Pituitary Tumors

American Brain Tumor Association

FOCUSING ON TUMORS

A Word About ABTA

Founded in 1973, the not-for-profit **American Brain Tumor Association** has a proud history of funding research, providing patient services, and educating people about brain tumors. Our mission is to eliminate brain tumors through research and to meet the needs of brain tumor patients and their families.

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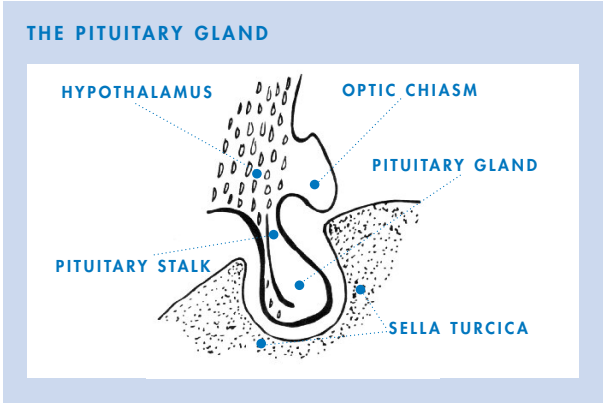
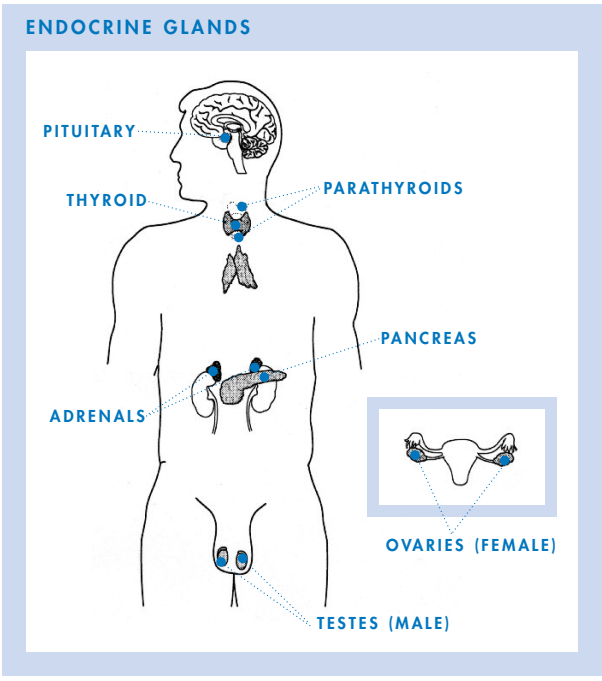
Introduction

The pituitary gland is a bean-sized organ located in the midline at the base of the brain, just behind the bridge of the nose, in a bony pouch called the "sella turcica."



The pituitary itself is known as the "master gland" because it helps to control the secretion of hormones from a number of other glands and organs in the body. These include the thyroid, the adrenals, testes, and ovaries. The pituitary gland releases hormones into the blood stream, where they are carried to distant glands or organs in the body. Those distant glands release other hormones which, in turn, feed back to the brain through the bloodstream. Once back in the brain, hormones cause the hypothalamus (a part of the brain near the pituitary) to signal the pituitary gland to secrete more hormones or slow down hormone production, depending on the needs of the body. A stem-like stalk connects the pituitary gland to the hypothalamus and it is through this stalk that the hypothalamus sends signals to control the activity of the pituitary gland.

The medical term for pituitary tumors is "pituitary adenoma" — *adeno* means gland, *oma* means tumor. Most pituitary adenomas develop in the front two-thirds of the pituitary gland. That area is called the adenohypophysis, or the anterior pituitary. Pituitary tumors rarely develop in the rear one-third of the pituitary gland, called the



neurohypophysis or the posterior pituitary. The tumors are almost always benign and are very treatable. Some tumors can be treated effectively with medications while others require surgery. Because the pituitary gland is important in the function of other glands in the body, treating a pituitary tumor requires an active coordinated multi-disciplinary health care approach, support, and follow-up.

Incidence

Pituitary tumors account for 9 – 12% of all primary brain tumors, making them the third most common primary brain tumor in adults following meningiomas and the gliomas. Abnormalities including small tumors and benign cysts within the pituitary are quite common. Although exact statistics are not yet available on these tumors (registries only recently began counting pituitary tumors in their data) it is estimated that 20-25% of the general population may have small, symptomless pituitary tumors. It appears that 10% of the general population will have an abnormality big

enough to see on magnetic resonance imaging (MRI). These abnormalities most often do not cause symptoms and generally do not require medical or surgical therapy.

Pituitary tumors can be found in every age group, but their incidence tends to increase with age. Functioning (also called secreting) tumors most frequently occur in younger adults. Non-functioning (non-secreting) tumors tend to occur in older adults. Women are diagnosed with pituitary tumors slightly more often than men. This may be due to the tumors' interference with the female menstrual cycle, which sometimes makes symptoms more obvious.

Causes

Pituitary tumors, similar to tumors located elsewhere in the body, develop from one single abnormal cell that multiplies into many abnormal cells, eventually forming a tumor. Stimulation from the hypothalamus may also contribute to tumor growth. If your doctor determines you have a tumor, the next step is learning the "type" of pituitary tumor.

Types of Tumors

Pituitary tumors may be classified and named by:

- the hormones they secrete, if any
- their size
- the appearance of the tumor cells under a microscope

HORMONES

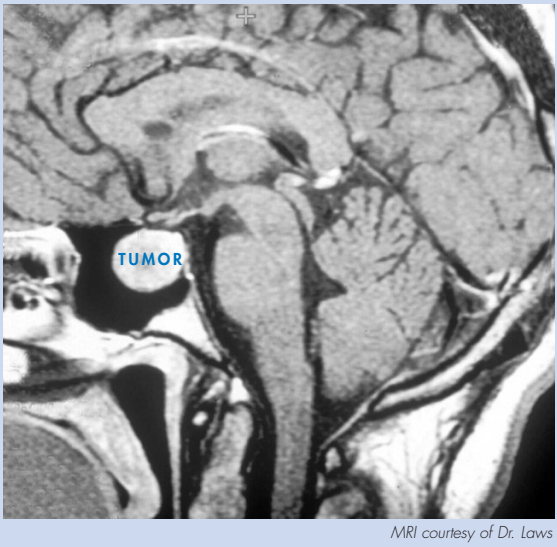
Some pituitary tumors inappropriately secrete excessive amounts of a particular hormone. These are known by several names including functioning adenomas, hormonally active adenomas, and secretory adenomas. "Functioning" or "secreting" tumors may cause the pituitary gland to ignore the signals from the hypothalamus, allowing the pituitary gland to secrete excessive amounts of

hormones such as prolactin (PRL), growth hormone (GH), adrenocorticotropic hormone (ACTH), or thyroid-stimulating hormone (TSH). Sometimes these tumors secrete more than one type of hormone. Other pituitary tumors do not oversecrete any active hormone at all and may even cause a slow down or a stoppage in hormone production (a condition called hypopituitarism). These tumors are commonly called nonfunctioning adenomas (NFAs). Other names for these non-secreting tumors include "hormonally inactive" or "non-secretory adenomas."

SIZE

Pituitary tumors are also classified by their size. Tumors appearing to be less than 10 mm (about 3/4") in diameter on an MRI scan are called microadenomas. Those larger than 10 mm are called macroadenomas.

MRI OF AN INTRASELLAR MACROADENOMA



MRI courtesy of Dr. Laws

MICROSCOPIC APPEARANCE

If your tumor is surgically removed, it will be examined by a pathologist — a doctor specially trained to look at tumor cells using a microscope. The pathologist will examine the sample of tumor tissue, and provide a report to your doctor. The

"pathology report" describes the hormone content, structure, and the cells that gave rise to your tumor. It usually takes about a week for your doctor to receive the surgical pathology report. All of this information is then used to determine your tumor type, form a treatment plan, and predict the possible future activity of the tumor.

Symptoms

Since almost 70% of pituitary tumors are functional, or secreting, tumors the most common symptoms are related to excess hormone production. Lack of menstrual periods (amenorrhea), production of breast milk without pregnancy (galactorrhea), excessive growth (acromegaly or gigantism), Cushing's Syndrome, and/or a hyperactive thyroid may be clues to the presence of a tumor in this gland. Headache, vision changes, sleep and eating disorders, and excessive thirst and urination (diabetes insipidus) may also be noted.

DO YOU THINK YOU HAVE A PITUITARY TUMOR?

If you have symptoms causing you concern, begin by making an appointment to see your family doctor or your primary care physician. Explain your symptoms, and be a good historian. How long have you been experiencing the symptoms? Have they changed any since your symptoms first began? Share your concerns and your medical history. Your doctor can order a few basic tests (blood work and/or a scan) that will help determine the next step. There are many diseases that can cause hormone-related symptoms; not all of them are tumors. Your primary care physician can help you sort through your symptoms, and make the appropriate referrals if necessary. An endocrinologist may be very helpful in clarifying the diagnosis and managing medical therapy.

Diagnosis

If your doctor suspects you have a tumor, several tests are available that can help determine the diagnosis. Special blood tests can determine your hormone levels and whether the pituitary gland is the source of any excess hormone.

Hormone	Responsible for…	Normal Blood Levels in Adults
ACTH adrenocorticotropic hormone	Controls the production of cortisol — a natural steroid needed to control blood pressure, sugar and salt levels	9 to 52 pg/ml
GH growth hormone	Controls bone growth; height; body proportion in the extremities and jaw	0 to 3 ng/ml
PRL prolactin	Controls milk production in women, impacts sex drive and sperm counts in men	Males and non-pregnant women: 0 to 20 ng/ml In pregnancy: 10 to 300 ng/ml
TSH thyroid stimulating hormone	Controls thyroid functions such as metabolism, heart rate and appetite	0.2 to 4.7 mcU/ml

Following a neurological exam and endocrine screening (blood hormone levels), an MRI scan with contrast dye is used to obtain images of the pituitary gland, the sella and the area around it. In some circumstances, scans of the chest or abdomen may be necessary to verify that the hormone imbalances are caused by the pituitary gland. An ophthalmologist — a doctor specializing in visual problems — may examine your eyes if the tumor affects your eyesight, and impairs peripheral vision

Sometimes, pituitary tumors are found "incidentally." This usually means the tumor was seen on an MRI scan ordered for another, unassociated medical reason such as a sports-related accident. These symptomless tumors require careful evaluation, but may not always need immediate treatment.

MRI OF A MACROADENOMA

MRI courtesy of Dr. Laws

Specific Tumors

PROLACTINOMAS, OR PROLACTIN-PRODUCING ADENOMAS

Prolactinomas represent about 30-40% of all pituitary tumors, making them the most common of these tumors. Prolactinomas are most often found in women of childbearing age. In men, prolactinomas are more frequent in the fourth and fifth decade of life. About half of these tumors are microadenomas, which are small tumors.

In women, high prolactin levels may cause menstruation to stop (amenorrhea) or inappropriate production of breast milk (galactorrhea) may develop. In men, prolactin-secreting tumors may cause a decreased sex drive and impotence. Men also tend to develop larger tumors which may cause headaches or vision problems.

GROWTH HORMONE-PRODUCING ADENOMAS

These tumors represent about 20% of pituitary adenomas. Growth hormone-producing tumors

are more common in men than in women. Often macroadenomas, these tumors may extend toward the cavernous sinus, an area of the brain located next to the pituitary. Mixed prolactin and growth hormone-secreting tumors are not uncommon.

Growth hormone-secreting tumors may cause gigantism in children and adolescents. In adults who have reached their full height, the hands, feet, and lower jaw become enlarged. This is called acromegaly. Excessive growth hormone can aggravate other medical conditions such as diabetes mellitus, hypertension, and heart disease.

ACTH-PRODUCING ADENOMAS

These tumors represent about 16% of the pituitary adenomas. They are much more common in women than in men. ACTH (adrenocorticotropic hormone) stimulates the adrenal gland to make and secrete glucocorticoids, which are natural steroids. Excess glucocorticoids cause Cushing's disease. Some of the symptoms of Cushing's disease are a moon-shaped face, excess hair growth on the body, bruising, menstrual irregularities, and high blood pressure.

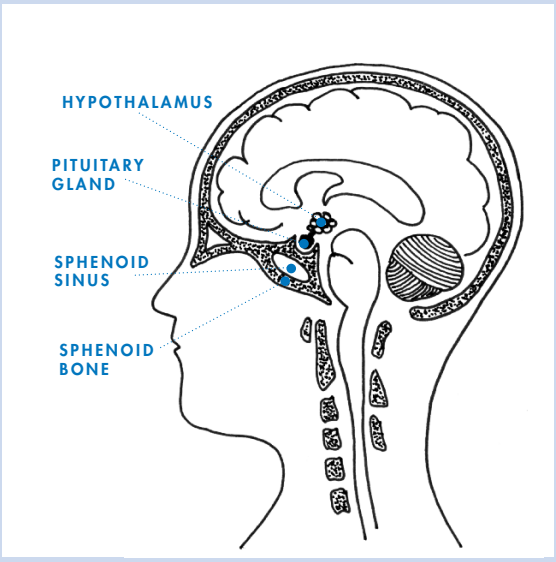
OTHER HYPERSECRETING PITUITARY ADENOMAS

This group represents less than 1% of pituitary adenomas. Some of these tumors excrete increased amounts of thyrotropin (thyroid-stimulating hormone). Others may secrete follicle stimulating hormone/luteinizing hormone (which controls the ovaries and testes) or alpha subunit (a glycoprotein hormone).

NON-SECRETING TUMORS

Also called non-functioning tumors, these represent about 25% of the pituitary adenomas. Null cell adenomas, oncocytomas, silent corticotroph adenomas, and gonadotroph adenomas fall into this group. These tumors grow slowly and generally cause minimal symptoms. They may be sizable

LATERAL VIEW OF THE BRAIN



before their presence is suspected. When they expand outside the sella turcica, they may press on the nearby optic nerves causing vision loss and headache. Such tumors can also compress the pituitary gland itself so it cannot produce and deliver its normal output of hormones. Called hypopituitarism, this symptom is associated with general weakness and fatigue, a pale complexion, loss of sexual function and apathy.

PITUITARY CARCINOMA

Cancers, or true malignancies of the tissue of the pituitary gland, are very rare. Instead, a "pituitary carcinoma" is usually defined as a tumor that begins in the pituitary gland and then metastasizes, or spreads, within the brain or outside the central nervous system. These tumors are generally macroadenomas which are resistant to therapy, recur locally and ultimately metastasize to the spinal canal or other organs of the body. The majority of pituitary carcinomas are functional tumors, secreting prolactin or ACTH.

Treatment

Treatment of a pituitary tumor depends on the hormonal activity of the tumor, the size and location of the tumor, as well as the age and overall health of the person with the tumor. The goals of treatment may be to remove the tumor, to reduce or control tumor size, and/or to re-balance hormone levels.

SURGERY

If your doctor recommends surgery, the goal will be to remove as much tumor as possible. A "transsphenoidal approach" — literally meaning "across the sphenoid sinus" — is the most common. During this surgery, extremely small instruments, microscopes and endoscopes are used to remove the tumor from within the nose (endonasal) or under the lip and above the teeth (sublabial). Less often, a craniotomy may be done, during which a portion of the skull bone is temporarily removed to gain access to the pituitary gland. Some surgeons use endoscopes (a long, thin tube-like instrument) to reach the tumor.

Your neurosurgeon will speak with you about the surgery planned for your tumor, the risks and benefits of the procedure, and your follow-up care. We offer a free publication, titled *Surgery*, which explains the different surgical tools available, how procedures are planned, and questions to ask your physician about the procedure recommended for you. If you would like a copy, please call us at 800-886-2282.

RADIATION THERAPY OR RADIOSURGERY

Radiation therapy is often used as a second, or adjuvant, treatment for pituitary tumors. It may be given in addition to surgery and/or drug therapy, Radiation therapy may be used to treat tumors that have re-grown, or it may be used for aggressive tumors. The goal of radiation therapy for pituitary tumors is to reduce or control tumor size; however, it may take several months or longer before the effects of this treatment cause a change in your

hormone levels or your MRI scan. There are several different types of radiation therapy; your doctor will decide which is best for your tumor. Conventional external beam radiation is "standard" radiation given 5 days a week for 5 or 6 weeks. Stereotactic radiosurgery is focused radiation therapy.

The Gamma Knife, LINAC (modified linear accelerators), CyberKnife, and proton beam radiation are all forms of stereotactic radiosurgery. Conformal photon radiation, also known as intensity-modulated radiation therapy, shapes radiation beams to the contours of the tumor. If your doctor thinks your tumor would be best treated with radiation, he/she can initiate an appropriate referral, and can speak with you about the type of therapy suggested and the effects of that particular treatment.

DRUG THERAPY

There are several drugs used to treat pituitary tumors. The drug chosen will depend on the hormone functions of the tumor.

Dopamine agonists, such as bromocriptine (Parlodel) or cabergoline are used to control the production of prolactin. The drugs reduce tumor size by reducing the amount of prolactin made by the pituitary gland. Most people with prolactin-secreting tumors require long term therapy to control the size of their tumor; generally, if the medication is stopped prolactin levels begin to increase. In a small percent of people with very small tumors, treatment may be stopped after a year or so to see if the tumor re-grows. Those with very high prolactin levels may find that drug therapy decreases their hormone levels but does not relieve all of their symptoms. In these situations, the drug successfully lowers the prolactin level but the level may still be higher than "normal," thus causing symptoms.

Somatostatin analogues such as octreotide (Sandostatin or Sandostatin LAR, or Lanreotide) can reduce growth hormone levels and relieve the

associated symptoms. These drugs may also be used to control the production of thyroid-stimulating hormone in thyrotrophic tumors. Dopamine agonists, such as those listed above, can be given to treat growth-hormone secreting tumors, but their effect is primarily relief of symptoms rather than reduction of hormone levels to normal.

A growth hormone receptor antagonist drug called pegvisomant is highly effective in normalizing a protein called IGF-1, found in elevated levels in people with acromegaly, but is not yet in general use. New drugs are being tested in research studies called clinical trials; information about those trials can be found through your endocrinologist.

Ketoconazole (Nizoral) is used to treat ACTH secreting tumors that produce Cushing's disease. This drug lowers cortisol (natural steroid) production, but generally does not reduce the size of the tumor or inhibit its activity, and is not suitable for long-term treatment.

Followup

Once you recover from surgery or radiation therapy, or begin medical treatment, your doctors will determine a schedule for follow-up MRI scans and endocrine testing. These are used to monitor the effectiveness of therapy, assure normal hormonal balance, and watch for possible tumor re-growth.

Many people with pituitary tumors are followed regularly by an endocrinologist — a physician specially trained in the treatment of disorders of the hormone-producing glands. Hormone imbalances can be caused by the tumor itself, or may result from the treatment necessary to control the tumor. The endocrinologist will monitor your hormone blood levels, outline a treatment plan, and make drug adjustments when needed. The endocrinologist becomes an active member of your healthcare team, working closely with your neurosurgeon and your primary care physician.

Although pituitary tumors are almost always benign, they can recur, and therefore periodic followup MRI scans are necessary. Your doctor will tell you how often those scans should be done. If you do not know when your next MRI should be scheduled, call your doctor's office to ask.

A Next Step

Support groups and pen-pal programs allow you to share experiences with others who have experienced a pituitary tumor. Internet resources, such as online chat rooms and e-mail groups, can be of benefit to those who do not have access to a local support group. Social workers can help you find these support networks. Nurses will provide you with information about how to monitor your health, especially when your treatment plan involves hormone therapies. Dieticians, working along with your endocrinologist, can design a healthy eating plan to ensure your body receives balanced nutrients. We can help you locate these resources in your area.

Our Web site — *www.abta.org* — offers extensive brain tumor information, treatment and research updates, and patient/family stories. The thread that runs through each of our services and programs is hope. Become involved — join us in some way, to make sure there is a cure, and ultimately, a way to prevent brain tumors.

We hope that the information in this pamphlet helps you communicate better with the people who are caring for you. Our purpose is not to provide answers; rather, we encourage you to ask questions.

This publication is not intended as a substitute for professional medical advice and does not provide advice on treatments or conditions for individual patients. All health and treatment decisions must be made in consultation with your physician(s), utilizing your specific medical information. Inclusion in this publication is not a recommendation of any product, treatment, physician, or hospital.

Questions I Want to Ask

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Publications & Services

BUILDING KNOWLEDGE
Dictionary for Brain Tumor Patients
Living with a Brain Tumor
A Primer of Brain Tumors

FOCUSING ON TUMORS
Ependymoma
Glioblastoma Multiforme and Anaplastic Astrocytoma
Medulloblastoma
Meningioma
Metastatic Brain Tumors
Oligodendroglioma and Oligoastrocytoma
Pituitary Tumors

FOCUSING ON TREATMENT
Chemotherapy
Conventional Radiation Therapy
Stereotactic Radiosurgery
Steroids
Surgery
Physician Resource List: Physicians Offering Clinical Trials for Brain Tumors

FOR & ABOUT CHILDREN
Adolescent and Young Adult Resources
Alex's Journey: The Story of a Child with a Brain Tumor *(DVD)*
Educating Children and Teenagers
Resources For Talking with Children When a Parent is Ill
When Your Child Returns to School

SUPPORT RESOURCES
Bibliography
Care Options
Caregiver Resources
Emergency Alert Wallet Cards
Employment Rights and Job Retraining Resources
End-of-Life Care
Financial Assistance Resources
Health Insurance Resources
Housing During Treatment Resources
Neuropsychology Resources
Scholarship & Educational Financial Aid Resources
Searching Medical Journals
Social Security Disability Resources
Spanish-Language Resources
Therapeutic Recreation Resources
Transportation Assistance Resources
Wig and Head Covering Resources
Wish Fulfillment Resources

NEWSLETTER
Message*line* Newsletter
ABTA Brain Tumor E-News

FOCUSING ON SUPPORT
Listing of Brain Tumor Support Groups
Listing of Bereavement (Grief) Support Groups
Organizing and Facilitating Support Groups
Pen Pal Programs
  Connections (program for patients and family members)
  Bridges (program for those who have lost someone to a brain tumor)
Reaching Out For Support
Resources for Online Support
TLC (Tips for Living and Coping) e-bulletin

Single copies of our publications are available free of charge.

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2720 River Road, Suite 146 Des Plaines, Illinois 60018

800.886.2282 TEL info@abta.org EMAIL
847.827.9918 FAX www.abta.org WEB

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