Focus on Tumors

Pituitary Tumors

American Brain Tumor Association
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 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 including the thyroid, the adrenals, testes, and ovaries. The pituitary gland releases hormones into the bloodstream, 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. They 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 multi-disciplinary health care approach, support, and follow-up.

**Incidence**

Pituitary tumors account for 7–15% 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). Those abnormalities most often do not cause symptoms and do not require medical or surgical therapy.
Pituitary tumors can be found in every age group, but their incidence increases with age up to age 74, at which it seems to peak. Functioning (also called secreting) tumors tend to 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 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 doctors refer to these tumors as “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”) on a scan are called microadenomas. Those larger than 10 mm are called macroadenomas.

**MICROSCOPIC APPEARANCE**

If your tumor is surgically removed, it will be sent to 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 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 water imbalances (diabetes insipidous) 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.

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.

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.

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.

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

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 the 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 14% 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 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 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 began in the pituitary gland then metastasized, or spread, within the brain or outside the central nervous system. These tumors are generally macroadenomas which are resistant to therapy, recur locally several times, then 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 bone”— is the most common. During this surgery, extremely small instruments and microscopes 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 an endoscope, a long, thin tube-like instrument, to reach the tumor.

Transsphenoidal approach: To the left is a profile view in the midline. The nasal speculum (only half seen) is placed inside the nose up to the level of the sphenoid sinus and points toward the tumor. The bone in front of the sphenoid sinus must be removed as well as the bone overlying the tumor. To the right are two common incisions made within the nose. The incisions cannot be seen and no incision is made on the face.

Diagram courtesy of Dr. Jane.
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 in detail 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. Intracavitary and interstitial radiation place the radioactive source directly into the tumor during surgery. If your doctor thinks your tumor would be best treated with radiation, he/she 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), pergolide (Permax), or cabergoline (Dostinex), 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), are used to both 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, are given to treat growth-hormone secreting tumors but their effect is primarily relief of symptoms rather than reduction of hormone levels. 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.
Followup

Once you begin treatment, your doctors will determine a schedule for follow-up MRI scans and endocrine testing. These are used to monitor the effectiveness of therapy and watch for possible tumor re-growth.

Many people with functioning pituitary tumors are followed 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.
Questions I Want to Ask


Questions I Want to Ask
Questions I Want to Ask
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
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Oligodendroglioma and Oligoastrocytoma
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FOCUSING ON TREATMENT
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Conventional Radiation Therapy
Stereotactic Radiosurgery
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Physician Resource List: Physicians Offering Clinical Trials for Brain Tumors

FOR & ABOUT CHILDREN
Alex’s Journey: The Story of a Child with a Brain Tumor (Video or DVD)
Education Packet (Parent or Teacher)
When Your Child Returns to School

SUPPORT RESOURCES
Bibliography
Care Options
Emergency Alert Wallet Cards
Employment Information
End-of-Life Care
Financial Aid Resources
Health Insurance Resources
Housing During Treatment Resources
Net-Working Links
Neuropsychology Resources
Scholarship & Educational Financial Aid Resources
Social Security Disability Resources
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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)
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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