, hearing loss, sometimes discharge from the ear, or even swelling around the ear. If the cancer affects nearby nerves, you might experience facial weakness or jaw pain. It’s good to report any new or worsening symptoms so we can manage them promptly.

Patient: How did this happen? What causes it?

Doctor: Several factors can contribute, such as chronic ear infections, prolonged sun exposure to the ear skin, and prior radiation treatments. Sometimes, viral infections like HPV may play a role. But it’s important to focus on treatment now.

Patient: What are the treatment options?

Doctor: Treatment usually involves surgery to remove the tumor and some surrounding tissues to ensure all cancer cells are cleared. This can include removing parts of the ear canal or nearby structures if needed. After surgery, radiation therapy is commonly used to target any remaining cancer cells. Chemotherapy is considered in some cases, depending on the tumor’s extent.

Patient: What about the risks from surgery? And the recovery?

Doctor: Surgery in this area can affect your hearing or facial nerve function, so we take great care to minimize risks. After treatment, you’ll need some time to heal, and we will monitor your recovery closely. Rehabilitation and supportive therapies can help with any side effects.

Patient: What is my outlook? Can this be cured?

Doctor: Prognosis depends on how early we diagnose and treat it. If caught early and completely removed, the chances of control and cure are higher. If the tumor is more advanced, treatment aims to control the cancer and maintain quality of life. We will do our best to tailor treatment to your situation.

Patient: That’s reassuring. What should I do next?

Doctor: We’ll schedule the surgery soon, and I will explain the details and answer any questions you have. Meanwhile, please note any symptoms and keep in touch if you notice changes. We’ll also involve specialists like radiation oncologists and therapists to support you.

Patient: Thank you, doctor, for explaining everything clearly.

Doctor: You’re very welcome. We are here to support you every step of the way.

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**Thrush**

ALTERNATIVE NAMES

## Oral Candidiasis

**DEFINITION / DESCRIPTION**

Thrush is a fungal (yeast) infection that can grow in your mouth, throat and other parts of your body. With oral thrush (oral candidiasis), you may develop white, raised, cottage cheese-like lesions (spots) on your tongue and cheeks. Thrush can quickly become irritated and cause mouth pain and redness.

Thrush happens when there’s an overgrowth of Candida, a type of fungus. Another name for thrush in your mouth or throat is oropharyngeal candidiasis.

Healthcare providers treat thrush with antifungal medication. If your immune system is healthy, thrush is a minor problem that goes away a couple of weeks after you start treatment.

#### **Who can get thrush?**

While thrush can affect anyone, some people are more at risk, including:

* Babies under 1 month old.
* Toddlers.
* Adults aged 65 or over.
* People with weakened immune systems (where symptoms are harder to control).

**CAUSES**

Most people have small amounts of the Candida fungus in their mouth, digestive tract and skin. When illnesses, stress or medications disturb this balance, the fungus grows out of control and causes thrush.

Medications that can make yeast flourish and cause infection include:

* Corticosteroids.
* Antibiotics.
* Birth control pills.

#### **Is thrush contagious?**

Thrush can be contagious to those at risk (like people with weakened immune systems or who take certain medications). In people with healthy immune systems, it’s unusual to pass thrush through kissing or other close contact. In most cases, thrush isn’t particularly contagious (meaning, it doesn’t spread from person to person), but it is transmittable (meaning, you can catch it in other ways).

If you’re worried about getting thrush from another person who has it, avoid coming into contact with their saliva (spit). It’s smart to wash your hands as often as possible if you’re near someone who has thrush.

#### **What are the risk factors for thrush?**

Candida infection is more likely to develop in babies and people with:

* Diabetes.
* Anemia.
* HIV/AIDS (thrush in your esophagus — or swallowing tube — is common in this group).
* Cancer.
* Dry mouth (xerostomia).
* Pregnancy (due to the hormonal changes that occur).
* Smoking.
* Ill-fitting dentures.

**SIGNS / SYMPTOMS**

Thrush usually develops suddenly. A common sign is the presence of creamy white, slightly raised lesions in your mouth — usually on your tongue or inner cheeks. You may also have lesions on the roof of your mouth, gums, tonsils or back of your throat.

Other symptoms may include:

* Redness and soreness inside and at the corners of your mouth.
* Loss of sense of taste (ageusia).
* Cottony feeling in your mouth.

The lesions can hurt and may bleed a little when you scrape them or brush your teeth. In severe cases, the lesions can spread into your esophagus and cause:

* Pain or difficulty swallowing.
* A feeling that food gets stuck in your throat or mid-chest area.
* Fever, if the infection spreads beyond your esophagus.

Thrush can spread to other parts of your body, including your lungs, liver and skin. This happens more often in people with cancer, HIV or other conditions that weaken the immune system.

**DIAGNOSIS METHODS**

A healthcare provider can usually tell right away if you have thrush by looking for the distinctive white lesions on your mouth, tongue or cheeks. Lightly brushing the lesions away reveals a reddened, tender area that may bleed slightly. A microscopic exam of tissue from a lesion can confirm whether or not you have thrush.

If thrush extends into your esophagus, your healthcare provider might:

* Take a throat culture (swab the back of your throat with sterile cotton and study the microorganisms under a microscope).
* Perform an endoscopy of your esophagus, stomach and small intestine (examine the lining of these areas with a lighted camera mounted on the tip of a flexible tube).
* Take X-rays of your esophagus.

**TREATMENT OPTIONS**

The typical treatment for thrush is antifungal medications:

#### **Antifungal medications**

Healthcare providers usually prescribe antifungals (like nystatin) to treat thrush. These medicines are available in tablets, lozenges or liquids that are “swished” around in your mouth before swallowing. Usually, you need to take these medications for 10 to 14 days. Your healthcare provider will recommend specific treatment based on your age and the cause of the infection.

Kids and adults with healthy immune systems typically respond well to antifungal treatment. But thrush symptoms may be more severe and harder to treat in those with weakened immune systems.

### **How soon after treatment will I feel better?**

Antifungals can clear up thrush in one to two weeks. You may need to continue the medication for a few more days to kill any fungus that’s left behind.

**PREVENTION TIPS**

You can do these things to reduce your risk for thrush:

* Practice good oral hygiene. Brush your teeth at least twice a day and floss at least once a day.
* Avoid certain mouthwashes or sprays. Some of these products can destroy the normal balance of microorganisms in your mouth. Talk to your dentist or doctor about which ones are safe to use.
* See your dentist regularly. This is especially important if you have diabetes or wear dentures.
* Limit the amount of sugar and yeast-containing foods you eat. Foods such as bread, beer and wine encourage Candida growth.
* Avoid smoking and other tobacco use. Ask your healthcare provider about ways to help you quit smoking.

**OUTLOOK / PROGNOSIS**

With treatment, thrush usually goes away within one to two weeks. But if your symptoms linger or get worse, let your healthcare provider know.

**POSSIBLE COMPLICATIONS**

Thrush rarely causes complications in people with healthy immune systems. But if you have a weakened immune system, Candida can enter your bloodstream and spread to other areas of your body, such as your eyes, brain or heart. This type of infection is serious and may lead to septic shock, a life-threatening condition

**WHEN TO SEE A DOCTOR / RED FLAG**

If you develop signs or symptoms of thrush — such as soreness, bleeding or raised white areas inside your mouth — schedule an appointment with a healthcare provider.

If you’ve already taken antifungals for thrush but your symptoms return, call your provider right away. It could indicate a more serious infection

**DIFFERENTIAL DIAGNOSIS**

* Aphthous Ulcers
* Blastomycosis
* Histiocytosis
* Lymphohistiocytosis (Hemophagocytic Lymphohistiocytosis)
* Pediatric Candidiasis
* Pediatric Cytomegalovirus Infection
* Pediatric Diphtheria
* Pediatric Echovirus
* Pediatric Enteroviral Infections
* Pediatric Esophagitis
* Pediatric Herpes Simplex Virus Infection
* Pediatric HIV Infection
* Pediatric Pharyngitis
* Pediatric Syphilis

**GENOMIC DATA**

* *Candida albicans* has a diploid genome of about 14 megabases (Mb) distributed across 8 chromosomes. It contains roughly 6,100 genes.
* The *C. albicans* genome was first fully sequenced and published in 2004, revealing a highly heterozygous genome with two copies of each chromosome exhibiting genetic variation.
* Genomic studies have identified mutations, gene variations, and chromosomal features that contribute to its ability to survive both as a harmless commensal organism in humans and as an opportunistic pathogen causing infections like thrush.
* Genomic resources such as the Candida Genome Database (CGD) provide continuously updated, chromosome-level assemblies and annotations of the *C. albicans* genome to support ongoing research.
* Specific clinical isolates of *C. albicans* from oral candidiasis patients have been sequenced to study strain variability contributing to pathogenicity and antifungal resistance.
* Unique features include the peculiar translation of the codon CUG as serine instead of leucine, which is rare among fungi, and extensive gene families related to adhesion, biofilm formation, and antifungal resistance, important for thrush pathogenesis.

**TREATMENT DRUG INFORMATION AND THEIR SIDE EFFECTS**

## Common Antifungal Drugs for Thrush

1. Nystatin (Topical)
   * Form: Oral suspension or lozenges
   * Use: Mild to moderate oral thrush
   * How to use: Swish and swallow or dissolve lozenges in the mouth
   * Side effects:
     + Nausea or upset stomach
     + Mouth irritation or burning
     + Rare allergic reactions
   * Advantage: Minimal systemic absorption, generally well tolerated
2. Clotrimazole (Topical)
   * Form: Lozenges (troches), oral gel
   * Use: Mild to moderate thrush
   * Side effects:
     + Taste disturbances
     + Mouth irritation, burning, or soreness
     + Rare allergic reactions
   * Note: Clotrimazole cream can affect latex contraception; mouth forms usually do not
3. Miconazole (Topical)
   * Form: Buccal tablets (Oravig), oral gel
   * Use: Mild to moderate thrush
   * Side effects:
     + Oral irritation or burning
     + Unpleasant taste
     + Potential drug interactions if swallowed
4. Fluconazole (Systemic)
   * Form: Oral tablets or suspension; also IV form for severe cases
   * Use: Moderate to severe thrush, or cases not responding to topical treatment; esophageal candidiasis
   * Dosage: Typically 100-200 mg once daily for 7-14 days
   * Side effects:
     + Headache
     + Nausea, abdominal pain
     + Diarrhea
     + Rare liver toxicity (monitor liver function)
     + Skin rash (possible rare severe reactions)
   * Advantages: Effective systemic therapy, good bioavailability
5. Itraconazole (Systemic)
   * Use: Alternative systemic agent, especially for refractory cases
   * Side effects:
     + Gastrointestinal upset
     + Headache
     + Liver enzyme elevation
     + Potential for significant drug interactions
6. Other agents (for severe or refractory cases):
   * Amphotericin B (IV)
   * Echinocandins (caspofungin, micafungin) – IV use in invasive candidiasis
   * Side effects of these are more serious and usually reserved for hospitalized patients

**EPIDEMIOLOGY**

### United States statistics

As many as 37% of newborns may develop thrush during the first months of life.

### International statistics

Thrush is universal and is more common in poorly nourished populations.

### Sex- and age-related demographics

Thrush occurs equally in males and females.

Thrush is rare during the first week of life. Incidence peaks around the fourth week of life; thrush is uncommon in infants older than 6-9 months. Thrush can occur, however, at any age in predisposed patients.

**PREDEFINED Q & A SETS**

### **Are there any home remedies for oral thrush?**

You’ll need antifungal medication to clear up thrush. But you might try some of these home remedies to ease your symptoms:

* Swish with warm saltwater.
* Take probiotics.
* Eat yogurt that contains healthy bacteria.

Q1: What is thrush?  
A1: Thrush is an infection of the mouth caused by an overgrowth of a fungus called *Candida*, most commonly *Candida albicans*. It appears as white patches or plaques inside the mouth and on the tongue.

Q2: Who can get thrush?  
A2: Thrush can affect anyone but is more common in infants, older adults, people with weakened immune systems (such as those with HIV or cancer), people using inhaled corticosteroids, or after antibiotic use.

Q3: What are the symptoms of thrush?  
A3: Symptoms include white, creamy patches in the mouth that can be scraped off, redness or soreness underneath, difficulty swallowing, a burning sensation, and sometimes loss of taste.

Q4: How is thrush diagnosed?  
A4: Thrush is usually diagnosed based on clinical appearance during an oral exam. Sometimes, a sample of the white patches may be examined under a microscope to confirm the presence of *Candida*.

Q5: What causes thrush?  
A5: Thrush develops when the balance of the normal flora in the mouth is disrupted, allowing *Candida* to grow excessively. Causes include immune system weakness, antibiotic or steroid use, diabetes, dry mouth, or denture use.

Q6: How is thrush treated?  
A6: Treatment usually involves antifungal medications. For mild cases, topical antifungals like nystatin or clotrimazole are used. More severe or persistent infections may require systemic antifungals such as fluconazole.

Q7: Can thrush be prevented?  
A7: Prevention includes good oral hygiene, controlling underlying conditions like diabetes, rinsing the mouth after inhaled steroid use, limiting antibiotic use when possible, and maintaining proper denture care.

Q8: Are there any complications from thrush?  
A8: Thrush is generally not serious but can cause discomfort. In people with weakened immunity, it may spread to other parts of the body like the esophagus and cause more serious infections.

Q9: When should I see a doctor about thrush?  
A9: If you have white mouth patches that don't go away or you have pain, difficulty swallowing, or recurrent thrush, you should see a healthcare provider for evaluation.

Q10: Is thrush contagious?  
A10: Thrush itself is not usually considered contagious, but *Candida* can be transmitted. However, infections generally only occur if the person’s immune defenses are weakened or the local environment favors fungal growth.

**DOCTOR-PATIENT CONVERSATIONS**

Doctor: Hello! What brings you in today?

Patient: Hi, doctor. I've been noticing white patches inside my mouth and it feels sore and uncomfortable. It sometimes burns when I eat or drink.

Doctor: That sounds like it might be thrush, which is a common fungal infection caused by a yeast called Candida. Have you had any recent illnesses, antibiotic use, or are you using inhalers with steroids?

Patient: Yes, I recently finished a course of antibiotics for a chest infection, and I do use an inhaler for asthma.

Doctor: Both of those can increase the risk of thrush by disrupting the normal balance of microbes in your mouth. The white patches are typical and can sometimes be scraped off, leaving a red, sore area underneath. Thrush can cause soreness, burning, and sometimes affect your taste.

Patient: Is it serious? How do you treat it?

Doctor: Thrush is usually not serious and can be treated effectively. Mild cases often respond well to antifungal medications like nystatin or clotrimazole, which you apply inside your mouth. It’s important to continue good oral hygiene and rinse your mouth after using your inhaler. For more severe or persistent cases, oral antifungal tablets like fluconazole may be needed.

Patient: Are there any side effects from the treatment?

Doctor: Topical antifungals might cause minor mouth irritation or an unpleasant taste, but they are generally safe. Oral antifungals can sometimes cause nausea or, rarely, liver issues, so we monitor patients closely. Most people tolerate treatment well.

Patient: How long will it take to get better?

Doctor: With treatment, symptoms usually improve within a week or two. Make sure to complete the full course to prevent recurrence. If symptoms persist or worsen, come back for a follow-up.

Patient: Can thrush come back?

Doctor: Yes, especially if your risk factors like inhaler use or antibiotics continue. Good oral care and managing underlying conditions help reduce this risk.

Patient: Thank you, doctor. That makes me feel more at ease.

Doctor: You’re welcome! If you have any questions or new symptoms, please don’t hesitate to reach out.

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**Tongue cancer**

ALTERNATIVE NAMES

**DEFINITION / DESCRIPTION**

Tongue cancer occurs when cells on your tongue start to grow and divide uncontrollably. Your tongue starts at your hyoid bone (located where your chin meets your neck) and ends at the floor of your mouth.

When cancer affects the front part of your tongue (the part you can see), healthcare providers call it oral cancer or oral tongue cancer. If you develop cancer on the back portion of your tongue (the part you can’t see), providers call it oropharyngeal cancer or base of tongue cancer.

#### **Types of tongue cancer**

Several types of cancer can affect your tongue, but the most common is squamous cell carcinoma (SCC). Squamous cell carcinoma starts in the squamous cells in the outer layer of your skin. Typically, SCC affects the parts of your body most often exposed to the sun. But it can also affect mucous membranes like the inside of your mouth.

#### **How common is tongue cancer?**

Tongue cancer is rare overall, making up less than 1% of new diagnoses in the United States. But it’s one of the most common types of head and neck cancers.

Tongue cancer is twice as common in males. It’s also more common in people age 40 and over.

**CAUSES**

Experts know that tongue cancer occurs when abnormal cells grow out of control. But they’re still learning why some people get tongue cancer and others don’t — and why these cells grow out of control in the first place. Many researchers believe that DNA mutations (changes) that affect your tongue can cause tongue cancer.

#### **Tongue cancer risk factors**

Experts have identified several risk factors for tongue cancer. A risk factor is something that increases your chances of getting a certain condition. The two most significant risk factors include heavy smoking and high alcohol consumption (alcohol use disorder). Your risk goes up drastically if both pertain to you.

Other tongue cancer risk factors include:

* HPV (human papillomavirus). A history of HPV is a leading cause of base of tongue cancer. Less commonly, it can cause oral tongue cancer.
* Chewing tobacco.
* Betel nut.
* Family history of oral or oropharyngeal cancers.
* Personal history of cancer (especially squamous cell carcinoma).

**SIGNS / SYMPTOMS**

First, it’s important to note that signs of tongue cancer aren’t always apparent. What cancer looks like and whether you can “see” it depends on the location of the tumor and how far the disease has progressed.

The most common visual tongue cancer symptoms include:

* Red (erythroplakia) or white (leukoplakia) patches on your tongue.
* A lump on the side of your tongue that bleeds easily.
* A red or grayish ulcer on your tongue that doesn’t go away.

Additional tongue cancer symptoms may include:

* Thickening of the skin in your mouth.
* A sore throat that doesn’t go away (chronic pharyngitis).
* Neck pain.
* Ear pain.
* Feeling like there’s something stuck in your throat.
* Numbness in your tongue or other areas of your mouth.
* A burning sensation on your tongue.
* Difficulty or pain chewing.
* Difficulty or pain swallowing.
* Hoarseness.
* Jaw swelling.
* Loose teeth.
* Sudden changes in the way your dentures fit.

**DIAGNOSIS METHODS**

Dentists are often the first to detect tongue cancer during routine exams or oral cancer screenings. Or your primary care physician (PCP) may notice signs during an exam for another condition.

To gather more information, your provider may:

* Shine a fluorescent light into your mouth to see if it reveals any abnormal tissue.
* Use an endoscope (a thin, flexible tube with a camera and light) to look at your mouth and throat.
* Check for swollen lymph nodes.

If a healthcare provider sees visual signs of tongue cancer, they’ll recommend a biopsy. During this procedure, they’ll take a small sample of affected tissue. They’ll send it to a lab, where a pathologist will examine the tissue and look for cancer cells.

Your provider may also order imaging tests like:

* CT (computed tomography) scans.
* MRI (magnetic resonance imaging) scans.
* PET (positron emission tomography) scans.

**TREATMENT OPTIONS**

Tongue cancer treatments depend on a few factors, including the size and location of the tumor and how far the cancer cells have spread. Tongue cancer treatments include:

* Tongue cancer surgery. This usually includes glossectomy (partial or total tongue removal) and, in some cases, neck dissection (lymph node removal) to reduce the risk of metastasis (when cancer spreads from the primary tumor site to another area of your body).
* Radiation therapy. Healthcare providers might use radiation therapy before surgery to shrink a tumor or, more commonly, after surgery to kill any remaining cancer cells. In some cases, oncologists use it as a stand-alone treatment, especially if cancer has already spread to other areas.
* Drug therapies. These may include chemotherapy, immunotherapy and targeted therapy.

**PREVENTION TIPS**

You can’t prevent tongue cancer altogether. But there are things you can do to significantly reduce your risk:

* Avoid tobacco and alcohol. The best way to prevent tongue cancer is to avoid tobacco and alcohol, as they’re the two most significant risk factors. But stopping after many years of use can still greatly lower your risk for all types of oral and oropharyngeal cancers.
* Get the HPV vaccine. HPV is a leading cause of oropharyngeal cancers. It’s common and it rarely causes symptoms. The HPV vaccine protects against tongue cancer and other cancers linked to HPV, including cancers of the cervix, penis, vagina, vulva and anus.
* Eat plenty of fruits, vegetables and whole grains. Try to get your nutrition from fresh, unprocessed foods and limit your intake of red meat, sugary drinks and highly processed foods. Ask your healthcare provider about a nutrition plan that works for you.
* Visit your dentist regularly. Your dentist can do routine oral cancer screenings during these appointments. In addition, good oral health promotes whole-body health and well-being.

**OUTLOOK / PROGNOSIS**

Tongue cancer can be curable with early diagnosis and treatment. That’s why it’s so important to tell your healthcare provider if you develop any new lumps, spots on your tongue or sores that don’t go away.

Speech therapy and reconstructive surgery can help you redevelop language skills and swallowing function after tongue cancer surgery.

#### **Tongue cancer survival rate**

The survival rate for tongue cancer depends on whether the cancer has spread to other areas.

Survival rates compare people who have a certain type and stage of cancer to those in the general population. For instance, if the five-year survival rate for a particular type of cancer is 93%, that means that 93% of people with that type and stage of cancer will still be alive five years after their diagnosis.

| **Tongue cancer stage** | **How far the cancer spread** | **Five-year survival rate** |
| --- | --- | --- |
| Localized | The cancer hasn’t spread beyond your tongue. | 84% |
| Regionalized | The cancer has spread to nearby lymph nodes or structures. | 70% |
| Distant | The cancer has spread to distant areas of your body. | 41% |

Keep in mind that survival rates are estimates. They can’t tell you how long you’ll live, or which treatments will work best in your situation. To learn more about survival rates and what they mean for you, talk to your healthcare provider.

**WHEN TO SEE A DOCTOR / RED FLAG**

Schedule an appointment with your healthcare provider any time you notice a new lump, bump, spot, ulcer or discoloration on your tongue, especially if symptoms linger for more than two weeks.

In addition, be sure to visit your dentist regularly for cleanings and checkups. For most people, that means every six months. But your dentist will tell you how often you should come in based on your specific needs. If there’s something suspicious going on, they’re likely to detect it during a routine appointment.

**DIFFERENTIAL DIAGNOSIS**

* Squamous cell carcinoma
* Leukoplakia
* Carcinoma in situ
* Sarcoma
* Rhabdomyosarcoma
* Lymphoma
* Rhabdomyoma
* Neurofibroma
* Pyogenic granuloma
* Papilloma
* Vascular or lymphatic malformation
* Lingual thyroid
* Dermoid cyst
* Epidermoid cyst

**Staging**

The American Joint Committee on Cancer uses the TNM system for staging tumors of the head and neck region by the site. It was recently updated to incorporate the prognostic impact of the presence of extranodal extension. Accordingly, clinically evident extranodal extension upstages the N-stage of a tumor to N3b.

T-stage is classified as follows:

* T0: No evidence of primary tumor
* Tis: Carcinoma in situ
* T1: Tumor 2cm or less with a depth of invasion (DOI) less than or equal to 5mm
* T2: Tumor 2cm or less with DOI more than 5mm OR tumor more than 2cm but less than or equal to 4cm
* T3: Tumor more than 2cm but less than or equal to 4cm with DOI more than 10mm OR tumor more than 4cm with DOI less than or equal to 10mm
* T4a: Tumor more than 4cm with DOI more than 10mm OR invades adjacent structures only (skin of the face, maxillary sinus, cortical bone of maxilla or mandible)
* T4b: Tumor invades masticator space, pterygoid plates, skull base, and/or encases the internal carotid artery.

N-stage or clinical nodal stage is classified as follows:

* N0: No regional nodes metastasis.
* N1: Metastasis in a single ipsilateral lymph node 3cm or less in the greatest dimension - negative extranodal extension (ENE)
* N2a: Metastasis in a single ipsilateral lymph node more than 3cm but not larger than 6cm in the greatest dimension, negative ENE
* N2b: Metastasis in multiple ipsilateral lymph nodes, none more than 6cm in greatest dimension, negative ENE
* N2c: Metastasis in bilateral or contralateral lymph node(s), none more than 6cm in greatest dimension, negative ENE
* N3a: Metastasis in any lymph node more than 6cm in the greatest dimension, negative ENE
* N3b: Metastasis in a lymph node with clinically overt positive ENE

M-stage is classified as follows:

* MX: Distant metastasis cannot be assessed
* M0: No distant metastasis
* M1: Distant metastasis

**POSSIBLE COMPLICATIONS**

Complications of surgery are multiple and include bleeding, infection, and orocutaneous fistula. When a free or pedicled flap is employed, complications include donor site infection, bleeding, and partial or complete flap loss or infection. Depending on the extent of resection and adequacy of reconstruction, there may be function-related complications such as impaired speech, dysphagia, and odynophagia. A gastrostomy is frequently needed at the time of surgery if speech and swallow rehabilitation are anticipated to be problematic or prolonged. A tracheostomy is also frequently employed if significant oral cavity and pharyngeal airway edema are anticipated in the immediate postoperative period.

Radiation therapy complications include oral/pharyngeal mucositis, skin damage, cellulitis, dysgeusia, xerostomia, fibrosis, and oral mucosal neuropathy. One of the most troublesome complications of radiation therapy in this region is mandibular osteoradionecrosis (ORN). This may require surgical intervention for curettage, and in severe cases, segmental mandibulectomy of the affected region and reconstruction. Although hyperbaric oxygen therapy is sometimes given to treat mandibular ORN, some surgeons argue this may increase the probability of malignancy recurrence or may catalyze the growth of microscopic persistent disease. No high-level evidence studies to date have conclusively demonstrated this correlation, but anecdotal evidence exists, and the decision remains clinician-dependent.

Common radiation side effects in the neck region include post-radiation fibrosis, frozen neck, neuropathy, and hypothyroidism, among others. Symptomatic esophageal stenosis due to post-radiation fibrosis is not uncommon and may manifest even years after radiation therapy.

One of the most feared complications of advanced-stage head and neck cancer is carotid blowout. This usually presents with life-threatening hemorrhage and used to be almost uniformly fatal. However, mortality today is close to 60% due to major surgical and endovascular advances. Nonetheless, periprocedural morbidity is high and may include re-bleeding and stroke

**EPIDEMIOLOGY**

Classically, tongue carcinoma is a condition of older males with a history of smoking and/or drinking alcohol. It has a slight male predominance, and the estimated frequency of disease varies widely with geographic location. In the past, there had been a steady decrease in the incidence of the disease, perhaps attributable to a worldwide overall decrease in smoking.

However, studies suggest an alarming increasing incidence of both oral and base of the tongue squamous cell carcinoma during the last decades, specifically in women and younger patients without the traditional risk factors of alcohol or tobacco use.This is, in part, believed to be related to the dramatic spike in HPV-associated oropharyngeal squamous cell carcinoma.Additional genetic etiologic factors that may help explain this changing demographic profile of the disease are under investigation.

**PREDEFINED Q & A SETS**

PLS EXPLAIN THE PATHOLOGY REPORT?

1. Explanation of Your Pathology Report:

Your pathology report is the detailed examination of the tissue sample taken from your tongue lesion. It confirms the diagnosis of cancer and provides important information that helps guide treatment. Key aspects usually included are:

* Diagnosis: Usually, tongue cancer is a squamous cell carcinoma (SCC), meaning cancer arises from the mucosal lining cells.
* Tumor Grade: This describes how much the cancer cells look like normal cells. Grades can be well, moderately, or poorly differentiated. Well-differentiated tumors tend to grow slower, while poorly differentiated are more aggressive.
* Depth of Invasion (DOI): This measures how deeply the tumor penetrates the tongue tissue. It influences staging and prognosis. For example, tumors deeper than 5 mm may represent a higher stage.
* Pattern of Invasion: This shows how the cancer cells spread at the tumor edges, either in cohesive large groups or as dispersed small groups/single cells—non-cohesive patterns usually indicate more aggressive behavior.
* Margins: Whether the cancer was completely removed with a clear edge (margin) or if cancer cells are close to or at the margin, which may necessitate further treatment.
* Lymphovascular Invasion: Whether cancer cells are found in blood vessels or lymphatic channels, possibly raising recurrence risk.
* Perineural Invasion: Cancer involving nerves indicates a more aggressive tumor and might affect treatment decisions.
* Lymph Node Status: If lymph nodes are involved, this affects staging and treatment.

You can request a copy of your pathology report directly from your healthcare provider or hospital. It is your right to have a copy for your records or to share with other doctors.

2. Will You Need More Tests?

Additional tests may be necessary to:

* Stage the cancer fully: imaging such as CT scan, MRI, PET scan are commonly used to check the tumor size, involvement of surrounding tissues, and lymph node or distant spread.
* Evaluate your overall health before treatment: blood tests, dental evaluation, and anesthesia clearance if surgery is planned.
* Sometimes, further biopsies or sentinel lymph node biopsy may be needed.

3. Treatment Options for Tongue Cancer:

Treatment depends on the tumor size, location, depth, lymph node involvement, your overall health, and preferences. Common options include:

* Surgery: Removal of the tumor with a margin of healthy tissue. May include partial glossectomy (part of tongue removal) and neck dissection if lymph nodes are involved.
* *Benefits:* Can remove cancer completely if detected early  
  *Risks:* Impact on speech, swallowing, taste; surgical complications, need for reconstruction
* Radiation Therapy: High-energy X-rays target cancer cells. May be primary treatment or after surgery to reduce recurrence.
* *Benefits:* Preserves tongue structure if surgery is not feasible  
  *Risks:* Mucositis, dry mouth, taste changes, damage to nearby tissue
* Chemotherapy: Often used with radiation (chemoradiation) in advanced cases or if surgery isn’t possible.
* *Benefits:* Enhances radiation effects, treats microscopic disease  
  *Risks:* Side effects like nausea, fatigue, immune suppression
* Targeted Therapy/Immunotherapy: For certain advanced or resistant cancers, drugs targeting specific molecules or immune checkpoints may be options.

Your multidisciplinary team will tailor treatment to your circumstances.

4. Benefits and Risks Summary:

| **Treatment** | **Benefits** | **Risks/Side Effects** |
| --- | --- | --- |
| Surgery | Potential cure, direct tumor removal | Speech/swallowing difficulties, infection, bleeding |
| Radiation | Non-invasive, preserves anatomy | Mucositis, dry mouth, fibrosis |
| Chemotherapy | Treats microscopic disease | Systemic side effects (nausea, fatigue, infection) |
| Targeted/Immuno | New options for resistant tumors | Variable, immune-related side effects |

5. Is There a Recommended Treatment?

The best option depends on your individual case. Generally:

* Early-stage (small, localized tumors) are often treated with surgery alone or surgery plus radiation.
* Advanced tumors may require combined chemoradiation or surgery plus adjuvant therapies.
* Your medical team considers tumor biology, functional outcomes, and your preferences.

6. What Would I Recommend to a Loved One in the Same Situation?

I would recommend:

* Seeking care at a specialized center with a multidisciplinary head and neck cancer team.
* Thoroughly understanding the diagnosis and treatment options.
* Choosing treatment that balances cancer control with quality of life.
* Open communication with the care team and family support.

7. Should You Get a Second Opinion?

Getting a second opinion from a head and neck cancer specialist is often helpful and encouraged, especially for complex cases. It can:

* Confirm your diagnosis and treatment plan
* Offer access to specialized expertise or clinical trials

Costs and Insurance:

* Many insurance plans cover second opinions—check with your insurer.
* Costs vary by healthcare system and provider; some hospitals offer free or low-cost second opinions.
* Your current doctor or hospital typically can facilitate the referral.

**DOCTOR-PATIENT CONVERSATIONS**

Doctor: Hello, thank you for coming in today. I want to talk with you about the biopsy results from your tongue lesion. The report shows that you have squamous cell carcinoma of the tongue.

Patient: I’m shocked. What exactly is that?

Doctor: Squamous cell carcinoma is a type of cancer that starts in the flat cells lining the surface of your tongue. It can cause a persistent sore or lump that doesn’t heal, and can affect your ability to speak and swallow if not treated.

Patient: How did I get this? What causes tongue cancer?

Doctor: Several factors increase the risk, including tobacco and alcohol use, chronic irritation like from a rough tooth, viral infections like HPV, and sometimes genetic susceptibility. But it’s important now to focus on managing the cancer effectively.

Patient: What are the treatment options for this cancer?

Doctor: The main treatments are surgery to remove the tumor, radiation therapy, chemotherapy, or a combination depending on the stage and specifics of your cancer. Surgery usually involves removing part of your tongue and possibly some lymph nodes in your neck to prevent spread.

Patient: What are the risks and benefits of surgery?

Doctor: Surgery can potentially cure the cancer if it’s caught early and completely removed. Risks include changes in speech, swallowing difficulties, and healing complications. But we work closely with specialists like speech therapists to help recovery.

Patient: Will I need radiation or chemotherapy too?

Doctor: That depends on how advanced the tumor is and if it has spread. Sometimes radiation is used after surgery to lower the risk of recurrence. Chemotherapy is generally reserved for more advanced cases or if surgery isn’t possible.

Patient: What is my outlook? Can this be cured?

Doctor: Early-stage tongue cancers have a good chance of cure with treatment. The prognosis depends on factors like tumor size, depth, lymph node involvement, and your overall health. We will tailor your treatment to maximize the chance of cure and maintain quality of life.

Patient: Should I get a second opinion?

Doctor: It’s perfectly reasonable and often helpful to get a second opinion, especially at a specialized center with experience in head and neck cancers. Many patients find this gives them confidence in their treatment plan.

Patient: Can I have copies of my pathology and other test results?

Doctor: Absolutely. It’s your right to have copies of all your medical records. I can help you obtain them to keep or share with other doctors if you like.

Patient: Thank you, I appreciate you explaining this clearly.

Doctor: You’re very welcome. We’re here to support you throughout treatment. Please don’t hesitate to ask any questions as we move forward.

**GENOMIC DATA**

* TP53 mutations are the most common genetic alteration found in tongue cancer, occurring in approximately 40-80% of cases in different studies, especially affecting the DNA-binding domain between exons 5 and 8. These mutations often result in loss of normal p53 tumor suppressor function, leading to unchecked cell growth and poor prognosis in some cases.
* Other frequently mutated genes include CDKN2A (p16), NOTCH1, CASP8, FAT1, and TERT promoter (TERTp) mutations. These genes generally impact cell cycle regulation, apoptosis, and cellular signaling pathways critical for tumor growth.
* Novel mutations identified in familial and sporadic oral tongue cancers include VAV2 and IQGAP1, which influence tumorigenesis-associated pathways, indicating possible hereditary predispositions in some cases.
* Mutations in DNA damage repair genes such as ATR, ATM, CHEK1, and CHEK2 have also been reported, with some novel mutations proposed as potential biomarkers. Frameshift deletions, nonsynonymous single nucleotide variants (SNVs), and stop gain mutations contribute to genetic instability in these tumors.
* Mutations in oncogenes and tumor suppressors like MET, PIK3CA, STK11, and less commonly BRAF have been found but with varying prevalence based on population cohorts.
* The mutational burden may vary with age of onset, with early onset oral tongue cancers showing fewer somatic mutations compared to typical onset cases

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**Tracheal stenosis**

ALTERNATIVE NAMES

**DEFINITION / DESCRIPTION**

Tracheal stenosis involves narrowing of your trachea (windpipe) that makes it harder to breathe. Your trachea is a tube made of cartilage and soft tissue. Air travels through your trachea on the way from your nose and mouth to your lungs. When you have tracheal stenosis, inflammation, injury or scar tissue in your trachea makes it harder for air to flow through.

“Stenosis” is the medical term for atypical narrowing in a body passage. It can develop in different parts of your throat. For example, a related condition called laryngotracheal stenosis involves narrowing in both your trachea and larynx (voice box). Subglottic stenosis is narrowing in the area above your trachea and below your vocal cords.

Regardless of the part of your throat that’s affected, you should contact a healthcare provider if you’re having difficulty breathing.

#### **Types**

There are two types of tracheal stenosis:

* Acquired tracheal stenosis is acquired (developed) during your lifetime because of an injury or illness. The most common type, it affects both adults and children.
* Congenital tracheal stenosis (CTS) is present at birth. A rare and potentially fatal condition, it affects 1 out of every 64,500 babies. Most are diagnosed at birth or within the first few months of being born.

Tracheal stenosis is life-threatening in infants. Seek emergency care if your newborn is showing signs of tracheal stenosis, like struggling for air.

That said, both forms of tracheal stenosis make it harder to breathe, which can affect your quality of life. That’s why seeing a healthcare provider is so important if you or your child has this condition.

## **Symptoms and Causes**

### **What are the symptoms of tracheal stenosis?**

Many tracheal stenosis symptoms are the same for children and adults. Common symptoms include:

* Difficulty breathing after everyday activities like climbing stairs or walking
* Stridor (sounds like a wheeze or whistle when you breathe both in and out)
* Persistent cough
* Difficulty coughing up phlegm
* Frequent colds, bouts of pneumonia or other respiratory infections
* Persistent asthma that isn’t better after treatment
* Chest congestion
* Gaps in breathing (apnea) and sleep apnea

Children are more likely to have additional symptoms:

* Infants might have difficulty breastfeeding or bottle feeding. They might also seem unusually tired after feeding.
* Older children might choke or have difficulty breathing while they eat.
* Older children’s skin around their noses and their gums might appear blue (cyanosis).

### **What causes tracheal stenosis?**

With congenital tracheal stenosis (CTS), the cartilage that makes up an infant’s trachea doesn’t form correctly, causing their windpipe to be too narrow.

Long-term intubation is the most common cause of acquired tracheal stenosis. Intubation is a lifesaving treatment that involves inserting a tube into your trachea so you can breathe. Sometimes, intubation causes damage that leads to stenosis. Children may develop tracheal stenosis if they were born with premature lungs and their provider used breathing tubes to help them get air.

You might also develop tracheal stenosis if you:

* Have an autoimmune disorder, like granulomatosis with polyangiitis (GPA)
* Have an inflammatory condition, like pulmonary sarcoidosis
* Have an infectious disease, like tuberculosis, or other bacterial and viral infections that affect your respiratory system
* Have a benign or malignant tumor pressing on your trachea
* Inhaled a substance that damaged your trachea, including chemicals or poisonous gases
* Had radiation therapy directed toward your neck or chest
* Had a tracheostomy (emergency surgery that creates a hole in your trachea so that you can breathe)

## **Diagnosis and Tests**

Otolaryngologists (ENTs) use several tests to diagnose tracheal stenosis and decide how to treat it. Tests may include:

* Endoscopic procedures. Bronchoscopy is the primary procedure for diagnosing tracheal stenosis. Your healthcare provider may also perform a laryngoscopy.
* Imaging procedures. A computed tomography (CT) scan of your chest and neck is the most common imaging procedure that shows tracheal stenosis. Sometimes, healthcare providers recommend magnetic resonance imaging (MRI) to help plan treatment.
* Pulmonary function test. Providers will ask you to complete several breathing tests. This helps them check things like how your trachea size and lung function affect your breathing.

You may need other tests to check for what’s causing your condition. Tests may include a blood test to check for inflammation or infection or a biopsy to see if unusual growths in your airway are cancerous.

## **Management and Treatment**

Surgeries and procedures that widen your trachea are the most common treatments for tracheal stenosis. Treatment options include:

* Tracheal dilation. Healthcare providers place a balloon or tracheal dilator in your trachea. The balloon or dilator stretches your trachea so that you can breathe.
* Laser bronchoscopy. Providers direct a laser beam at scar tissue in your trachea. The laser burns away the tissue, opening up your airways.
* Trachea airway stent. A provider places a small, plastic or metal tube called a stent that holds your trachea open. (Stenting may or may not be an option, depending on the location of the stenosis.)
* Tracheal resection and reconstruction. A provider cuts away (resects) the tissue that’s causing the narrowing. Then, they join the two remaining ends of your trachea together. This procedure reconstructs your trachea to create an unobstructed airway.

The best treatment for tracheal stenosis depends on lots of things, including where the narrowing is and how severe it is. Your provider will explain how these factors inform which procedures will work best for you.

## **Outlook / Prognosis**

You’ll likely need surgery or a nonsurgical procedure if you have tracheal stenosis. Each treatment option has different recovery times and outcomes.

For example, tracheal resection and reconstruction surgeries are invasive. But they’re more likely to eliminate the narrowing in your trachea in the long term. Nonsurgical procedures, like tracheal dilation, are less invasive. They may be the only treatment you need, or you may repeat procedures.

Regardless of the procedure, your healthcare provider will monitor you to check for recurrence. Tracheal stenosis sometimes comes back because treatment can cause new scar tissue to form. Your provider will explain how likely it is that your condition will return.

## **Living With**

You might start by asking your healthcare provider how your surgery will affect you. Every procedure to treat tracheal stenosis will require different at-home care. Your healthcare provider will have information about your next steps. They may advise you on:

* What you can eat. For the first 24 hours, you may need to stick with soft foods that are easy to swallow. It may be a good idea to limit yourself to bland foods that won’t upset your stomach.
* How you should sleep. For the first few days, you may need to keep your upper body elevated as you try to sleep.
* How to manage pain. Your provider can recommend over-the-counter (OTC) medications or prescribe pain medicines as needed.
* How active you should be. You may have activity restrictions for the first week or so following surgery.

### **When should I see my healthcare provider?**

Your healthcare provider will schedule follow-up appointments to check on your recovery after treatment. At first, you may need to see your provider every few weeks or so. If you’re healing well, your provider may extend follow-up visits to every few months until they’re confident the stenosis won’t return.

Contact your provider anytime you’re experiencing shortness of breath. While next steps vary depending on your condition, it can be dangerous to put off getting help when you’re having difficulty breathing. It’s essential to seek care.

You should go to the emergency room if you can’t breathe or have other tracheal stenosis symptoms. The symptoms might be a sign your tracheal stenosis has come back.

If you’re caring for a newborn or infant who’s having difficulty breathing, get them to an emergency room immediately.

**DIFFERENTIAL DIAGNOSIS**

* Subglottic or laryngotracheal stenosis: Narrowing just below the vocal cords may be mistaken for tracheal stenosis.
* Tracheomalacia: Weakness and collapse of the tracheal walls causing airway obstruction.
* Vocal cord paralysis or paresis: Can cause breathing difficulty and stridor similar to stenosis.
* Congenital cysts or masses: Such as cysts in the airway spaces that block airflow.
* Foreign body aspiration: Especially in children, causing acute or chronic airway narrowing.
* Infections and inflammatory diseases: Tuberculosis, fungal infections, granulomatosis with polyangiitis (Wegener’s), sarcoidosis, and amyloidosis may cause tracheal narrowing.
* External compression: Mediastinal masses or thyroid enlargement compressing the trachea externally.
* Neoplasms: Tumors arising in or invading the trachea.
* Angioedema: Sudden swelling of airway tissues can mimic stenosis symptoms.
* Gastroesophageal reflux disease (GERD): Can cause laryngeal inflammation and mimic airway obstruction.

**EPIDEMIOLOGY**

* The overall incidence of tracheal stenosis in patients undergoing tracheostomy ranges widely but is reported between approximately 6% to 31% depending on diagnostic methods and patient populations. However, only a smaller fraction of these cases become symptomatic (around 6%-7%) and clinically significant.
* Post-intubation and post-tracheostomy stenosis is the most common etiology, and studies report an incidence rate of 0.6% to 21% following these airway interventions.
* In large datasets from the U.S., tracheal stenosis related to tracheostomy accounted for over 28% of tracheostomy-related emergency and hospital readmissions. Among patients with tracheostomies, about 1% to 7% may be readmitted with tracheal stenosis after initial treatment.
* Risk factors influencing incidence include the duration of intubation/tracheostomy, technique, presence of infection, and critical illness severity.
* The condition affects both sexes and all adult age groups, usually those who have undergone intensive care airway management.
* Recent cohorts during the COVID-19 pandemic showed increased incidence of tracheal stenosis in ICU patients, likely due to prolonged intubation periods.
* The mortality associated with severe tracheal stenosis requiring hospitalization varies; for example, about 3.2% in-hospital mortality was reported among patients hospitalized with tracheal stenosis due to tracheostomy in one large study, increasing to around 22.8% for those requiring readmission and prolonged inpatient care

**PREDEFINED Q & A SETS**

Q1: What is tracheal stenosis?  
A1: Tracheal stenosis is the narrowing or constriction of the windpipe (trachea) that can make breathing difficult. It may result from scar tissue, inflammation, injury, or other causes.

Q2: What causes tracheal stenosis?  
A2: Common causes include prolonged intubation or tracheostomy, trauma to the airway, infections, tumors, autoimmune diseases, or congenital anomalies.

Q3: What are the symptoms of tracheal stenosis?  
A3: Symptoms often include shortness of breath, noisy breathing (stridor), wheezing, persistent cough, difficulty swallowing, and sometimes recurrent respiratory infections.

Q4: How is tracheal stenosis diagnosed?  
A4: Diagnosis includes physical examination, imaging such as CT scans or MRI, and direct visualization with bronchoscopy or laryngoscopy to assess the degree and location of narrowing.

Q5: What treatment options are available?  
A5: Treatment depends on severity and cause and may include:

* Observation for mild cases
* Endoscopic procedures such as balloon dilation to widen the narrowed segment
* Surgical resection and reconstruction for localized stenosis
* Use of airway stents or other medical therapies like corticosteroids
* Innovative therapies like cryotherapy or photodynamic therapy in selected cases

Q6: What are the benefits and risks of the treatments?  
A6:

* Balloon dilation can provide immediate relief but may require repeat procedures.
* Surgery can offer a definitive cure but carries risks like infection, complications from anesthesia, and impact on airway function.
* Stents help keep the airway open but might cause irritation or require removal later.
* Medical therapies reduce inflammation but may not resolve structural narrowing.

Q7: Can tracheal stenosis heal on its own?  
A7: No, tracheal stenosis usually requires medical or surgical intervention to prevent worsening symptoms and airway obstruction.

Q8: What follow-up care is needed?  
A8: Regular follow-up with your healthcare provider is important to monitor airway patency, assess for symptom recurrence, and manage any complications.

Q9: When should I seek urgent medical help?  
A9: Seek immediate care if you experience severe difficulty breathing, inability to speak, or bluish discoloration of lips or face.

Q10: Can lifestyle changes help manage symptoms?  
A10: Avoiding irritants like smoke or pollutants, maintaining good hydration, and doing breathing exercises can help improve symptoms and lung function.

**Medications Used in Tracheal Stenosis Treatment**

Corticosteroids (e.g., Budesonide, Triamcinolone)

Role: Reduce inflammation and swelling in the trachea, either through inhalation, systemic administration, or local injection into the stenotic area.

Side Effects: Throat irritation, oral candidiasis, voice changes, systemic effects (if oral or intravenous) including immunosuppression, osteoporosis, and adrenal suppression with prolonged use.

Antibiotics (e.g., Penicillin, Erythromycin)

Role: May be used especially if infection is a contributing factor. Erythromycin additionally has anti-inflammatory and immunomodulatory effects shown to reduce granulation tissue and fibrosis in some studies.

Side Effects: Allergic reactions, gastrointestinal upset, photosensitivity (especially with erythromycin), antibiotic resistance risk.

Antifibrotic Agents (e.g., Nintedanib)

Role: Experimental use shown to reduce fibrosis and tissue proliferation by modulating molecular pathways including histone deacetylase 2 (HDAC2) activation.

Side Effects: Possible liver enzyme elevations, gastrointestinal symptoms (diarrhea, nausea), fatigue.

Mitomycin C (topical)

Role: Applied locally during endoscopic procedures to inhibit fibroblast proliferation and reduce scar formation.

Side Effects: Local irritation, potential tissue necrosis if misapplied.

Methotrexate (low-dose)

Role: Used as an adjunct in recurrent or resistant laryngotracheal stenosis to reduce inflammation and fibrosis.

Side Effects: Immunosuppression, liver toxicity, gastrointestinal upset, marrow suppression (requires monitoring).

**GENOMIC DATA**

* Congenital tracheal stenosis is often caused by mutations (pathogenic variants) in genes important for tracheal development and cartilage formation. For example, mutations in the TBX5 gene have been identified in patients with congenital tracheal stenosis and associated congenital heart defects. TBX5 plays a role in mesenchymal tissue development in the trachea.
* Other implicated genes in tracheal development that when mutated can lead to tracheal stenosis or related malformations include SOX9, FGF10, GLI3, and components of key developmental signaling pathways such as Sonic Hedgehog (SHH) and Bone Morphogenetic Protein (BMP) pathways. These regulate cartilage formation, tracheal specification, and epithelial-mesenchymal interactions.
* Mutations or disruptions in these genes can cause abnormal cartilage ring formation (such as complete O-shaped rings instead of normal C-shaped rings), leading to fixed narrowing of the tracheal lumen characteristic of congenital stenosis.
* In acquired tracheal stenosis (such as post-intubation or post-tracheostomy), recent preliminary research suggests that genetic differences—especially in genes related to wound healing and scarring—may influence individual susceptibility to abnormal scar formation causing stenosis. Certain genetic variants have been linked with increased risk of laryngotracheal stenosis, with possible differences among ethnic backgrounds.

**DOCTOR-PATIENT CONVERSATIONS**

Doctor: Hello, thank you for coming in today. I want to talk with you about your breathing difficulties. Based on your tests, you have a narrowing of your windpipe called *tracheal stenosis*.

Patient: What exactly is tracheal stenosis?

Doctor: Tracheal stenosis means that part of your trachea—the airway that carries air to your lungs—is narrower than usual. This narrowing can make it harder for you to breathe, especially when exercising or during a respiratory infection.

Patient: What causes this to happen?

Doctor: There are several causes. The most common is scarring or inflammation from previous breathing tube placement or surgery, infections, trauma, or sometimes autoimmune conditions. In some cases, it can be present from birth due to developmental reasons.

Patient: What symptoms should I expect or watch for?

Doctor: You may experience shortness of breath, noisy or wheezing breathing (called stridor), persistent cough, or difficulty with exercise. If the narrowing worsens, these symptoms may become more severe.

Patient: How do you diagnose it?

Doctor: Diagnosis usually involves imaging like a CT scan to see the airway anatomy, and a bronchoscopy, where we insert a thin camera into your airway to directly view the narrowing.

Patient: What treatments are available?

Doctor: Treatment depends on the severity. For mild narrowing, sometimes supportive care and monitoring are enough. For more significant cases, options include:

* Endoscopic procedures like balloon dilation, where we widen the narrowed area using a small balloon.
* Surgical procedures to remove the narrowed segment and reconstruct the airway if needed.
* Use of airway stents to keep the airway open in some cases.
* Medications like corticosteroids to reduce inflammation.

Sometimes multiple treatments are needed for best results.

Patient: What are the risks of these treatments?

Doctor: Procedures like dilation are less invasive but may need repeating. Surgery is more definitive but carries risks such as infection, bleeding, and impact on breathing during recovery. Stents can sometimes cause irritation or need removal later on.

Patient: What happens after treatment? Will I need follow-up?

Doctor: Yes, close follow-up is very important to monitor your airway and catch any recurrence early. Initially, you may need frequent visits, which may gradually space out as you heal.

Patient: Is there anything I can do on my own to help?

Doctor: Avoiding irritants like smoke, dust, and pollutants is important. Breathing exercises and maintaining good lung health can help. Your care team may also recommend speech therapy if your voice or breathing is affected.

Patient: When should I seek emergency help?

Doctor: If you experience severe difficulty breathing, inability to speak, or bluish discoloration of your lips or face, seek emergency care immediately.

Patient: Thank you, doctor. That helps me understand what’s going on.

Doctor: You're welcome. We're here to support you throughout treatment. Please reach out anytime with questions or concerns.

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**Tracheitis**

**ALTERNATIVE NAMES**

* Bacterial Tracheitis
* Acute Tracheitis
* Bacterial croup
* Laryngotracheobronchitis
* Tracheobronchitis
* Infectious Tracheitis
* Laryngotracheitis (when inflammation involves both the larynx and trachea)

**DEFINITION / DESCRIPTION**

Tracheitis is an infection of the windpipe (trachea) that makes breathing difficult. A rare but serious complication of an upper respiratory tract infection, it is more common in children—their smaller tracheas are more easily blocked by swelling.

Tracheitis symptoms include a cough similar to croup, fever, and wheezing. It may also cause a blue tinge to lips, nails, and skin—a dangerous sign that the body is not getting enough oxygen.

Tracheitis is a medical emergency. Children with tracheitis may need a temporary breathing tube (intubation), treatment in the intensive care unit (ICU), and intravenous (IV) antibiotics.

**CAUSES**

Tracheitis often begins as a viral infection of the upper airways, which sets the stage for a secondary bacterial infection. Bacteria most commonly involved in tracheitis include:

* *Staphylococcus aeureus*
* *Streptococcus pneumoniae*
* *Moraxella catarrhalis*
* *Haemophilus influenzae type B (HiB)*
* *Haemophilus influenzae*

Tracheitis is rare: It only occurs in about one in 1 million upper respiratory infections. It is most common in children between ages 3 to 8 and occurs more often in boys

This is because children have a smaller windpipe that is more easily blocked by swelling

Tracheitis is much less common in adults, but the risk is higher in those who have HIV/AIDS, a weak immune system, or diabetes. It can occur from a tracheostomy, a procedure that creates a breathing hole in the trachea through the neck, and mechanical ventilator use

While bacteria is often the culprit, fungal infections, such as *Aspergillus* and *Zygomycetes*, can causetracheostomy-associated tracheitis. Most adult cases of tracheitis occur in patients who are already hospitalized

**SIGNS / SYMPTOMS**

Symptoms of tracheitis vary from person to person and may include

* Cough that sounds “croupy,” and the child may cough up secretions
* Breathing difficulties
* Stridor when breathing in (common)
* Wheezing when breathing out (uncommon)
* High fever, usually greater than 102 F
* Blue lips (a sign of cyanosis or low oxygen levels), if the ability to breathe is deteriorating

**Tracheitis is a life-threatening condition that requires medical attention. If your child is experiencing stridor or cyanosis, seek emergency medical treatment or call 9-1-1.**

Tracheitis symptoms in adults are similar to those in children and may include:

* Coughing up blood
* A croupy cough
* Difficulty breathing
* A high fever
* A hoarse voice
* Sore throat
* Stridor (high-pitched, whistling when taking in a breath)
* Wheezing

### Difference Between Tracheitis and Croup

A “croupy cough” is a hallmark symptom of viral croup that may also be present in tracheitis. It is often described as “barking,” “brassy,” or resembling a seal’s bark.

This cough makes it easy to confuse the two illnesses. The following features can help distinguish tracheitis from croup

Croup

* Affects children ages 3 months to 5 years
* Symptoms progressively worsen
* Cough does not produce phlegm
* Fever is usually mild though may be as high as 104 F
* Stridor may occur with activity
* Responds well to treatment with humidified oxygen and inhaled racemic epinephrine

Tracheitis

* Affects children ages 3 to 8 years
* Symptoms come on quickly
* Cough may or may not be productive
* Fever of 102 F or higher
* Stridor occurs at rest
* Does not respond well to treatment with humidified oxygen and inhaled racemic epinephrine

**DIAGNOSIS METHODS**

Tracheitis is a diagnosis based on symptom history, physical examination, and testing. Diagnostic tests that your healthcare provider may perform include

* The measurement of oxygen levels in the blood (pulse oximetry)
* A culture of sputum from the trachea collected during a laryngoscopy, which can then be analyzed in the lab to determine the infectious agent causing your symptoms
* An X-ray of the lungs and breathing passageways

During these procedures, your healthcare provider will be using the findings to help differentiate from epiglottitis, another relatively rare disorder that may require rescue interventions for breathing

**TREATMENT OPTIONS**

Aggressive treatment early on is important in properly managing tracheitis. Initial treatments will usually require admission into an ICU and may include the following:

### Laryngoscopy

Laryngoscopy is an exam of the back of your throat. This can be done while awake using a tool with a small mirror and a light

In some cases, a more advanced laryngoscopy may be needed. This is done under general anesthesia and uses a tube called a laryngoscope.

During this procedure, your healthcare provider will clear secretions and membranes from the airway to prevent obstruction

### Intubation

A breathing tube may be placed (intubated) to improve oxygen intake in people with tracheitis. A breathing tube will allow the ICU nurses to perform aggressive airway suctioning to keep breathing as comfortably as possible

### Antibiotics

Tracheitis requires treatment with IV antibiotics for five to six days, followed by oral antibiotics for 10 to 14 days of treatment. Antibiotic regimens commonly used to treat tracheitis include

* Amoxicillin-clavulanic acid
* Ampicillin-sulbactam
* Ceftriaxone plus nafcillin or vancomycin
* Clindamycin plus a third-generation cephalosporin

People who are allergic to those antibiotics may be treated with vancomycin or clindamycin plus levofloxacin or ciprofloxacin. If cultures come back resistant to these antibiotics, they may be changed to a different regimen.

### Antifungals

In the case of fungal tracheitis infections like *Aspergillus* or *Zygomycetes*, treatment includes IV antifungal medications, such as Vfend IV (voriconazole).

**OUTLOOK / PROGNOSIS**

* Most patients recover fully without long-term complications once the acute phase is managed effectively with airway support and antibiotics. Recovery often begins within 5 days of treatment initiation, with hospital stays typically ranging from 3 to 12 days depending on severity.
* The main risks during the acute phase are related to upper airway obstruction and complications from respiratory distress, which can be life-threatening if not addressed promptly. Mortality rates historically ranged from 4% to 20% but have decreased dramatically with modern medical care and are now very rare, especially in developed healthcare settings.
* Common complications during illness include pneumonia, septicemia, toxic shock syndrome, acute respiratory distress syndrome, and rarely cardiorespiratory arrest. Long-term morbidity is minimal but can include subglottic stenosis related to airway inflammation or endotracheal intubation.
* Intubation for airway stabilization is often required (reported rates vary from 38% to 100%), and smaller endotracheal tubes are used due to airway swelling. Duration of intubation averages around 3 days but varies with clinical response.
* Early recognition and aggressive treatment with broad-spectrum antibiotics and airway management greatly improve prognosis. Unlike viral croup, steroids and racemic epinephrine have not shown significant efficacy in bacterial tracheitis

**POSSIBLE COMPLICATIONS**

* Airway obstruction: The most critical complication, caused by swelling and pus blocking the airway, which can lead to respiratory distress and can be life-threatening if not managed promptly.
* Pneumonia: Reported in 19-60% of cases due to spread of infection into the lungs.
* Septicemia (blood infection) and sepsis: Systemic spread of bacteria from the trachea.
* Toxic shock syndrome, particularly when caused by *Staphylococcus aureus*.
* Acute Respiratory Distress Syndrome (ARDS): Severe lung inflammation causing respiratory failure.
* Cardiorespiratory arrest: Due to severe hypoxia from airway obstruction or systemic illness.
* Anoxic encephalopathy: Brain damage from lack of oxygen during severe airway compromise.
* Post-intubation complications: Including endotracheal tube plugging or accidental extubation, postextubation stridor, and subglottic stenosis (narrowing of the airway due to inflammation or scarring).
* Other less common complications include acute renal failure, pulmonary edema, pneumothorax, pneumomediastinum, and lobar atelectasis

**WHEN TO SEE A DOCTOR / RED FLAG**

* Recent upper respiratory infection followed by sudden high fever
* Cough that worsens or becomes severe and deep
* Difficulty breathing or noisy breathing such as stridor (a high-pitched wheeze when inhaling)
* Signs of airway obstruction like chest retractions, nasal flaring, or cyanosis (blue tinge to skin)
* Excessive drooling, inability to swallow, or rapid breathing
* General worsening of symptoms with risk of respiratory distress

**DIFFERENTIAL DIAGNOSIS**

* Viral croup (laryngotracheobronchitis): Presents with a barking cough and stridor but usually has a milder course and responds to steroids and nebulized epinephrine. It is mostly caused by parainfluenza virus. Unlike bacterial tracheitis, viral croup usually does not produce purulent secretions or high fever.
* Epiglottitis: Characterized by sudden onset of high fever, drooling, difficulty swallowing, and a muffled "hot potato" voice. It tends to have a more rapidly progressive airway obstruction and patients often sit leaning forward to breathe. Caused commonly by Haemophilus influenzae type b (Hib).
* Pharyngitis and tonsillitis: These cause sore throat, fever, and sometimes cough, but lack the severe airway obstruction and stridor seen in tracheitis. Usually caused by viruses or Group A streptococcus.
* Retropharyngeal abscess: Presents with fever, neck pain, difficulty opening the mouth, and sometimes stridor, due to a deep neck space infection. Imaging often reveals abscess formation.
* Diphtheria: Rare now due to vaccination but can cause membranous pharyngitis and airway obstruction; differentiation depends on clinical and microbiological findings.
* Subglottic stenosis: Usually congenital or acquired, presents with chronic stridor and respiratory symptoms but lacks acute infection features.
* Angioneurotic edema: Sudden airway swelling usually due to an allergic reaction, without infectious symptoms like fever or purulent cough

**EPIDEMIOLOGY**

### United States statistics

Tan and Manoukian reported that 500 children were hospitalized for croup at one pediatric hospital over a 32-month period.Approximately 98% had viral croup, and 2% had bacterial tracheitis. Cases usually occur in the fall or winter months, mimicking the epidemiology of viral croup.

A study that described the frequency and severity of complications in hospitalized children younger than 18 years with seasonal influenza (during 2003-2009) and 2009 pandemic influenza A(H1N1) (during 2009-2010) reported that out of 7293 children hospitalized with influenza, less than 2% had complications from tracheitis. However, along with other rare complications, tracheitis was associated with a median hospitalization duration of more than 6 days, with 48%-70% of children requiring intensive care.

### International statistics

Bacterial tracheitis remains a rare condition, with an estimated incidence of approximately 0.1 cases per 100,000 children per year.

### Sex- and age-related demographics

In most epidemiologic studies, male cases are preponderant. Gallagher et al reported a male-to-female predominance of 2:1.

Bacterial tracheitis may occur in any pediatric age group. Gallagher et al reported 161 cases of patients younger than 16 years.The age range was from 3 weeks to 16 years, with a mean age of 4 years. This is in contrast to viral laryngotracheobronchitis, which occurs in patients aged 6 months to 3 years.

* The incidence is approximately 0.1 cases per 100,000 individuals worldwide. Specifically, studies from regions such as North West England and Victoria, Australia, report incidence rates around 0.08 to 0.09 per 100,000 children per year.
* It mostly affects children under 10 years, with a median age of about 5.2 years, and peak incidence between ages 3 to 8 years.
* Males are more commonly affected than females, with a male-to-female ratio approximately 1.3:1 to 2:1 in various studies.
* There is no specific racial or regional predilection reported.
* Tracheitis incidence tends to rise in winter months, correlating with viral epidemics such as influenza and respiratory syncytial virus (RSV), which often precede bacterial tracheitis.
* Historically, the mortality rate was high (10% to 40%), but with improved understanding, prompt airway management, and use of antibiotics, mortality is now very rare or near zero in developed healthcare settings.
* Bacterial tracheitis is an uncommon cause of hospital admissions in pediatric populations, with reported incidences around 0.4 per 1000 pediatric admissions in some centers.

**PREDEFINED Q & A SETS**

Q1: What is tracheitis?  
A1: Tracheitis is inflammation and infection of the trachea (windpipe). It is often caused by bacterial or viral infections and can result in symptoms like severe cough, stridor, and difficulty breathing. It is particularly serious in children due to their narrower airways.

Q2: What causes tracheitis?  
A2: Common causes include bacterial infections (most often Staphylococcus aureus, Streptococcus species, Moraxella catarrhalis, Haemophilus influenzae), viral upper respiratory infections, and irritants such as smoke or chemical inhalation. It often follows a viral cold or flu.

Q3: Who is most at risk of tracheitis?  
A3: It primarily affects children aged 3 to 8 years. Immunocompromised individuals and those with tracheostomies are also at risk. The incidence often increases during colder months when viral infections are more common.

Q4: What are the typical symptoms?  
A4: Symptoms include severe dry cough, high fever, noisy breathing (stridor), chest pain with coughing, difficulty breathing, hoarseness or low-pitched voice, and sometimes thick purulent secretions from the airway.

Q5: How is tracheitis diagnosed?  
A5: Diagnosis is clinical, supported by neck or chest x-rays showing subglottic narrowing or irregular tracheal margins. Tracheal cultures may identify the causative organisms. It is important to distinguish it from similar conditions like viral croup and epiglottitis.

Q6: What is the treatment for tracheitis?  
A6: Treatment includes supportive care and antibiotics targeting typical bacteria. Empiric therapy often includes vancomycin or clindamycin combined with a third-generation cephalosporin or ampicillin-sulbactam. Severe cases may require airway management or intensive care.

Q7: Can tracheitis be life-threatening?  
A7: Yes, bacterial tracheitis can cause airway obstruction and is potentially life-threatening if not treated promptly. However, with modern medical care, most patients recover fully.

Q8: How can tracheitis be prevented?  
A8: Reducing exposure to respiratory infections, maintaining good hygiene, avoiding smoking and heavy alcohol use, and vaccinations (like Hib vaccine) can help lower the risk

**DOCTOR-PATIENT CONVERSATIONS**

Doctor: Good morning. What brings you in today?

Patient (or Parent): My child has had a severe cough for a few days and is having trouble breathing. There's also a high fever.

Doctor: I see. Has the cough been barking or harsh? Any noisy breathing like a wheeze or stridor?

Patient: Yes, it sounds like a barking cough, and sometimes there’s a noisy inhale.

Doctor: Has your child had any recent cold or flu symptoms before this started?

Patient: Yes, about a week ago they had a cold.

Doctor: These symptoms could be due to tracheitis, which is an inflammation of the windpipe often caused by a bacterial infection following a viral illness. I’ll need to examine your child’s throat and listen to the breathing.

(The doctor performs a physical exam.)

Doctor: I can hear some airway narrowing and the symptoms are consistent with bacterial tracheitis. This can sometimes cause airway obstruction, so it’s important we start treatment quickly.

Patient: What treatment is needed?

Doctor: We will start intravenous antibiotics to target the infection. Depending on how severe the breathing difficulty is, your child may need supportive care such as oxygen or humidified air. In some cases, airway management may be necessary if swelling obstructs breathing.

Patient: Is this dangerous?

Doctor: It can be serious, especially in children, but with early treatment, most recover well. It’s important to watch for worsening symptoms like difficulty breathing, increased fever, or lethargy, and seek emergency care if these occur.

Patient: How long will the treatment last?

Doctor: Typically, antibiotics are given for 7–10 days, sometimes initially in hospital. We will monitor your child closely to ensure they improve.

Patient: What can we do at home to help?

Doctor: Keep the child well hydrated, maintain humidified air, avoid irritants like smoke, and ensure proper rest. Follow up with us regularly.

**GENOMIC DATA**

* Bacterial tracheitis is often polymicrobial, with common pathogens including *Staphylococcus aureus*, *Haemophilus influenzae*, *Moraxella catarrhalis*, and occasionally *Klebsiella pneumoniae*. Genomic studies of these bacteria aim to identify strain-level diversity and resistance genes but detailed tracheitis-specific pathogen genomics are still limited.
* Metagenomic whole genome shotgun sequencing methods have been applied to airway samples to identify bacterial taxa at genus level and viral diversity in respiratory infections that may precede or coexist with tracheitis. These techniques provide insights into microbial communities in the respiratory tract but often lack strain-specific or functional genomic details relevant to tracheitis pathogenesis.
* Metagenomic next-generation sequencing (mNGS) is emerging as a promising tool for diagnosing lower respiratory tract infections, including those leading to tracheitis, by detecting a broad spectrum of pathogens—bacteria, viruses, and fungi—from respiratory samples without prior culturing.
* There is no known specific human genetic or genomic susceptibility linked directly to the development of tracheitis; rather, it is primarily a secondary bacterial infection often initiated by preceding viral infections that damage airway mucosa.

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### **Tracheomalacia**

## **Alternative Names for Tracheomalacia**

· Congenital Tracheomalacia  
 When the condition is present from birth.

· Acquired Tracheomalacia  
 When tracheomalacia develops later due to injury, inflammation, or external compression.

· Tracheal Cartilage Softening  
 Describes the underlying pathology of weakened tracheal cartilage.

· Dynamic Tracheal Collapse  
 Refers to the airway collapse during breathing due to tracheal wall weakness.

· Tracheal Collapse Syndrome  
 Emphasizes the functional consequence of the malacia.

· Airway Malacia  
 A broader term that includes tracheomalacia and bronchomalacia (softening of the bronchi).

· Tracheobronchomalacia (TBM)  
 When both the trachea and bronchi are involved.

**DEFINITION AND DESCRIPTION**

Tracheomalacia (*TRAY-kee-oh-muh-LAY-shia*) is when you have weak or floppy cartilage in your trachea (windpipe). The walls of your windpipe can collapse or fall in, causing symptoms like high-pitched breathing. It can also trap mucus in your lungs, making it difficult to clear them out. In severe cases, tracheomalacia may be life-threatening, but it’s curable with treatment.

Tracheomalacia usually affects newborns, but anyone can develop it. Babies born with the condition often develop symptoms when they’re 1 to 2 months old. Then, symptoms improve over the first three years of life. That’s because cartilage strengthens as your baby’s windpipe grows. But severe cases may need surgery.

#### **Types of this condition**

There are two types of tracheomalacia:

· **Congenital tracheomalacia**: “Congenital” means you’re born with the condition. It happens when a baby’s windpipe didn’t form properly during fetal development.

· **Acquired tracheomalacia**: “Acquired” means the condition developed after you were born. It happens when your windpipe breaks down or gets damaged. Injuries, surgeries or prolonged mechanical ventilation can cause this. Although it’s uncommon, acquired tracheomalacia can occur at any age.

Some people with tracheomalacia also have weak bronchi (the tubes that run from your windpipe to your lungs). Healthcare providers call this condition tracheobronchomalacia.

Congenital tracheomalacia is somewhat rare. Even so, it’s the most common birth defect affecting the windpipe. Approximately 1 in 2,100 children are born with the condition.

Acquired tracheomalacia (which can occur at any age) is also very uncommon.

### **causes tracheomalacia**

Congenital tracheomalacia happens when the cartilage in your baby’s windpipe doesn’t develop the way it should. The walls of their windpipe are floppy instead of rigid.

Acquired tracheomalacia causes include:

· Chronic acid reflux (GERD).

· Damage from surgery or other medical procedures.

· Emphysema.

· Polychondritis (inflammation of the cartilage) in your windpipe.

· Tracheoesophageal fistula repair (fixing an abnormal connection between your esophagus and trachea).

· Tracheostomy, or long-term use of a breathing tube.

· Upper respiratory infections, like bronchitis.

Tracheomalacia may be associated with other conditions like:

· Developmental delays.

· Ehlers-Danlos syndrome.

· Heart defects.

### 

### **Risk Factors**

· **Age:** Tracheomalacia is more common in infants and young children, particularly those born prematurely.

· **Gender:** Some studies suggest a higher prevalence in males.

· **Geographic Location:** Exposure to environmental pollutants may vary by location, influencing the risk.

· **Underlying Conditions:** Individuals with chronic respiratory diseases, such as asthma or chronic obstructive pulmonary disease (COPD), may be at higher risk.

### **symptoms of tracheomalacia**

The most common tracheomalacia symptom is high-pitched or noisy breathing (stridor). Other symptoms include:

· Breathing issues that get worse when feeding, crying or coughing.

· Choking.

· Chronic coughing.

· Cyanosis (a condition where your skin, lips or nails turn blue from a lack of oxygen).

· Difficulty swallowing.

· Hoarseness.

· Recurring (returning) airway infections like pneumonia or bronchitis.

· Shortness of breath.

· Wheezing or rattling sounds.

## **Diagnosis and Tests**

A healthcare provider will do a physical exam and ask about your symptoms. They’ll also use laryngoscopy or bronchoscopy to look at your windpipe. To do this, they’ll guide a lighted scope with a camera down your throat.

Your healthcare provider may need to run more tests to diagnose tracheomalacia, like:

· Airway fluoroscopy.

· Barium swallow (esophagram).

· Chest X-ray.

· CT (computed tomography) scan.

· Lung function tests.

· MRI (magnetic resonance imaging).

## **Management and Treatment**

Healthcare providers can treat tracheomalacia with nonsurgical therapies, medications or surgery. What’s right for you depends on the extent of the condition.

#### **Nonsurgical therapies**

The following treatments help keep your airways open and your lungs clear:

· **Breathing humidified air**. Using a humidifier can thin out mucus so you can breathe comfortably.

· **Chest physical therapy**. A physical therapist can teach you breathing exercises to clear mucus from your lungs.

· **Continuous positive airway pressure (CPAP)**. Using a CPAP machine can keep your windpipe from collapsing too much during sleep.

#### **Medications**

Healthcare providers use several different medications to treat tracheomalacia, including:

· **Antibiotics** to treat bacterial infections.

· **Bronchodilators** to relax the muscles that help you breathe.

· **Corticosteroids** to reduce inflammation.

· **Mucolytics** to thin out mucus.

#### **Surgery**

Severe tracheomalacia cases may need surgery. Some options include:

· **Aortopexy**. A surgeon moves your aorta up and away from your airway and attaches it to your breastbone. This keeps your windpipe from collapsing.

· **Stenting**. This involves placing a stent (hollow tube) in your airway to keep it open.

· **Tracheopexy**. A surgeon connects part of your windpipe to a nearby structure (like your breastbone or a ligament at the top of your spine). This keeps your windpipe open and prevents collapse.

### **complications of tracheomalacia**

Without treatment, tracheomalacia can cause:

· Airway obstruction.

· Aspiration pneumonia.

· Growth faltering.

· Upper respiratory infections that linger or come back often

## **Outlook / Prognosis**

After treatment, most people with tracheomalacia go on to live healthy lives with no complications.

Treatment isn’t always necessary. Congenital tracheomalacia usually improves on its own by age 3. As your baby’s tracheal cartilage grows stronger, their symptoms will likely improve. But in some cases, your baby may need medication and/or surgery.

People with tracheomalacia need close monitoring if they develop respiratory infections. Even minor colds can cause complications. Your healthcare provider may recommend treatments or medications to manage your symptoms.

**Prevention**

There’s nothing you can do to reduce your risk for tracheomalacia. But you can manage it with the help of your healthcare provider.

**When should I see my healthcare provider?**

Schedule an appointment with your healthcare provider if your baby shows tracheomalacia symptoms. This includes things like frequent cough, noisy breathing or prolonged respiratory infections.

Tracheomalacia in adults can cause exercise intolerance and frequent respiratory infections. Tell your provider if you notice these things. They can confirm the diagnosis and recommend appropriate treatment.

## **Diagnostic Considerations**

The differential diagnosis of tracheomalacia includes laryngomalacia, subglottic stenosis, congenital cysts, vocal cord paralysis, and hypocalcemic tetany. Complications include problems with acute airway obstruction and perioperative morbidity and mortality.

According to a 2005 study by Boogaard, when pediatric pulmonologists diagnosed airway malacia (on the basis of symptoms, history, and lung function) prior to bronchoscopy, a correct diagnosis was made in 74% of the cases.However, in 52% of the diagnoses of airway malacia, the diagnosis was not suspected prior to bronchoscopy. Children with tracheomalacia present with atypical and variable clinical features; considerable overlap occurs with features of allergic asthma

## **Differential Diagnosis for Tracheomalacia**

## **Structural Airway Abnormalities**

· Laryngomalacia  
 Softening and collapse of supraglottic structures causing inspiratory stridor, often coexisting with tracheomalacia in infants.

· Bronchomalacia  
 Weakness and collapse of bronchial walls, causing wheezing and recurrent infections.

· Vascular Rings and Slings  
 Congenital vascular anomalies (e.g., double aortic arch, pulmonary sling) compressing the trachea externally.

· Tracheal Stenosis  
 Fixed narrowing of the trachea due to congenital or acquired causes (e.g., prolonged intubation, trauma).

· Tracheal Web or Atresia  
 Congenital membrane or absence of tracheal segments causing obstruction.

## **Infectious and Inflammatory Causes**

· Tracheitis (Bacterial or Viral)  
 Infection causing airway inflammation and edema mimicking airway collapse.

· Granulomatous Diseases  
 Such as tuberculosis or sarcoidosis causing airway narrowing.

## **Neuromuscular Disorders**

· Conditions causing poor airway muscle tone leading to dynamic airway collapse (e.g., muscular dystrophies).

## **Foreign Body Aspiration**

· Partial airway obstruction causing wheezing and respiratory distress.

## **Tumors or Masses**

· Intratracheal or extrinsic masses compressing or invading the airway.

## **Chronic Obstructive Pulmonary Disease (COPD) and Emphysema**

· In adults, airway collapse due to loss of elastic recoil and cartilage weakening.

## **Epidemiology**

All types of tracheomalacia are extremely rare; no definite incidence rates are available.

In a total of 512 bronchoscopies, airway malacia was diagnosed in 160 children (94 males) at a median age of 4.0 years (range, 0-17 y). Airway malacia was classified as primary in 136 children and as secondary in 24 children. The incidence of primary airway malacia was estimated to be at least 1 in 2100.

**medications used, their purposes, and potential side effects**

Medications Used in Tracheomalacia Treatment

| Medication Type | Examples | Purpose / Mechanism | Common Side Effects |
| --- | --- | --- | --- |
| Nebulized Medications | Saline solution | Helps humidify airways, loosen mucus, ease cough | Minimal; possible mild irritation |
| Anticholinergic Bronchodilators | Ipratropium bromide (Atrovent) | Opens airways by relaxing bronchial muscles, reduces cough and airway collapse | Dry mouth, throat irritation, cough, dizziness |
| Bronchodilators | Albuterol (short-acting beta-agonist) | Relaxes airway smooth muscle to improve airflow | Tremor, nervousness, palpitations |
| Corticosteroids | Inhaled or systemic steroids (e.g., prednisone) | Reduce airway inflammation that may worsen symptoms | Oral thrush (inhaled), hoarseness, immune suppression (systemic) |
| Antibiotics | Various (if bacterial infection present) | Treat respiratory infections that worsen airway obstruction | GI upset, allergic reactions |

## **Additional Non-Drug Therapies**

· Humidified air and chest physiotherapy to clear secretions and reduce airway irritation.

· Noninvasive positive pressure ventilation (CPAP or BiPAP) to stent open airways during breathing, especially at night.

· Surgical interventions (for severe cases) such as aortopexy, tracheopexy, tracheostomy, airway stenting, or tracheal reconstruction.

## **Important Notes on Medication Use**

· Medications like ipratropium bromide and bronchodilators help improve airway tone but do not cure tracheomalacia; they manage symptoms.

· Corticosteroids are used cautiously due to side effects and are typically reserved for cases with significant airway inflammation.

· Treating respiratory infections promptly with antibiotics is critical, as infections can worsen airway collapse.

· Many infants improve as their tracheal cartilage strengthens with growth, reducing the need for long-term medication.

#### **PREDEFINED Q AND A**

## **1. How severe is the condition?**

Severity is usually assessed by the degree of airway collapse during breathing, especially exhalation:

· Mild: 25–50% airway lumen narrowing

· Moderate: 50–75% narrowing

· Severe: >75% narrowing, sometimes near complete collapse  
 Severity also depends on symptoms and whether collapse occurs during quiet breathing or only with coughing or agitation. Flexible bronchoscopy is the gold standard for diagnosis and severity assessment, but interpretation can vary between clinicians.

## **2. What caused the condition?**

Tracheomalacia can be:

· Congenital: due to underdeveloped or soft tracheal cartilage present from birth

· Acquired: caused by prolonged intubation, trauma, inflammation, external compression (e.g., vascular rings), chronic infections, or chronic obstructive pulmonary disease (COPD) in adults  
 Risk factors include obesity and gastroesophageal reflux disease (GERD).

## **3. What treatment do you recommend?**

Treatment depends on severity and symptoms:

· Mild cases often improve with conservative management (humidified air, airway clearance, treating infections)

· Medical therapies may include bronchodilators, corticosteroids, and antibiotics if infections occur

· Noninvasive ventilation (CPAP/BiPAP) can help stent open airways during breathing

· Severe cases may require surgical interventions like aortopexy, tracheopexy, or airway stenting.

## **4. Will surgery be necessary?**

Surgery is usually reserved for:

· Severe tracheomalacia causing significant airway obstruction or respiratory distress

· Failure of conservative and medical management

· Associated anatomical abnormalities causing external airway compression  
 The decision depends on clinical symptoms, airway collapse severity, and response to other treatments.

## **5. What can I do to ease symptoms?**

· Use humidified air to keep airways moist

· Avoid respiratory irritants like smoke and pollutants

· Promptly treat respiratory infections with antibiotics if needed

· Practice airway clearance techniques as advised

· Use prescribed inhalers or medications properly

· Monitor for worsening symptoms and seek medical attention if breathing difficulty increases.

## **6. How soon do I need treatment?**

· Mild cases may only require monitoring and supportive care.

· Moderate to severe cases with symptoms like stridor, recurrent infections, or respiratory distress need prompt evaluation and treatment.

· If symptoms worsen rapidly (e.g., severe breathing difficulty, cyanosis), emergency care is necessary

#### **What conditions might be confused for tracheomalacia?**

The following conditions may be confused for tracheomalacia:

· **Laryngomalacia**: This condition refers to floppy tissue above your voice box. Your voice box sits above your windpipe.

· **Tracheobronchomalacia**: This describes weak bronchi (the tubes that run from your windpipe to your lungs). Some people with tracheomalacia also have tracheobronchomalacia.

**GENOMIC DATA**

· Tracheomalacia can be associated with genetic syndromes and congenital anomalies. It often occurs alongside conditions such as Ehlers-Danlos syndrome, Marfan syndrome, Mounier-Kuhn syndrome, and other connective tissue disorders that affect cartilage strength and elasticity.

· Some cases of congenital tracheomalacia are linked to mutations affecting cartilage development and extracellular matrix proteins, though specific causative genes for isolated tracheomalacia are not well defined.

· A rare autosomal recessive condition involving mutations in the DCHS1 gene (encoding protocadherin-16) has been associated with tracheomalacia among other systemic features like intellectual disability. However, this is not a common cause of isolated tracheomalacia.

· Tracheomalacia frequently coexists with other congenital malformations such as tracheoesophageal fistula, vascular rings, and esophageal atresia, which have complex embryological and genetic backgrounds.

· Most cases of tracheomalacia are considered multifactorial or developmental anomalies rather than single-gene disorders. There is no routine genetic testing for isolated tracheomalacia, and it is often sporadic without clear inheritance patterns

**DOCTOR PATIENT CONVERSATION**

Doctor: Hello, I understand your child has been having noisy breathing and some coughing. After reviewing the tests, it looks like your child has a condition called tracheomalacia. Have you heard of this before?

Parent: No, I haven’t. What exactly is tracheomalacia?

Doctor: Tracheomalacia means that the cartilage in the windpipe, or trachea, is softer than usual. This softness can cause the airway to partially collapse when your child breathes out, leading to noisy breathing, coughing, or sometimes difficulty breathing.

Parent: Is this serious? Will it get worse?

Doctor: In many cases, especially in infants and young children, tracheomalacia improves as the cartilage strengthens with growth. Mild to moderate cases often get better over time without surgery. However, if the airway collapse is severe, it can cause breathing difficulties that need closer monitoring or treatment.

Parent: What causes tracheomalacia?

Doctor: It can be present from birth due to how the trachea developed, or it can develop later from injury, infections, or pressure from nearby blood vessels. Sometimes it occurs along with other conditions like vascular rings or certain genetic syndromes.

Parent: What treatments are available?

Doctor: Treatment depends on how severe the symptoms are. For mild cases, we usually recommend supportive care like humidified air, avoiding irritants, and monitoring. If infections occur, they are treated promptly. In more severe cases, medications such as inhalers or breathing support like CPAP might be needed. Surgery is rarely necessary but can be considered if symptoms are severe or don’t improve.

Parent: What should I watch for at home?

Doctor: Watch for signs of increased breathing difficulty, bluish color around lips or face, poor feeding, or frequent respiratory infections. If any of these happen, seek medical care promptly.

Parent: How long will this last?

Doctor: Many children improve by the time they are 1 to 2 years old as their airway cartilage becomes stronger. We will follow your child closely and adjust treatment as needed.

Parent: Thank you, doctor. This helps me understand what’s going on.

Doctor: You’re welcome. Please feel free to ask any questions anytime, and we’ll work together to support your child’s breathing health.

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**Tinnitus**

ALTERNATIVE NAMES

**DEFINITION / DESCRIPTION**

“Tinnitus” (pronounced “tin-NITE-us” or “TIN-ne-tus”) is the medical term for ringing in the ears. If you have tinnitus, sounds fill your head that no one else hears — like ringing, clicking, pulsing, humming or rushing. These sounds can be so soft you barely notice them or so loud that they seem to block out sounds in your environment. People with severe tinnitus might have trouble concentrating or sleeping. Over time, this can lead to frustration and depression.

Tinnitus isn’t a disease. It’s a symptom of several health conditions like ear injuries or age-related hearing loss. According to research, about 15% of the world’s population has it — including more than 50 million people in the United States. Anyone can get tinnitus, but it’s most common in people between the ages of 40 and 80.

**CAUSES**

Experts don’t know exactly why some people have ringing in their ears and others don’t. But tinnitus is a common symptom of certain medical conditions.

Common tinnitus causes include:

* Age-related hearing loss. Your brain is used to a certain level of sound stimulation. As a result, your hearing nerve is used to a certain level of activity. When hearing loss occurs, your brain receives less stimulation, but your hearing nerve may still fire at the rate it always has. This can cause you to hear sounds that aren’t really there. About 1 in 3 adults over age 65 develop ringing in their ears.
* Noise-induced hearing loss (NIHL). Exposure to loud noises can cause hearing loss and tinnitus. This can happen over time or from a single incident, like an explosion or close-range gunshot.
* Ear injuries and trauma. These injuries may affect nerves or areas of your brain that help you hear. People who get tinnitus after an injury usually only have ringing in one ear.
* Ear conditions. Earwax blockages or ear infections can cause temporary hearing loss, resulting in ear ringing.
* Medications. Certain drugs can cause tinnitus, including some antibiotics, antidepressants, cancer drugs and nonsteroidal anti-inflammatory drugs (NSAIDs).

Less common tinnitus risk factors include:

* Eustachian tube dysfunction.
* Ménière’s disease.
* Temporomandibular joint (TMJ) disorders.
* Vestibular schwannoma (a noncancerous tumor that forms on the vestibular nerve).
* Otosclerosis (abnormal bone growth inside your ears).
* Blood vessel disorders, which can cause pulsatile tinnitus — a rare form of ear ringing that happens in time with your heartbeat.
* Autoimmune diseases like lupus or rheumatoid arthritis.

People with tinnitus may experience flare-ups after exposure to certain triggers like stress or sleep deprivation. As you can imagine, this can result in a frustrating cycle.

**Symptoms**

Tinnitus is most often described as a ringing in the ears, even though no external sound is present. However, tinnitus can also cause other types of phantom noises in your ears, including:

* Buzzing
* Roaring
* Clicking
* Hissing
* Humming

Most people who have tinnitus have subjective tinnitus, or tinnitus that only you can hear. The noises of tinnitus may vary in pitch from a low roar to a high squeal, and you may hear it in one or both ears. In some cases, the sound can be so loud it interferes with your ability to concentrate or hear external sound. Tinnitus may be present all the time, or it may come and go.

In rare cases, tinnitus can occur as a rhythmic pulsing or whooshing sound, often in time with your heartbeat. This is called pulsatile tinnitus. If you have pulsatile tinnitus, your doctor may be able to hear your tinnitus when he or she does an examination (objective tinnitus).

**DIAGNOSIS METHODS**

Your doctor will typically diagnose you with tinnitus based on your symptoms alone. But in order to treat your symptoms, your doctor will also try to identify whether your tinnitus is caused by another, underlying condition. Sometimes a cause can't be found.

To help identify the cause of your tinnitus, your doctor will likely ask you about your medical history and examine your ears, head and neck. Common tests include:

* **Hearing (audiological) exam.** During the test, you'll sit in a soundproof room wearing earphones that transmit specific sounds into one ear at a time. You'll indicate when you can hear the sound, and your results will be compared with results considered normal for your age. This can help rule out or identify possible causes of tinnitus.
* **Movement.** Your doctor may ask you to move your eyes, clench your jaw, or move your neck, arms and legs. If your tinnitus changes or worsens, it may help identify an underlying disorder that needs treatment.
* **Imaging tests.** Depending on the suspected cause of your tinnitus, you may need imaging tests such as CT or MRI scans.
* **Lab tests.** Your doctor may draw blood to check for anemia, thyroid problems, heart disease or vitamin deficiencies.

Do your best to describe for your doctor what kind of tinnitus noises you hear. The sounds you hear can help your doctor identify a possible underlying cause.

* **Clicking.** This type of sound suggests that muscle contractions in and around your ear might be the cause of your tinnitus.
* **Pulsing, rushing or humming.** These sounds usually stem from blood vessel (vascular) causes, such as high blood pressure, and you may notice them when you exercise or change positions, such as when you lie down or stand up.
* **Low-pitched ringing.** This type of sound may point to ear canal blockages, Meniere's disease or stiff inner ear bones (otosclerosis).
* **High-pitched ringing.** This is the most commonly heard tinnitus sound. Likely causes include loud noise exposure, hearing loss or medications. Acoustic neuroma can cause continuous, high-pitched ringing in one ear.

**TREATMENT OPTIONS**

Treatment for tinnitus depends on whether your tinnitus is caused by an underlying health condition. If so, your doctor may be able to reduce your symptoms by treating the underlying cause. Examples include:

* **Earwax removal.** Removing an earwax blockage can decrease tinnitus symptoms.
* **Treating a blood vessel condition.** Underlying blood vessel conditions may require medication, surgery or another treatment to address the problem.
* **Hearing aids.** If your tinnitus is caused by noise-induced or age-related hearing loss, using hearing aids may help improve your symptoms.
* **Changing your medication.** If a medication you're taking appears to be the cause of tinnitus, your doctor may recommend stopping or reducing the drug, or switching to a different medication.

### **Noise suppression**

Many times, tinnitus can't be cured. But there are treatments that can help make your symptoms less noticeable. Your doctor may suggest using an electronic device to suppress the noise. Devices include:

* **White noise machines.** These devices, which produce a sound similar to static, or environmental sounds such as falling rain or ocean waves, are often an effective treatment for tinnitus. You may want to try a white noise machine with pillow speakers to help you sleep. Fans, humidifiers, dehumidifiers and air conditioners in the bedroom also produce white noise and may help make tinnitus less noticeable at night.
* **Masking devices.** Worn in the ear and similar to hearing aids, these devices produce a continuous, low-level white noise that suppresses tinnitus symptoms.

### **Counseling**

Behavioral treatment options aim to help you live with tinnitus by helping you change the way you think and feel about your symptoms. Over time, your tinnitus may bother you less. Counseling options include:

* **Tinnitus retraining therapy (TRT).** TRT is an individualized program that is usually administered by an audiologist or at a tinnitus treatment center. TRT combines sound masking and counseling from a trained professional. Typically, you wear a device in your ear that helps mask your tinnitus symptoms while you also receive directive counseling. Over time, TRT may help you notice tinnitus less and feel less distressed by your symptoms.
* **Cognitive behavioral therapy (CBT) or other forms of counseling.** A licensed mental health professional or psychologist can help you learn coping techniques to make tinnitus symptoms less bothersome. Counseling can also help with other problems often linked to tinnitus, including anxiety and depression. Many mental health professionals offer CBT for tinnitus in individual or group sessions, and CBT programs are also available online.

### **Medications**

Drugs can't cure tinnitus, but in some cases they may help reduce the severity of symptoms or complications. To help relieve your symptoms, your doctor may prescribe medication to treat an underlying condition or to help treat the anxiety and depression that often accompany tinnitus.

### **Potential future treatments**

Researchers are investigating whether magnetic or electrical stimulation of the brain can help relieve symptoms of tinnitus. Examples include transcranial magnetic stimulation (TMS) and deep brain stimulation.

**Lifestyle and home remedies**

Often, tinnitus can't be treated. Some people, however, get used to it and notice it less than they did at first. For many people, certain adjustments make the symptoms less bothersome. These tips may help:

* **Use hearing protection.** Over time, exposure to loud sounds can damage the nerves in the ears, causing hearing loss and tinnitus. To keep your tinnitus from getting worse, take steps to protect your hearing. If you use chain saws, are a musician, work in an industry that uses loud machinery or use firearms (especially pistols or shotguns), always wear over-the-ear hearing protection.
* **Turn down the volume.** Listening to music at very high volume through headphones can contribute to hearing loss and tinnitus.
* **Use white noise.** If tinnitus is especially noticeable in quiet settings, try using a white noise machine to mask the noise from tinnitus. If you don't have a white noise machine, a fan, soft music or low-volume radio static also may help.
* **Limit alcohol, caffeine and nicotine.** These substances, especially when used in excess, can affect blood flow and contribute to tinnitus.

**Alternative medicine**

There's little evidence that alternative medicine treatments work for tinnitus. However, some alternative therapies that have been tried for tinnitus include:

* Acupuncture
* Ginkgo biloba
* Melatonin
* Zinc supplements

**PREVENTION TIPS**

Because tinnitus is a symptom of other health conditions, you can’t always prevent it. But you can take steps to protect your hearing, which can help reduce your risk. Hearing protection may be warranted:

* When working in construction or factory settings.
* In certain performance arts or music careers.
* In some exercise classes. Many gyms play loud music for motivation.
* At concerts and movie theatres. Noise levels at these types of events can be too loud and may harm your hearing over time.
* When using power tools, mowing the lawn or utilizing other loud equipment.
* With the use of firearms like recreational shooting or hunting.

You should also be mindful any time you wear headphones or earbuds. Protect your ears by keeping the volume low.

Audiologists can fit custom hearing protection. Consider seeing an audiologist for this service if you participate in one or more of these activities frequently.

**POSSIBLE COMPLICATIONS**

Tinnitus affects people differently. For some people, tinnitus can significantly affect quality of life. If you have tinnitus, you may also experience:

* Fatigue
* Stress
* Sleep problems
* Trouble concentrating
* Memory problems
* Depression
* Anxiety and irritability
* Headaches
* Problems with work and family life

Treating these linked conditions may not affect tinnitus directly, but it can help you feel better.

**WHEN TO SEE A DOCTOR / RED FLAG**

You should schedule a visit with a healthcare provider if you have:

* Ringing in your ears that lasts for more than one week.
* Hearing loss.
* Dizziness.
* Vertigo.
* Tinnitus that sounds like your heartbeat.

If you develop sudden hearing loss in addition to tinnitus, schedule an appointment with a healthcare provider immediately. Treatment for sudden hearing loss is most effective within the first three days.

**DIFFERENTIAL DIAGNOSIS**

Differential diagnosis for tinnitus includes:

* Cytomegalovirus
* Hypercholesterolemia
* Lyme disease
* Measles
* Meningitis
* Neoplasm
* Neurosyphilis
* Rubella
* Sickle cell anemia
* Small vessel disease
* Stroke
* Tumour

**RECENT GUIDELINES OR UPDATES**

Guidelines for the assessment and management of chronic tinnitus. The major recommendations include the following:

* Conditions that if promptly managed may relieve tinnitus should be identified during the initial evaluation of a patient with presumed primary tinnitus
* Patients with tinnitus that is unilateral and associated with hearing difficulties or that persists for 6 months or more should undergo a comprehensive audiologic examination.
* Imaging studies of the head and neck are not recommended specifically to evaluate tinnitus unless one or more of the following is present: tinnitus that localizes to 1 ear, pulsatile tinnitus, focal neurologic abnormalities, and/or asymmetrical hearing loss
* Distinguish patients with bothersome tinnitus from those with non-bothersome tinnitus; educate patients with persistent, bothersome tinnitus about management strategies
* Patients with hearing loss and persistent, bothersome tinnitus should be offered a hearing aid evaluation, sound therapy, and cognitive behavior therapy
* Antidepressants, anticonvulsants, anxiolytics, or intratympanic medications should not be offered as a primary treatment for persistent, bothersome tinnitus
* Ginkgo biloba, melatonin, zinc, or other dietary supplements or transcranial magnetic stimulation should not be recommended for treating patients with persistent, bothersome tinnitus

**EPIDEMIOLOGY**

Almost everyone experiences tinnitus at some point in their lifetime. The American Tinnitus Association estimates that 10 million people suffer from tinnitus. It is also common in non-industrialized countries. Military personnel have a high incidence of tinnitus due to loud explosions and gunfire. It is also seen in the movie and stage workers who prepare scenes with explosions and gunfire. Musicians exposed to loud noise get tinnitus, such as drummers and those who perform in front of loudspeakers. Children may have tinnitus, but it is mostly unrecognized because they do not recognize the disorder. The usual history is of a worker exposed to loud factory noises, where workers had to shout to be heard. Many workers develop high-tone hearing loss, but only a small percentage also have tinnitus. Hyperacusis also can accompany tinnitus. In these cases, certain ordinary sounds, like closing doors, moving chairs, and dropping books, are so loud and strong that they are extremely uncomfortable or sometimes unbearable.

**PREDEFINED Q & A SETS**

### **Can you live a long life with tinnitus?**

Yes, you can. Tinnitus itself doesn’t shorten your life expectancy. But some underlying conditions, like heart disease, could affect your lifespan.

### **What is tinnitus a warning of?**

It’s not always apparent what causes tinnitus. But there are common links in some people:

| **If you have tinnitus in addition to these symptoms:** | **You might have:** |
| --- | --- |
| Pain, ear drainage. | Ear infection. |
| Dizziness, balance issues. | Ménière’s disease or a neurological condition. |
| Jaw pain, headaches, facial pain. | Temporomandibular joint (TMJ) disorder. |
| Noises that sound like your heartbeat (pulsatile tinnitus). | High blood pressure. |
| Headaches, migraines, neck pain, changes in tinnitus with head/neck movements. | Cervical spine conditions. |

Having the above symptoms doesn’t necessarily mean that you have the corresponding conditions. If you notice something isn’t quite right, you should tell your healthcare provider so they can give you a proper diagnosis.

### **Will tinnitus go away?**

It depends on the cause. If your ears are ringing after you leave a loud concert, it’s probably temporary. But if you still have tinnitus after a week or two, there’s probably something else going on.

There’s no standard tinnitus cure. But providers can often treat underlying conditions and help you manage your symptoms.

### **What should I avoid?**

If you have ringing in your ears, try to avoid:

* Total silence (this can make symptoms worse because the ringing is more noticeable).
* Exposure to loud noises.
* Common tinnitus triggers like poor sleep quality and extreme stress.

#### **Foods to avoid with tinnitus**

Some experts believe that nutrition plays a role in whether people experience tinnitus. There’s limited research about this topic, but recent studies suggest that these foods and beverages may increase your risk for tinnitus:

* Caffeine.
* Salt.
* Saturated fats.

**DOCTOR-PATIENT CONVERSATIONS**

Doctor: Good morning. What brings you in today?

Patient: I've been hearing ringing in my ears for a while now, and it's really bothering me.

Doctor: I see. Can you describe the sound you hear? For example, is it ringing, buzzing, clicking, or something else?

Patient: It’s mostly a high-pitched ringing, and sometimes it feels like a buzzing.

Doctor: How long have you been experiencing this? Is it constant, or does it come and go?

Patient: It’s been going on for several months now, and it’s pretty much constant.

Doctor: Is it in one ear or both?

Patient: Both ears.

Doctor: Have you noticed any hearing loss, dizziness, or problems with your balance?

Patient: No dizziness or balance problems, but I do feel like my hearing isn’t as sharp as before, especially in noisy places.

Doctor: Do you have any other symptoms like ear pain, drainage, or any history of ear infections?

Patient: No pain or drainage.

Doctor: Have you been exposed to loud noises recently, or do you take any medications that could affect your ears?

Patient: I work in a noisy environment, but I don’t take any medications regularly.

Doctor: Thank you for sharing this. The next steps will include a hearing test called an audiological exam to assess your hearing levels and help identify any underlying causes of your tinnitus. I may also ask you to move your jaw or neck to see if that changes your tinnitus, as sometimes muscle or joint issues can contribute. Depending on the findings, we might consider imaging or blood tests to rule out less common causes.

Patient: Will there be any treatment?

Doctor: Treatment for tinnitus focuses mostly on managing symptoms and any underlying conditions. Common approaches include sound therapy, cognitive behavioral therapy to help with coping, and sometimes hearing aids if hearing loss is significant. There is no single cure, but many people find relief with these methods.

Patient: Is tinnitus dangerous?

Doctor: Most tinnitus is not dangerous but can affect quality of life. If your tinnitus were associated with sudden hearing loss, neurological symptoms, or pulsatile sounds synchronized with your heartbeat, that would warrant more urgent investigation. Since you have chronic tinnitus with gradual hearing changes, we'll follow standard evaluation and management steps.

Patient: What can I do at home to help?

Doctor: Avoid loud noises and use ear protection when needed. Try to reduce stress and avoid caffeine or nicotine if they worsen your tinnitus. Maintaining good sleep hygiene also helps. We’ll work together to find the best approach for you.

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**Thyroid eye disease**

**DEFINITION / DESCRIPTION**

Thyroid eye disease (TED) is an inflammatory disorder that affects the tissues around your eyes, causing swelling, discomfort and other symptoms. It happens in some people who have autoimmune diseases that attack their thyroid gland. Most often, it happens in people who have Graves’ disease. In this case, your healthcare provider may call it Graves’ eye disease.

Although autoimmune diseases are lifelong, TED has distinct phases. It may be active for two years or more before the inflammation subsides. During this time, symptoms may come and go and may be mild to severe. TED is mild for most people. But when it’s severe, it can cause permanent damage, with lasting cosmetic and vision changes.

Ophthalmopathy and orbitopathy are two other terms for eye disease. Your healthcare provider may describe TED as:

* Thyroid ophthalmopathy
* Thyroid orbitopathy
* Graves’ ophthalmopathy
* Graves’ orbitopathy

**CAUSES**

Thyroid eye disease is an autoimmune disease. This means your immune system attacks your thyroid and eye tissues. Graves’ disease is a common cause, but other autoimmune thyroid diseases, like Hashimoto’s disease, can also cause it. Even people with normal thyroid function can develop TED.

In these diseases, your immune system creates antibodies that mimic thyroid hormones and attach to thyroid hormone receptors. These receptors are mostly within your thyroid itself, but some are in the tissues behind your eyes. So, the same antibodies that affect your thyroid can also affect your eyes.

#### **Thyroid eye disease risk factors**

You may be more likely to get thyroid eye disease (Graves’ eye disease) if you:

* Are female. Females are five times more likely to get Graves’ disease and Graves’ eye disease. (But when males get it, it’s more likely to be severe.)
* Have high or low thyroid hormone levels. Abnormal thyroid hormone levels can contribute to TED. These hormones stimulate the same receptors in your eye tissues that the antibodies do.
* Have had radioiodine therapy. RAI therapy is a standard treatment for overactive thyroid (hyperthyroidism), which Graves’ disease causes. But it may contribute to TED or make it worse.
* Smoke. Smoking and secondhand smoke exposure significantly raise your risk of thyroid eye disease. Smoke exposure also makes you more likely to have more severe symptoms that last longer. If you have TED, the best thing you can do for your disease is quit smoking

**SIGNS / SYMPTOMS**

Typical thyroid ophthalmopathy symptoms include:

* Bulging eyes (proptosis)
* Eye irritation
* Swollen and inflamed eyelids (blepharitis)
* Dry eyes or teary eyes
* Frequent blinking
* Light sensitivity (photophobia)
* Eye pain and headaches
* Difficulty moving your eyes
* Double vision (diplopia)

Symptoms normally affect both eyes, but sometimes, you may only notice symptoms in one eye.

**DIAGNOSIS METHODS**

A healthcare provider will be able to diagnose thyroid eye disease by doing a physical eye exam. They’ll be able to examine both your eyelids and your eyes.

If your healthcare provider thinks that you have thyroid eye disease, they’ll order blood tests to check if your thyroid hormone levels and antibodies are too high or too low.

Other tests your provider may request include:

* Ultrasound of the eyes
* Computed tomography (CT)
* Magnetic resonance imaging (MRI)

## **Management and Treatment**

The treatment you’ll need will depend on what stage the disease is in and how severe it is. Supportive treatments can help ease your symptoms while thyroid eye disease is active. These might include home remedies, over-the-counter or prescription medications. After the active phase has ended, you might need or want cosmetic surgery or vision correction surgery.

Supportive treatments for thyroid eye disease may include:

* Eye drops. During the active phase of TED, eye drops can help relieve your symptoms. Different types of eye drops can help with different symptoms, like dryness, redness or pain.
* Selenium supplements. Selenium deficiency increases TED symptoms.
* Scleral lenses. Similar to contact lenses, these lenses cover more of the surface of your eye, offering protection from the elements. You can also use them to apply medication.
* Vision aids. If you have double vision, your eye care provider may suggest wearing an eye patch or special eyeglasses with prisms to help correct it while thyroid eye disease is active.
* Thionamides. If you have hyperthyroidism (Graves’ eye disease), you may need medications to reduce your thyroid hormone levels. Examples include methimazole and propylthiouracil.
* Corticosteroids. When TED symptoms are more severe, healthcare providers prescribe a short course of corticosteroids to bring the inflammation down. You may take them by mouth or IV (intravenously, through a vein).
* Teprotumumab (Tepezza®). This newer drug became the first medication specifically approved to treat thyroid eye disease in the U.S. in 2020. It’s an alternative anti-inflammatory drug within the biologics category. More drugs like it are in development.
* Radiation therapy. Your eye doctor may recommend that you see a radiation oncologist to consider radiation, which reduces inflammation by killing the immune cells that are active behind your eyes.

Your healthcare team can offer alternative treatments if these don’t work for you. Rarely, some people may need surgery during the active phase of Graves’ eye disease. But in most cases, you and your team will consider surgery after your condition has stabilized. At this point, it’ll be easier to tell which appearance and vision changes are likely to be permanent.

#### **Surgery for Graves’ eye disease**

Surgery for thyroid eye disease may include:

* Orbital decompression surgery. This eye surgery may be necessary in the rare event that inflammation compresses your optic nerve. A surgeon will remove bone to reduce the compression. This surgery may also be done to reduce eye bulging after your condition has stabilized.
* Thyroidectomy. This treats severe hyperthyroidism. If your healthcare team can’t control your thyroid hormone levels with medications, they might need to remove your thyroid gland.
* Eyelid surgery. If your eyelids have retracted, you might need eyelid surgery to reposition them. This can improve your comfort as well as appearance and protect your eyes from exposure.
* Eye muscle surgery. You might need surgery on your eye muscles if they’re scarred and restricting your eye movement, or you have double vision because your eyes are misaligned.
* Oculoplastic surgery. Oculoplastic surgery adjusts the soft tissues and/or bones around your eyes. It can address cosmetic concerns like bulging eyes and bags around your eyes.
* Corneal transplant. If your cornea was badly damaged by exposure during the active phase of Graves’ eye disease, you might consider replacing it with a transplant to improve your vision.

**OUTLOOK / PROGNOSIS**

The outlook is good for most people with thyroid eye disease. Most have mild symptoms, which often resolve on their own. People over the age of 50 tend to have more severe symptoms. If you have a serious case of TED, you might need a combination of treatments to manage it. Some people have lasting changes to their eyes, which may require surgery

**POSSIBLE COMPLICATIONS**

Appearance and vision changes that occur with thyroid eye disease may improve after the active phase is over, but they don’t always. Scarring can prevent the tissues around your eyes from healing and returning to their normal shape. Sometimes, parts of your eyes that affect your vision are permanently damaged. Surgery can treat some of these effects.

Lasting appearance changes may include:

* Eyelid retraction
* Protruding eyes
* Baggy eyes
* Red eyes

Lasting vision changes may include:

* Blurry vision
* Double vision
* Severe vision loss

**WHEN TO SEE A DOCTOR / RED FLAG**

When you have thyroid eye disease, it’s important to let your healthcare team know if your symptoms worsen. You should also let them know if you notice any changes in your vision. Some changes might need urgent treatment. Call right away if you notice:

* Your field of vision has narrowed or closed off
* Colors appear differently than they used to
* Sudden, severe eye pain

**DIFFERENTIAL DIAGNOSIS**

Non-specific orbital inflammatory disease (NSOID): Bilateral proptosis with lacrimal gland enlargement. EOM enlargement involves the muscle belly and the tendinous origin. The lacrimal gland enlarges and prolapses out of the lacrimal fossa. It can be associated with systemic autoimmune diseases like polymyositis, dermatomyositis, and IGG4-related diseases. Serology and soft tissue biopsy are diagnostic.

Lymphoma: Bilateral proptosis with lymphadenopathy. Hemogram and blood counts are usually normal. Orbital CT scan shows bilateral diffuse soft tissue enlargement with the erosion of the bony orbit. Histopathology and immunohistochemistry features on the soft tissue biopsy are diagnostic.

Blow-out fracture of the orbit: Altered globe position and limitation of EOM movements. History of trauma is followed by enophthalmos and restriction of elevation. An orbital CT scan shows the bony defect with entrapped soft tissues.

Amyloidosis: Bilateral proptosis with neuropathy. Soft tissue biopsy shows eosinophilic material on histopathology with birefringence on polarised light microscopy

**EPIDEMIOLOGY**

The prevalence of thyroid eye disease is approximately 50% among GD patients in the Caucasian population. The annual incidence is 16.0 in 100,000 for females and 2.9 in 100,000 for males in the US population.

**Risk Factors**

1. Ethnicity: The African-American population exhibits the maximal risk, followed by the White race and Asian populations.
2. Age: TED shows a bimodal peak incidence. It occurs in age groups of 40 to 44 years and 60 to 64 years in females, and ages of 45 to 49 years and 65 to 69 years in males. It is more severe in older patients with higher chances of restrictive myopathy and dysthyroid optic neuropathy (DON).
3. Gender: There is a female preponderance due to a higher risk of autoimmune diseases. Males cases have more severe ocular involvement and worse outcomes.
4. Genetics: CTLA-4, HLA-DRB-1, and TNF-a genes - are most often associated with TED.
5. Systemic associations: Autoimmune disorders like pernicious anemia, systemic lupus erythematosus, Addison’s disease, vitiligo, coeliac disease, and rheumatoid arthritis - a higher risk of TED.
6. Environmental factors: Smoking is strongly associated with the TED incidence.
7. Dysthyroid status: At the time of diagnosis, 90% of TED cases are hyperthyroid, 6% euthyroid, 3% have Hashimoto thyroiditis, and 1% are hypothyroid
8. Radioactive iodine therapy (RAIT): causes exacerbation in 24% of TED cases.
9. Stress: Psychological stress can aggravate TED by rebound immune hyperactivity following prolonged corticosteroid-induced immune suppression.
10. Pregnancy: New onset or worsening of TED occurs in 30% of GD cases in the post-partum period.
11. Others: Trauma can be a stimulus for activating an autoimmune cascade in the orbit. High serum cholesterol may also be a risk factor for TED

**PREDEFINED Q & A SETS**

Q1: What is thyroid eye disease?  
A1: Thyroid eye disease (TED) is an autoimmune condition causing inflammation and swelling of the tissues around the eyes, including the muscles and fat within the orbit. It is often associated with Graves’ disease (an autoimmune thyroid disorder), but can also occur in people with normal or low thyroid hormone levels.

Q2: What are common symptoms of TED?  
A2: Symptoms include bulging eyes (proptosis), eyelid swelling and retraction, eye redness, gritty or irritated eyes, double vision, light sensitivity, pain or pressure around the eyes, and sometimes blurred vision. Changes can affect one or both eyes.

Q3: How is TED diagnosed?  
A3: Diagnosis is based on clinical eye examination, thyroid function tests, and antibody testing. Imaging studies such as CT or MRI of the orbits help identify muscle enlargement and inflammation.

Q4: Who is at risk for TED?  
A4: TED primarily affects adults with Graves’ disease but can also occur in euthyroid (normal thyroid levels) or hypothyroid patients. Risk factors include smoking, previous radioactive iodine treatment for thyroid disease, and male gender (associated with more severe disease).

Q5: Can TED develop without thyroid hormone abnormalities?  
A5: Yes, some people have “euthyroid” TED with typical eye symptoms but normal thyroid function tests. They require regular monitoring as thyroid dysfunction may develop later.

Q6: How is TED treated?  
A6: Treatment depends on disease severity and activity:

* Mild cases may improve with lubricating eye drops, sunglasses, and selenium supplements.
* Moderate to severe inflammation is treated with corticosteroids (often intravenous).
* A newer drug, Teprotumumab (Tepezza), targets molecular pathways to reduce inflammation and eye bulging.
* Surgery may be needed for severe proptosis, eyelid problems, or vision-threatening optic nerve compression.

Q7: Will controlling thyroid levels cure TED?  
A7: Managing thyroid hormone levels is important for overall health but does not usually cure TED. Eye symptoms may persist or worsen even when thyroid function is normalized.

Q8: What is the outlook for people with TED?  
A8: Most cases are mild and improve over time. Severe cases risk vision loss if untreated but modern therapies including steroids, biologics, and surgery improve outcomes. Smoking cessation significantly reduces risk and severity.

Q9: When should I see a doctor?  
A9: Prompt evaluation is needed if you notice eye bulging, pain, vision changes, double vision, or redness, especially if you have thyroid disease.

**DOCTOR-PATIENT CONVERSATIONS**

Doctor: Good morning. What brings you in today?

Patient: I’ve noticed that my eyes look swollen and seem to be bulging out. Sometimes they feel dry and irritated, and I’m starting to have double vision.

Doctor: Thank you for sharing that. These symptoms can be related to an autoimmune condition called thyroid eye disease, or TED, which often occurs along with thyroid problems like Graves’ disease. Have you had any thyroid issues diagnosed previously?

Patient: No, I haven’t had any thyroid problems diagnosed so far.

Doctor: That’s okay. TED can sometimes be the first sign of thyroid dysfunction. We’ll start by checking your thyroid hormone levels and ordering some imaging of your eyes to see if the muscles and tissues around your eyes are inflamed or swollen.

Patient: What causes this disease? Is it serious?

Doctor: TED is caused by the immune system mistakenly attacking the tissues around your eyes, leading to inflammation, swelling, and sometimes pressure on the optic nerve. If untreated, it can affect your vision, and the changes in your eyes can be uncomfortable. The severity varies—a lot of patients have mild symptoms that improve, but others may need treatment to reduce inflammation or protect vision.

Patient: What kind of treatments are available?

Doctor: For mild cases, we often recommend lubricating eye drops, protecting your eyes from dryness and irritants, and sometimes selenium supplements. For more active or severe inflammation, corticosteroids or newer medications like Teprotumumab can reduce swelling and bulging. Occasionally, surgery is needed to relieve pressure, correct eyelid problems, or improve appearance.

Patient: How long will it take to get better?

Doctor: The course can vary. Some symptoms improve over months to years. With treatment, we aim to control inflammation early, prevent complications, and preserve your vision. We’ll monitor your progress closely and adjust your treatment plan as needed.

Patient: Is there anything I can do to help myself?

Doctor: Yes, quitting smoking is very important because smoking worsens TED. Also, keep your eyes well lubricated, wear sunglasses to reduce light sensitivity, avoid rubbing your eyes, and follow up regularly. Managing your thyroid function with your primary doctor or endocrinologist is also key to overall control.

Patient: Thank you, doctor. I appreciate the information and help.

Doctor: You’re welcome. Don’t hesitate to reach out if you notice worsening vision, pain, or increased eye bulging. We will work together to manage this.

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**Temporo-mandibular joint (TMJ) pain**

ALTERNATIVE NAMES

**DEFINITION / DESCRIPTION**

The temporomandibular (tem-puh-roe-man-DIB-u-lur) joint, also called TMJ, acts like a sliding hinge. It connects the jawbone to the skull. There is one joint on each side of the jaw. TMJ disorders — a type of temporomandibular disorder or TMD — can cause pain in the jaw joint and in the muscles that control jaw movement.

The exact cause of TMJ disorder is often hard to determine. The pain may be due to a mix of factors, including habits such as teeth clenching, gum chewing and nail biting; stress; and painful conditions that occur along with TMJ disorder such as fibromyalgia, osteoarthritis or jaw injury. The habit of teeth clenching or grinding also is known as bruxism.

Most of the time, the pain and discomfort related to TMJ disorders lasts for only a limited time. Self-managed home care, physical therapy for the jaw and the use of a mouth guard can be effective in treating symptoms of TMJ disorder. Surgery is usually a last resort after conservative measures have failed. But surgical treatments may help some people with TMJ disorders.

**CAUSES**

The temporomandibular joint combines a hinge action with sliding motions. The parts of the bones that work together in the joint are covered with cartilage and separated by a small shock-absorbing disk. This disk usually keeps the movement smooth.

Painful TMJ disorders can happen if:

* The disk erodes or moves out of its proper relationship between the ball and socket of the joint.
* Sprain or strain occurs in the ligaments or soft tissues associated with TMJ disorders.
* Arthritis damages the joint's cartilage.
* A blow or another impact damages the joint.
* Jaw muscles are associated with TMJ spasms.

Many times, the cause of TMJ disorder has a variety of causes and is hard to identify.

**RISK FACTORS**

Factors that may raise the risk of getting TMJ disorders include:

* Different types of arthritis, such as rheumatoid arthritis or osteoarthritis.
* Jaw injury.
* Habits such as gum chewing, nail biting, and grinding or clenching of teeth.
* Certain connective tissue diseases.
* Stress, post-traumatic stress disorder, anxiety or depression.
* Conditions such as fibromyalgia, ankylosing spondylitis and sleep disturbances.
* Smoking.

**SIGNS / SYMPTOMS**

Symptoms of TMJ disorders may include:

* Pain or tenderness in the jaw.
* Pain in one or both temporomandibular joints.
* Aching pain in and around the ear.
* Having a hard time chewing or pain while chewing.
* Aching facial pain.
* Locking of the joint, making it hard to open or close your mouth.
* Headache.
* Neck pain.
* Eye pain.
* Tooth pain that occurs along with jaw tenderness.

TMJ disorders also can cause a clicking sound or grating sensation when you open your mouth or chew. But if there's no pain or limitation of movement related to your jaw clicking, it's likely that you won't need treatment.

**DIAGNOSIS METHODS**

Your healthcare professional will likely discuss your symptoms and study your jaw by:

* Listening to and feeling your jaw when you open and close your mouth.
* Watching the range of motion in your jaw.
* Pressing on areas around your jaw to find sites of pain or discomfort.

If your healthcare professional thinks there is an issue, you may need:

* Dental X-rays to look at your teeth and jaw.
* CT scan to show detailed images of the bones involved in the joint.
* MRI to show issues with the joint's disk or surrounding soft tissue.

TMJ arthroscopy is sometimes used to diagnose a TMJ disorder. During TMJ arthroscopy, your healthcare professional inserts a small thin tube called a cannula into the joint space. Then a small camera called an arthroscope is inserted to look at the area and help find a diagnosis.

TMJ arthroscopy also is sometimes used to treat TMJ disorder. The procedure can help with therapy, such as releasing scar tissue and removing inflammatory soft tissue and byproducts to improve TMJ symptoms and help the jaw move without pain.

**TREATMENT OPTIONS**

Sometimes the symptoms of TMJ disorders may go away without treatment. If your symptoms don't go away, your healthcare professional may recommend treatment options, often more than one to be done at the same time.

### **Medications**

Along with other treatments that don't involve surgery, these medicine options may ease the pain related to TMJ disorders:

* **Pain relievers and anti-inflammatories.** If pain medicines available without a prescription aren't enough to relieve TMJ pain, your healthcare professional may prescribe stronger pain relievers for a limited time, such as prescription-strength ibuprofen (Advil, Motrin IB, others).
* **Tricyclic antidepressants.** These medicines, such as nortriptyline (Pamelor), are used mostly for depression. But in low doses, they're sometimes used to ease pain, control bruxism and help with sleeplessness.
* **Muscle relaxants.** These medicines are sometimes used for a few days or weeks to ease pain caused by TMJ disorders that muscle spasms create.

### **Therapies**

Therapies for TMJ disorders that don't involve drugs include:

* **Self-care.** To ease symptoms, apply heat or ice, depending on the type of TMJ symptoms. Avoid teeth clenching, gum chewing and nail biting. Practice good resting jaw posture with the tongue gently rested on the palate, teeth apart and jaw in a relaxed position.
* **Oral splints or mouth guards.** Often, people with jaw pain will benefit from wearing a soft or firm device inserted over their teeth. The reasons why these devices help are not well understood.
* **Physical therapy.** Along with exercises to stretch and strengthen jaw muscles, treatments might include ultrasound and transcutaneous electrical nerve stimulation, also known as TENS. Also, moist heat and stretching are effective in repeated applications throughout the day.
* **Counseling.** Education and counseling can help you learn more about the factors and behaviors that may worsen your pain, so you can change your behavior. Examples include teeth clenching or grinding, leaning on your chin, or biting fingernails.

### **Surgical or other procedures**

When other methods don't help, your healthcare professional might suggest procedures such as:

* **Arthrocentesis.** Arthrocentesis (ahr-throe-sen-TEE-sis) is a minimally invasive procedure that involves inserting small needles into the joint so that fluid can flow through the joint to remove debris and inflammatory byproducts.
* **Injections.** In some people, corticosteroid injections into the joint may help. Rarely, injecting botulinum toxin type A (Botox, others) into the jaw muscles used for chewing may ease pain related to TMJ disorders.
* **TMJ arthroscopy.** Sometimes arthroscopic surgery can be as effective for treating various types of TMJ disorders as open-joint surgery. A small thin tube called a cannula is placed into the joint space. Then an arthroscope is inserted, and small surgical instruments are used for surgery. TMJ arthroscopy has fewer risks and complications than open-joint surgery does. But it has some limitations too.
* **Modified condylotomy.** Modified condylotomy (kon-dih-LOT-uh-mee) addresses the TMJ indirectly, with surgery on the mandible but not in the joint itself. It may ease pain and locking.
* **Open-joint surgery.** If your jaw pain does not go away with more-conservative treatments and a structural issue in the joint appears to be causing the pain, your healthcare professional may suggest open-joint surgery to repair or replace the joint. But open-joint surgery involves more risks than other procedures do. Think about this procedure very carefully after discussing the pros and cons with your healthcare professional.

If your healthcare professional recommends surgery or other procedures, be sure to talk about the possible benefits and risks. Also, ask about all your options.

**Lifestyle and home remedies**

Become more aware of habits linked with stress — clenching your jaw, grinding your teeth or chewing pencils — so you won't do them as often. These tips may help you reduce symptoms of TMJ disorders:

* **Avoid overuse of jaw muscles.** Eat soft foods. Cut food into small pieces. Steer clear of sticky or chewy food. Don't chew gum.
* **Practice gentle stretching and massage.** A physical therapist or other healthcare professional can show you how to do exercises that stretch and strengthen your jaw muscles and how to massage the muscles yourself.
* **Apply heat or cold.** Applying warm, moist heat or an ice pack to the side of your face may ease pain. Acute pain is best treated with an ice pack. Chronic dull pain is best treated with heat therapy. Apply heat or cold for 15 to 20 minutes several times a day. This approach, combined with stretching, has been found to be very effective.
* **Adjust your diet.** Eating soft foods or smaller pieces of food, not opening your mouth too wide, and eating foods using utensils such as a spoon or fork may ease symptoms. Chewing foods with both sides of the mouth and not just one side also may ease symptoms.

**Alternative medicine**

Complementary and alternative medicine techniques may help manage ongoing pain often related to TMJ disorders. Examples include:

* **Acupuncture.** A specialist trained in acupuncture treats ongoing pain by inserting hair-thin needles at specific locations on your body.
* **Relaxation techniques.** Consciously slowing your breathing and taking deep, regular breaths can relax tense muscles. In turn, this can lessen pain.
* **Biofeedback.** Electronic devices that track the tightness of specific muscles can help you practice effective relaxation techniques.

**PREVENTION TIPS**

You can’t always prevent TMD because some risk factors are unavoidable. But there are things you can do to reduce your risk:

* Wear a mouth guard at night if you clench or grind your teeth.
* Wear a mouth guard while playing contact sports.
* Practice good posture.
* Practice meditation, mindfulness or other stress reduction techniques.

**OUTLOOK / PROGNOSIS**

TMJ dysfunction affects everyone differently. For some, it’s a temporary issue that goes away in a week or two. For others, it’s a chronic condition that negatively impacts quality of life.

If you have frequent jaw pain, facial pain, headaches or other TMJ symptoms, tell a healthcare provider. They can help you find an effective treatment.

#### **How long does TMJ dysfunction last?**

It depends. In some cases, TMD goes away on its own in one to two weeks. But in severe instances, it can last several months or even years. TMJ disorder can be short term (acute) or long term (chronic).

### **Can TMJ disorder be cured?**

Yes, it’s possible to cure TMJ dysfunction with appropriate treatment and management.

**WHEN TO SEE A DOCTOR / RED FLAG**

If you have persistent TMJ pain, popping jaws or other symptoms, schedule a visit with a healthcare provider. They can help you find out why your jaws are aching and determine what kind of treatment you need.

**common drug types with their typical side effects:**

1. Nonsteroidal Anti-Inflammatory Drugs (NSAIDs):

Examples: ibuprofen (Advil, Motrin), naproxen, aspirin, diclofenac, celecoxib (Celebrex)

Purpose: Reduce pain and inflammation in the TMJ and surrounding muscles.

Side Effects: Gastrointestinal upset (nausea, ulcers), increased risk of bleeding, kidney function impairment, elevated blood pressure in some cases. Long-term use may exacerbate hypertension and renal issues.

2. Tricyclic Antidepressants (TCAs):

Examples: amitriptyline, nortriptyline, desipramine

Purpose: Used in low doses to manage chronic TMJ pain, help control bruxism (teeth grinding), and improve sleep disturbances.

Side Effects: Constipation, urinary retention, dizziness, orthostatic hypotension (low blood pressure when standing), sedation. TCAs are contraindicated in elderly patients and those with certain cardiac conditions.

3. Muscle Relaxants:

Purpose: To alleviate muscle spasms contributing to TMJ pain, typically used short-term (days to weeks).

Side Effects: Drowsiness, dizziness, dry mouth, and potential dependency with prolonged use.

4. Corticosteroids:

Delivery: Sometimes given as intra-articular (into the joint) injections or orally.

Purpose: Potent anti-inflammatory effect to reduce severe inflammation and pain.

Side Effects: With systemic oral use—weight gain, increased blood sugar, osteoporosis, immunosuppression. Injections generally have fewer systemic effects but risks include local infection or tissue damage.

5. Other Medications:

Selective Serotonin Reuptake Inhibitors (SSRIs) and Serotonin-Norepinephrine Reuptake Inhibitors (SNRIs): Sometimes prescribed, but in TMJ patients, SSRIs may increase bruxism and exacerbate pain. Side effects include nausea, dry mouth, insomnia, sexual dysfunction, and anxiety.

Opioids: Used only in severe, refractory cases due to risk of dependence. Side effects include sedation, constipation, nausea, and addiction potential.

Vitamin B Complex: May have analgesic effects when combined with NSAIDs.

Beta-blockers (e.g., propranolol): Emerging evidence suggests they may help manage TMD pain by modulating sympathetic nerve activity

**GENOMIC DATA**

* Several genes have been identified as associated with TMD, including IL1B (interleukin 1 beta), IL6, CCL2, VEGFA (vascular endothelial growth factor A), and ESR1 (estrogen receptor 1). These genes relate largely to *inflammation*, *immune response*, and *bone/joint biology*, indicating that inflammatory pathways are crucial in TMD.
* A systematic review found over 100 genes associated with TMD, emphasizing it as a complex, multifactorial disorder with a genetic contribution to its pathology.
* Specific single nucleotide polymorphisms (SNPs) have also been linked to TMD pain and susceptibility, such as variants in the COMT gene (related to pain sensitivity) and other genes including MTRR. Some variants may exhibit sex-specific effects on TMD risk.
* Genetic differences in people influence how the temporomandibular joint develops morphologically and responds to injury or stress, with certain genes regulating morphogenesis and growth of the TMJ complex.
* Overall, genetic research suggests that TMD is not just a mechanical or environmental disorder but has important genetic and molecular bases involving immune, inflammatory, and pain pathways. These findings may help develop better diagnostic biomarkers and personalized treatment in the future.
* The percentage of TMD cases estimated to have a genetic component is substantial; for example, about 27% of TMJ pain cases have a hereditary basis, indicating a meaningful role for inherited factors

**DIFFERENTIAL DIAGNOSIS**

## Diagnostic criteria for pain-related temporomandibular disorders

Disorders in this category include the following:

* Myalgia
* Local myalgia
* Myofascial pain
* Myofascial pain with referral
* Arthralgia

For diagnosis, both the history and physical examination criteria must be met. With all those disorders, the two criteria for history are pain in the jaw, temple, ear, or in front of the ear and modification of the pain with jaw movement, function, or parafunction. Examination must confirm that the pain is located in the temporalis and/or masseter muscles. Additional essential exam criteria vary as follows:

* Myalgia: The patient reports familiar pain on provocation testing with palpation of the temporalis and/or masseter muscles, or on maximum unassisted or assisted opening movement(s)
* Local myalgia: The patient reports familiar pain on palpation of the temporalis and/or masseter muscles that is localized to the site of palpation
* Myofascial pain: The patient reports familiar pain on palpation of the temporalis and/or masseter muscles; the pain spreads beyond the site of palpation but remains within the boundary of the muscle
* Myofascial pain with referral: The patient reports familiar pain on palpation of the temporalis and/or masseter muscles and reports pain at a site beyond the boundary of the muscle being palpated

Unlike the above disorders, which involve pain of muscle origin, arthralgia is pain of joint origin. The history criteria are the same as for those disorders, but the exam criteria are that the exam must confirm that the pain is located in the area of the temporomandibular joint (TMJ) and that the patient reports familiar pain in the TMJ with palpation of or around the lateral pole and/or with maximum unassisted or assisted opening, right or left lateral movements, or protrusive movement(s).

The final pain-related TMD is headache attributed to TMD. The patient must already have an established diagnosis of a pain-related TMD. History criteria are headache of any type in the temple, in the last 30 days, and modification of the headache with jaw movement, function, or parafunction. The exam criteria require confirming the location of the headache in the area of the temporalis muscle(s) and the patient reports that palpation of the temporalis muscle(s) and/or maximum unassisted or assisted opening, right or left lateral, or protrusive movement(s) provokes the familiar headache in the temple area.

## **Diagnostic criteria for intra-articular temporomandibular disorders**

Disorders in this category include the following:

* Disk displacement disorders (four types)
* Degenerative joint disease
* Subluxation

Diagnosis of subluxation is based on history alone; with the other intra-articular TMD, both the history and exam criteria must be met. When any of those diagnoses need to be confirmed, imaging criteria apply.

*Disk displacement with reduction:*

* History: In the last 30 days, any TMJ noise with jaw movement or function, or patient report of any noise during the exam
* Exam: Clicking, popping, and/or snapping noise during both opening and closing movements, detected with palpation during at least one of three repetitions of jaw opening and closing; or clicking, popping, and/or snapping noise detected with palpation during at least one of three repetitions of opening or closing movements and at least one of three right or left lateral or protrusive movement(s)
* Imaging (reference standard for this diagnosis): TMJ MRI shows that in the maximum intercuspal position, the posterior band of the disk is located anterior to the 11:30 position and the intermediate zone of the disk is located between the condylar head and the articular eminence.

*Disk displacement with reduction with intermittent locking:*

* History: In the last 30 days, any TMJ noise with jaw movement or function, or patient report of any noise during the exam and the jaw locks with limited mouth opening, even for a moment, and then unlocks
* Exam: Clicking, popping, and/or snapping noise during both opening and closing movements, detected with palpation during at least one of three repetitions of jaw opening and closing; or clicking, popping, and/or snapping noise detected with palpation during at least one of three repetitions of opening or closing movements and at least one of three right or left lateral or protrusive movement(s)
* Imaging: Criteria are the same as for disk displacement with reduction, if intermittent locking is not present at the time of imaging. If locking occurs during the imaging study, it renders an imaging-based diagnosis of disk displacement without reduction, and clinical confirmation of reversion to intermittent locking is needed.

*Disk displacement without reduction with limited opening:*

* History: Jaw locked so that the mouth will not open all the way and limitation in jaw opening severe enough to limit jaw opening and interfere with the ability to eat
* Exam: Maximum assisted opening (passive stretch) movement including vertical incisal overlap < 40 mm
* Imaging: On TMJ MRI, in the maximum intercuspal position, the posterior band of the disk is located anterior to the 11:30 position and the intermediate zone of the disk is anterior to the condylar head and on full opening, the intermediate zone of the disk is located anterior to the condylar head

*Disk displacement without reduction without limited opening:*

* History: In the past, the jaw locked so that the mouth will not open all the way and limitation in jaw opening was severe enough to limit jaw opening and interfere with the ability to eat
* Exam: Maximum assisted opening (passive stretch) movement including vertical incisal overlap ≥ 40 mm
* Imaging: TMJ MRI criteria are the same as for disk displacement without reduction with limited opening

*Degenerative joint disease:*

* History: In the last 30 days, any TMJ noise present with jaw movement or function, or patient report of any noise present during the exam.
* Exam: Crepitus detected with palpation during at least one of the following: opening, closing, right or left lateral, or protrusive movement(s)
* Imaging (reference standard for this diagnosis): TMJ MRI criteria positive for at least one of the following: subchondral cyst(s), erosion(s), generalized sclerosis or osteophyte(s). Note that flattening and/or cortical sclerosis are considered indeterminant findings for this diagnosis.

*Subluxation:*

* History: In the last 30 days, jaw locking or catching in a wide-open mouth position, even for a moment, so the patient could not close from the wide-open position, and inability to close the mouth from a wide-open position without a self-maneuver
* Exam: Not required for this diagnosis, but when the disorder is present clinically, exam is positive for inability to return to a normal closed-mouth position without the patient performing a manipulative maneuver
* Imaging: Condyle positioned beyond the height of the articular eminence, with the patient unable to close his/her mouth

## 

## **Differential Diagnoses**

* Acute Otitis Media
* Cluster Headache
* Dental Abscess
* Giant Cell Arteritis (Temporal Arteritis)
* Gout and Pseudogout
* Lyme Disease
* Mandible (TMJ) Dislocation
* Mandibular Fracture Management in the ED
* Migraine Headache
* Myopathies
* Rheumatoid Arthritis (RA)
* Tension Headache
* Trigeminal Neuralgia

**EPIDEMIOLOGY**

### Frequency

United States

Currently, an estimated 10 million people have TMJ disorders, and roughly 25% of the population have symptoms at some point in their lives.

### Mortality/Morbidity

The morbidity of the disorder is related to significant pain on movement of the jaw. While some patients' symptoms may resolve within weeks, others may have chronic symptoms that persist even with extensive therapy.

One study by Rammelsberg et al followed 235 patients over 5 years.In this study, roughly one third of patients had complete resolution of pain, one third had continuous pain over the 5 years, and one third had recurrent episodes with periods of remission.

### Race-, Sex-, and Age-related Demographics

See the list below:

* No apparent association with race exists.
* Female-to-male ratio is roughly 4:1.
* Highest incidence of TMJ syndrome is in adults aged 20-40 years
* TMJ syndrome is found infrequently in the pediatric population

**PREDEFINED Q & A SETS**

Q1: What is a TMJ disorder?  
A1: TMJ disorder refers to problems affecting the temporomandibular joint — the hinge connecting the jawbone to the skull — and the muscles controlling jaw movement. It can cause jaw pain, difficulty chewing, clicking or popping sounds, and limited jaw movement.

Q2: What causes TMJ disorders?  
A2: Causes include jaw injury or trauma, arthritis, teeth grinding or clenching (bruxism), stress leading to muscle tension, poor bite alignment, or connective tissue diseases. Sometimes, the cause may be unknown.

Q3: What are the common symptoms of TMJ disorders?  
A3: Symptoms often include:

* Jaw pain or tenderness
* Clicking, popping, or grating sounds when opening or closing the mouth
* Difficulty or discomfort when chewing
* Limited jaw movement or locking
* Earache or ear fullness
* Headaches or facial pain
* Swelling on the side of the face

Q4: How is TMJ disorder diagnosed?  
A4: Diagnosis is based on medical history and physical examination of the jaw, face, neck, and head. Imaging tests such as X-rays, CT scans, or MRI may be used to evaluate joint structure and rule out other conditions.

Q5: Are TMJ disorders permanent?  
A5: Many TMJ disorders improve with conservative treatment and do not cause permanent damage. However, some cases can become chronic if untreated.

Q6: What treatment options are available for TMJ disorders?  
A6: Treatment usually starts with conservative approaches such as:

* Pain relievers and anti-inflammatory medications
* Muscle relaxants
* Use of oral splints or mouthguards to reduce teeth grinding
* Physical therapy and jaw exercises
* Stress management  
  More severe cases may require corticosteroid injections, dental treatments to correct bite problems, or rarely, surgery.

Q7: Can lifestyle changes help with TMJ symptoms?  
A7: Yes. Avoiding hard or chewy foods, practicing good posture, applying warm or cold compresses, reducing stress, and avoiding extreme jaw movements can help reduce symptoms.

Q8: When should I see a doctor about TMJ pain?  
A8: Seek medical advice if you have persistent jaw pain, difficulty opening or closing your mouth, jaw locking, or if symptoms worsen despite self-care.

Q9: Can TMJ disorder cause headaches or ear problems?  
A9: Yes. TMJ disorders can cause referred pain leading to headaches, earaches, tinnitus (ringing in the ears), or a feeling of fullness in the ears.

**DOCTOR-PATIENT CONVERSATIONS**

Doctor: Good morning. What brings you in today?

Patient: I’ve been having pain and clicking in my jaw, especially when I chew or open my mouth wide. It sometimes feels stiff and sore.

Doctor: Thank you for telling me. How long has this been going on? Is the pain constant or does it come and go?

Patient: It started a few months ago and tends to get worse when I’m stressed or eat hard foods, but sometimes it’s just a dull ache.

Doctor: Do you notice any other symptoms like headaches, earaches, or difficulty opening your mouth fully?

Patient: Yes, I do have headaches around my temples and sometimes my ears feel full and a bit painful.

Doctor: Have you had any recent injuries to your jaw or dental work?

Patient: No injuries, but I do grind my teeth at night.

Doctor: That’s helpful to know. I’m going to examine your jaw now by feeling the joint and muscles, checking how wide you can open your mouth, and listening for any noises like clicking or popping.

(Doctor performs the physical exam.)

Doctor: I did notice some tenderness and clicking in the joint, and your jaw opening is a bit limited. Based on your symptoms and exam, it looks like you have a temporomandibular joint disorder, or TMJ disorder.

Patient: What causes TMJ disorder, and is it serious?

Doctor: TMJ disorders can be caused by muscle tension, jaw joint inflammation, teeth grinding, misalignment, or sometimes arthritis. It’s usually not serious, but it can cause discomfort and affect your quality of life if untreated.

Patient: What can be done to improve it?

Doctor: Treatment usually starts conservatively:

* Avoid hard or chewy foods
* Apply warm compresses
* Use anti-inflammatory medications or muscle relaxants if needed
* Consider a nightguard to prevent teeth grinding
* Physical therapy and jaw exercises may help improve movement and reduce pain

If symptoms persist, we may consider imaging like an MRI to look at the joint structure or referral to a specialist. Surgery is rarely needed.

Patient: How long will it take to get better?

Doctor: Many people improve within weeks to months with conservative care. I will provide you with exercises and advice, and we will follow up to see how you’re doing.

Patient: Thank you, doctor.

Doctor: You’re welcome. Please contact us if your symptoms worsen or if you have trouble opening your mouth.

REFERENCES:

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**Turbinate hypertrophy**

**ALTERNATIVE NAMES**

* Inferior turbinate hypertrophy (most common, since inferior turbinates are usually involved)
* Nasal turbinate hypertrophy
* Nasal turbinate enlargement
* Inferior turbinate enlargement
* Nasal cavity turbinate hypertrophy
* Hypertrophic turbinates
* Turbinate swelling
* Enlarged turbinates

**DEFINITION / DESCRIPTION**

Turbinate hypertrophy refers to an excessive growth or enlargement of the bony structures located inside of the nose. These structures are covered with a special skin called mucosa and they help filter, warm, and humidify the air as you breath. The mucosa naturally swells during the normal nasal cycle, when you lie down and in response to allergens or other irritants. If these turbinates become enlarged, breathing can become more difficult.

**CAUSES**

Turbinate hypertrophy is caused when the mucosa becomes enlarged or swollen. This can be chronic or acute and can be caused by conditions including:

* Hormonal changes
* Medications
* Rhinitis
* Sinus infections
* The common cold

**SIGNS / SYMPTOMS**

Symptoms of turbinate hypertrophy can include:

* Congestion or congestion when lying down
* Difficulty breathing
* Noisy or loud breathing
* Stuffiness and increased nasal drainage

**DIAGNOSIS METHODS**

* Physical Examination and Rhinoscopy:  
  The specialist visually inspects the nasal cavities using rhinoscopy, which involves inserting a rhinoscope or a small endoscope into the nostrils to directly view the turbinates and nasal passages. This is a minimally invasive procedure lasting about 5 to 10 minutes and allows assessment of the size and swelling of the turbinates.
* Nasal Endoscopy (Fiber Optic Scope):  
  A more detailed evaluation may involve using a fiber optic flexible scope with a small camera to view deeper into the nasal cavity and sinus openings. This helps the doctor assess turbinate size, mucosal condition, and rule out other causes of nasal obstruction.
* Imaging Studies:  
  In some cases, especially if surgery is being considered or to evaluate related sinus issues, imaging such as CT scans may be ordered. CT scans provide detailed images of the nasal structures and can help identify turbinate hypertrophy versus other structural causes of obstruction.
* Additional Tests:  
  Depending on clinical suspicion, allergy testing might be done to identify allergic causes contributing to turbinate swelling. Measurement of airflow or nasal obstruction through objective tests (e.g., rhinomanometry) may also be performed but is less common in routine diagnosis.
* Novel Diagnostic Approaches:  
  Advanced imaging like Narrow Band Imaging (NBI) endoscopy is a newer method showing promise in assessing vascular patterns of hypertrophic turbinates, aiding in distinguishing chronic enlargement.

**TREATMENT OPTIONS**

Treating turbinate hypertrophy will depend on the specific cause. See your primary care provider or one of our ENT specialists to diagnose what may be causing the turbinate hypertrophy and come up with an appropriate treatment plan.

### **Medicine**

Medications are often the first step to help reduce your symptoms. Medications can include:

* Allergy medications if your symptoms are caused by allergic rhinitis
* Nasal antihistamine sprays
* Nasal saline sprays or rinses
* Steroid sprays

### **Surgery**

If medications do not help improve your symptoms, surgery may be used to decrease the size of the turbinates. The surgery is usually performed through the nostrils and can be performed under local or general anesthesia as an outpatient procedure. The procedure may be combined with other procedures to reduce nasal obstruction like a septoplasty to address a deviated septum.

**PREVENTION TIPS**

* Minimizing exposure to environmental allergens and irritants:
  + Remove or reduce dust, pet dander, pollen, and mold from your home environment by regular cleaning, vacuuming, and using dust-proof covers on mattresses and pillows.
  + Use HEPA air filters indoors, especially in bedrooms, to reduce airborne allergens.
  + Avoid smoking and exposure to secondhand smoke, particularly indoors.
  + Control indoor humidity to prevent mold growth.
* Managing allergies effectively:
  + Use allergy medications such as antihistamines (e.g., cetirizine, loratadine) as recommended by your doctor to reduce allergic inflammation.
  + Allergy immunotherapy (allergy shots) may be considered for long-term immune modulation.
* Avoiding overuse of nasal decongestant sprays:
  + Nasal decongestants should be used only for short periods (no more than 3 days), as prolonged use can worsen turbinate swelling (rebound congestion).
* Nasal hygiene:
  + Regular use of saline nasal sprays or rinses to clear irritants from the nasal passages can help maintain turbinate health.
* Prompt treatment of upper respiratory infections and rhinitis:
  + Managing infections early may reduce inflammation that contributes to turbinate hypertrophy.

**OUTLOOK / PROGNOSIS**

* Without treatment Untreated turbinate hypertrophy can lead to worsening symptoms such as severe difficulty breathing through the nose, which can impact restful sleep. It can also increase the frequency of sinus infections .
* With treatment Many people experience significant symptom reduction with appropriate medical or surgical treatments .
  + Medical Management Initial treatment often involves steroid nasal sprays and/or antihistamines . Silver nitrate cauterization has also shown effectiveness in improving nasal breathing with fewer complications .
  + Surgical Interventions If conservative treatments are not effective, turbinate reduction surgery may be necessary .
    - Success Rates The success rate for turbinate reduction surgery is generally high, with approximately 80-90% of patients experiencing significant relief from nasal obstruction and other symptoms .
    - Recovery Recovery times vary depending on the type of procedure. Less invasive procedures have quick recoveries, often within three weeks . More invasive surgeries may take three to six months for full healing .
    - Long-Term While it is possible for turbinate tissue to regrow after surgery, many patients find the results satisfactory . Some studies indicate sustained benefit from procedures like radiofrequency volumetric tissue reduction (RFVTR) for at least two years post-treatment . Ongoing management, such as sublingual immunotherapy, may be recommended, especially if allergies were a contributing factor, to prevent future inflammation and maintain surgical benefits

**WHEN TO SEE A DOCTOR / RED FLAG**

* Difficulty breathing through your nose or nasal obstruction
* Nasal congestion that does not improve over time or feels severe
* Nasal stuffiness, especially if it alternates sides or worsens when lying down
* Prolonged nasal congestion that feels like a cold that won’t go away
* Noisy breathing or mouth breathing due to inability to breathe well through the nose
* Frequent or worsening sinus infections or facial pressure
* Snoring or sleep disturbances related to nasal blockage
* Increased nasal drainage or nosebleeds
* Symptoms affecting quality of life or daily functioning

D**IFFERENTIAL DIAGNOSIS**

* Allergic Rhinitis: Chronic or intermittent allergic inflammation causing bilateral turbinate swelling, sneezing, rhinorrhea, and nasal congestion due to mucosal edema and cellular changes.
* Vasomotor (Nonallergic) Rhinitis: Nasal congestion and turbinate swelling without allergic sensitization, triggered by irritants, weather changes, or autonomic nervous system dysregulation.
* Septal Deviation with Compensatory Turbinate Hypertrophy: Structural nasal septum deviation causing airflow obstruction; turbinates on the opposite side may enlarge to compensate, usually unilateral.
* Chronic Sinusitis: Persistent sinus inflammation leading to mucosal swelling and turbinate enlargement, often accompanied by sinus pain, pressure, or purulent nasal discharge.
* Infectious Rhinitis or Upper Respiratory Infection: Acute swelling of turbinates from viral or bacterial infection, usually transient with systemic infection signs.
* Nasal Polyps: Benign mucosal growths that can coexist with turbinate hypertrophy and cause obstruction but are distinguishable on endoscopy.
* Environmental or Medication-Induced Rhinitis: Prolonged use of nasal decongestants leading to rebound congestion (rhinitis medicamentosa) or exposure to irritants causing turbinate swelling.
* Nasal Neoplasms (Rare): Tumors in the nasal cavity may mimic hypertrophy but typically present with persistent unilateral symptoms, bleeding, or mass effect.

**EPIDEMIOLOGY**

* Studies surveying patients with sinonasal complaints show that inferior turbinate hypertrophy is present in about 72% to 77% of such patients. For example, one large US-based study of 1,906 patients reported a prevalence of 72% for inferior turbinate hypertrophy among those presenting with nasal obstruction.
* Among patients with severe or extreme nasal obstruction symptoms, the prevalence of inferior turbinate hypertrophy rises slightly, up to 77% in some cohorts.
* Turbinate hypertrophy commonly coexists with other anatomical contributors such as septal deviation (present in 76% of patients) and nasal valve collapse (67% prevalence).
* Inferior turbinate hypertrophy is often refractory to medical therapy and is a common indication for surgical intervention such as turbinate reduction procedures.
* It is associated with allergic rhinitis in a large proportion of patients, with some studies reporting over 70% of allergic rhinitis patients having hypertrophy of the inferior turbinate mucosa.

**PREDEFINED Q & A SETS**

Q1: What is turbinate hypertrophy?  
A1: Turbinate hypertrophy is an enlargement or swelling of the turbinates, which are structures inside the nose that help filter, warm, and humidify the air we breathe. When swollen, they can block nasal airflow and cause congestion.

Q2: What causes turbinate hypertrophy?  
A2: Common causes include allergies, infections (like colds or sinus infections), chronic inflammation, nasal structural issues such as a deviated septum, environmental irritants (smoke, pollutants), medication overuse (nasal decongestant sprays), and hormonal changes.

Q3: What are the typical symptoms?  
A3: Symptoms mainly include nasal congestion or stuffiness, difficulty breathing through the nose, nasal obstruction often worse on one side, runny nose, postnasal drip, snoring, and sometimes reduced sense of smell.

Q4: How is turbinate hypertrophy diagnosed?  
A4: Diagnosis involves a physical examination using rhinoscopy or nasal endoscopy to visually assess the turbinates, patient history, and sometimes imaging like CT scans for more detailed views. Allergy testing may also be done if allergic rhinitis is suspected.

Q5: What treatment options are available?  
A5: Initial treatment usually includes medical therapy such as nasal corticosteroid sprays, antihistamines, saline nasal rinses, and avoiding triggers. If medical treatment fails, minimally invasive procedures like radiofrequency turbinate reduction or surgery may be recommended to reduce turbinate size.

Q6: Can turbinate hypertrophy be prevented?  
A6: Prevention focuses on managing allergies and avoiding environmental irritants, not overusing nasal decongestants, maintaining good nasal hygiene with saline sprays, and treating infections promptly.

Q7: What is the prognosis?  
A7: Many people experience symptom relief with medical treatment. Surgical reduction typically has a high success rate (80-90%) with sustained improvement, though turbinates can sometimes regrow. Ongoing management of underlying causes is important.

Q8: When should I see a doctor?  
A8: If you have persistent or worsening nasal congestion, difficulty breathing, recurrent sinus infections, or symptoms impacting your quality of life, you should see an ENT specialist for evaluation.

**DOCTOR-PATIENT CONVERSATIONS**

Doctor: Good morning. What brings you in today?

Patient: I've been having a really stuffy nose for a while now. Sometimes I feel like I can't breathe well through one side or both, especially when I lie down.

Doctor: I see. Does your nasal congestion get worse at any particular time, like at night or during certain seasons?

Patient: Yes, it gets worse at night and sometimes when the weather changes. I also sneeze a lot and have a runny nose sometimes.

Doctor: Have you had any history of allergies, sinus infections, or used any nasal sprays recently?

Patient: I do have seasonal allergies, and I sometimes use nasal decongestant sprays when my nose is really blocked.

Doctor: Okay. Based on your symptoms, it sounds like you may have turbinate hypertrophy, which means the structures inside your nose called turbinates are swollen, causing the blockage. I’ll perform a nasal examination to have a look.

(Doctor performs nasal endoscopy or rhinoscopy.)

Doctor: Your turbinates are indeed enlarged and swollen, which is common with allergies or irritation. This is narrowing your nasal passages and causing the congestion.

Patient: Is this serious? What can be done to help?

Doctor: It’s usually not dangerous but can definitely be uncomfortable and impact your breathing and sleep. Treatment generally begins with medical options like nasal steroid sprays, antihistamines for allergies, and saline rinses to reduce swelling and inflammation. It’s also important to avoid overusing nasal decongestant sprays, as they can worsen swelling over time.

Patient: Will I need surgery?

Doctor: Most patients respond well to medical therapy. However, if symptoms persist despite treatment, minimally invasive procedures like radiofrequency turbinate reduction or other turbinate reduction surgeries can be considered to reduce the size and improve airflow.

Patient: Are there any risks with surgery?

Doctor: Surgery is generally safe, but like all procedures, it carries some risks such as bleeding, infection, or recurrence of turbinate swelling. But we usually try all medical options first before considering surgery.

Patient: What can I do at home to help?

Doctor: Managing your allergies, avoiding irritants like smoke, using a humidifier, and doing regular saline nasal rinses can help keep your nasal passages clear. Also, keep track of when your symptoms worsen and let me know.

Patient: Thank you. That helps a lot.

Doctor: You’re welcome. Let’s plan to follow up in a few weeks after you try the medications and see how you feel.

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**Uvulitis**

ALTERNATIVE NAMES

**DEFINITION / DESCRIPTION**

Uvulitis is another word for a swollen uvula. Your uvula is that little fleshy piece of tissue that hangs from the roof of your mouth. It can swell because of things like infections, allergies or trauma. You might develop a swollen uvula when you have inflammation in nearby areas, like your throat or tonsils.

Uvulitis is common. Treating the cause usually eases your symptoms.

**CAUSES**

Some of the most common swollen uvula causes include:

* Allergies. Pet dander, dust, pollen or certain foods can cause your uvula to swell.
* Dehydration. Not getting enough fluids, like water, can cause a swollen uvula.
* Environmental irritants. Vaping, smoking or inhaling certain chemical substances can irritate your uvula and lead to inflammation.
* Genetics. Some genetic conditions can result in uvulitis, like hereditary angioedema.
* GERD (chronic acid reflux). Severe acid reflux can irritate your throat and uvula.
* Infections. Some people get swollen uvulas as a symptom of viral or bacterial infections like strep throat, the flu, or the common cold.
* Medications. Some drug side effects can cause a swollen uvula.
* Snoring. Many people with obstructive sleep apnea will also have swollen or sore uvulas. (Snoring can be a cause or a symptom of uvulitis.)
* Sexually transmitted infections (STIs). Some STIs like gonorrhea and syphilis could cause your uvula to swell. (But uvulitis itself isn’t an STI.)
* Trauma. Surgical procedures, like tonsillectomy, can sometimes injure your uvula. Uvulitis could also happen from intubation during surgery with general anesthesia.

#### **Is a swollen uvula contagious?**

Uvulitis itself isn’t contagious. But the viruses and bacteria that cause it can spread from person to person.

If you have a contagious infection, be sure to wash your hands frequently and stay home if you have a fever.

**SIGNS / SYMPTOMS**

Swollen uvula symptoms might include:

* Coughing, gagging or choking
* Difficulty swallowing
* Excess saliva production
* Feeling like something’s stuck in your throat
* Fever
* Sore throat
* Snoring
* Trouble breathing
* White spots on your uvula or throat

**DIAGNOSIS METHODS**

A healthcare provider will do a physical exam and ask about your symptoms. They may also request certain tests to see what caused the inflammation. These tests may include:

* Allergy tests. If something in your environment causes a swollen uvula, these tests can determine the culprit.
* Throat cultures. This quick, painless test can tell you what kind of germs caused your uvula to swell.
* Blood tests. These tests can detect germs in your blood, like viruses, bacteria and funguses related to uvulitis.

**TREATMENT OPTIONS**

The quickest way to get rid of a swollen uvula is to treat the underlying cause. To do this, your healthcare provider may recommend:

* Antihistamines. Medications like loratadine (Claritin®) and diphenhydramine (Benadryl®) may reduce uvular swelling related to allergies.
* Corticosteroids. These drugs can reduce swelling if you have severe inflammation.
* Medications to treat infections. Your healthcare provider might prescribe antibiotics, antifungals or antivirals if you have an infection.
* Surgery. If less invasive treatments don’t work, or if you have chronic uvulitis, you may need surgery to remove part or all of your uvula (uvulectomy).

#### **Swollen uvula home remedies**

You may be able to ease mild uvulitis symptoms at home with these remedies:

* Drink lots of water.
* Eat ice chips.
* Gargle with warm saltwater.
* Get lots of rest.
* Make hot tea sweetened with honey.
* Suck on throat lozenges.
* Take over-the-counter pain relievers.
* Use a humidifier.

**PREVENTION TIPS**

You can’t always prevent a swollen uvula. But there are things you can do to reduce your risk:

* Avoid allergens as much as possible.
* Avoid chemical irritants and other environmental triggers.
* If you smoke, consider quitting.
* Practice safe sex (since uvulitis is linked to some STIs)

**OUTLOOK / PROGNOSIS**

Uvular swelling often goes away in a couple of days. But if you have severe or lingering symptoms, you should schedule an appointment with your healthcare provider.

**WHEN TO SEE A DOCTOR / RED FLAG**

You should schedule an appointment with your healthcare provider if you have a swollen uvula that doesn’t improve after five days.

If you’re choking or you feel like you can’t breathe, call 911 or head to your nearest emergency room.

**DIFFERENTIAL DIAGNOSIS**

* Infectious uvulitis: Often bacterial causes such as *Streptococcus pyogenes*, *Haemophilus influenzae*, *Streptococcus pneumoniae*, and *Fusobacterium*, or viral infections (e.g., viral pharyngitis). Candida albicans (fungal) is also a rare cause. Uvulitis frequently occurs alongside pharyngitis, tonsillitis, or epiglottitis.
* Angioedema (Quincke's disease): This is a non-infectious allergic or hereditary swelling of the uvula, typically caused by type 1 hypersensitivity reactions, medication reactions (e.g., ACE inhibitors), or inhalation irritants. It is important to differentiate this from infectious uvulitis because it requires different management.
* Tonsillitis: Inflammation of the tonsils can cause overlapping symptoms and uvular swelling, but the primary site of infection is the tonsils.
* Pharyngitis: Inflammation of the pharynx that can involve the uvula secondarily.
* Epiglottitis: Inflammation of the epiglottis can be associated with uvular swelling and is a medical emergency due to airway obstruction risk.
* Peritonsillar or retropharyngeal abscess: Deep neck space infections can cause uvular deviation and swelling, and require prompt diagnosis and often surgical drainage.
* Trauma or irritation: Mechanical trauma from endotracheal intubation, endoscopy, prolonged suctioning, or foreign body ingestion can cause uvular swelling.
* Environmental or chemical irritation: Exposure to smoke (tobacco, marijuana), inhaled irritants, or acid reflux-induced uvular inflammation.

**EPIDEMIOLOGY**

* The incidence of infectious uvulitis has decreased significantly since the introduction of the Hib vaccine in the 1990s, which greatly reduced invasive Hib infections that previously caused severe uvulitis in children.
* Nowadays, Group A Streptococcus (GAS) is the predominant bacterial cause of uvulitis in immunized populations, with occasional fungal and viral causes also reported, including rare cases of viral uvulitis caused by parainfluenza virus.
* Uvulitis may occur in the setting of upper respiratory infections, especially in regions or seasons with high rates of streptococcal pharyngitis or influenza, suggesting incidence fluctuates with respiratory infection prevalence.
* Some studies from regions with high rates of traditional uvulectomy (a cultural practice involving surgical removal of the uvula) show pooled prevalences of uvula-related conditions around 28-41% in children, particularly in rural African populations where traditional medicine is common.
* Although mainly infectious, genetic predisposition may influence susceptibility to uvulitis through variations in genes regulating inflammation (e.g., IL-1B and IL-6), but this aspect remains poorly understood and under-researched.

**PREDEFINED Q & A SETS**

Q1: What is uvulitis?  
A1: Uvulitis is inflammation and swelling of the uvula, the small, fleshy tissue that hangs at the back of your throat. It can cause discomfort, difficulty swallowing, and sometimes breathing problems.

Q2: What causes uvulitis?  
A2: Causes include bacterial or viral infections (such as strep throat), allergic reactions, irritants like smoke or toxins, trauma to the uvula, and acid reflux. Sometimes the exact cause is unknown.

Q3: What are the symptoms of uvulitis?  
A3: Symptoms include a swollen, red uvula, sore throat, difficulty swallowing, feeling like something is stuck in the throat, snoring, gagging, and rarely difficulty breathing.

Q4: How is uvulitis diagnosed?  
A4: Diagnosis is usually made by physical examination of the throat by a healthcare provider. Sometimes throat swabs, blood tests, or allergy tests are done to identify the cause.

Q5: How is uvulitis treated?  
A5: Treatment depends on the cause:

* Bacterial infections are treated with antibiotics.
* Allergic reactions may require antihistamines, steroids, or epinephrine.
* Reflux-related uvulitis might be helped by reflux medications.
* Home care includes rest, staying hydrated, gargling with warm salt water, and over-the-counter pain relief.

Q6: How long does uvulitis last?  
A6: Mild uvulitis often improves within 1 to 2 days with or without treatment. More severe cases may take longer and need medical care.

Q7: When should I see a doctor?  
A7: See a doctor if you have trouble breathing or swallowing, severe pain, high fever, symptoms lasting more than a few days, or if symptoms worsen despite treatment.

Q8: Can uvulitis cause serious complications?  
A8: Severe swelling may obstruct the airway and cause choking, which is a medical emergency requiring immediate care.

Q9: How can I care for uvulitis at home?  
A9: Rest, drink plenty of fluids, avoid irritants like smoking, use throat lozenges or sprays for pain, and follow your doctor’s instructions carefully if medicines are prescribed.

**medications for uvulitis and their possible side effects:**

1. Antibiotics
   * Used if uvulitis is caused by a bacterial infection (often similar organisms as in streptococcal pharyngitis, e.g., *Streptococcus pyogenes*). Common antibiotics include penicillin, amoxicillin, co-amoxiclav, or ceftriaxone (for more severe infections).
   * Side effects: Allergic reactions (rash, anaphylaxis), gastrointestinal upset (nausea, diarrhea), yeast infections, and antibiotic resistance with improper use.
   * Important: Complete the full course even if symptoms improve.
2. Corticosteroids
   * Used to reduce inflammation and swelling, particularly if uvulitis is due to allergic reactions or severe inflammation. Steroids may be given orally or intravenously depending on severity.
   * Side effects: Increased blood sugar, mood changes, immunosuppression (higher infection risk), gastric irritation, and long-term side effects if used extensively.
3. Antihistamines
   * Often prescribed if uvulitis is related to allergies (e.g., angioedema/Quincke’s disease). They reduce itching, swelling, and other allergic symptoms.
   * Side effects: Drowsiness, dry mouth, dizziness, and sometimes gastrointestinal discomfort.
4. Epinephrine
   * Used in emergency cases of severe allergic uvulitis causing airway obstruction (anaphylaxis). It rapidly reduces swelling and opens airways.
   * Side effects: Palpitations, increased heart rate, anxiety, tremors, and elevated blood pressure; used only in urgent scenarios.
5. Reflux medications
   * If uvulitis is related to acid reflux irritation, proton pump inhibitors or H2 blockers may be used to reduce acid exposure. Side effects vary by medication class but are generally mild.
6. Supportive Care
   * Pain relievers such as acetaminophen or ibuprofen can help reduce throat discomfort.
   * Gargling with warm salt water, hydration, and avoiding irritants like smoking are also important.
7. Surgery
   * Rarely, if swelling persists and causes airway risk, partial uvula removal (uvulectomy) may be considered.

**DOCTOR-PATIENT CONVERSATIONS**

Doctor: Good morning. What brings you in today?

Patient: Hi, Doctor. I’ve noticed my uvula feels swollen and sore. My throat hurts when I swallow, and it feels like something is stuck in the back of my throat.

Doctor: Thank you for sharing that. How long have you had these symptoms? Is the swelling constant or does it come and go?

Patient: It started about two days ago and seems to be getting worse. It’s sore all the time now.

Doctor: Any difficulty breathing, choking sensation, or voice changes?

Patient: No breathing problems yet, but my voice sounds a bit hoarse.

Doctor: Have you had any recent infections, allergies, or were you exposed to irritants like smoke or allergens?

Patient: I had a mild cold last week, and I sometimes smoke socially.

Doctor: I see. I’m going to examine your throat now to see the uvula and surrounding tissues.

(Doctor examines the throat and observes a swollen, red uvula.)

Doctor: Your uvula is indeed swollen and inflamed, which is called uvulitis. It’s likely related to the recent infection or irritation. Sometimes bacterial infections can cause this or allergies can be involved.

Patient: Is this serious? What do I need to do?

Doctor: Most cases improve quickly with treatment. If it’s bacterial, you may need antibiotics. For inflammation or allergies, antihistamines or corticosteroids can help reduce the swelling. Avoid smoking and irritants, stay hydrated, and gargle with warm salt water.

Patient: What should I watch out for?

Doctor: If you develop difficulty breathing, severe pain, high fever, or trouble swallowing, seek emergency care immediately, as these could indicate serious complications.

Patient: How long will it take to get better?

Doctor: Usually in a few days to a week with appropriate treatment. We’ll monitor your progress and adjust treatment if necessary.

Patient: Thank you, Doctor.

Doctor: You’re welcome. If symptoms worsen or don’t improve, please come back for evaluation.

REFERENCES:

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**Undifferentiated pleomorphic sarcoma**

ALTERNATIVE NAMES

**DEFINITION / DESCRIPTION**

Undifferentiated pleomorphic sarcoma (UPS) is a rare type of cancer that begins mostly in the soft tissues of the body. Soft tissues connect, support and surround other body structures.

UPS usually occurs in the arms or legs. Less often it can happen in the area behind the abdominal organs (retroperitoneum).

The name undifferentiated pleomorphic sarcoma comes from the way the cancer cells appear under the microscope. Undifferentiated means the cells don't look like the body tissues in which they develop. The cancer is called pleomorphic (plee-o-MOR-fik) because the cells grow in multiple shapes and sizes.

Treatment for UPS depends on the location of the cancer, but often involves surgery, radiation and drug treatments.

UPS used to be called malignant fibrous histiocytoma.

**CAUSES**

It's not clear what causes undifferentiated pleomorphic sarcoma.

Doctors know this cancer begins when a cell develops changes in its DNA. A cell's DNA contains the instructions that tell a cell what to do. The changes tell the cell to multiply rapidly, creating a mass of abnormal cells (tumor). The cells can invade and destroy nearby healthy tissue. In time, the cancer cells can break away and spread (metastasize) to other parts of the body, such as the lungs and bones.

**RISK FACTORS**

Factors that may increase the risk of undifferentiated pleomorphic sarcoma include:

* **Older age.** This cancer tends to occur in adults older than 50, though it can occur at any age.
* **Previous radiation therapy.** Rarely, this cancer can develop in an area of the body that was previously treated with radiation therapy.

Most people who develop undifferentiated pleomorphic sarcoma have no known risk factors, and many people who have risk factors never develop the cancer.

**SIGNS / SYMPTOMS**

Undifferentiated pleomorphic sarcoma symptoms depend on where the cancer occurs. It most often happens in the arms and legs, but it can happen anywhere in the body.

Signs and symptoms may include:

* Growing lump or area of swelling.
* If it grows very large, there may be pain, tingling and numbness.
* If it occurs in an arm or leg, there may be swelling in the hand or foot of an affected limb.
* If it occurs in the abdomen, there may be pain, loss of appetite and constipation.
* Fever.
* Weight loss

**DIAGNOSIS METHODS**

Diagnosis for undifferentiated pleomorphic sarcoma usually starts with a review of your symptoms and a physical examination. This cancer is often diagnosed after other types of cancer have been ruled out.

Tests and procedures may include:

* **Physical exam.** Your doctor will ask you questions about when your symptoms began and if they've changed over time. He or she will examine the area to better understand the size and depth of the growth, whether it's connected to nearby tissues, and whether there are any signs of swelling or nerve damage.
* **Imaging tests.** Your doctor may recommend imaging tests to create pictures of the affected area and understand more about your condition. Imaging tests may include X-rays, CT, MRI and positron emission tomography (PET) scans.
* **Removing a sample of tissue for testing (biopsy).** To make a definitive diagnosis, your doctor collects a sample of the tumor tissue and sends it to a lab for testing. Depending on your particular situation, the tissue sample may be collected with a needle inserted through your skin or during an operation.  
  In the lab, doctors trained in analyzing body tissues (pathologists) examine the sample to determine the types of cells involved and whether the cells are likely to be aggressive. This information helps rule out other types of cancer and guides your treatment.

Determining the type of biopsy needed and the specifics of how it should be performed requires careful planning by the medical team. Doctors need to perform the biopsy in a way that won't interfere with future surgery to remove the cancer. For this reason, ask your doctor for a referral to a team of experts with extensive experience in treating soft tissue sarcomas before the biopsy.

**TREATMENT OPTIONS**

Treatment for undifferentiated pleomorphic sarcoma usually involves surgery to remove the cancer cells. Other options include radiation therapy and drug treatments (systemic therapies), such as chemotherapy, targeted therapy and immunotherapy. Which treatments are best for you will depend on the size and location of your cancer.

### **Surgery**

When possible, doctors try to remove the sarcoma completely with surgery. The goal is to remove the cancer and a margin of healthy tissue around it with as minimal an impact as possible.

When the cancer affects the arms and legs, surgeons prefer to use limb-sparing operations. However, in some cases it may be necessary to amputate the affected arm or leg. Other treatments, such as radiation therapy and chemotherapy, might be recommended before surgery to shrink a cancer so that it's easier to remove without amputating the affected limb.

### **Radiation therapy**

Radiation therapy uses high-powered beams of energy, such as X-rays or protons, to kill cancer cells. Radiation therapy can be given as:

* **External beam radiation.** This type of radiation comes from a machine that moves around you as you lie on a table. The machine directs the radiation to precise points on your body.  
  Radiation may be used before surgery to shrink a sarcoma and make it easier to remove. It may also be used after surgery to kill any cancer cells that remain.
* **Intraoperative radiation therapy (IORT).** IORT is used during surgery just after the cancer is removed. The radiation is directed to the area around where the cancer used to be. IORT might be recommended if the cancer is located in an area that makes it difficult to remove the cancer completely during surgery.

### **Chemotherapy**

Chemotherapy is a drug treatment that uses chemicals to kill cancer cells. It can be administered by pill or through a vein (intravenously), or both.

Chemotherapy is most often used to treat undifferentiated pleomorphic sarcoma that comes back after initial treatment or that spreads to other areas of the body.

Sometimes chemotherapy is used before surgery to shrink the cancer so that it's easier to remove during an operation.

Chemotherapy may also be combined with radiation.

### **Targeted drug therapy**

Targeted drug treatments focus on specific abnormalities present within cancer cells. By blocking these abnormalities, targeted drug treatments can cause cancer cells to die.

For undifferentiated pleomorphic sarcoma, targeted therapy drugs may be combined with chemotherapy.

Some targeted therapies only work in people whose cancer cells have certain genetic mutations. Your cancer cells may be tested in a laboratory to see if these drugs might help you.

### **Immunotherapy**

Immunotherapy uses your immune system to fight cancer. Your body's disease-fighting immune system may not attack your cancer because the cancer cells produce proteins that help them hide. Immunotherapy works by interfering with that process.

Immunotherapy treatments are generally reserved for people with advanced cancer.

**Alternative medicine**

No alternative treatments have been found helpful in treating undifferentiated pleomorphic sarcoma. But some complementary and alternative treatments may relieve the symptoms you experience due to cancer or cancer treatment.

Alternative treatments that may help relieve symptoms include:

* Acupuncture.
* Exercise.
* Massage.
* Meditation.
* Music therapy.
* Relaxation exercises.

**OUTLOOK / PROGNOSIS**

Life expectancy varies depending on the size and stage of the sarcoma. The five-year survival rate for high-grade UPS is about 60%. That means that around 60% of people diagnosed with this condition are still alive five years later. The 10-year survival rate is 48%.

Survival rates are just estimates. They can’t tell you how long you’ll live or what kind of treatment will work for you. To learn more about UPS survival rates and how they affect you, talk to your oncology team.

#### **Is undifferentiated pleomorphic sarcoma curable?**

It’s possible to cure UPS, especially during the early stages. There still may be treatment options in later stages, which can prolong your quality of life. Like any type of cancer, early detection and treatment increase your chances for successful outcomes.

**POSSIBLE COMPLICATIONS**

**Undifferentiated Pleomorphic Sarcoma Complications**

UPS is a high-grade soft tissue malignancy associated with various complications that can arise from the tumor itself, its local and systemic effects, and the treatment modalities used.

Local complications primarily result from the tumor's aggressive and infiltrative behavior. Large or deeply situated tumors can compress or invade adjacent neurovascular structures, leading to pain, sensory deficits, or motor dysfunction. In the extremities, such involvement may result in impaired limb function or even necessitate amputation in cases where critical structures cannot be preserved during resection. In retroperitoneal locations, UPS may compress or infiltrate vital organs, leading to bowel obstruction, hydronephrosis, or vascular compromise.

UPS has a high propensity for hematogenous metastasis, most commonly to the lungs. Pulmonary metastases may be asymptomatic or present with cough, hemoptysis, pleuritic pain, or dyspnea. Less commonly, liver, bone, or brain metastases may occur, contributing to systemic symptoms and organ-specific dysfunction. High-grade tumors are also at risk for early recurrence, particularly if resection margins are inadequate.

**Treatment-Related Complications**

Treatment-related complications are also significant. Surgical resection, especially when involving large or anatomically complex tumors, can lead to wound healing problems, seroma or hematoma formation, infection, and long-term functional impairment. In the case of preoperative or postoperative radiation therapy, patients may experience fibrosis, joint stiffness, lymphedema, delayed wound healing, and secondary radiation-induced malignancies. Chemotherapy-related toxicity includes myelosuppression, cardiotoxicity (particularly with doxorubicin), nephrotoxicity, and fatigue, all of which may limit treatment intensity or duration.

Finally, psychosocial distress, decreased quality of life, and physical disability are essential but sometimes underrecognized complications, particularly in patients requiring extensive surgeries or facing recurrent or metastatic disease. As such, early engagement of rehabilitation, pain management, psychosocial support, and survivorship care is essential to comprehensive management

**WHEN TO SEE A DOCTOR / RED FLAG**

Make an appointment with a doctor if you develop any persistent signs or symptoms that worry you.

**DIFFERENTIAL DIAGNOSIS**

The most common differential diagnoses of UPS are other types of soft tissue sarcomas, which must be distinguished by clinical history, physical examination, and immunohistochemistry markers, including atypical fibroxanthoma, liposarcoma, leiomyosarcoma, angiosarcoma, fibrosarcoma, myxofibrosarcoma, dermatofibrosarcoma protuberans, osteosarcoma, and malignant peripheral nerve sheath tumor. Metastases, desmoplastic melanoma, and spindle-cell squamous cell carcinoma may also resemble UPS clinical or histopathological morphology.

**EPIDEMIOLOGY**

The incidence and prevalence of UPS are challenging to quantify due to historical misclassification (many tumors previously classified as UPS were likely other undifferentiated sarcomas) and the lack of specific tracking in national cancer registries (eg, the SEER database), where UPS is grouped within broader categories of high-grade or unclassified soft tissue sarcomas. Nevertheless, UPS is estimated to comprise approximately 5% to 10% of all adult soft tissue sarcomas, making it one of the most common high-grade sarcoma subtypes in this population. The estimated annual incidence of soft tissue sarcomas in the United States is roughly 13,000 cases, suggesting that approximately 650 to 1,300 of these may be UPS. Global incidence rates are likely similar, as no geographic or racial disparities have been reported.

UPS predominantly occurs in older adults, with peak incidence between the ages of 50 and 70, and is rare in pediatric populations. A slight male predominance has been observed, though sex differences in outcomes have not been established. Improved histopathologic classification and the adoption of advanced molecular diagnostics have led to a decline in UPS diagnoses, as many previously labeled cases have been reclassified into distinct, genetically defined sarcoma subtypes

**PREDEFINED Q & A SETS**

1. Other possible causes for your symptoms:  
Symptoms such as a growing lump, pain, or swelling could be caused by other soft tissue tumors (benign or malignant), infections, cysts, or inflammatory conditions. Diagnosis requires ruling out these and other cancers via imaging and biopsy.

2. Tests needed to confirm diagnosis and preparation:

* Physical exam to assess the tumor size, location, and involvement of nearby tissues.
* Imaging tests: X-rays, CT scans, MRI, and PET scans help visualize the tumor, its extent, and possible spread.
* Biopsy: A tissue sample is taken (via needle or during surgery) to confirm pathological diagnosis. Biopsy must be done carefully by experts to avoid jeopardizing future surgery.
* Preparation: Usually no special preparation for imaging other than informing about allergies or kidney function for contrast media; biopsy may require fasting or specific instructions from your medical team depending on procedure type. Ask your doctor for detailed instructions before tests.

3. Stage of the sarcoma:  
Staging depends on tumor size, depth, grade (how aggressive cells look under a microscope), lymph node involvement, and metastasis (spread to other organs). Early-stage means localized tumor without spread, while advanced stages involve larger tumors or distant metastases. Accurate staging requires imaging and biopsy results.

4. Treatment options and recommendations:

* Surgery: Preferred to completely remove the tumor with clear margins; surgical removal is often possible though depends on tumor size and location.
* Radiation therapy: Often used before (neoadjuvant) to shrink tumors or after (adjuvant) surgery to reduce local recurrence.
* Chemotherapy: Sometimes used depending on stage and tumor aggressiveness though effectiveness varies.
* Multidisciplinary approach: Treatment planning by a sarcoma specialist team optimizes outcomes.

5. Side effects from treatment:

* Surgery: Risks include infection, bleeding, pain, and functional impairment depending on tumor location.
* Radiation: Skin irritation, fatigue, tissue swelling, and potential long-term tissue damage.
* Chemotherapy: Nausea, hair loss, fatigue, immune suppression, and other systemic effects.  
  Side effects vary by treatment type, dose, and individual factors.

6. Alternatives to primary treatment:  
If surgery is not feasible or in advanced disease, options include palliative radiation, chemotherapy alone, experimental therapies or clinical trials. Your specialist will tailor alternatives based on your particular case.

7. Managing other health conditions:  
Discuss all your medical conditions with your sarcoma care team so treatments can be coordinated. Managing chronic diseases (diabetes, heart disease, etc.) during cancer therapy involves careful monitoring, possible medication adjustments, and supportive care.

8. Dietary or activity restrictions:

* Generally, maintain good nutrition with adequate protein and calories to support healing.
* Activity may be limited during treatment and recovery — your care team will guide safe levels based on surgery and therapy effects.
* Avoid smoking, alcohol, and other irritants to optimize outcomes.

9. Prognosis:  
Prognosis depends on tumor size, grade, location, completeness of surgical removal, and presence of metastases. Early-stage, completely resected UPS has a better prognosis, but it is an aggressive cancer requiring close follow-up.

11. Role of radiation therapy before or after surgery:  
Radiation therapy is commonly used either before surgery (to shrink the tumor and make resection easier) or after surgery (to reduce local recurrence risk). Your sarcoma team will decide timing based on your individual case.

12. Experience of your surgeon:  
It is crucial your surgeon has specialized experience in soft tissue sarcoma surgery because intricate planning and technique affect outcomes considerably. Ask your doctor about surgeon qualifications and experience with UPS specifically.

**STAGING**

1. Tumor size and extent (T):
   * T1: Tumor ≤ 5 cm in greatest dimension
   * T2: Tumor > 5 cm but ≤ 10 cm
   * T3: Tumor > 10 cm but ≤ 15 cm
   * T4: Tumor > 15 cm
2. Nodal involvement (N):
   * N0: No regional lymph node involvement
   * N1: Regional lymph node metastasis present
3. Distant metastasis (M):
   * M0: No distant metastasis
   * M1: Distant metastasis present (e.g., lungs, bone)
4. Grade of the tumor (G):
   * G1: Low grade (well differentiated)
   * G2: Intermediate grade
   * G3: High grade (poorly differentiated, aggressive behavior)

Because most UPS tumors are high-grade (G3), the grade is a crucial prognostic factor. The tumor size and presence of metastasis influence the overall stage:

* Stage I: Small tumors (≤5 cm), low grade, no nodal involvement or metastasis
* Stage II: Larger tumors (>5 cm), low to intermediate grade, no nodal involvement or metastasis
* Stage III: Large tumors (>5 cm) with high grade but no metastasis
* Stage IV: Any tumor with regional lymph node involvement or distant metastasis

The stage reflects tumor aggressiveness and guides treatment planning.

**DOCTOR-PATIENT CONVERSATIONS**

Doctor: Good morning. I understand you have some concerns about a lump and possible diagnosis of undifferentiated pleomorphic sarcoma. Can you tell me more about your symptoms?

Patient: Yes, Doctor. I've noticed a growing lump in my thigh that doesn’t hurt much but seems to be getting bigger over the last few weeks.

Doctor: Thank you. UPS often presents as a painless, enlarging mass. We need to do some tests to confirm the diagnosis and check the extent of the tumor. Have you had any imaging done so far, like an MRI or CT scan?

Patient: Not yet. What kind of tests will I need?

Doctor: Typically, we start with an MRI to get detailed images of the soft tissues and see how large the tumor is and whether it's close to nerves or blood vessels. We’ll also do a biopsy, where a small sample of the tumor tissue is taken to look at under the microscope. This helps us confirm the diagnosis and determine how aggressive the cancer is.

Patient: Does the biopsy hurt? Will I need to prepare for it?

Doctor: The biopsy is usually done under local anesthesia and is a minor procedure. Sometimes for deeper tumors, sedation or a small surgical procedure might be needed. We will give you instructions beforehand, such as avoiding blood-thinning medications if applicable.

Patient: How do you stage this sarcoma? What does staging mean?

Doctor: Staging tells us the size of the tumor, whether it has spread to lymph nodes or distant organs, and how aggressive the tumor cells are. UPS staging usually uses measurements from imaging and biopsy results. This helps us decide the best treatment.

Patient: What treatments are available?

Doctor: The mainstay of treatment is surgical removal of the tumor with clean margins to reduce recurrence risk. Depending on the size and location, we often recommend radiation therapy either before or after surgery to help control the tumor locally. Chemotherapy may be considered in some cases, especially if the tumor is large or has spread. Treatment is planned by a team of sarcoma specialists to tailor it to you.

Patient: What kind of side effects should I expect from surgery and radiation?

Doctor: Surgery side effects depend on the tumor location but can include pain, swelling, and temporary or sometimes permanent changes in function or sensation. Radiation might cause skin irritation, fatigue, and swelling in the treatment area, usually improving after finishing. We will support you through these.

Patient: What if I have other health problems? Will this affect my treatment?

Doctor: It's very important to discuss all your health conditions so we can adjust treatments and monitor closely. Coordinated care ensures your other medical conditions are managed safely during cancer treatment.

Patient: How long will recovery take? Will I need to change my diet or activity?

Doctor: Recovery times vary depending on the surgery. Initially, you’ll have activity restrictions to allow healing, but we encourage gradual mobilization. Nutrition should be balanced and support healing—our team can provide guidance.

Patient: What is my prognosis?

Doctor: Prognosis depends on tumor size, grade, stage, and how completely we can remove the tumor. UPS is aggressive, but with early and appropriate treatment, many patients achieve good control. Close follow-up is essential.

Patient: Can you provide me any brochures or websites to learn more?

Doctor: Absolutely. I will give you printed materials and recommend reliable websites like the Mayo Clinic and Moffitt Cancer Center for detailed information.

Patient: Is the surgeon experienced in tumors like this?

Doctor: Yes, your surgical oncologist specializes in soft tissue sarcomas and works in a multidisciplinary center experienced with UPS. This expertise improves outcomes significantly.

Patient: Thank you, Doctor. That helps me understand what to expect.

Doctor: You’re welcome. We will guide you step-by-step and ensure you have support throughout your treatment journey. Please ask me any questions anytime.

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**VAGAL PARAGANGLIOMAS**

ALTERNATIVE NAMES: Alternative names for vagal paragangliomas include glomus vagale tumors, vagal body tumors, vagal chemodectomas, and glomus vagale. These tumors are also sometimes referred to as paragangliomas of the vagus nerve.

***OVERVIEW***

A paraganglioma is a growth of cells that can happen in different places in the body. The growth, called a tumor, forms from an important type of nerve cell that's found throughout the body.

Paragangliomas most often start in the head, neck, stomach area or pelvis. A paraganglioma is rare. And most often, it's not cancer. When a tumor isn't cancer, it's called benign.

Sometimes a paraganglioma is cancerous. A cancerous tumor can spread to other parts of the body.

Paragangliomas often don't have a clear cause. Some paragangliomas are caused by DNA changes that pass from parents to children.

Paragangliomas can form at any age. Healthcare professionals most often find them in adults between the ages of 20 and 50.

Paraganglioma treatment typically involves surgery to remove the tumor. If the paraganglioma is cancerous and spreads to other areas of the body, more treatments may be needed.

**DEFINITION / DESCRIPTION**

Vagal paragangliomas (previously known as glomus vagale tumors) are tumors that occur along the path of the vagus nerve (CN X). They are a subset of extra-adrenal neuroendocrine tumors that are derived from the nonchromaffin paraganglion cells.

***Terminology***

The term "glomus" was historically used to describe certain types of neuroendocrine tumors arising from paraganglia. The term is, however, imprecise and can be confused with the glomus bodies and tumors that arise from them.

It can also be mixed up with glomus tumors of the subcutaneous skin, also referred to as glomangioma.

***Clinical presentation***

Typically presents as a painless mass behind the carotid artery. Vocal cord paralysis is a relatively frequent finding (~47%) 3.

***Pathology***

For a general discussion on the pathology of these tumors, please refer to the parental article on paragangliomas.

***Location***

Although they could occur at a similar position to carotid body tumors they tend to be more rostral in location and do not widen the carotid bifurcation. They displace the internal and external carotid arteries anteriorly, and the internal jugular vein posteriorly 1.

Vagal paraganglioma is a rare finding that develops from paraganglionic tissue found around the vagus nerve; it has a prevalence of 0.012% of all tumors.

It is the third most common paraganglioma of the head and neck but still accounts for less than 5% of these tumors, and it has a well-established female prevalence.

It is a difficult tumor to identify early based on its symptoms alone and only a thorough investigation can help solidify its diagnosis. In this report, we discuss a presentation of this phenomenon that is not only unique in its manifestation but also a very difficult diagnosis due to its deceptive presentation and multiple extensions. These masses need a good surgical regime to be removed properly and postoperative complications are very frequent in most of these cases.

***Paraganglioma***

A paraganglioma (also known as an extra-adrenal pheochromocytoma) is a rare neuroendocrine tumor (NET) that forms near your carotid artery (the major blood vessels in your neck), along nerve pathways in your head and neck and in other parts of your body.

The tumor is made of a certain type of cell called chromaffin cells, which produce and release certain hormones known as catecholamines.

Your adrenal glands, the two glands that are on top of each kidney, make several hormones. Among these are hormones called catecholamines that help control the following important bodily functions:

* Heart rate.
* Blood pressure.
* Blood sugar (blood glucose).
* The way your body responds to stress.

The primary catecholamines include:

* Dopamine.
* Epinephrine (adrenaline).
* Norepinephrine (noradrenaline).

Even though paragangliomas don’t form in your adrenal glands, they’re made of tissue that’s found in your adrenal glands. Paragangliomas may release extra catecholamines into your blood, causing certain signs and symptoms.

***Paraganglioma and pheochromocytoma***

Paraganglioma and pheochromocytoma are both rare tumors that form from the same type of cells known as chromaffin cells. The difference is where they form in your body.

Pheochromocytomas form in the center of your adrenal gland (adrenal medulla), and paragangliomas form outside your adrenal gland, usually along the arteries or nerves in your neck. Paragangliomas are also called extra-adrenal pheochromocytomas.

***Is paraganglioma cancer?***

Paragangliomas can be benign (not cancer) or malignant (cancer). Approximately 20% of paragangliomas are malignant.

It can be very challenging for healthcare providers to tell if a paraganglioma is cancerous or not — even after they’ve looked at the tumor tissue under a microscope after it’s been removed. Because of this, a paraganglioma is often considered cancer if it has:

* Spread to nearby tissues (regional spread of paraganglioma).
* Spread to distant areas like your lungs or bone (metastasized).
* Come back after initial treatment (recurred).

There’s no standard staging system for paraganglioma if it’s cancerous. Instead, it’s described as the following:

* Localized paraganglioma: The tumor is in one area only.
* Regional paraganglioma: Cancer has spread to lymph nodes or other tissues near its original location.
* Metastatic paraganglioma: Cancer has spread to other parts of your body, such as your liver, lungs, bone or distant lymph nodes. Approximately 35% to 50% of malignant paragangliomas may spread to other parts of your body (metastasize).
* Recurrent paraganglioma: Cancer has recurred (come back) after it has been treated. It may come back in the same place or in another part of your body.

***How fast does a paraganglioma grow?***

Paragangliomas usually grow very slowly. But this could vary from case to case.

***Who does paraganglioma affect?***

Anyone at any age can get a paraganglioma, but they occur most often in people between 30 and 50 years of age. Approximately 10% of cases occur in children.

***How common is paraganglioma?***

Paraganglioma is a rare tumor. It’s estimated that only 2 out of every 1 million people have paraganglioma.

**CAUSES**

Paragangliomas often don't have a clear cause. Sometimes these tumors run in families. They can be caused by DNA changes that are passed from parents to children. But many people with paragangliomas don't have a family history of these tumors and the cause is not known.

A paraganglioma is a growth of cells. It forms from a type of nerve cell called a chromaffin cell. Chromaffin cells perform key roles in the body, including controlling blood pressure.

A paraganglioma starts when chromaffin cells develop changes in their DNA. A cell's DNA holds the instructions that tell the cell what to do. In healthy cells, the DNA gives instructions to grow and multiply at a set rate. The instructions also tell the cells to die at a set time.

In paraganglioma cells, the DNA changes give different instructions. The changes tell the paraganglioma cells to make many more cells quickly. The cells keep living when healthy cells die. This causes a growth of cells called a tumor.

Most paragangliomas stay where they started. They don't spread to other parts of the body. But sometimes cells can break away from a paraganglioma and spread. When this happens, it's called metastatic paraganglioma. When a paraganglioma spreads, it most often spreads to nearby lymph nodes. It also can spread to the lungs, liver and bones.

A paraganglioma is closely related to another rare tumor called a pheochromocytoma. A pheochromocytoma is a tumor that starts in chromaffin cells in the adrenal glands. The adrenal glands are two glands that sit on top of the kidneys.

Vagal paragangliomas (VPGL) are rare neuroendocrine tumors that originate from the paraganglion associated with the vagus nerve. The causes of VPGL are not fully understood, but several factors and genetic changes have been identified.

Genetic mutations play a significant role, with the most common being mutations in the SDHx genes, which are associated with hereditary forms of the disease.

However, a rare case has been reported where a germline variant in the FH gene, specifically FH p.S249R, was found to be associated with VPGL. This variant, along with the loss of the wild-type allele of the FH gene in the tumor, suggests that biallelic gene damage may promote tumor initiation.

In addition to genetic factors, other potential causes and risk factors for VPGL include familial inheritance, genetic alterations in susceptibility genes, female predominance, young age, Carney’s triad, and possibly chronic hypoxia.

Chronic hypoxia, which can result from residing at high altitudes or having conditions such as cyanotic congenital heart disease or chronic obstructive lung disease, may also contribute to the development of these tumors.

While the exact cause of VPGL is often unknown, the role of genetic predispositions and environmental factors such as hypoxia is increasingly recognized in the pathogenesis of these tumors.

In most cases of paraganglioma, the exact cause is unknown, and it occurs randomly. Approximately 25% to 35% of people who have paraganglioma have a hereditary condition (passed through the family) that’s linked to paraganglioma, including:

* Multiple endocrine neoplasia 2 syndrome, types A and B (MEN2A and MEN2B).
* Von Hippel-Lindau (VHL) disease.
* Neurofibromatosis type 1 (NF1).
* Hereditary paraganglioma syndrome.
* Carney-Stratakis dyad (paraganglioma and gastrointestinal stromal tumor [GIST]).
* Carney triad (paraganglioma, GIST, and pulmonary chondroma).

**RISK FACTORS**

The risk of paraganglioma is higher in people who have a family history of this tumor. Some paragangliomas are caused by DNA changes that pass from parents to children. Having a family history of paraganglioma might be a sign that certain DNA changes run in your family.

Some other health conditions caused by DNA changes that pass from parents to children raise the risk of paragangliomas. These conditions include:

* Multiple endocrine neoplasia, type 2. Multiple endocrine neoplasia, type 2, also called MEN 2, can cause tumors in one or more of the glands that make hormones, including the thyroid and the parathyroid glands. There are two types of MEN 2 — type 2A and type 2B. Both raise the risk of paragangliomas.
* Von Hippel-Lindau disease. Von Hippel-Lindau disease can cause tumors and cysts to form in many parts of the body. Possible sites include the brain, spinal cord and kidneys.
* Neurofibromatosis 1. Neurofibromatosis 1 causes tumors called neurofibromas to form in the skin. The condition also can cause tumors of the optic nerve. The optic nerve is the nerve at the back of the eye that connects to the brain.
* Hereditary paraganglioma syndromes. Hereditary paraganglioma syndromes can cause pheochromocytomas or paragangliomas. People with these syndromes often have more than one paraganglioma.
* Carney-Stratakis dyad. Carney-Stratakis dyad causes tumors of the digestive tract and paragangliomas.

Paragangliomas are rare neuroendocrine tumors that can occur in various parts of the body, including the vagal region. Several risk factors have been identified for the development of these tumors.

Genetic predispositions play a significant role, with conditions such as multiple endocrine neoplasia 2 syndrome (MEN2A and MEN2B), von Hippel-Lindau syndrome (VHL), neurofibromatosis type 1 (NF1), and hereditary paraganglioma-pheochromocytoma syndrome being associated with an increased risk.

Additionally, specific gene mutations, such as those in the SDHB, SDHC, SDHD, and MAX genes, are linked to familial forms of paragangliomas.

Environmental factors also contribute to the risk. For instance, residing at high altitudes or having cyanotic congenital heart disease and chronic obstructive lung disease can elevate the risk of carotid body paragangliomas.

Hypoxia, which is common in these conditions, may activate hypoxia-inducible factors (HIFs), which are involved in the development of paragangliomas in some cases.

Other risk factors include a family history of paragangliomas or other neuroendocrine tumors, the presence of extra-adrenal tumors, multiple and/or metastatic tumors, and elevated levels of dopamine and methoxytyramine.

Age is also a factor, with paragangliomas occurring most frequently in individuals between 30 and 50 years of age, although they can occur at any age.

In summary, the risk factors for vagal paragangliomas include genetic syndromes and mutations, environmental factors such as hypoxia, family history, and specific clinical features like extra-adrenal tumors and elevated catecholamine levels.

**SIGNS / SYMPTOMS**

Vagal paragangliomas (VPGLs) are rare neuroendocrine tumors that arise from paraganglia along the vagus nerve. The signs and symptoms of VPGLs can vary depending on the tumor's location and size. Common symptoms include:

* Neck mass: A painless, rubbery, compressible slow-growing mass in the upper neck, with limited vertical but free lateral mobility in 60% of cases.
* Pulsatile tinnitus: A rhythmic thumping in the ear, often associated with the tumor's proximity to blood vessels.
* Hoarseness and dysphagia: Caused by compression of the vagus nerve or other cranial nerves, leading to vocal cord paralysis or difficulty swallowing.
* Horner’s syndrome: Characterized by a constricted pupil, drooping of the upper eyelid, and absence of sweating on the face, which can occur due to compression or invasion of the cervical sympathetic chain.
* Cranial nerve deficits: VPGLs can cause paralysis or dysfunction of cranial nerves, particularly the vagus nerve (cranial nerve X), which is the most commonly affected.
* Dysphagia and odynophagia: Difficulty or pain in swallowing, which may occur if the tumor compresses the esophagus or other structures in the neck.
* Tinnitus: Ringing or buzzing in the ear, which can be a symptom of VPGLs located near the ear or in the parapharyngeal space.
* Syncope: Rarely, the tumor may compress the carotid sinus or internal carotid artery, leading to fainting.
* Hypertension and other catecholamine-related symptoms: In cases where the tumor secretes catecholamines, patients may experience high blood pressure, palpitations, excessive sweating, and anxiety.

It is important to note that VPGLs are often asymptomatic and may be discovered incidentally during imaging or other procedures. However, when symptoms do occur, they are typically related to the tumor's size, location, and potential for hormonal secretion.

Signs and symptoms of paraganglioma happen when the tumor releases too much adrenaline or noradrenaline into your blood. However, some paraganglioma tumors don’t make extra adrenaline or noradrenaline and don’t cause symptoms (are asymptomatic). Common symptoms of paraganglioma include episodes of:

* High blood pressure (hypertension).
* Headaches.
* Excessive sweating for no known reason.
* A pounding, fast or irregular heartbeat.
* Feeling shaky.

Less common symptoms of pheochromocytoma include:

* Being much paler than you usually are.
* Nausea and/or vomiting.
* Diarrhea.
* Constipation.
* Elevated blood glucose levels (hyperglycemia).
* An extreme drop in blood pressure upon standing suddenly (orthostatic hypotension).
* Unexplained weight loss.

Some people who have a paraganglioma may experience symptoms infrequently or in bursts.

Paraganglioma symptoms may depend on where the tumor starts. Paragangliomas most often start in the head, neck, stomach area or pelvis.

Symptoms of a paraganglioma in the head or neck can include:

* A rhythmic pulsing or whooshing sound in the ears, called pulsatile tinnitus.
* Trouble swallowing.
* Hoarse voice.
* Hearing loss.
* Blurred vision.
* Dizziness.

Symptoms of a paraganglioma in the head and neck area might happen as the tumor grows larger. The tumor can press on nearby structures.

When paragangliomas form in other places in the body, the symptoms are more likely to be caused by hormones that the paraganglioma makes.

The hormones, called catecholamines, play a role in how the body responds to stress. They include adrenaline, also known as the fight-or-flight hormone.

Symptoms of paragangliomas that make hormones include:

* High blood pressure.
* Feelings of having a fast-beating, fluttering or pounding heart.
* The sudden loss of color in the face.
* Sweating.
* Headache.
* Uncontrolled shaking in the hands or arms.
* General weakness.

These symptoms may come and go.

Some people with paragangliomas don't have any symptoms. They may learn they have these tumors when imaging tests done for other reasons happen to spot the tumors.

**DIAGNOSIS METHODS**

Preoperative diagnosis of vagal paraganglioma, even though difficult, can be made by combining clinical findings and radiographic studies.

These tumors are easily diagnosed by MRI and, in some cases, with the addition of MR angiography. Contrast-enhanced CT and MRI depict these highly vascular, soft tissue masses equally well. Specific imaging characteristics, like salt-and-pepper appearance due to flow voids in contrast to MRI, are crucial to differentiate carotid body paragangliomas from other tumors of the parapharyngeal space.

CT is indicated for tumors invading the skull base to better delineate the details of the bony erosion and the extent of involvement.

MRI is indicated in most cases and is complementary to CT. It will be evident radiologically that vagal paragangliomas are located behind the internal carotid artery, unlike carotid body paragangliomas, which are found at the carotid bifurcation.

Vagal paragangliomas characteristically displace both the internal and external carotid arteries anteriorly, while the internal jugular vein is compressed and displaced posteriorly.

Our patient also underwent similar investigations and a mass was found posterior to the right carotid bifurcation, external carotid artery, and internal carotid artery. Splaying of the internal carotid and external carotid arteries along with its anterior displacement was found.

However, the presentation of this tumor was rather deceptive, as it was a relatively large tumor with multiple extensions that included the internal carotid artery completely superiorly and partially inferiorly, which made the exact diagnosis tricky,

Finally, scintigraphy with octreotide and PET-CT can be useful devices to aid in identifying multicentric locations, as these tumors express large numbers of somatostatin receptors. However, due to a lack of somatic symptoms, this was also not performed.

Since paraganglioma is a rare tumor and is sometimes asymptomatic, it can be difficult to diagnose. Healthcare providers sometimes find paragangliomas when they order a test or procedure for another reason.

A provider may suspect a diagnosis of paraganglioma after reviewing the following factors:

* A detailed medical history, including previous pheochromocytoma or paraganglioma cases in your family.
* A thorough physical and medical evaluation.
* Certain symptom characteristics, such as high blood pressure that’s unresponsive to standard treatment.

***What tests are used to diagnose paraganglioma?***

Your healthcare provider may use the following tests and procedures to diagnose paraganglioma:

* Physical exam: Your provider will perform a physical exam of your body to check general signs of health, such as your blood pressure. They will also ask you about your medical history, including your family’s medical history as it relates to endocrine issues.
* 24-hour urine test: This type of urine (pee) test involves collecting samples of your urine for 24 hours to measure the amounts of adrenal hormones called catecholamines in your urine. Substances that result from the breakdown of these hormones are also measured. Higher-than-normal amounts of certain catecholamines in your urine may be a sign of paraganglioma.
* Blood catecholamine tests: These tests measure the level of catecholamines in your blood. Substances that result from the breakdown of these hormones are also measured. Higher-than-normal amounts of certain catecholamines in your blood may be a sign of paraganglioma.
* PET scan (positron emission tomography scan): A positron emission tomography (PET) scan uses a safe injectable radioactive chemical called a radiotracer and a device called a PET scanner to produce images of your organs and tissues. The scanner detects metabolically active cells and tumors that absorb large amounts of the radiotracer, which indicates a potential health problem. This imaging test is particularly good for determining the location of paragangliomas.
* CT scan (computer tomography scan): A CT scan is an imaging procedure that takes a series of X-ray images from different angles to provide detailed pictures of areas inside your body. Your provider may recommend a CT scan so that they can determine where the tumor is (most commonly in the neck area).
* MRI (magnetic resonance imaging): An MRI is an imaging procedure that uses a magnet, radio waves and a computer to make a series of detailed pictures of areas inside your body. Your provider may recommend an MRI so that they can look at the region of your body where the tumor is located.

After your provider has diagnosed you with paraganglioma, they’ll likely perform additional tests to see if it has spread to other parts of your body.

***Genetic testing for paraganglioma***

If you’re diagnosed with paraganglioma, your provider will likely recommend genetic counseling to find out your risk for having an inherited syndrome and associated cancers.

Your healthcare provider may recommend genetic testing if any of the following situations apply to you:

* You have a personal or family history of traits linked with inherited pheochromocytoma or paraganglioma syndrome.
* You have signs or symptoms of higher-than-normal catecholamine levels in your blood or cancerous paraganglioma.
* You’ve been diagnosed with paraganglioma before age 40.

If your genetic counselor finds certain gene changes in your testing results, they will likely recommend that your family members who are at risk but don't have signs or symptoms be tested as well.

**TREATMENT OPTIONS**

The treatment options for vagal paragangliomas include surgical resection, radiation therapy, and, in some cases, watchful waiting. Surgical resection is typically the primary treatment, especially for symptomatic or malignant tumors, while radiation therapy is used for malignant or unresectable cases.

Surgical resection involves careful dissection to remove the tumor while preserving cranial nerves, although sacrificing the vagus nerve may be necessary, leading to potential deficits.

Radiation therapy, including stereotactic radiosurgery and fractionated radiotherapy, is an alternative for patients with extensive tumors or those who are not suitable for surgery.

For patients with multifocal disease or those who are not candidates for surgery, a "watch-and-wait" approach may be considered, especially in elderly patients. Additionally, newer treatment modalities such as targeted therapies and radiopharmaceuticals are being explored for advanced or metastatic cases.

In some cases, embolization may be used to reduce tumor size and bleeding before surgery. The choice of treatment depends on factors such as tumor size, location, patient age, and overall health.

Treatment options for paraganglioma depend on several factors, including:

* The size of the tumor.
* If the tumor is benign (not cancer) or malignant (cancer).
* If you have symptoms caused by higher-than-normal levels of catecholamines.
* If the tumor is in one area only or has spread to other places in your body (metastasized).
* If the tumor has been diagnosed for the first time or has come back (recurred).

If you have a paraganglioma that causes symptoms due to excess adrenal hormones, your healthcare provider will likely recommend medication to manage the symptoms.

Medications may include:

* Medication that keeps your blood pressure regulated, such as alpha-blockers.
* Medication that keeps your heart rate normal, such as beta-blockers.
* Medication that blocks the effect of the excess hormones released by your adrenal gland(s).

Treatment options for paraganglioma include:

* Surgery and tumor removal.
* Radiation therapy.
* Chemotherapy.
* Ablation therapy.
* Targeted therapy.

Together, you and your healthcare team will determine a treatment plan that works best for you and your situation.

***Surgery and tumor removal***

Surgery is the main form of treatment for paragangliomas. During the surgery to remove the tumor, your surgeon will check the surrounding tissue and lymph nodes to see if the tumor has spread. If it has, your surgeon will remove the affected tissue(s) as well, if possible.

Most paragangliomas can be removed using minimally invasive techniques such as laparoscopic surgery, which involves making a few small incisions in your skin and removing the tumor with special instruments.

However, traditional open surgery may be needed for large tumors.

After surgery, your provider will check your catecholamine levels in your blood or urine. Normal catecholamine levels are a sign that all the paraganglioma cells were removed.

***Radiation therapy***

Radiation therapy is a cancer treatment that focuses strong beams of energy to destroy cancer cells or keep them from growing while sparing as much surrounding healthy tissue as possible.

There are two types of radiation therapy:

* External radiation therapy: This therapy uses a machine outside your body to send radiation toward cancer.
* Internal radiation therapy: This therapy uses a radioactive substance sealed in needles, seeds, wires or catheters that a healthcare provider places directly into or near cancer.

The type of radiation therapy your provider may recommend depends on whether your cancer is localized, regional, metastatic or recurrent. Providers most often use external radiation therapy and/or 131I-MIBG therapy to treat malignant paraganglioma.

The treatment 131I-MIBG is a radioactive substance infusion that collects in certain kinds of tumor cells, killing them with the radiation that it gives off.

***Chemotherapy***

Chemotherapy is the standard therapy for treating metastatic paraganglioma. It’s a cancer treatment that uses drugs to stop the growth of cancer cells by killing the cells or by preventing them from dividing and multiplying. Chemotherapy is usually given through a vein (intravenously). It’s usually an effective treatment, but it can cause side effects.

***Ablation therapy***

Ablation therapy is a minimally invasive treatment option that uses very high or very low temperatures to destroy tumors. Ablation therapies that can help kill cancer cells and abnormal cells include:

* Radiofrequency ablation: This is a therapy that uses radio waves to heat and destroy cancer cells and abnormal cells. The radio waves travel through electrodes (small devices that carry electricity).
* Cryoablation: This therapy uses liquid nitrogen or liquid carbon dioxide to freeze and destroy cancer cells and abnormal cells.

***Targeted therapy***

Targeted therapy is a treatment option that uses medications or other substances to attack specific cancer cells without harming healthy cells. Healthcare providers use targeted therapies to treat metastatic and recurrent paraganglioma.

Researchers are currently studying sunitinib, a type of tyrosine kinase inhibitor, for treatment for metastatic paraganglioma. Tyrosine kinase inhibitor therapy is a type of targeted therapy that prevents tumors from growing.

**PREVENTION TIPS**

There are no specific prevention tips mentioned in the provided context for vagal paragangliomas. However, general prevention strategies for paragangliomas and other health conditions can be inferred from related medical information.

* Genetic counseling: If there is a family history of paragangliomas or related hereditary conditions, genetic counseling may help identify risks and guide preventive measures.
* Regular medical check-ups: Routine health screenings and medical evaluations can help detect paragangliomas early, especially in individuals with a family history of the condition.
* Healthy lifestyle choices: Maintaining a healthy lifestyle, including a balanced diet, regular exercise, and avoiding tobacco and excessive alcohol consumption, may reduce the risk of various health issues, although specific links to paragangliomas are not mentioned.

Unfortunately, you can’t prevent developing a paraganglioma. However, if you’re at risk for developing a paraganglioma due to certain inherited syndromes and genes, genetic counseling can help screen for paraganglioma and potentially catch it in its early phases.

Talk to your healthcare provider if you have any first-degree relatives (siblings and parents) that have been diagnosed with paraganglioma or pheochromocytoma and/or any of the following genetic conditions:

* Multiple endocrine neoplasia 2 syndrome.
* Von Hippel-Lindau (VHL) disease.
* Neurofibromatosis type 1.
* Hereditary paraganglioma syndrome.
* Carney-Stratakis dyad.
* Carney triad.

**OUTLOOK / PROGNOSIS**

A paraganglioma is a rare neuroendocrine tumor that usually forms along major blood vessels and nerve pathways in the neck and head. The outlook or prognosis for paragangliomas depends on several factors, including whether the tumor is benign or malignant, its location, and whether it has spread to other parts of the body.

For paragangliomas, the prognosis is generally favorable if the tumor is localized and can be completely removed through surgery. However, if the tumor is malignant or has spread to other parts of the body, the prognosis may be less favorable.

In such cases, additional treatments such as chemotherapy, radiation therapy, targeted therapy, ablation, and embolization may be used.

It is important to note that the prognosis can also be influenced by the presence of certain genetic syndromes that are associated with paragangliomas. These syndromes can increase the risk of developing paragangliomas and may affect the overall prognosis.

In summary, the outlook for paragangliomas is generally good, especially for localized and benign tumors. However, the prognosis can vary depending on the specific characteristics of the tumor and the presence of any underlying genetic conditions. It is important to discuss the specific prognosis with a healthcare provider who can provide more detailed information based on the individual case.

***What is the prognosis (outlook) for paraganglioma?***

The prognosis (outlook) for paraganglioma varies depending on certain factors, including:

* Where the tumor is in your body and how big it is.
* If it’s cancer and has spread to other parts of your body.
* If the tumor was able to be surgically removed and if so, how much of the tumor was removed during surgery.

People who have a small paraganglioma that has not spread to other parts of their body (has not metastasized) have a five-year survival rate of about 95%.

People who have paraganglioma that has come back after initial treatment (recurred) or spread to other parts of their body (metastasized) have a five-year survival rate between 34% and 60%.

There are also cases of aggressive paraganglioma tumors that haven’t metastasized but have invaded local tissue to the point where surgery can’t fully remove it. In these cases, the excess release of adrenaline and noradrenaline can be dangerous and difficult to treat.

If paragangliomas are left untreated, whether benign or malignant, they can potentially cause serious, life-threatening complications due to the excess amounts of adrenaline and noradrenaline they can secrete. Complications can include:

* Heart muscle disease (cardiomyopathy).
* Inflammation of your heart muscle (myocarditis).
* Uncontrolled bleeding in your brain (cerebral hemorrhaging).
* Accumulation of fluid in your lungs (pulmonary edema).
* Heart attack (myocardial infarction).
* Stroke.
* Coma.
* Death.

**POSSIBLE COMPLICATIONS**

Possible complications for vagal paragangliomas include surgical morbidity such as cranial nerve dysfunction, which is common after surgery for cervical paragangliomas and has been reported to occur almost always after the excision of tumors originating from the vagus nerve.

These complications can include hoarseness, difficulty in swallowing, and deviation of the angle of the mouth. Additionally, the surgical resection can lead to cardiopulmonary failure and aspiration pneumonia.

Other potential complications include neurologic sequelae from radiation therapy and the risk of recurrence even after successful surgery. If left untreated, paragangliomas can cause serious, life-threatening complications due to the excess amounts of adrenaline and noradrenaline they can secrete, such as heart muscle disease, inflammation of the heart muscle, uncontrolled bleeding in the brain, accumulation of fluid in the lungs, and heart attack.

**WHEN TO SEE A DOCTOR / RED FLAG**

If you’ve been diagnosed with paraganglioma and experience concerning symptoms, contact your healthcare provider.

If you’re experiencing symptoms of paraganglioma, such as high blood pressure and headaches, talk to your provider. Even though paraganglioma is rare and the likelihood of having it is low, it’s important to treat high blood pressure regardless.

If you’ve recently found out that one of your first-degree relatives (siblings and parents) has a genetic syndrome, such as multiple endocrine neoplasia 2 syndrome or von Hippel-Lindau (VHL) disease, that puts you at a higher risk of developing a paraganglioma, ask your provider about genetic testing.

**DIFFERENTIAL DIAGNOSIS**

The differential diagnosis in our patient involved vagal paraganglioma, schwannoma, neuroma, jugular meningioma, carotid body paraganglioma as well as jugular and tympanic paragangliomas. Due to their similar clinical presentation, they were high in our differentials.

However, schwannoma shows moderate enhancement while paragangliomas reveal avid enhancement on imaging. Histologic characteristics of schwannomas include alternating Antoni A and B regions, which are distinctly different from paragangliomas.

Displacement of the common or internal carotid arteries is also characteristic of these tumors. They do not demonstrate flow voids. Hence our initial differentials also revolved around this list and investigations, an anatomical position once exposed intraoperatively, and postoperative histological confirmation.

**RECENT GUIDELINES OR UPDATES**

Paragangliomas are rare tumors that originate from paraganglionic tissue, and they can occur in various locations, including the head and neck region. Vagal paragangliomas specifically arise from the vagus nerve and are characterized by their vascular nature and potential to cause cranial nerve deficits.

Recent guidelines and updates for the management of vagal paragangliomas focus on a multidisciplinary approach involving surgery, imaging, and sometimes embolization.

The primary goal of treatment is to achieve complete resection while preserving cranial nerve function. Surgical techniques such as microsurgical approaches and pericapsular dissection are commonly used to minimize complications and ensure safe removal of the tumor.

In terms of diagnosis, advanced imaging techniques like MRI and CT scans are crucial for identifying the tumor's location, size, and relationship with surrounding structures.

Diagnostic carotid angiography may also be employed to assess the vascular supply of the tumor, which is essential for planning pre-operative embolization if necessary.

For patients with multifocal disease or those who are not suitable candidates for surgery, an expectant policy may be adopted, with regular follow-up and monitoring.

The decision to proceed with surgery is based on factors such as the patient's age, overall health, and the potential for tumor growth or complications.

It is important to note that while the majority of paragangliomas are benign, approximately 20% can be malignant, and these cases require more aggressive management and follow-up.

In summary, the management of vagal paragangliomas involves a comprehensive evaluation and personalized treatment plan, with a focus on preserving neurological function and minimizing surgical risks.

**EPIDEMIOLOGY**

Vagal paragangliomas are rare tumors that develop from paraganglionic tissue around the vagus nerve, with a prevalence of 0.012% of all tumors.

They are the third most common paraganglioma of the head and neck but account for less than 5% of these tumors.

These tumors have a well-established female prevalence, with a female-to-male ratio ranging from 2.7 to 6. Vagal paragangliomas are even rarer, with an incidence of 0.33% among head and neck paragangliomas.

They typically occur in individuals between 30 and 50 years of age, although they can affect people of any age. Approximately 10% of cases occur in children.

**PREDEFINED Q & A SETS**

Q1: What is a vagal paraganglioma?  
A: A vagal paraganglioma is a rare, typically slow-growing neuroendocrine tumor that arises from paraganglia associated with the vagus nerve. These tumors commonly originate in the parapharyngeal space in the neck area.

Q2: What symptoms do vagal paragangliomas cause?  
A: Symptoms may include a painless neck mass, hoarseness, dysphagia, or lower cranial nerve palsies. Occasionally, symptoms related to hormone secretion can occur but are rare in vagal paragangliomas.

Q3: How are vagal paragangliomas diagnosed?  
A: MRI is the preferred imaging modality and can identify the tumor location and extent. Angiography may be added for vascular assessment. Diagnosis may also include biopsy in selected cases.

Q4: What treatment options are available?  
A: Management options include:

* Watchful waiting for small or asymptomatic tumors, especially in older patients.
* Surgical excision is the mainstay for tumors causing symptoms or growth, but it often requires sacrificing the vagus nerve, which can cause permanent nerve deficits. Surgery should be performed by a multidisciplinary skull base team to optimize outcomes and rehabilitation.
* Radiation therapy can stabilize tumor growth and preserve nerve function, often used in elderly patients or those with higher surgical risks.
* Observation might be considered in select cases without symptoms or risks from tumor progression.

Q5: What are the risks and complications of surgery?  
A: Surgical removal usually results in vagal nerve paralysis and may cause deficits in adjacent lower cranial nerves. Rehabilitation following surgery is crucial. Risks must be balanced against benefits, and surgery may be deferred if bilateral nerve damage is a concern.

Q6: Can vagal paragangliomas be malignant?  
A: Malignant transformation is rare but documented. Malignant cases require more aggressive surgical and adjuvant radiation therapy.

Q7: Are vagal paragangliomas hereditary?  
A: About 20% of cases show familial links, with higher rates of multifocal tumors in familial forms. Genetic screening is advised in patients with family history or multifocal disease.

Q8: What is the prognosis?  
A: With appropriate management, prognosis is generally good, but cranial nerve morbidity and quality of life considerations are significant. Long term follow-up is recommended.

If you or a loved one has been diagnosed, discussing these aspects with a multidisciplinary team experienced in skull base tumors is essential for personalized care tailored to tumor size, symptoms, patient age, and risks.

**DOCTOR-PATIENT CONVERSATIONS**

Doctor: Good morning. I understand you have a lump in your neck and perhaps some other symptoms that brought you in today?

Patient: Yes, Doctor. I noticed a painless swelling on the side of my neck. Sometimes when I touch it, I get a coughing fit. I'm a bit worried about what this might be.

Doctor: Thank you for sharing that. The lump you describe could be a vagal paraganglioma, which is a rare tumor that arises from the nerve tissue along your vagus nerve in the neck. These tumors typically present as painless neck masses and can sometimes cause symptoms like a cough or hoarseness due to irritation or involvement of surrounding nerves.

Patient: How do you diagnose this tumor? Do I need special tests?

Doctor: Yes, to diagnose a vagal paraganglioma, we would perform imaging studies such as an MRI or CT scan of your neck, which give detailed pictures of the tumor and its relationship to nearby structures. Sometimes an angiogram or special PET scans are also used. A biopsy may be done carefully, but often imaging is enough to strongly suggest the diagnosis.

Patient: What are my treatment options?

Doctor: Treatment depends on the tumor size, location, and whether it is causing symptoms or growing. For many patients, careful observation with regular imaging is an option if the tumor is small and stable. If the tumor causes symptoms, grows, or shows signs of malignancy, surgery is often recommended to remove it. Surgery can be complex because the tumor lies close to important nerves and blood vessels, and there is a risk of nerve injury, which could affect your voice, swallowing, or shoulder movement. In some cases, radiation therapy may be used to control the tumor, especially if surgery is not possible or the tumor has spread.

Patient: What are the risks of surgery? Will I have any lasting effects?

Doctor: Because the tumor is near the vagus nerve, surgery can lead to vocal cord paralysis causing hoarseness, difficulty swallowing, or changes in voice. There might also be temporary or permanent nerve deficits. However, surgeons use careful techniques to preserve as much nerve function as possible. Recovery varies, and in many cases, patients regain good function over time.

Patient: Can this tumor be cancerous? Will it spread to other parts of my body?

Doctor: Vagal paragangliomas are usually benign, but about 17% to 20% can show malignant behavior and metastasize. We carefully evaluate your tumor to estimate this risk and plan treatment accordingly.

Patient: Are there any lifestyle or activity restrictions I should follow now or after treatment?

Doctor: For now, there are no strict lifestyle restrictions, but avoid activities that cause neck strain or trauma. After treatment, depending on your recovery, you may need voice therapy or swallowing exercises if nerve function is affected. We will guide you on rehabilitation if needed.

Patient: What kind of follow-up do I need?

Doctor: Regular follow-up with clinical exams and imaging is important to monitor for tumor recurrence or growth. We also often recommend genetic testing because paragangliomas can sometimes be hereditary.

Patient: Is this a common tumor? I haven’t heard much about it before.

Doctor: It's a very rare tumor, making up less than 5% of all head and neck paragangliomas. Because of its rarity, it's best managed by a multidisciplinary team experienced in these tumors.

Patient: Thank you for explaining. What should I do next?

Doctor: We'll arrange the necessary imaging tests and a consultation with a head and neck surgeon. We'll discuss the findings together and decide the best individualized treatment plan for you. If you have questions along the way, don’t hesitate to ask.

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