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Pituitary Tumors: adenoma, craniopharyngioma, cyst basic level

Overview

Pituitary tumors grow from the pituitary gland, the master gland of the body, located deep in the skull. These tumors can affect the whole body by interfering with normal hormone production. As the tumor grows, it can cause a variety of symptoms including compression of nearby nerves, resulting in vision problems. There are various kinds of pituitary tumors: adenomas, craniopharyngiomas, and Rathke's cleft cysts. Most are benign (not cancerous) and are often curable. Treatment options aim to remove the tumor or control its growth and restore normal hormone function. You may need medications to correct hormone levels.

The pituitary and sellar region

The pituitary gland is a small, bean-shaped organ that sits at the base of the brain, behind the bridge of the nose (Fig. 1 and 2). The gland is divided into a larger anterior lobe (adenohypophysis) and a smaller posterior lobe (neurohypophysis). It sits in a small pocket of bone in the base of the skull called the sella turcica. Within this sellar region, the pituitary gland is connected to the hypothalamus of the brain by the pituitary stalk. The pituitary gland is bordered on either side by the cavernous sinuses and below by the sphenoid sinus. Unlike the sphenoid sinus, which is an air-filled sinus that drains into the nose, the cavernous sinuses are a network of veins. The internal carotid arteries and the nerves that control eye movement lie within the cavernous sinuses. Directly above the pituitary gland is the optic chiasm, which is responsible for vision (see Anatomy of the Brain). When pituitary tumors grow they can compress the above-mentioned structures and cause problems.

Known as the master gland, the pituitary controls the other endocrine glands in the body. It secretes hormones that control sexual development, promote bone and muscle growth, respond to stress, and fight disease. A healthy pituitary gland releases secretions into the bloodstream and provides feedback to the hypothalamus. The hypothalamus then regulates pituitary hormone levels depending on the needs of the body.

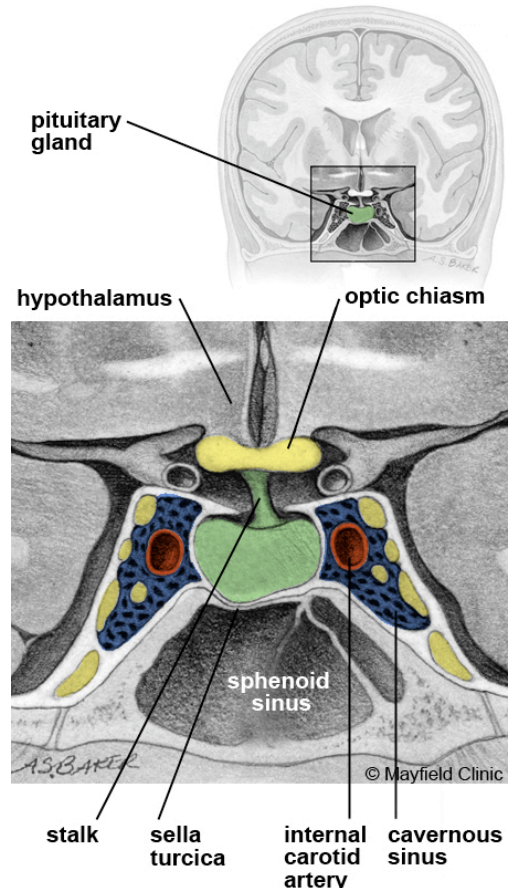


Figure 1A. Front view. A cross-section of the pituitary gland (green) shows its relationship to the optic chiasm above, the sphenoid sinus below, and the cavernous sinuses on each side.

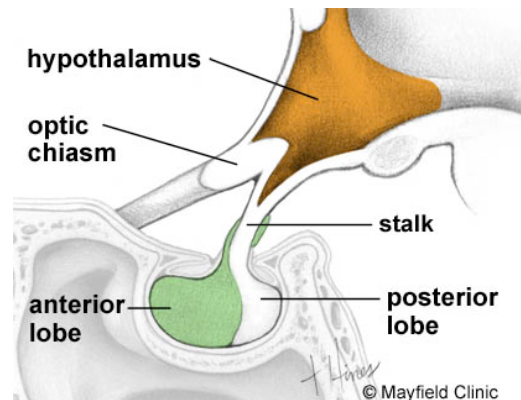


Figure 1B. Side view. A cross-section of the pituitary gland inside the bony sella turcica.

Hormones made by the pituitary gland include:

- Prolactin hormone: causes a woman's breasts to make milk after pregnancy.
- Growth hormone: helps control body growth and the metabolism of sugar and fat.
- Adrenocorticotrophic hormone: causes the adrenal glands to make the hormone cortisol. Cortisol helps control the use of sugar, protein, and fats in the body, and helps the body deal with stress.
- Thyroid-stimulating hormone: causes the thyroid gland to make other hormones that control growth, temperature, and heart rate.
- Antidiuretic hormone (ADH or vasopressin): regulates water balance. Too little of this hormone can cause diabetes insipidus. Too much of this hormone can cause syndrome of inappropriate ADH. Both of these conditions affect the kidneys.
- Luteinizing hormone and follicle-stimulating hormone: control the menstrual cycle in women and sperm production in men.

What is a pituitary tumor?

A tumor that grows from the pituitary gland is called an adenoma. Adenomas most often arise in the anterior lobe of the pituitary gland. Closely related to pituitary adenomas are craniopharyngiomas and Rathke's cleft cysts. These tumors grow from embryonic remnant cells of the pituitary. Craniopharyngiomas typically grow from the pituitary stalk upward into the third ventricle and cause similar symptoms to pituitary adenomas. Tumors of the pituitary gland occur in 15 to 20% of adults and are classified as functional or nonfunctional.

Functional tumors secrete abnormal levels of hormones and interfere with the normal hormone regulation process. These tumors behave according to their cell of origin and are named for the specific hormone they produce. For example, if a tumor originates in a prolactin-producing cell, you may develop a prolactin-secreting pituitary tumor.

Nonfunctional tumors do not secrete hormones. Instead, they grow until their size and mass effect cause headache, vision loss, nausea, vomiting, or fatigue. Based on size, pituitary tumors can be either microadenomas (less than 10mm) or macroadenomas (larger than 10mm). Large tumors can press on the optic nerves and invade the cavernous sinuses, which house the carotid arteries and the nerves involved in eye movement.

What are the symptoms?

Symptoms of a pituitary tumor vary depending on its size and hormone secretion function; many are asymptomatic. About 26% of pituitary tumors are nonfunctional; 74% are functional. Of hormonally active adenomas, about 50% secrete prolactin, 20% secrete growth hormone, 20% secrete

adrenocorticotrophic hormone, and 10% secrete multiple hormones.

Prolactin-producing tumors. The most common pituitary tumor, a prolactinoma, causes an overproduction of prolactin, a hormone that helps control sexual function. In women, the tumor can cause menstruation to stop (amenorrhea) or inappropriate production of milk by the breasts (galactorrhea). In men, prolactin-producing tumors may cause enlarged breasts (gynecomastia), erectile dysfunction or impotence, infertility, decrease in body hair, and low sex drive.

Growth hormone-producing tumors. Most commonly found in men, these pituitary tumors are larger and may extend toward the third ventricle of the brain and/or the cavernous sinus. These tumors may cause gigantism in children or adolescents. Adults may develop acromegaly – an enlargement of bones in the hands, feet or face. Other symptoms include excess sweating, high blood pressure, heart disease, diabetes, and arthritis.

Adrenocorticotrophic hormone (ACTH)-producing tumors. ACTH-producing adenomas occur more often in women. The ACTH hormone stimulates the adrenal gland to secrete cortisol. Excess cortisol causes Cushing's syndrome – a fatty hump between the shoulders; weight gain in the face, neck, and trunk of the body; and pink or purple stretch marks on the skin. Cushing's can also cause diabetes, menstrual irregularities, excessive hair growth, bruising, hypertension, and bone fractures from calcium depletion.

Thyroid-stimulating hormone-producing tumors. TSH-secreting tumors can cause hyperthyroidism. Hyperthyroidism can accelerate your body's metabolism, causing sudden weight loss, a rapid or irregular heartbeat, and nervousness or irritability.

Non-secreting tumors. Non-functioning pituitary tumors have few symptoms and are difficult to recognize until they grow quite large. This excessive growth can press against nearby optic nerves, causing headaches or vision loss. It can also cause the pituitary gland to decrease secretion of hormones, which can cause fatigue, weakness, loss of body hair, and pale skin.

In rare instances, a pituitary tumor can suddenly bleed (hemorrhage) – a condition called pituitary apoplexy. This loss of blood supply can cause tumor cell death, bleeding, and swelling. Symptoms include sudden onset of a severe headache and vision changes, including vision loss, double vision, or drooping of an eyelid. Pituitary apoplexy requires emergency treatment, usually with corticosteroids and possibly surgery.

What are the causes?

Like most brain tumors, the cause of pituitary tumors is unknown. Some researchers believe they occur when a cell in the pituitary gland becomes abnormal or mutates. The cell then reproduces additional mutations until a tumor is formed.

Who is affected?

Pituitary tumors affect 15 to 20% of adults; some do not cause symptoms. They can occur in every age group, but are more common after puberty.

How is a diagnosis made?

If you have symptoms that suggest a pituitary tumor, your physician will work with a team of specialists to confirm the diagnosis. This team may include a neurosurgeon, otolaryngologist (ear, nose, and throat surgeon), endocrinologist, ophthalmologist, pathologist, and radiologist.

First, the doctor will obtain your personal and family medical history and perform a physical examination. In addition to checking your general health, the doctor performs a neurological exam to check mental status and memory, cranial nerve function (sight, hearing, smell, tongue, and facial movement), muscle strength, coordination, reflexes, and response to pain. Additional tests may include:

- Magnetic Resonance Imaging (MRI) scan is a noninvasive test that uses a magnetic field and radiofrequency waves to give a detailed view of the soft tissues of the brain. It views the brain 3-dimensionally in slices that can be taken from the side or from the top as a cross-section. A dye (contrast agent) may be injected into your bloodstream. MRI is very useful to evaluate brain lesions and their effects on surrounding brain (Fig. 3).
- An endocrine evaluation measures hormone levels in your blood or urine to detect abnormal levels caused by pituitary tumors.
- A visual field acuity test is performed by a neuro-ophthalmologist to detect vision loss and missing areas in your field of view. This test measures both central vision (how much is seen when looking straight ahead) and peripheral vision (how much is seen in all other directions while staring straight ahead). The eyes are tested one at a time.

What treatments are available?

Treatment options vary depending on the type, grade, size and location of the tumor; whether it has spread; and your age and general health. Medication, surgery, and radiation either alone or in combination, are used to treat pituitary tumors and return hormone levels to normal.

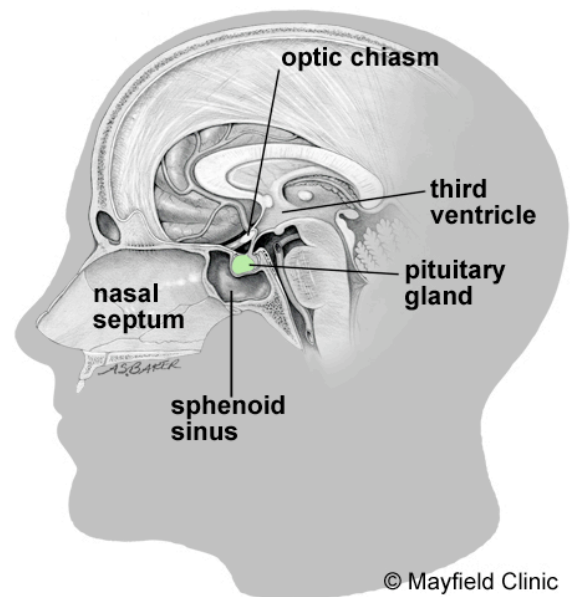


Figure 2. Side view. The pituitary gland (green) is located deep within the skull in an area called the sella. The pituitary gland is related to the optic chiasm above and the sphenoid sinus below.

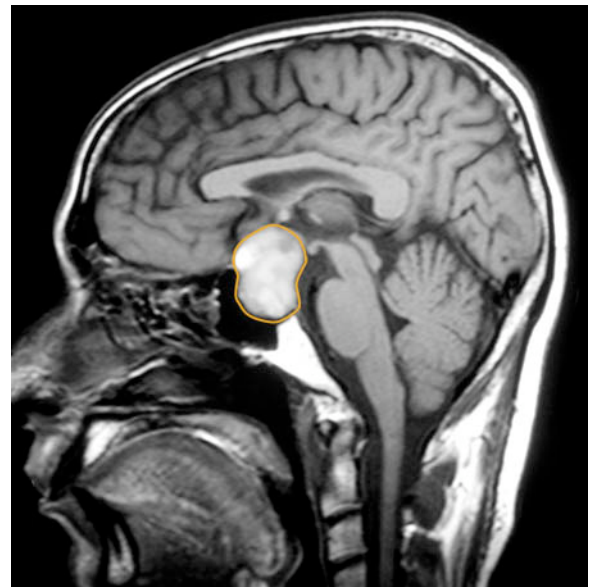


Figure 3. An MRI scan of a pituitary macroadenoma (orange line) that has expanded upward to compress the optic chiasm.

Medication

The goal of medication is to block the tumor from making abnormal quantities of hormones. Specific drugs are used to control specific types of hormone secretion. The production of prolactin can be controlled with the drugs cabergoline (Dostinex) or bromocriptine (Parlodel), which reduces the size of the tumor while maintaining normal prolactin levels. About 80% of patients will have prolactin levels restored to normal with cabergoline. Many will have marked tumor shrinkage, making surgery unnecessary.

The production of growth hormone can be controlled with the drugs octreotide (Sandostatin) or pegvisomant (Somavert) and is used in conjunction with surgical removal. These drugs are also used on recurrent tumors.

To control the production of ACTH, the drugs mitotane (Lysodren), ketoconazole (Nizoral), and aminoglutethimide (Cytadren) may be prescribed.

Surgery

Surgical treatment of a pituitary tumor may be performed as a minimally invasive or open transsphenoidal approach, or a craniotomy. The best option varies for each patient depending on the tumor size, type, and location. Removing the tumor surgically may be the only treatment necessary. For growth hormone-producing and ACTH-producing tumors, surgery is the treatment of choice to reverse the endocrine problems. A neurosurgeon and ear, nose, and throat (ENT) surgeon work as a team to remove pituitary tumors. Sometimes only part of the tumor is removed if it is near critical areas. A partial removal can still relieve symptoms. Radiation or chemotherapy may be used on the remaining tumor cells.

- **Endoscopic transsphenoidal.** This minimally invasive approach uses no skin incision. An ENT surgeon inserts an endoscope through the nostril to reach the sphenoid sinus. An endoscope is a thin, tube-like instrument with a light and a camera. Video from the camera is viewed on a monitor. An opening is made in the sphenoid sinus to access the sella and pituitary gland. Next, the neurosurgeon removes the tumor by passing long instruments through the nostril while watching the monitor (Fig. 4). A large tumor may be difficult to remove with this procedure, especially if it invades nearby nerves or brain tissue.

Using an endoscope to remove pituitary tumors reduces the trauma to the nasal tissues and the recovery time for the patient. The endoscopic technique is replacing the traditional transsphenoidal approach. The traditional technique uses an incision made under the upper lip (sublabial) and removal of the nasal septum to access the sphenoid sinus and sella.

- **Craniotomy.** A skin incision is made in the forehead or eyebrow. A small bone flap above the eye (supraorbital craniotomy) is cut and removed to access the brain (Fig. 5). The brain is gently retracted to locate and remove the tumor. The bone flap is replaced and secured with tiny plates and screws. This craniotomy approach is used for large tumors that have invaded nearby tissues and cannot be removed through a transsphenoidal approach.

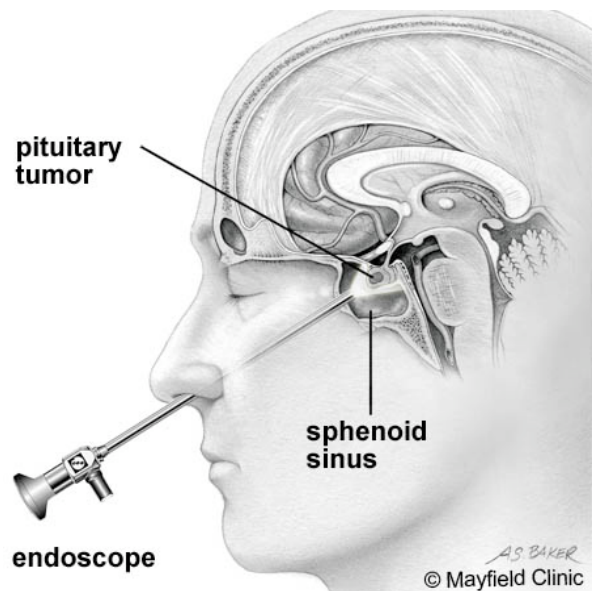


Figure 4. Endoscopic transsphenoidal approach through the nose to a pituitary microadenoma. An opening is made in the sphenoid sinus to access the sella and remove the pituitary tumor.

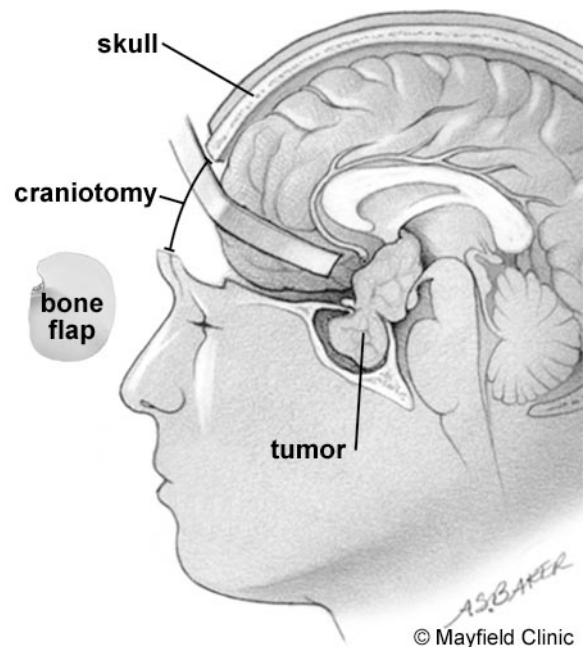


Figure 5. Craniotomy approach to a pituitary macroadenoma that extends down into the sphenoid sinus and up into the third ventricle. A bone flap is cut above the eye (supraorbital) to expose the brain. Retractors gently open a corridor to the tumor.

Interventional MRI or CT is a specially designed operating room in which the patient can undergo an MRI or CT scan before, during, and after surgery. This enables the surgeon to have real-time images of the patient's brain and to know exactly how much tumor has been removed prior to ending the procedure.

Radiotherapy

Radiation therapy uses controlled high-energy rays to treat pituitary tumors. Radiation works by damaging the DNA inside cells making them unable to divide and reproduce. The goal of radiation therapy is to maximize the dose to abnormal tumor cells and minimize exposure to normal healthy cells. The benefits of radiation are not immediate but occur over time. Gradually, the lesion will stop growing, shrink, and in some cases, completely disappear. Aggressive tumors, whose cells divide rapidly, typically respond more quickly to radiation.

External beam radiation is delivered from outside the body by a machine that aims high-energy rays (x-rays, gamma rays) at the tumor. There are two ways to deliver radiation: a single high dose (radiosurgery) or multiple low doses (radiotherapy).

- Stereotactic radiosurgery (SRS) delivers a high dose of radiation during a single session. Although it is called surgery, no incision is made. Because a single radiosurgery dose is more damaging than multiple fractionated doses, the target area must be precisely located and completely immobilized with a stereotactic head frame or mask.
- Fractionated stereotactic radiotherapy (FSR) delivers a low dose of radiation over many visits. A facemask is used to precisely locate the tumor and accurately reposition the patient for each treatment session. Patients return daily over several weeks to receive the complete radiation dose.

Observation (growth monitoring)

Sometimes the best treatment is observation. Small, slow growing tumors that have few symptoms may be observed with routine MRI scans until their growth or symptoms necessitate surgery. Observation may be the best option for older patients with other health conditions. You and your doctor can weigh the risk of symptoms developing versus the risk of treatment intervention.

Sources & links

If you have more questions, please contact the Mayfield Clinic at 800-325-7787 or 513-221-1100. For information about the University of Cincinnati Neuroscience Institute's Brain Tumor Center, call 866-941-8264.

Links

Pituitary Network Association, www.pituitary.org

National Brain Tumor Society, www.brainumor.org
American Brain Tumor Association, www.abta.org

For local support groups, contact The Wellness Community of Greater Cincinnati at 513-791-4060.

Glossary

acromegaly: enlargement of the hands, feet or face in adults due to overproduction of growth hormone; often from a growth hormone-secreting pituitary tumor.

adenoma: a tumor that grows from a gland.

apoplexy: sudden bleeding inside an organ.

Cushing's syndrome: an endocrine disease caused by increased levels of cortisol in the body; often from an adrenocorticotropic hormone (ACTH)-secreting pituitary tumor. Hallmark signs include a fatty hump between the shoulders, a rounded face, and pink or purple stretch marks on the skin. Also caused by excessive use of corticosteroid medication.

diabetes insipidus: a disorder in which there is an abnormal increase in urine output, fluid intake, and often thirst. Caused by a decrease in vasopressin hormone due to damage of the posterior pituitary lobe.

gigantism: excessive growth and height in children caused by overproduction of growth hormone; often from a growth hormone-secreting pituitary tumor.

hormone: a chemical substance produced in the body that controls and regulates the activity of certain cells or organs.

hyperthyroidism: increased heart rate, weight loss, nervousness, and sleeplessness caused by excess thyroid hormone; can be caused by thyroid-stimulating hormone (TSH)-producing pituitary tumor.

hypothalamus: a part of the brain that regulates pituitary hormone responses by secreting releasing factors or inhibiting factors, depending on the needs of the body.

prolactinoma: a benign pituitary tumor that overproduces the hormone prolactin. Too much prolactin causes abnormal milk production in the breasts, lack of menstruation, infertility, and decreased sex drive.

sphenoid sinus: an air-filled, mucous-lined cavity in the skull located behind the nose and between the eyes.

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