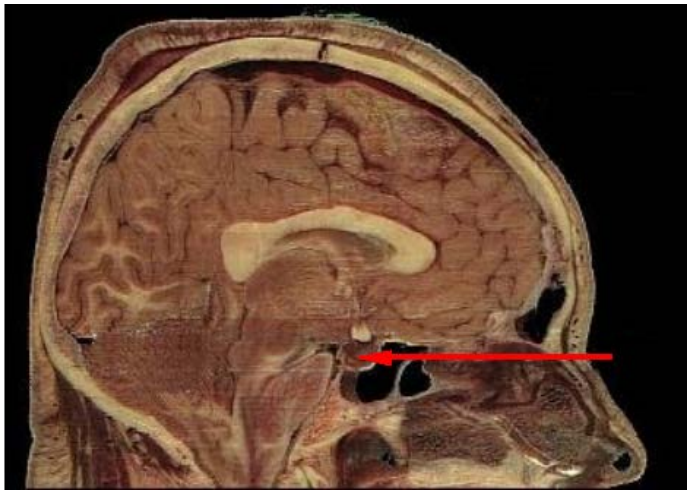
# NANOS Patient Brochure

# Pituitary Tumor

# Pituitary Tumor

## What is the pituitary gland?

Your pituitary gland controls many different hormones in your body. It sits at the bottom of the skull, between your eyes, positioned underneath the brain, just behind your sinuses. It is one of the many organs in your body that help regulate your body functions. Because so many hormones are controlled by the pituitary gland, it is often referred to as the "master gland."



"Pituitary gland" edited by Jim Thomas, uploaded to Wikipedia by Jomegat. Licensed under CC BY-SA 3.0 via Commons https://commons.wikimedia.org/wiki/File:Pituitary_gland.png#/media/File:Pituitary_gland.png

Your pituitary gland produces many different hormones, which have very different effects on your body. **Thyroid stimulating hormone (TSH)** signals your thyroid gland to produce *thyroid hormone*, which helps to regulate your metabolism. **Adrenocorticotropin hormone (ACTH)** activates the release of *cortisol*, the stress hormone. Steroid medications that are used for treating inflammation are based on cortisol. **Growth hormone (GH)** regulates the growth of your bones and other structures in your body. **Follicle stimulating hormone (FSH)** controls ovulation (egg production and release) in women, and sperm production in men. **Luteinizing hormone (LH)** signals menstruation and stimulates estrogen production in women, and regulates testosterone production in men. **Prolactin** signals milk production (lactation), and **anti-diuretic hormone (ADH)** controls urine production and water retention.

Many major blood vessels and nerves surround the pituitary gland. The **internal carotid arteries** are the major vessels that supply blood to the brain, eyes, and other organs in the skull. There is an internal carotid artery on either side of the pituitary gland. The **optic nerves** are the nerves that supply visual information from the eyes to the brain. They join together briefly just above the pituitary gland, called the **optic chiasm**, before splitting again to connect to the brain. There are also **nerves that control eye movements and open your eyelids** that travel next to the pituitary gland, as well as **nerves that sense touch and pain** from the face.

## What is a pituitary tumor?

**Pituitary tumors** are non-cancerous (benign) growths of the pituitary gland. They are typically overgrowths of normal gland tissue (called an **adenoma**). Depending on what type(s) of cells are overgrowing, the growth can overproduce one or more hormones, causing significant hormonal imbalances. These are called **secreting adenomas**, and can be of any size. **Nonsecreting adenomas** do not directly release any hormones. However, if the tumor grows large enough to push on the other pituitary gland cells, it can cause a decrease in other hormones. Very small growths are called **microadenomas**, and very large tumors are called **macroadenomas**.

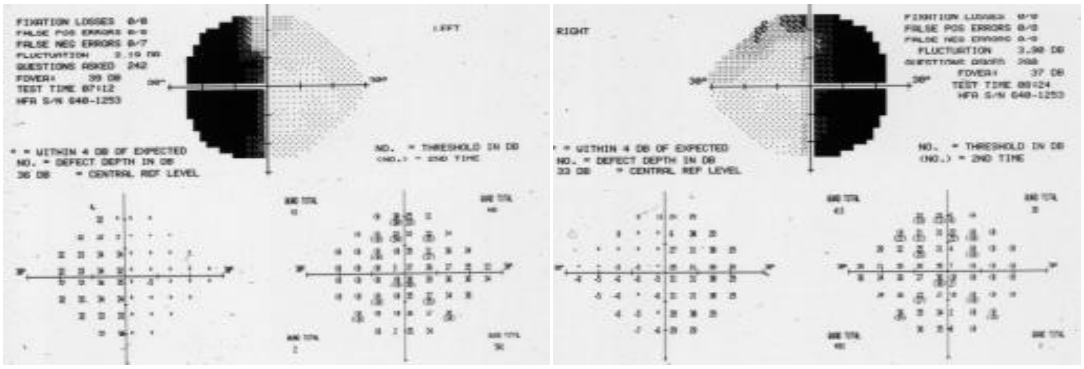
Pituitary tumors are not considered brain tumors, although they are located close to the brain. Most of the largest tumors do not affect the brain, but they can cause other problems (see below).

Pituitary tumors are actually very common! It is estimated that in upwards of 25% (1 out of 4) people have a pituitary tumor. Most people do not have any symptoms and may be unaware they had a tumor until a brain scan is performed for a different reason, such as for a head injury or headache. Pituitary tumors are typically slow-growing and may take years to become detectable. Secreting adenomas tend to be identified earlier, because of the effects the hormones have on the rest of the body.

## How can a pituitary tumor affect my vision?

The optic nerves meet behind the eye at the **optic chiasm** before dividing, which sorts the information from the right half of your vision in each eye from the left half of your vision. Because the optic chiasm sits just above the pituitary gland, a large pituitary tumor can push on the optic nerves or chiasm and cause vision loss. Depending on where the tumor pushes on those nerves, you may notice vision loss in one eye or both eyes. While it will typically affect your outer peripheral vision in either eye, you can also lose central reading vision. Changes to your side (peripheral) vision are often not detected until it has significantly affected your vision.

To help measure your vision, your doctor may perform several tests. A **visual acuity** is measured to determine the reading clarity of your vision, with and/or without glasses. A **visual field** is a test that helps map out your side vision. It is measured by counting fingers or by looking at different lights/objects. Your doctor may use a computer test to measure your visual field, which will provide a print out that may look similar to this:




In the above figures, the left image shows the vision in the left eye, and the right image shows the vision in the right eye, as though you are looking straight ahead. The dark areas represent blind spots. The left half of the vision in the left eye and the right half of the vision of the right eye are dark. When this happens, your ability to see objects or people to either side are limited. You may not be qualified to drive or do certain activities.

More rarely, pituitary tumors can involve other nerves and cause abnormal eye movements, pupil enlargement, droopy eyelids, facial numbness, or double vision that goes away when you cover one eye.

## What other symptoms can I have with a pituitary tumor?

Pituitary tumors often do not cause any symptoms. Headaches are one of the most common complaints, reported in approximately 45% of people with pituitary tumors. Depending on whether or not hormone levels are elevated or decreased, pituitary tumors may have other effects on the body. A hormone specialist (**endocrinologist**) is often consulted to help evaluate hormone levels.

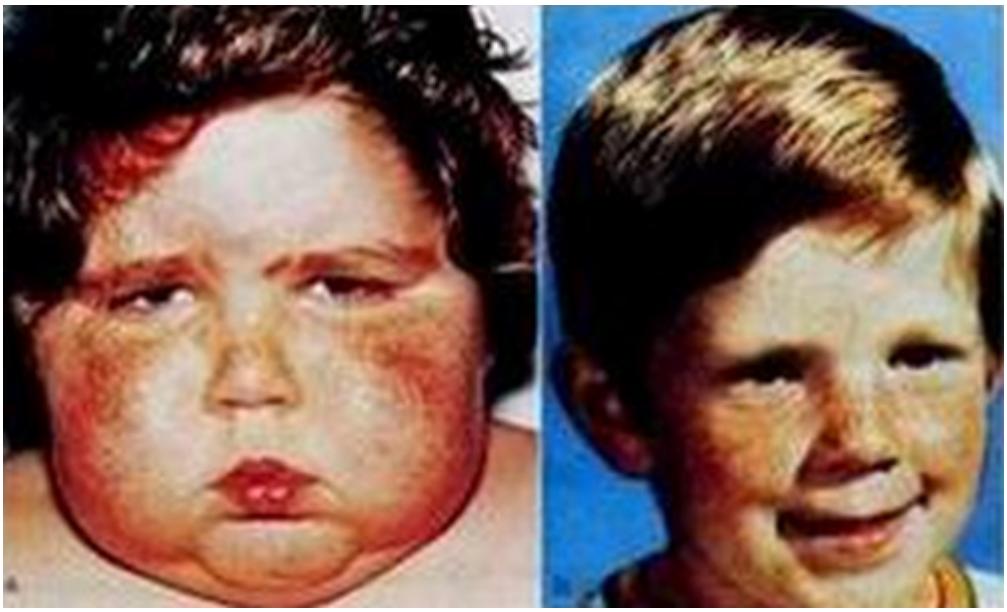
The most common hormone imbalance associated with a pituitary tumor is increased prolactin (**hyperprolactinemia**). Increased prolactin levels cause decreased menstruation, infertility, and increased milk production in women. In men, high prolactin levels cause impotence, decreased sex drive, and decreased sex organ function (**hypogonadism**). Breast tissue can enlarge in both men and women, which is called **gynecomastia**. Often these pituitary tumors are detected during testing for infertility.

Overproduction of growth hormone (GH) results in **gigantism** in children, and **acromegaly** in adults. Gigantism is a condition of excessive growth, with the person becoming extremely tall (7 to 9 feet tall). These people typically look normal except for their size, because growth hormone stimulates all parts of the body to grow symmetrically. Acromegaly is similar to gigantism, as these people will also become extremely large. However, because GH stimulates growth after the growth plates fuse, there are typically facial and skeletal deformities. Andre the Giant, famed wrestler and actor in *The Princess Bride*, is one of the best-known people who had acromegaly.

**Left:** Picture of Robert Wadlow, a person with gigantism.
**Image from:** http://www.ancient-wisdom.com/Images/giant%20robert_wadlow.jpg

**Right:** Picture of Andre the Giant, a person with acromegaly.
**Image from:** http://bashny.net/uploads/images/00/00/45/2014/03/12/473c6a1357.jpg

An overproduction of adrenocorticotropin hormone (ACTH) from a pituitary tumor results in **Cushing syndrome**, which is caused by excess cortisol (stress hormone). People with Cushing syndrome may have their weight redistributed from their arms and legs to their trunk, thinned skin and hair, facial rounding (a "chipmunk face"), fatigue, and/or bruise easily. Because of the effects of high levels of steroids, people with Cushing syndrome are more susceptible to infections; they retain fluid, lose bone density, have increased blood sugar (even without a previous diagnosis of diabetes), and can have increased stomach acid production, among many other symptoms. This phenomenon also occurs with long-term use of steroid anti-inflammatory medications.



**Left:** A child with Cushing's syndrome.
**Right:** The same child without Cushing's syndrome.
**Image from:** http://syndromepictures.com/wp-content/uploads/2011/11/Cushings-Syndrome-pictures.jpg

If thyroid stimulating hormone (TSH) is overproduced, your thyroid hormone levels may be elevated (**hyperthyroidism**). People with high thyroid hormone levels may have various symptoms such as rapid, sometimes irregular heartbeats, heat sensitivity, weight loss, increased appetite, increased feelings of anxiety and/or irritability, diarrhea, fatigue, thinning hair and skin, tremor, or sweating. Hyperthyroidism is typically caused by an overactive thyroid gland; pituitary tumors are a very rare cause.

Decreased levels of anti-diuretic hormone are rarely associated with pituitary tumors. When they are, they can cause a condition called **diabetes insipidus**, which is a type of diabetes not associated with elevated blood sugar or the need for insulin. It causes excessive thirst and increased urination, and can lead to severe dehydration.

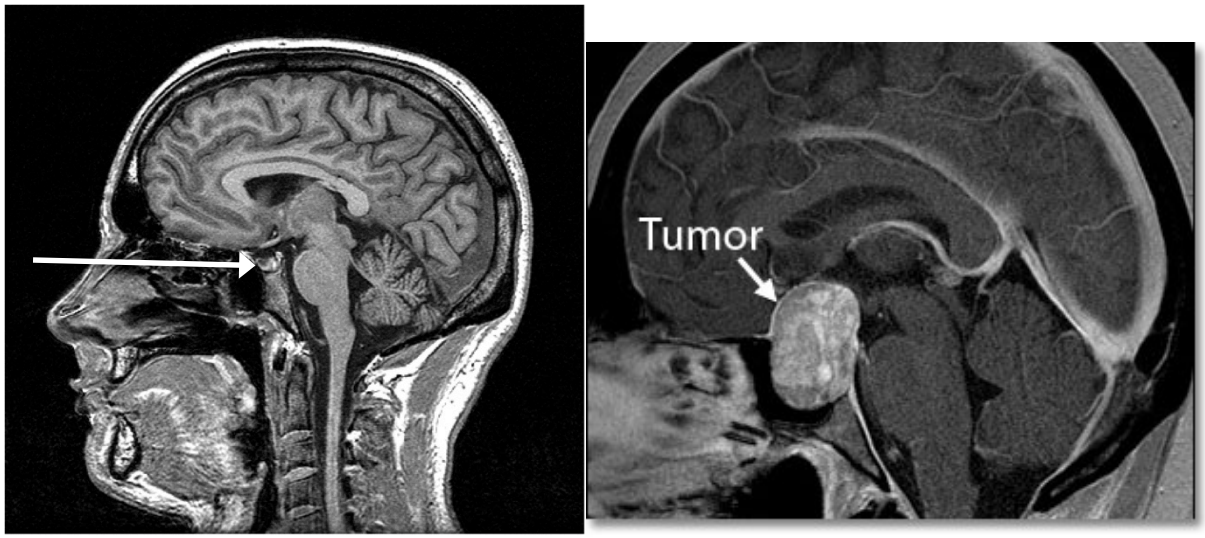
Sometimes pituitary tumors will cause decreased production of some hormones, called **hypopituitarism** (or **panhypopituitarism** if all hormone production is decreased), which can occur when the tumor pushes against the healthy cells and prevents those cells from releasing their hormones. If this happens, you may feel extremely ill, feverish, sluggish, or fatigued.

Very rarely, there can be rapid bleeding into the tumor, a condition known as **pituitary apoplexy**. Patients with pituitary apoplexy will have a very sudden-onset of a headache, often associated with double vision or blurred vision. They may also feel feverish or have flu-like symptoms. If you develop these symptoms, you should call your doctor to see if you need urgent medical attention.

## Could I have something other than a pituitary tumor?

Your doctor probably has or will order some tests to help determine what you have. This may include a brain scan, which is either a **CT scan** (which uses X-ray technology) or an **MRI scan** (which uses magnets and computers) to construct a 3-D image of your head. The images will help visualize the presence, size, and

characteristics of the tumor, and help determine how close it is to your optic nerves.




**Left:** MRI scan showing a normal pituitary gland (white arrow).
**Image from:** http://www.mylifewithabrokenbrain.co.za/wp-content/uploads/2014/01/sagittal_scan.jpg

**Right:** MRI scan showing a pituitary tumor (labeled arrow).
**Image from:** http://neurosurgery.ucla.edu/images/Pituitary%20Program/NonFunction_MRI.jpg

While tumors that grow from the pituitary gland itself are the most common cause for growths in that area of the head, there are some other rare causes that your doctor will also evaluate.

## Why do I need to see so many doctors?

Because pituitary tumors affect so many systems in your body, you may need to have many different specialists who will help coordinate the best care for you. An **ophthalmologist** or **neuro-ophthalmologist** monitors your central vision and peripheral vision and treats any double vision you may have. An **endocrinologist** monitors your hormones and treats any hormonal imbalances. A **neurologist** may monitor the tumor and treat the headaches. A **neurosurgeon** may need to perform surgery to treat the tumor, and a **radiation oncologist** may be asked to help direct any radiation therapy.

## Will I need treatment? What are the treatment options?

Not all tumors need to be treated! Tumors that are very small and do not cause any symptoms may be monitored without treatment. However, if you have symptoms of any kind, you will most likely require some form of treatment.

Hormonal imbalances are treated with **medications** prescribed by your endocrinologist. Some medications are designed to mimic normal hormones, which can cause tumor cells to shrink but not disappear. Because the tumor cells can regrow, these medications often need to be continued indefinitely. Other medications are used to control symptoms of excess hormone secretion (especially acromegaly), and are less effective at shrinking the tumor.

**Surgery** (performed by a neurosurgeon) may be needed if the tumor is too large to be controlled with medication alone and is dramatically affecting vision. The goal of surgery is typically to reduce the size of the tumor, called "debulking." Some tumor cells are almost always left behind in surgery. Because these cells tend

to grow slowly, additional treatment may not be necessary unless they grow enough to cause more symptoms. There are several techniques to the surgery; the way the surgery is performed is determined by your specific situation. The most common way is through the nose or under the lip, which can decrease scarring and healing time. This approach is called **transsphenoidal hypophysectomy** (TSH or **pituitectomy**). However, some tumors have to be removed through the scalp.

In some cases, **radiation therapy** may be recommended as part of your treatment plan. Radiation is targeted at the pituitary gland to "kill" tumor cells. While every effort is made to treat only the tumor cells, the surrounding tissue may be affected. Some techniques require multiple, small-dose treatments over consecutive days (**fractionated**), while other forms use a higher focal dose (**stereotactic radiosurgery**). **Gamma knife** and **LINAC** are examples of stereotactic treatments your doctor may recommend. If the pituitary tumor is pressing on the optic nerve, focal radiation cannot be used. Because radiation therapy may affect normal pituitary gland function, you may need hormone supplements after being treated. Radiation may help prevent further growth of the tumor but does not shrink it. For this reason, radiation is typically used with medication or surgery. In rare circumstances, radiation is used alone if surgery is not possible.

For people struggling with **double vision** as a result of a pituitary tumor, shrinking the tumor with medication or removing a large part of it with surgery can make the double vision go away. There can be some residual double vision or eye movement problems. Those symptoms can be temporarily relieved with a patch, putting Scotch tape over one of the lenses in your glasses, or with a temporary prism on your glasses. Once your doctor is sure your eye movements are not changing, more permanent treatments such as **prisms in glasses** or **eye muscle surgery** can reduce or eliminate double vision.

People with **pituitary apoplexy** may require hospital admission to receive close monitoring, hormone or steroid treatments, and sometimes surgery to help drain the blood from the tumor.

## What is my prognosis? Will my vision get better?

Pituitary tumors are typically benign (non-cancerous) and slow-growing. Most of the hormone imbalances are very treatable. Vision is one of the more unpredictable aspects of pituitary tumors, as it can often be difficult to determine what the chance is for visual improvement. As a general rule of thumb, the chance of visual improvement is best predicted by how long the damage to the optic nerve has been there. Your eye doctor (ophthalmologist or neuro-ophthalmologist) should be able to determine whether or not there is evidence of **optic atrophy**, a sign of permanent damage to the optic nerve. However, even when there are signs of permanent damage to the optic nerves, there can be good visual recovery if the pressure on the optic nerves is relieved with medication and/or surgery. For this reason, all people with a pituitary tumor and vision loss should be evaluated for treatment to relieve pressure on the optic nerves, regardless of how long the vision loss has been present.

## How often do I need to have checkups for my pituitary tumor?

Initially, if you do not have any vision loss, your ophthalmologist or neuro-ophthalmologist may want to check on you every 3-6 months to make sure you do not develop any vision problems. If you are starting to have vision problems, your eye doctor may see you more frequently. Your eye doctor will also need to examine you before and after any surgery or radiation treatments to measure your central reading vision and visual fields. Once your vision is stabilized and hopefully normalized, you and your doctor may choose to extend your follow-up appointments.

You will also need to be followed by your endocrinologist or primary care provider on a regular basis to monitor your hormone levels. You will probably have blood tests several times per year. If you are on hormone

replacement or medication to decrease hormone levels, your doctor will need to check up on you to make sure you do not need a change in the dose of the medications.

Because pituitary tumors continue to grow slowly, even after a successful surgery, recurrence is possible. To help monitor the size and growth of the tumor, a brain scan (typically an MRI) should be checked on a regular basis. How often you need to be checked is dependent on your situation and should be discussed with your doctors.

## Additional Reading/Resources

- **Pituitary Tumor, by the National Library of Medicine on MedlinePlus** (https://www.nlm.nih.gov/medlineplus/ency/article/000704.htm)
    - This article, written by the U.S. National Library of Medicine, is short but provides a lot of useful information about pituitary tumors. It also contains a video about the pituitary gland and how various hormones affect the body.
- **Pituitary Tumors, by The National Cancer Institute at the National Institutes of Health** (http://www.cancer.gov/types/pituitary)
    - Most pituitary tumors are not cancerous, but the National Cancer Institute has a good handout that describes pituitary tumors.
- **Pituitary Tumors, by the Mayo Clinic** (http://www.mayoclinic.org/diseases-conditions/pituitary-tumors/basics/definition/con-20028814)
    - The Mayo Clinic has a several-page web article about pituitary tumors. Less technical and more geared towards patients, this may be a helpful resource for more information from one of the leading medical centers in the United States.
- **Pituitary Tumors, by The American Cancer Society** (http://www.cancer.org/cancer/pituitarytumors/)
    - Although pituitary tumors are not typically cancerous, the American Cancer Society has a lot of information about pituitary tumors, how they are treated, and finding support.
- **Pituitary Tumors, by the Johns Hopkins Neurology and Neurosurgery Department** (http://www.hopkinsmedicine.org/neurology_neurosurgery/centers_clinics/pituitary_center/pituitary-tumor/)
    - Like the Mayo Clinic, Johns Hopkins has a several-page web article about pituitary tumors that provides some basic information about the condition and how it is treated. There are several links to support groups for people who have a pituitary tumor.
- **Pituitary Adenoma, by the American Academy of Ophthalmology's EyeWiki Project** (http://eyewiki.aao.org/Pituitary_Adenoma)
    - This article is a more technical description of pituitary tumors and may be too detailed for most readers, but contains up-to-date information about how doctors manage and treat pituitary tumors.
- **Pituitary Adenoma, by Wikipedia** (https://en.wikipedia.org/wiki/Pituitary_adenoma)
    - This may also be too technical for most readers, but may be less difficult to read than the EyeWiki article. Because Wikipedia is written and edited by the general public, it may be less accurate than the above websites, which are expert-written. Nevertheless, it is helpful when used in addition to other resources.

## Support Groups

(List found at the Johns Hopkins Neurology and Neurosurgery Pituitary Tumor page - http://www.hopkinsmedicine.org/neurology_neurosurgery/centers_clinics/pituitary_center/patient/support-groups.html)

- **The Pituitary Network Association** (http://pituitary.org)
  - This is an international non-profit organization founded by people with acromegaly whose mission is to provide support for people with pituitary tumors and promote awareness and advocacy for pursuing treatments and cures for pituitary tumors.
- **The Pituitary Society** (https://pituitarysociety.org/)
  - This group provides information about pituitary disease. The society, while geared towards medical professionals, researchers, and trainees, aims to provide up-to-date information about the understanding, diagnosis, and treatment of pituitary diseases.
- **The American Brain Tumor Association** (http://www.abta.org/)
  - Although pituitary tumors are not true brain tumors, the American Brain Tumor Association has a great guide for people who have just been diagnosed with a tumor, as well as providing useful educational information and links to support groups, sortable by location (as well as links to online groups).
- **The National Brain Tumor Society** (http://www.braintumor.org/)
  - The National Brain Tumor Society serves as an advocacy group that raises money for awareness and research into various brain tumors. Their website has some helpful information about different brain tumors, as well as getting involved in support groups and contributing to brain tumor research.
- **The Hormone Health Network** (http://www.hormone.org/)
  - The Hormone Health Network has handouts describing the different hormonal imbalances that can occur with pituitary tumors. They also have links to various support groups, depending on the hormone condition.